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Review Article

Gorlin-Goltz Syndrome: Surgical Resolution of Keratocysts in 3 Generations of a Family. Case Presentation

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Abstract

Gorlin Goltz syndrome (GGS) was reported in 1984. It is a genetic disease with autosomal dominant inheritance. Its 3 typical characteristics are: basal cell carcinomas, multiple maxillary keratocysts (OCK) and bifid rib syndrome. Its diagnosis is established when 2 major criteria are met or one major and 2 minor criteria originally defined by Evans., *et al*. in 1993 and subsequently modified by Kimonis., *et al*. in 1997 and Bree., *et al*. in 2011.

Its treatment requires a multidisciplinary approach. The dentist is a main actor when multiple osteolytic lesions are found in the jaws in panoramic radiographs requested during the consultation.

The case of a family where 3 generations, a mother, 5 children and her grandson present clinical manifestations of the syndrome is presented below. These patients were treated for maxillary keratocysts and are still under treatment.

All surgeries performed have been conservative approaches so far.

Keywords: *Basal Cell Nevus Syndrome; Odontogenic Cysts; Congenital Anomalies; Medulloblastoma; Keratocyst; Gorlin-Goltz Syndrome*

Introduction

Gorlin Goltz syndrome (GGS) or basal cell nevus carcinoma syndrome is a genetic disease of autosomal dominant inheritance that predisposes to the presence of developmental defects and the development of neoplasia's [1,2].

The syndrome has received different names since it was first reported by Jarish and White in 1894, until in 1960 Gorlin and Goltz described and gave a name to this entity, defining its 3 typical characteristics: basal cell carcinomas, multiple odontogenic keratocysts (OCK) and bifid rib syndrome [3-5].

It has an approximate average prevalence of 1:60,000 and affects men and women equally, without distinction of race [6]. The life expectancy of GGS patients is 73.4 years, significantly lower than that of the general population, which is approximately 80 years. The most important cause of premature death in these patients is medulloblastoma, a primary tumor of the central nervous system, which usually affects young adults aged 20 to 30 years and can originate in the brain or spinal cord [2].

The inheritance is caused by a mutation in the tumor suppressor gene PTCH1, located on the long arm of chromosome 9 (9q22.3-q31), a gene responsible for controlling normal tissue growth and development. This mutation is transmitted in an autosomal dominant manner with a high degree of penetrance (approximately 97%) and variable expressivity. These genetic defects can be transmitted from one generation to another and manifest in several family members [7].

GGS is characterized by clinically presenting a predisposition to the development of multiple neoplasia's and developmental abnormalities. The appearance of these generally occurs in childhood or adolescence [8]. Initially, the triad that characterizes the syndrome was described: multiple basal cell carcinomas, maxillary keratocysts and bifid ribs. In addition to this triad, calcifications of the falx cerebri, palmar and plantar pits, spinal and rib anomalies, relative macrocephaly, facial milia, frontal protuberances, ocular malformations, medulloblastomas, cleft lip and/or palate are also found [9,10]. This is why early diagnosis and multidisciplinary treatment are important [11,12].

Its diagnosis is established when 2 major criteria or one major and 2 minor criteria are met [13]. These criteria were originally defined by Evans., *et al*. in 1993 and later modified by Kimonis., *et al*. in 1997 and Bree., *et al*. in 2011 [14].

Major criteria

1. Excessive number of basal cell carcinomas disproportionate to previous sun exposure and skin type or < 20 years of age.
2. Odontogenic keratocysts of the jaws before age 20 years.
3. Palmar or plantar pits.
4. Lamellar calcification of the falx cerebri.
5. Medulloblastoma, typically desmoplastic.
6. First-degree relative with Gorlin-Goltz syndrome.

Minor criteria

1. Rib abnormalities.
2. Other specific skeletal malformations and radiologic changes (i.e., vertebral abnormalities, kyphoscoliosis, short fourth metacarpal, postaxial polydactyly).
3. Macrocephaly.
4. Cleft lip and/or cleft palate.

5. Ovarian/cardiac fibroma.
6. Lymphosenteric cysts.
7. Ocular abnormalities (i.e., strabismus, hypertelorism, congenital cataracts, glaucoma, coloboma).

Genetic testing for PTCH1 is suggested in the following situations:

1. Confirmation of diagnosis in patients lacking sufficient clinical diagnostic criteria.
2. Predictive testing for at-risk patients with an affected family member but who do not meet clinical criteria.
3. Prenatal testing in the presence of a known familial mutation [15].

Keratocyst

The keratocyst was first described by Philipsen in 1956; it comprises 10 to 12% of all odontogenic cysts. It is derived from the remains of the dental lamina (Serres remains) and is benign in origin [16] although it has an aggressive clinical behavior, is highly recurrent, and has specific histological characteristics [17-20].

The term "odontogenic keratocyst" was used in the classification of odontogenic lesions published by the World Health Organization in 1992. The 2005 classification reclassified this unique lesion as a neoplasm and renamed it "keratocystic odontogenic tumor" because of the high recurrence rate, aggressive clinical behavior, association with nevus basal cell carcinoma syndrome, and mutations in the PTCH tumor suppressor gene. The 2017 classification returned to the original and well-accepted terminology of odontogenic keratocyst because many articles demonstrated that the PTCH gene mutation could be found in non-neoplastic lesions including dentigerous cysts, and, furthermore, many investigators suggested that the cyst resolution after marsupialization was not compatible with a neoplastic process [21-26]. In the 2022 edition, it continues in the classification of cysts and has the longest section among the cysts of the jaw [26].

Most of these lesions are usually single, but exceptionally they can occur multiple, being called Gorlin-Goltz syndrome or also known as nevoid basal cell carcinoma syndrome [16]. A recurrence of 28-35% is reported as a solitary lesion and 2.5 - 62.5% related to nevoid basal cell carcinoma [20].

The keratocyst occurs most frequently in the second decade of life, has a 2:1 predilection for the male sex in relation to the female; It affects 60 - 80% of the mandible, mainly the region of the lower third molar and ascending ramus (28.8 - 30%), the region of the upper third molar (12.3 - 15%), the region of the first and second mandibular molars (8.6 - 9%), the region of the upper canine (12.3 - 15%) and the region of the lower premolars. Its size varies from 1 to 7 cm and can extend to several more centimeters, extending to the midline and base of the skull [1,27,28].

It is generally asymptomatic, with a long evolution, slow and expansive growth, but in 10% of cases it is accompanied by pain. It can be detected as a finding during a routine radiological examination; clinically, the first sign is expansion of the cortical bones, which causes an increase in volume, crepitation on palpation, due to thinning of the cortical bones, and facial asymmetry. There may be discharge of purulent material. The mucosa has a normal appearance and color. It can cause paresthesia in the lower lip if the affected area is the lower jaw [1,27,28].

Histopathologically, the OCK is characterized by the following features: 1) thin and uniform coating of parakeratinized squamous epithelium, usually 6 to 10 cells thick; 2) a wavy layer of parakeratin on its luminal surface; 3) a palisade layer of cuboidal or prismatic basal cells; 4) absence of papillae; and 5) the lumen of the cyst is characterized by having variable amounts of desquamated parakeratin [29,30]. Other features to take into account in the histopathology are the presence of dental lamina residues and the formation of microcysts or satellite cysts, which is usually related to the possible recurrence of the OCD. According to the literature, this can vary between 0% and 50% [27-32].

Radiographically it presents as a well-defined radiolucent area, with a thin mare sclerotic bone gene, if there is no added infectious process. It can be unilocular or multilocular and is associated in 25 - 40% to a retained tooth, 24% not associated to a tooth and 12% associated to a previous extraction area; in rare cases it has a "soap bubble" or "honeycomb" appearance; it can present displacement (divergence) of the roots, extrusion of adjacent teeth, rhizoclasia, tooth mobility in rare cases and occasionally paresthesia of the lower lip [1,27,33,34].

Regarding its differential diagnosis, there is a wide group of lesions that present similar characteristics, including some such as: dentigerous cyst, calcifying odontogenic cyst, ameloblastoma, adenomatoid odontogenic tumor, ameloblastic fibroma and myxoma. It is even more extensive when a solitary cystic lesion is not closely associated with the adjacent dentition; thus, traumatic bone cyst, radicular cyst, lateral periodontal cyst, residual cyst, nasopalatine cyst, central giant cell granuloma, cherubism, brown tumor of hyperparathyroidism, vascular malformations, benign bone tumors and plasmacytomas can be found [20].

There are several treatment modalities that can be classified into non-conservative (radical) treatments and conservative treatments accompanied by adjuvant methods. Among the non-conservative or radical treatments we find en bloc resection, which is the most aggressive way of treating a keratocyst; however, it has been shown to be the most effective in preventing recurrence of the lesion. Conservative treatments include marsupialization, decompression, and enucleation, with or without adjuvant therapy (cryotherapy, peripheral osteotomy, or application of Carnoy's solution) [16,29,32].

The choice of treatment is based on the histopathological diagnosis, location of the lesion in relation to its nearby bone structures, its aggressiveness and size, and possible involvement of dental structures, among others. It is important to highlight the multidisciplinary work for diagnosis and treatment planning [35,36].

Most recurrences of keratocysts can be diagnosed in the first two or three years of follow-up [37] however, there are reports of recurrence in the period of five to seven years after treatment. For this reason, annual follow-up with imaging tests is indicated during the first five years. After that period, tests should be performed every two or three years [38].

Presentation of Clinical Situations

For the present investigation, the rights of the patient were fundamentally protected, first, under the consent signed by the mother of the represented, by the patients of legal age and the authorization in the teaching area of the General Interzonal General Acute Hospital Perón, respecting the ethical principles based on the Declaration of Helsinki.

Note: To identify each one of them, and protect their identity, they will be named by situation and their corresponding age.

A 23-year-old female patient (patient age 23) was presented at the dentistry service of the "President Perón General Interzonal General Acute Hospital in Avellaneda", accompanied by her mother, referred from a peripheral ward, due to presenting an acute abscess in the lower right jaw, of 3 days' evolution, medicated for 6 days with cephalexin 500 mg every 12 hours.

At the time of the anamnesis, the patient reported hypothyroidism, with no current treatment, and transient obstructive hydrocephalus at birth.

During intraoral inspection, an acute abscess was observed in the lower maxillary vestibular groove between teeth 4.4; 4.3; 4.2, painful, accompanied by spontaneous purulent exudate through the gingival groove, bulging of the bone cortex, soft to the touch, depressible and crepitant, with normal-colored mucosa (Figure 1 and 2). Vitality tests were taken from teeth 4.5 to 3.5 inclusive, and were positive.

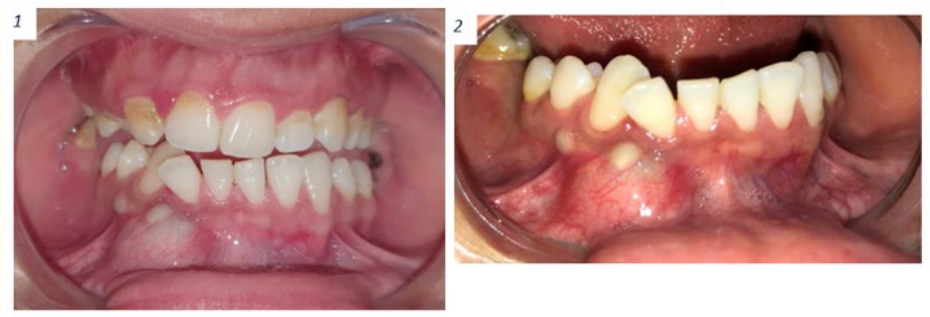


Figure 1 and 2: *Abscess in the right vestibular groove at the level of tooth 42 to 44 at the time of the first consultation of the patient 23.*

Extraoral cervicofacial inspection revealed marked hypertelorism, scoliosis, abnormal implantation of the left and right ear, and no cervical lymphadenopathy upon palpation (Figure 3A-3C).

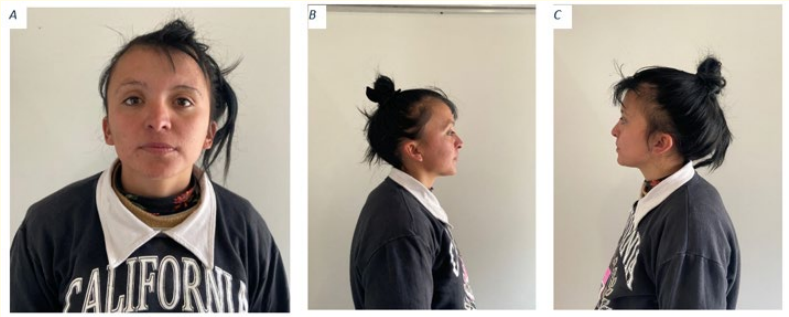


Figure 3A-3C: *Photograph of the front (A) and right (B) and left (C) profiles, where abnormal implantation of the ears, hypertelorism, macrocephaly, scoliosis can be observed.*

A panoramic x-ray (Figure 4) and a tomography of the craniofacial mass (Figure 5A-5C) were taken, where a multilocular, radiolucent lesion with indefinite radiopaque edges, measuring 8 x 3 cm, was observed in the lower jaw, extending from the distal part of tooth 35 to the distal part of tooth 45, with displacement of the same and from the alveolar ridge to the base of the mandible. It presents tooth 48 included in the right ascending ramus, with a radiolucent image at the coronal level, measuring 0.5 cm x 0.5 cm. In the upper jaw, a radiolucent image, 1.5 cm x 1.5 cm, associated with included tooth 13, unilocular, with defined radiopaque edges. A frontal chest x-ray showed spina bifida, scoliosis and fused vertebrae (Figure 6). Tomographic studies show a ventriculoperitoneal shunt valve placed in childhood due to hydrocephalus (nonfunctional as assessed by neurosurgery) and calcifications of the cerebral falx (Figure 7A and 7B).

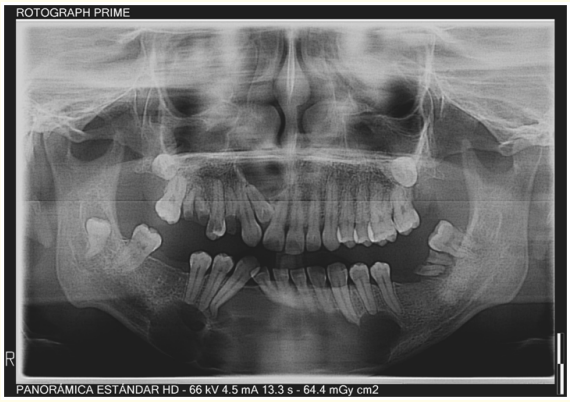


Figure 4: *Panoramic radiograph of the patient 23.*

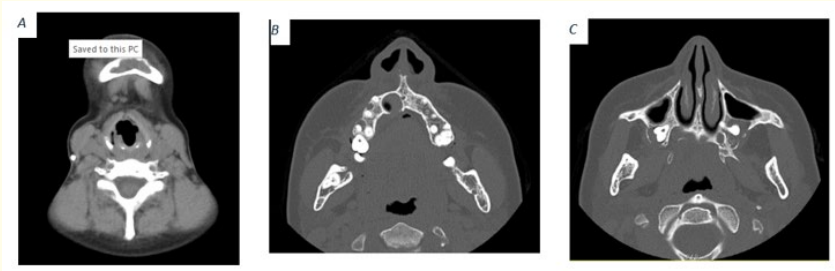


Figure 5A-5C: *Axial section showing a radiolucent lesion in the lower jaw (Figure A); axial section showing a radiolucent lesion in the upper jaw in the region of retained tooth 23 (Figure B); axial section showing a left and right radiolucent lesion associated with third molars (Figure C).*

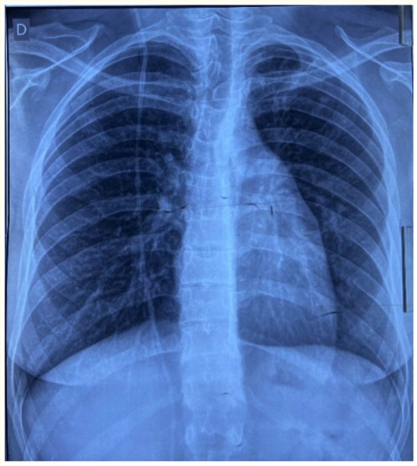


Figure 6: *Frontal chest X-ray, showing scoliosis, spina bifida and fused vertebrae.*

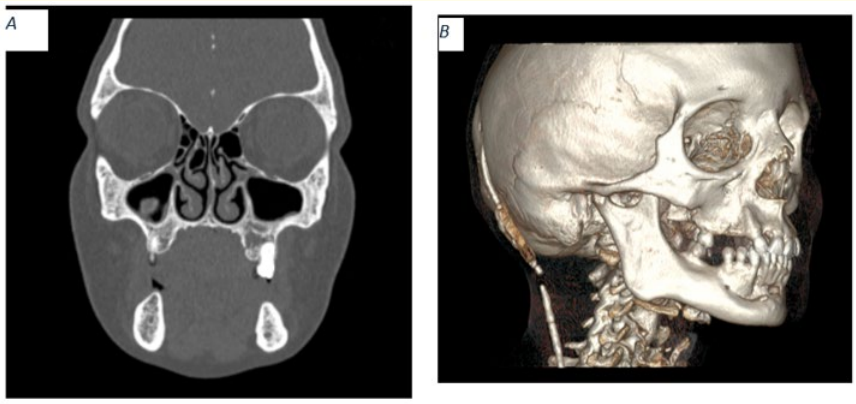


Figure 7A and 7B: *Axial section showing calcification of the cerebral falx (Figure A); 3D reconstruction showing the ventriculoperitoneal shunt valve (Figure B).*

Due to the presence of multiple radiolucent lesions, the mother of the patient present at the consultation was questioned. In the anamnesis, she reports being under treatment with the surgery and ophthalmology service since she was operated on in 2019 and 2020 for presenting facial basal cell carcinomas, on the back, torso and eyelids (Figure 8A and 8B).

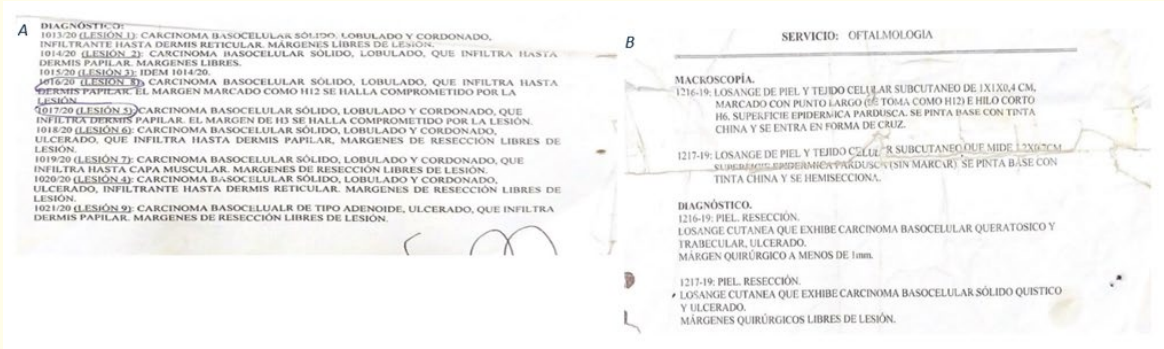


Figure 8A and 8B: *Pathological results of biopsies with a diagnosis of basal cell carcinoma performed by the surgery department (Figure A) and the ophthalmology department (Figure B).*

An intraoral evaluation was performed, which revealed poor oral condition, presence of root remains and poor hygiene. Extraoral inspection revealed hypertelorism, palmar and plantar pits (Figure 9), basal cell carcinomas on the hands (Figure 10), scars from basal cell carcinoma surgeries; palpation revealed no adenopathy.

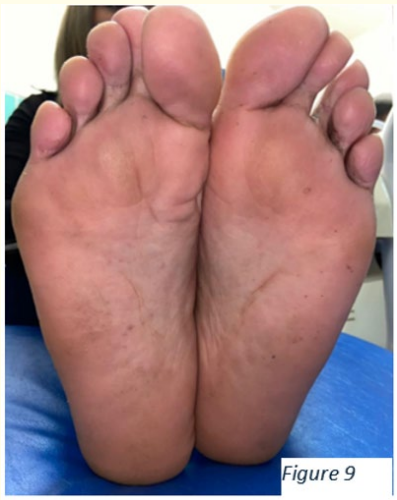


Figure 9: *Plantar feet.*

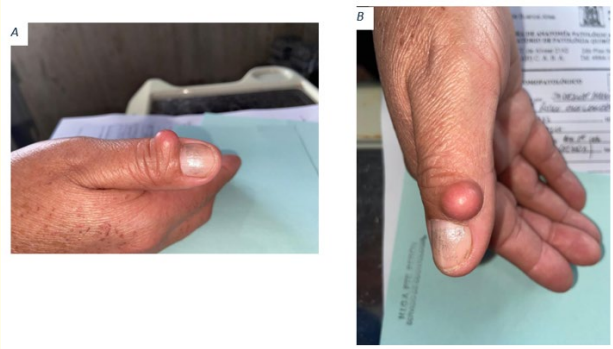


Figure 10A and 10B: *Basal cell carcinoma on the thumb of the mother's right hand.*

A panoramic radiograph was taken (Figure 11) and a CT scan of the craniofacial mass was requested (Figure 12A-12C), where a radiolucent lesion was observed in the ascending branch of the right maxilla, measuring 3.8 cm X 2 cm, unilocular with indefinite edges.

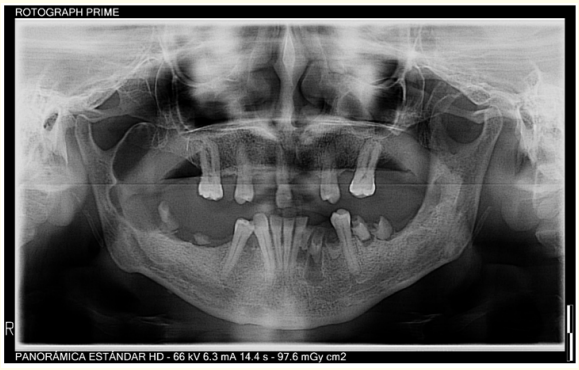


Figure 11: *Panoramic radiograph of the patient.*

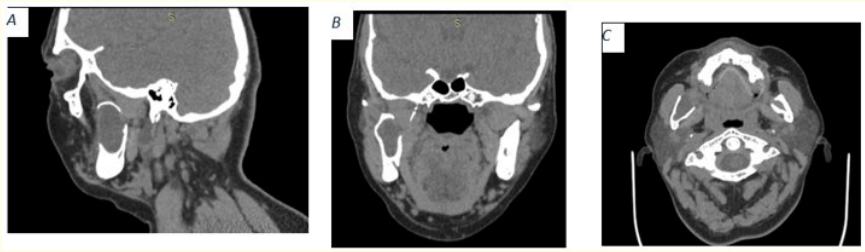


Figure 12A-12C: *Sagittal section (Figure A), coronal section (Figure B) and axial section (Figure C) of the mother's tomography.*

Given the findings of radiographic lesions, basal cell carcinoma, hydrocephalus, palmoplantar pits (mother and daughter), and hypertelorism (mother and daughter), Gorlin Goltz syndrome was immediately suspected, so the mother was asked to attend with the rest of her children for the relevant evaluation.

Three male and two female children from two marriages were evaluated clinically and radiographically. One of the patient's daughters did not present clinical or radiographic signs from an odontogenic point of view, but the dermatological evaluation detected basal cell nevi and ovarian fibroma; while the other female daughter did not present signs compatible with the syndrome in the clinical or radiographic evaluation. The 3 children, aged 5, 15, and 22, were found to have clinical and radiographic signs (Figure 13).

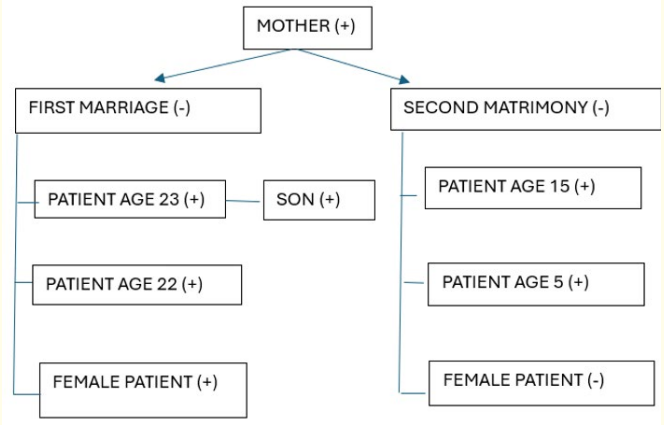


Figure 13: *Graph illustrating the positivity (+) and negativity (-) of signs in children and parents.*

In the 5-year-old patient, good oral condition is observed clinically, with mixed dentition. Radiographically (Figure 14) there is a unilocular radiolucent lesion measuring 2.6 x 2.4 cm, in the upper right jaw, at the level of the tuberosity, which displaces tooth 1.7 superiorly.

The 15-year-old patient reported in the anamnesis an atrial septal defect under treatment, pulmonary hypertension not detected in the last echodoppler.

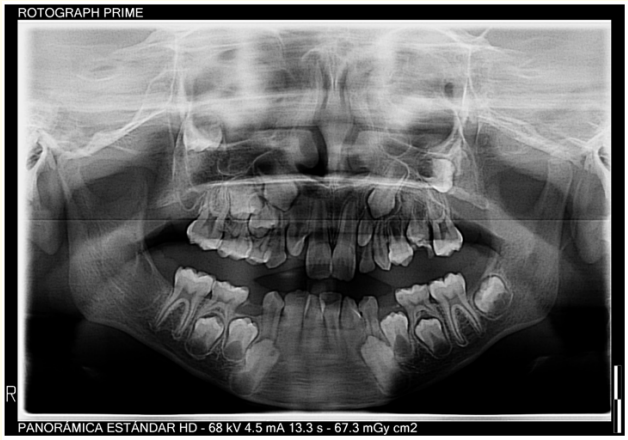
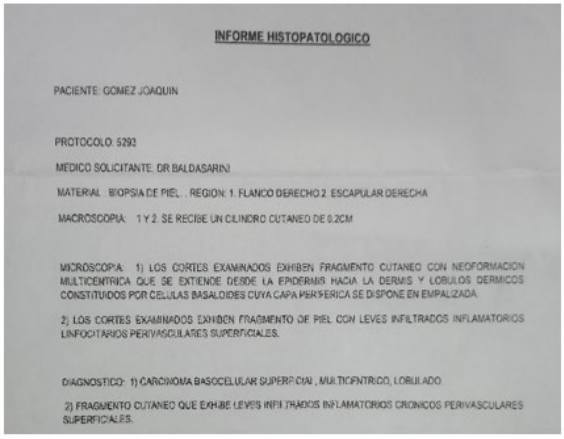


Figure 14: *Panoramic radiograph of the 5-year-old patient.*

During the extraoral inspection, he presented hypertelorism, palmoplantar pits, basal cell carcinomas in the upper limbs (Figure 15). Radiographically (Figure 16) and tomographically (Figure 17A-17F) a radiolucent, asymptomatic lesion with defined, radiopaque edges distal to tooth 37 was observed, which displaces tooth 38 towards the mandibular base, measuring 3 x 2 cm; an asymptomatic radiolucent lesion in the mandibular symphysis, associated with included tooth 43, multilocular, located between tooth 45 and 32, measuring 4 x 2 cm, causing their displacement, with positive vitality; and a radiolucent lesion associated with the crown of retained tooth 48. The frontal chest X-ray shows spina bifida and vertebral fusion (Figure 18).



INFORME HISTOPATOLOGICO

PACIENTE: GOMEZ JOAQUIN

PROTOCOLO: 5293

MEDICO SOLICITANTE: DR BALDASARINI

MATERIAL: BIOPSIA DE PIEL. . REGION: 1. FLANCO DERECHO 2. ESCAPULAR DERECHA

MACROSCOPIA: 1 Y 2. SE RECIBE UN CILINDRO CUTANEO DE 0.2CM

MICROSCOPIA: 1) LOS CORTES EXAMINADOS EXHIBEN FRAGMENTO CUTANEO CON NEOFORMACION MULTICENTRICA QUE SE EXTIENDE DESDE LA EPIDERMIS HACIA LA DERMIS Y LOBULOS DERMICOS CONSTITUIDOS POR CELULAS BASALOIDES CUYA CAPA PERIFERICA SE DISPONE EN EMPALIZADA.

2) LOS CORTES EXAMINADOS EXHIBEN FRAGMENTO DE PIEL CON LEVES INFILTRADOS INFLAMATORIOS LINFOCITARIOS PERIVASCULARES SUPERFICIALES.

DIAGNOSTICO: 1) CARCINOMA BASOCELULAR SUPERFICIAL, MULTICENTRICO, LOBULADO.

2) FRAGMENTO CUTANEO QUE EXHIBE LEVES INFILTRADOS INFLAMATORIOS CRONICOS PERIVASCULARES SUPERFICIALES.

Figure 15: *Result of biopsy performed by dermatology, with diagnosis of basal cell carcinoma.*

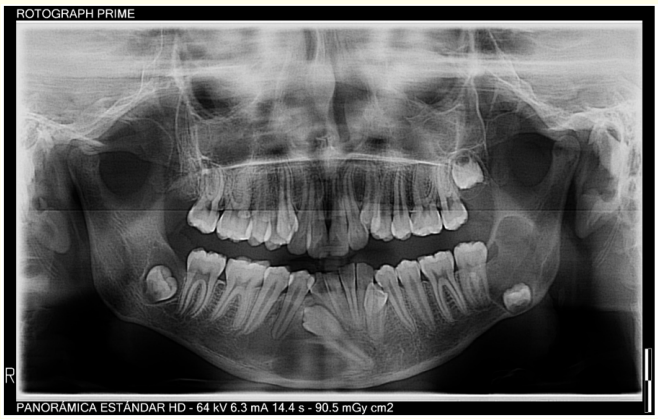


Figure 16: *Panoramic radiograph of the patient age 15.*

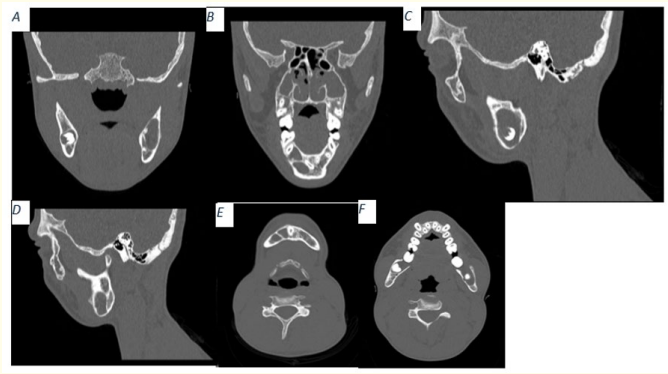


Figure 17: *Coronal section showing a radiolucent lesion in the left and right maxillary branches associated with impacted teeth (Figure A); coronal section showing a radiolucent lesion in the anteroinferior sector associated with an impacted tooth (Figure B); sagittal section of the left maxillary branch (Figure C); sagittal section of the right maxillary branch (Figure D); axial sections of the anteroinferior sector and the left right maxillary branch (Figure E and F).*

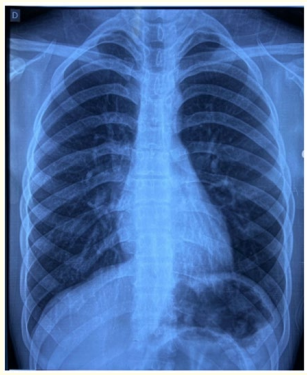


Figure 18: *Frontal chest X-ray, showing scoliosis, spina bifida and fused vertebrae.*

The 22-year-old patient also presented upon extraoral inspection hypertelorism, palmoplantar pits, and scars from basal cell carcinomas in the upper limbs (Figure 19A-19D). Radiographically (Figure 20) and tomographically (Figure 21), a unilocular radiolucent lesion with defined edges, radiopaque, measuring 2.7 x 2 cm, asymptomatic, associated with impacted and superiorly displaced tooth 23 was observed in the right upper jaw; in the left lower jaw, there was an asymptomatic unilocular radiolucent lesion in the mandibular ramus, with defined edges, associated with retained tooth 38; in the left maxillary body, a unilocular radiolucent lesion apical to teeth 44 and 45 measuring 1 x 0.8 cm; and a unilocular radiolucent lesion, 3.3 X 2.3 cm, in the body and maxillary branch, associated with tooth 45, 46, 47 (positive vitality) and 48. The frontal chest X-ray also shows spina bifida and Sprengel deformity (Figure 22).

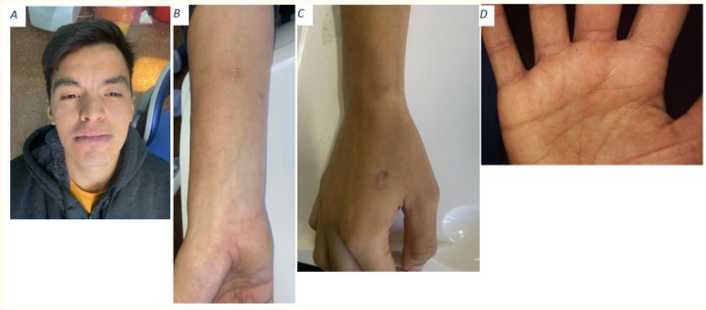


Figure 19A-19D: *Hypertelorism, macrocephaly, abnormal implantation of the ears are observed (Figure A); scars after removal of basal cell nevi (Figure B and C); palmar pits (Figure D).*

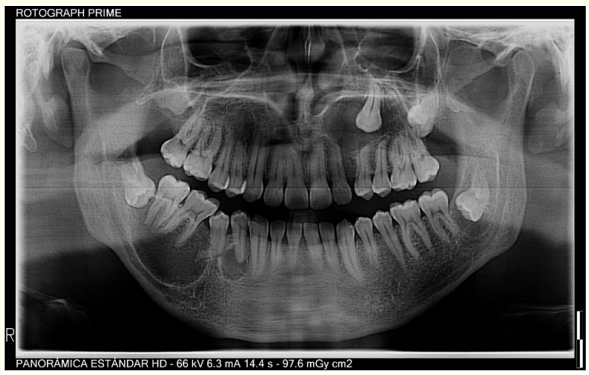
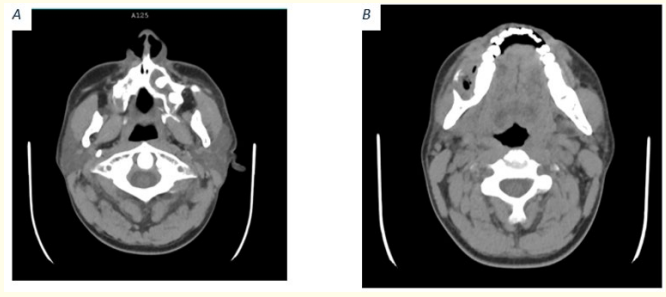


Figure 20: *Panoramic radiograph of the patient age 22.*



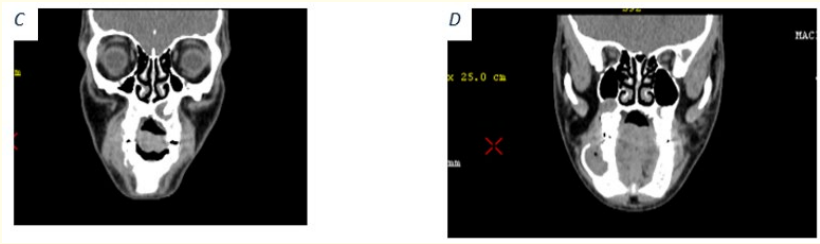


Figure 21: *Axial and sagittal sections of patient age 22 axial section of the lower jaw (Figure A); axial section of the lower jaw (Figure B); coronal section of the upper jaw (Figure C); coronal section of the lower jaw (Figure D). Both parents were evaluated clinically and radiographically and no clinical or radiographic signs were found.*

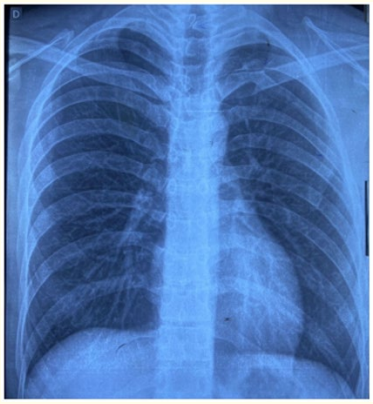


Figure 22: *Chest X-ray from the front showing spina bifida and vertebrae.*

The 23-year-old patient was asked to perform the clinical and radiographic evaluation of her 10-year-old son. No clinical or radiographic manifestations of the syndrome were found from the odontogenic approach. Extraoral inspection and general evaluation of the patient showed a marked Sprengel deformity, hypertelorism, scoliosis, abnormalities in the implantation of the ears, and pectum excavatum (Figure 23A-23D).

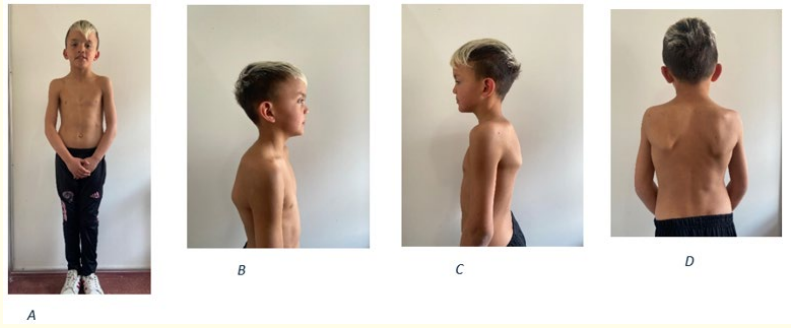


Figure 23A-23D: *Images from the front, right and left profile, and back, showing Sprengel deformity, hypertelorism, scoliosis, and ear implantation abnormality.*

After the clinical and radiographic evaluation, the patients were asked to undergo laboratory tests, a frontal chest X-ray, a pre-surgical cardiological and anesthetic evaluation, and a tomography of the craniofacial massif. Once these were completed, the corresponding biopsy was taken to arrive at a definitive diagnosis and plan treatment. Consultations were held with the neurology, ophthalmology, otorhinolaryngology, traumatology and dermatology services.

Surgical intervention

Patient age 23

Total surgical excision of the included tooth 13 and associated radiolucent lesion was performed under local anesthesia, and a sample was taken from the radiolucent lesion of the lower jaw together with the placement of a decompressive drainage tube.

Procedure

Intra- and extraoral field antisepsis with 10% povidone-iodine; placement of surgical fields; infiltrative anesthesia of the anterior dental nerve; nasopalatine and anterior palatine with 4% carticaine hydrochloride - adrenaline 1=100,000; intracrevicular incision from tooth 15 to tooth 21 with a Parker No. 3 Bard type scalpel with No. 15 blade; debonding of the vestibular and palatal mucoperiosteal flap; placement of a palatal tractor stitch; extraction of tooth 13 in the vestibule-palatine position with a straight elevator; total excision of associated pathology; treatment of the cavity with sterile physiological solution; Reposition and suture of the flap with 3/0 nylon with a simple stitch (Figure 24A-24D).

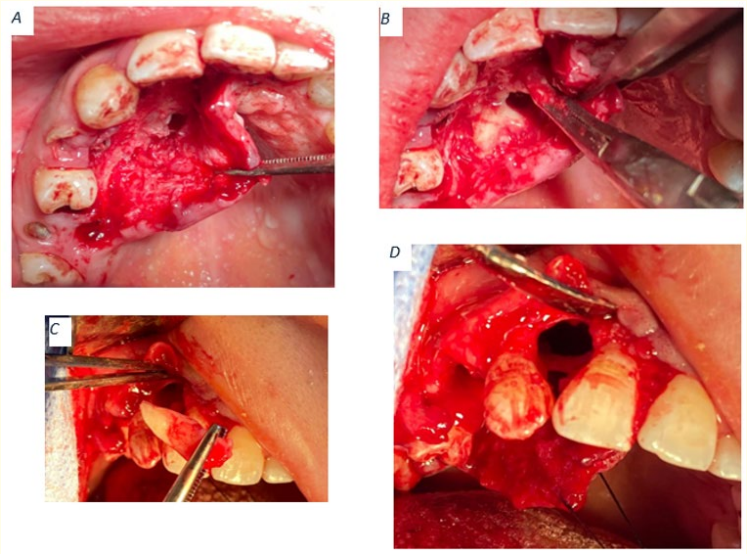


Figure 24A-24D: *Surgical sequence of the upper jaw. Intracrevicular incision and palatal flap detachment (Figure A); taking of excisional biopsy sample (Figure B); extraction of retained tooth 13 (Figure C); surgical lodge (Figure D).*

In the lower jaw, bilateral anesthesia of the lower dental and lingual nerve trunks and infiltration of the mental and bilateral incisive nerves; intracervicular incision from distal of tooth 45 to distal of tooth 35; full-thickness flap trimming; preservation of mental nerves; puncture and aspiration with a Luer-type syringe and a 40/30 needle to obtain whitish contents; ostectomy with a fenestrated cortical bone gouge to access the lesion; taking of an incisional biopsy with a cold scalpel; placement of a drainage tube from mesial of tooth 45 to mesial of tooth 35; toileting and hemostasis of the wound; reposition and suture of the flap with a simple interproximal stitch with 3/0

nylon; fixation of the drainage tube with a simple stitch; Placement of a compressive chin bandage (Figure 25A-25F). Amoxicillin 875 mg + clavulanic acid 125 mg was indicated; dexamethasone 8 mg intramuscularly; chlorhexidine gel 2%; lavages through the decompression tube with chlorhexidine 0.12%, physiological solution and iodine povidone 10%; placement of extraoral cold in the operating area; control for 24 hours; 72 hours and 10 days for suture removal.

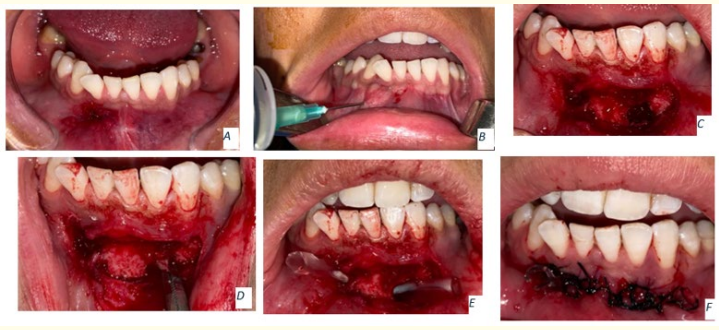


Figure 25A-25F: *Surgical sequence of first intervention in the lower jaw. Surgical site (Figure A), puncture and aspiration of the lesion (Figure B), detachment of the mucoperiosteal flap, ostectomy, sampling and placement of decompressive drainage (Figure C-E); repositioning and suturing of the flap and drainage (Figure F).*

The samples were fixed in 10% formalin and sent along with the corresponding protocol to the pathological anatomy service of the National University of Buenos Aires (UBA) School of Dentistry (FOUBA).

Pathological report (Figure 26)

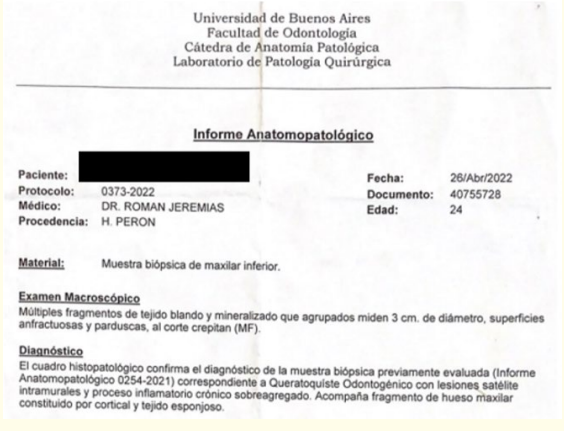
Macroscopic examination

Upper maxilla (1): Multiple soft tissue fragments, some of them with a membranous appearance, which together measure 1.5 cm in diameter, with an irregular and brownish surface, friable when cut (MF).

Lower maxilla (2): Soft tissue fragment with a membranous appearance that measures 1 x 0.7 x 0.1 cm, with an irregular and whitish surface, elastic consistency when cut (3F).

Diagnosis

The histopathological picture corresponds to an odontogenic keratocyst, with 1 intramural satellite lesion (MLP) observed in the biopsy sample.



Universidad de Buenos Aires
Facultad de Odontología
Cátedra de Anatomía Patológica
Laboratorio de Patología Quirúrgica

Informe Anatomopatológico

Paciente: [redacted]
Protocolo: 0373-2022
Médico: DR. ROMAN JEREMIAS
Procedencia: H. PERON
Fecha: 26/Abr/2022
Documento: 40755728
Edad: 24

Material: Muestra biópsica de maxilar inferior.

Examen Macroscópico
Múltiples fragmentos de tejido blando y mineralizado que agrupados miden 3 cm. de diámetro, superficies anfractuosas y parduscas, al corte crepitan (MF).

Diagnóstico
El cuadro histopatológico confirma el diagnóstico de la muestra biópsica previamente evaluada (Informe Anatomopatológico 0254-2021) correspondiente a Queratoquiste Odontogénico con lesiones satélite intramurales y proceso inflamatorio crónico sobreagregado. Acompaña fragmento de hueso maxilar constituido por cortical y tejido esponjoso.

Figure 26: *Lower jaw biopsy result from first intervention.*

Second surgical intervention

Total enucleation of the lesion in the lower jaw was performed under general anesthesia, one year after the placement of the drainage tube with periodic radiographic controls.

Under nasotracheal anesthesia, the surgical field was antisepsised with 10% povidone iodine; drape placement; pharyngeal pack placement; infiltrative anesthesia with lidocaine + epinephrine dilution 1/10; intracrevicular incision from piece 45 to 35; mucoperiosteal flap detachment and preservation of mental nerves; drainage tube removal; vestibular access ostectomy with a low-speed #8 round burr with profuse aqueous irrigation (sterile physiological solution); total excision of the lesion; cavity milling; placement of Carnoy's solution for 5 minutes and lavage with physiological solution; hemostasis and toileting of the wound (Figure 27); The flap was replaced and sutured with a simple 3/0 Vicryl stitch; the pharyngeal pack was removed and the patient was then transferred to the operating room.

Cephalothin 1g was administered intravenously every 6 hours, ketorolac 30 mg every 8 hours, dexamethasone 2 ml IV at 12 hours and a hydration plan of 28 drops per minute. Monitoring and discharge at 24 hours; outpatient monitoring at 24 hours, 72 hours and 7 days. The sample was fixed in 10% formalin and sent to FOUBA UBA. Cephalexin 1g was prescribed every 12 hours for 10 days; flurbiprofen 200 mg every 12 hours for mild pain; ketorolac 10 mg sublingual for acute pain; chlorhexidine 2% antiseptic gel.

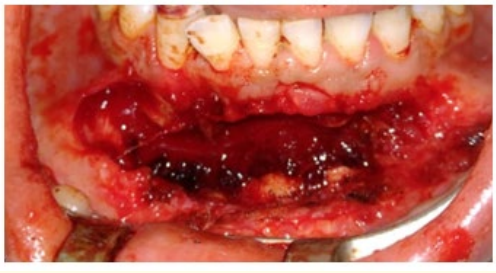


Figure 27: *Surgical Lodge after excision of cystic pathology.*

Result of the second intervention (Figure 28)

Multiple fragments of soft and mineralized tissue that together measure 3 cm in diameter, with an irregular and brownish surface, and crepitate (MF) when cut.

The histopathological picture confirms the diagnosis of the previously evaluated biopsy sample (anatomy-pathological report 0254-2021) corresponding to odontogenic keratocyst with intramural satellite lesions and chronic inflammatory process added. It is accompanied by a fragment of maxillary bone consisting of cortical and spongy tissue.

Informe Anatomopatológico

Paciente: [redacted] **Fecha:** 18/Mar/2021
Protocolo: 0254-2021 **H.Clínica/DNI:** 40755728
Médico: DRES. ROMAN J./MOSCA C. **Edad:** 23
Procedencia: H. PTE.PERON

Material: Muestras biópsicas de maxilar inferior.

Examen Macroscópico

1) Múltiples fragmentos de tejido blando algunos de ellos de aspecto membranoso que agrupados miden 1, 5 cm. de diámetro, superficies anfractuosas y parduscas, al corte friables (MF).
2) Fragmento de tejido blando de aspecto membranoso que mide 1 x 0,7 x 0,1 cm., superficies anfractuosas y blanquecinas, al corte consistencia elástica (3F).

Diagnóstico

1 y 2) El cuadro histopatológico corresponde a Queratoquiste Odontogénico, observándose en la muestra biópsica 1 lesiones satélite intramurales. (MLP)

Dra. María Luisa Paparella
Profesora Adjunta
Cátedra Anatomía Patológica

Figure 28: *Result of lower jaw biopsy from second intervention.*

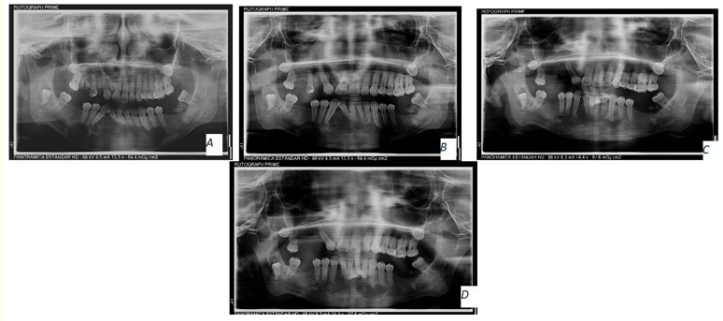


Figure 29A-29D: *Control radiographic sequence of patient age 23. Initial image 11/3/2021 (Figure A); Control X-ray after excision of lesion in the upper jaw and control of lower decompression 29/7/21 (Figure B); Control X-ray after surgery of the lower jaw 10/4/23 (Figure C); last control X-ray showing recurrence in sector III 16/1/24 (Figure D).*

Third surgical intervention

Right upper maxilla surgery

Excision of radiolucent lesion associated with tooth 17-18 and bilateral endoscopic median meatotomy of maxillary sinuses was performed; orotracheal intubation; antisepsis of operating field with 10% povidone-iodine, placement of surgical fields; placement of pharyngeal pack; infiltration with lidocaine + epinephrine dilution 1/10; incision with Parker No. 3 Bard scalpel blade No. 15, supracrestal distal to tooth 17 intracrevicular surrounding said tooth, crestal to distal of tooth 12 and compensatory vertical discharge; curettage of mucoperiosteal flap; extraction of tooth 17; lateral access ostectomy at the level of tooth 17 with piezosurgery; extraction of tooth 18 included in the lesion; Total excision of cystic pathology; wound cleansing; application of Carnoy's solution for 5 minutes and thorough washing with sterile physiological saline solution; hemostasis; the surgical lodge was closed in two planes using a pedicled flap of Bitchat fat ball and covering it with the vestibular curtain with 3/0 and 2/0 vicryl absorbable thread respectively (Figure 30A-30H).

Under endoscopic vision, an antrostomy and median meatotomy were performed, hemostasis was controlled and collateral meatoseptal plates were placed fixed with 4/0 nylon suture.

The sample was fixed in 10% formalin and sent to the F.O.U.B.A U.B.A. pathology laboratory.

The patient was discharged with antibiotic therapy of amoxicillin + clavulanic acid 1 g every 12 hours for 14 days, ketorolac 20 mg every 12 hours and control at 24, 48 and 72 hours and then at 14 days for removal of septal plaques.

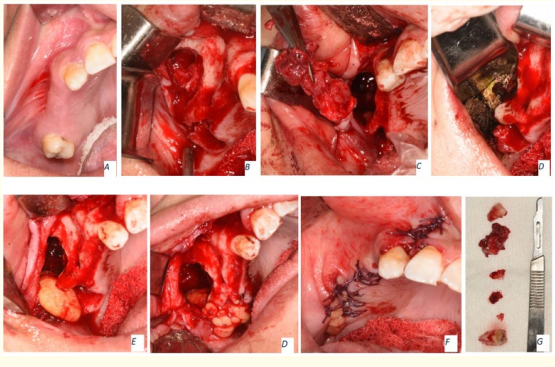


Figure 30A-30F: *Surgical site (Figure A); Extraction of tooth 17 and vestibular ostectomy (Figure B); excision of lesion (Figure C); topicalization with Carnoy's solution (Figure D); taking of Bichat fat ball (Figure E); suturing of fat ball to palatal mucosa (Figure F); closure of second plane with vestibular curtain (Figure G); sample sent to pathology (Figure H).*

Patient age 15

A sample was taken from a radiolucent lesion in the left maxillary branch and mental symphysis (Figure 31).

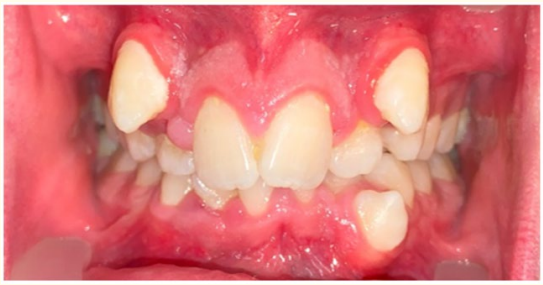


Figure 31: *Pre-surgical image.*

Procedure

Intra- and extraoral field antisepsis with 10% povidone-iodine; placement of surgical fields; anesthesia of the inferior dental nerve trunk and tongue, bilateral buccal and mental infiltration with 4% carticaine hydrochloride - adrenaline 1=100,000; incision in the anterior border of the maxillary branch with a Bard Parker scalpel No. 3, blade 15, mesial discharge at 37-point; mucoperiosteal stripping; puncture and aspiration with obtaining white, granular-consistency (ricotta) contents; access ostectomy with a No. 8 round bur; sample taking (Figure 32A-32C); hemostasis and wound cleaning; repositioning and suturing with a simple 3/0 nylon stitch.

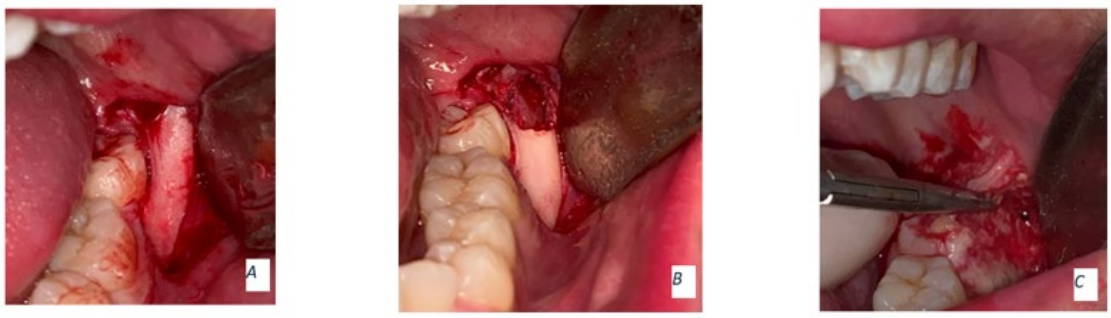


Figure 32A-32C: *Incisional biopsy of the lower jaw. Surgical site after full-thickness flap carving with mesial discharge to tooth 37 (Figure A); access ostectomy to the lesion (Figure B); taking of biopsy sample (Figure C).*

Intracrevicular incision from piece 45 to piece 32; mucoperiosteal stripping; puncture and aspiration with obtaining white, granular-consistency (ricotta) content; ostectomy with round burr #8; taking of biopsy sample from incisions; wound cleaning; flap repositioning and suturing with simple 3/0 nylon stitch.

Post-surgical instructions were given in written and verbal form. Amoxicillin 875 mg + clavulanic acid 125 mg was indicated; dexamethasone 8 mg intramuscularly; chlorhexidine gel 2%; placement of extraoral cold in the operative area; 24-hour control; 72 hours and 10 days later for suture removal.

Samples were fixed in 10% formalin and sent to FOUBA UBA along with the corresponding protocol for anatomopathological analysis.

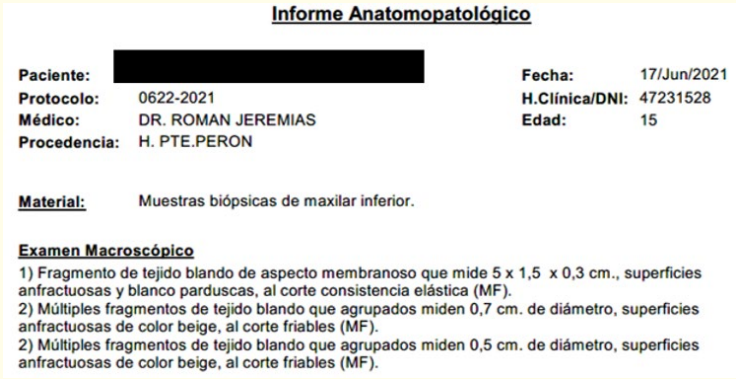
Pathological report (Figure 33)

Macroscopic examination

1) Soft tissue fragment with a membranous appearance measuring 5 x 1.5 x 0.3 cm, with an irregular, brownish-white surface and elastic consistency when cut (MF).
2) Multiple soft tissue fragments that grouped together measure 0.7 cm in diameter, with an irregular, beige-colored surface and friable when cut (MF).
3) Multiple soft tissue fragments that grouped together measure 0.5 cm in diameter, with an irregular, beige-colored surface and friable when cut (MF).

Diagnosis

1), 2) and 3) the histopathological picture is linked to an odontogenic keratocyst with intramural satellite lesions. Correlate with imaging studies.



Informe Anatomopatológico

Paciente: [redacted] **Fecha:** 17/Jun/2021
Protocolo: 0622-2021 **H.Clínica/DNI:** 47231528
Médico: DR. ROMAN JEREMIAS **Edad:** 15
Procedencia: H. PTE.PERON

Material: Muestras biópsicas de maxilar inferior.

Examen Macroscópico

1) Fragmento de tejido blando de aspecto membranoso que mide 5 x 1,5 x 0,3 cm., superficies anfractuosas y blanco parduscas, al corte consistencia elástica (MF).
2) Múltiples fragmentos de tejido blando que agrupados miden 0,7 cm. de diámetro, superficies anfractuosas de color beige, al corte friables (MF).
2) Múltiples fragmentos de tejido blando que agrupados miden 0,5 cm. de diámetro, superficies anfractuosas de color beige, al corte friables (MF).

Figure 33: *Pathological report.*

Second intervention

The total resection of the lesion in the lower jaw was performed under general anesthesia (Figure 34). The procedure was performed 2 years after the sample was taken due to the absence of the patient. Radiographic and tomographic studies were performed, which showed an unfavorable evolution of the lesion.

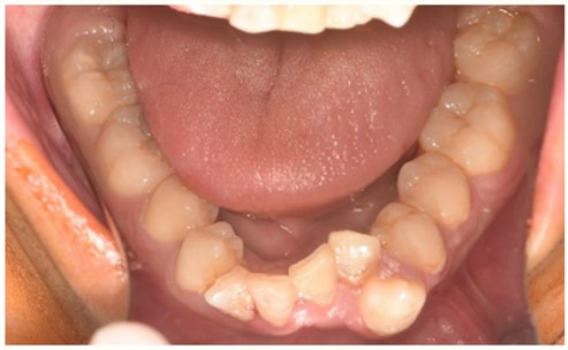


Figure 34: *Pre-surgical image of the lower jaw.*

Procedure

Under nasotracheal anesthesia, the surgical field was antisepsised with 10% povidone-iodine; drape placement; pharyngeal pack placement; infiltrative anesthesia with lidocaine + epinephrine dilution 1/10; incision in the anterior border of the right maxillary branch, intracrevicular from piece 47 to piece 35; full-thickness mucoperiosteal flap curettage; repair of the mental nerve; access ostectomy to the lesion in the mandibular symphysis with piezosurgery; Extraction of tooth 43 with a straight elevator (Figure 35A-35D); excision of cystic lesion; access ostectomy to lesion in the body and branch of the right maxilla; extraction of tooth 44, 45, 47, 48 in close relation to the lesion; excision of lesion in the body (Figure 36A-36D) and branch of the maxilla (Figure 37A and 37B); repair of the lower dental neurovascular bundle; drilling of cavities in the branch, body and symphysis; placement of Carnoy's solution for 5 minutes and thorough washing with sterile physiological saline solution; hemostasis (Figure 38A and 38B); wound cleaning; repositioning and simple suturing (Figure 39A and 39B). Removal of pharyngeal pack. The sample was fixed in 10% formalin and sent to FOUBA UBA (Figure 40). Transfer to the patient's room. Cephalothin 1g was administered intravenously every 6 hours, ketorolac 30 mg every 8 hours, dexamethasone 2 ml IV at 12 noon and a hydration plan of 28 drops per minute. Monitoring and discharge at 24 hours; outpatient monitoring at 24 hours, 72 hours and 7 days. Cephalexin 1g was prescribed every 12 hours for 10 days; flurbiprofen 200 mg every 12 hours for mild pain; ketorolac 10 mg sublingual for acute pain; chlorhexidine 2% antiseptic gel.

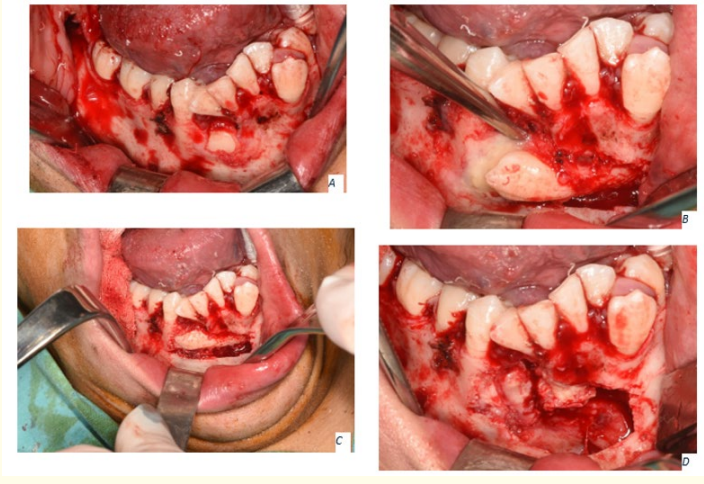


Figure 35A-35D: *Surgical sequence for excision of a lower maxillary keratocyst. Full-thickness incision and descaling from the anterior border of the right maxillary ramus, intracrevicular and distal discharge to tooth 35 (Figure A); dislocation of tooth 43 with a straight elevator (Figure B); vestibular ostectomy with piezosurgery (Figure C); tooth extraction and cystic pathology (Figure D).*

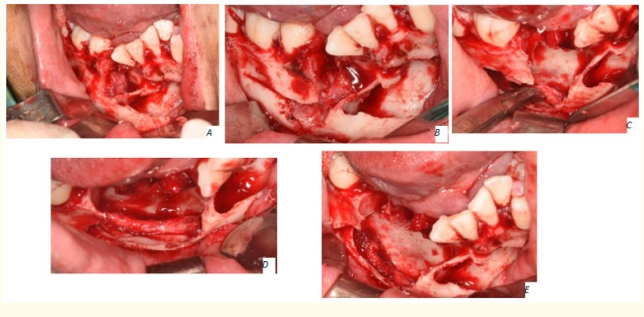


Figure 36: *Vestibular access ostectomy (Figure A); surgical lodge where cystic membrane is observed (Figure B); preservation of the lower dental vasculonervous bundle (Figure C); lower dental vasculonervous bundle (Figure D).*

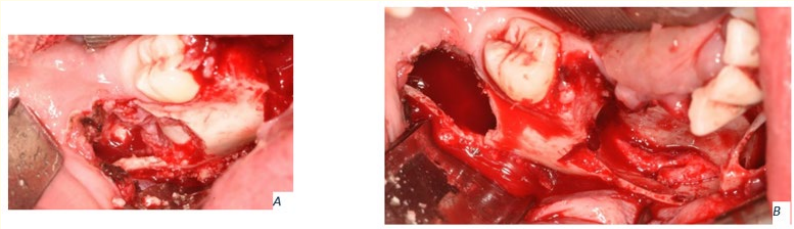


Figure 37A and 37B: *Vestibular ostectomy for tooth extraction of tooth 48 and cystic pathology (Figure A); surgical bed after extraction and enucleation of the lesion (Figure B).*

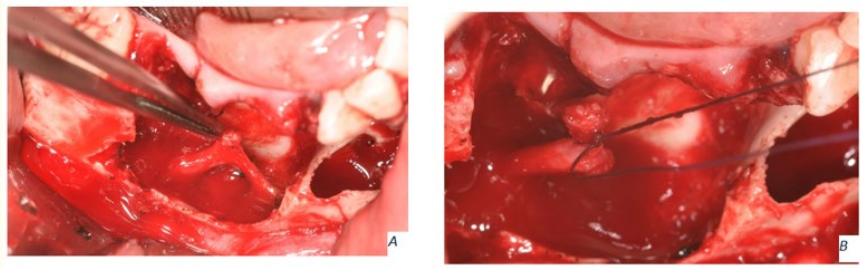


Figure 38A and 38B: *Hemostasis and ligation of the lower dental neurovascular bundle.*

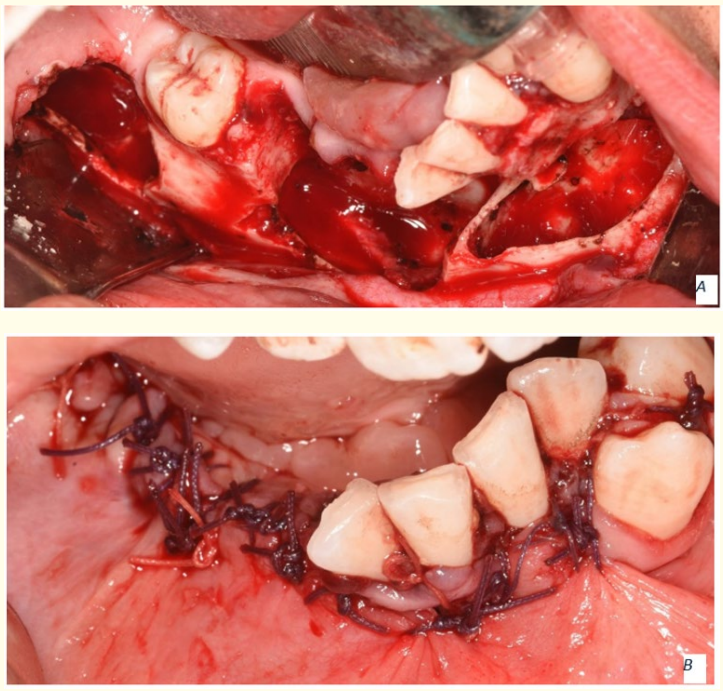


Figure 38A and 38B: *Hemostasis and ligation of the lower dental neurovascular bundle.*

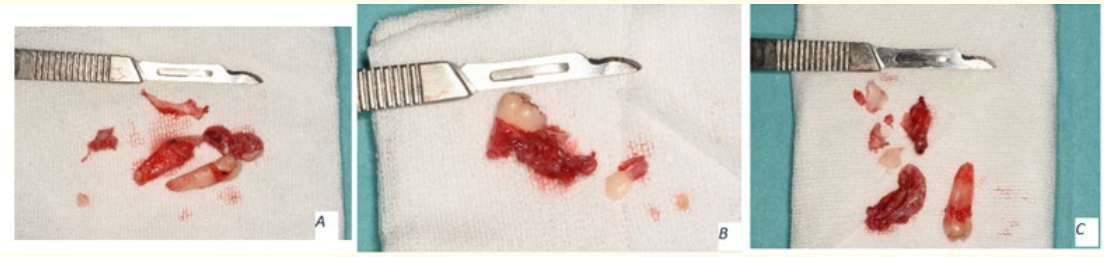


Figure 40A-40C: *Biopsy samples of the lower jaw.*

Pathological report (Figure 41)

Macroscopic examination

1) Teeth 4.3 and 4.4 intact and multiple soft tissue fragments, some of them with a membranous appearance, grouped together, measuring 3 cm in diameter, with an irregular and brownish white surfaces, and crepitating when cut (MF).
2) Teeth 4.5 and 4.6 intact and multiple mineralized and soft tissue fragments, some of them with a membranous appearance, grouped together, measuring 3 cm in diameter, with an irregular and brownish white surfaces, and crepitating when cut (MF).
3) Multiple tooth fragments and soft tissue fragments with a membranous appearance measuring 2.5 cm x 1 cm x 0.4 cm, with irregular and brownish surfaces, and crepitating when cut (MF).

Diagnosis

1), 2) and 3) the histopathological picture together with the evaluated radiographic image corresponds to an odontogenic keratocyst with intramural satellite lesions (Samples 1 and 2) and an added chronic inflammatory process.

Informe Anatomopatológico

Paciente: [redacted] **Fecha:** 25/Jul/2023
Protocolo: 0860-2023 **H.Clínica/DNI:** 47231528
Médico: DR. JEREMIAS ROMAN **Edad:** 16
Procedencia: H. PERON

Material: Muestras biópsicas de maxilar inferior.

Examen Macroscópico

1) Piezas dentarias 4.3 y 4.4 indemnes y múltiples fragmentos de tejido mineralizado y blando, algunos de ellos de aspecto membranoso, agrupados miden 3 cm. de diámetro, superficies anfractuosas y blanco parduscas; al corte crepitan (MF).
2) Piezas dentarias 4.5 y 4.6 indemnes y múltiples fragmentos de tejido mineralizado y blando, algunos de ellos de aspecto membranoso, agrupados miden 3 cm. de diámetro, superficies anfractuosas y blanco parduscas; al corte crepitan (MF).
3) Múltiples fragmentos dentarios y fragmento de tejido blando de aspecto membranoso que mide 2,5 x 1 x 0,4 cm., superficies anfractuosas y parduscas; al corte crepita (2F).

Diagnóstico

1, 2 y 3) El cuadro histopatológico junto con la imagen radiográfica evaluada corresponde a Queratoquiste Odontogénico con lesiones satélites intramurales (muestras 1 y 2) y proceso inflamamatorio crónico sobreagregado.

Figure 41: *Lower jaw biopsy report.*

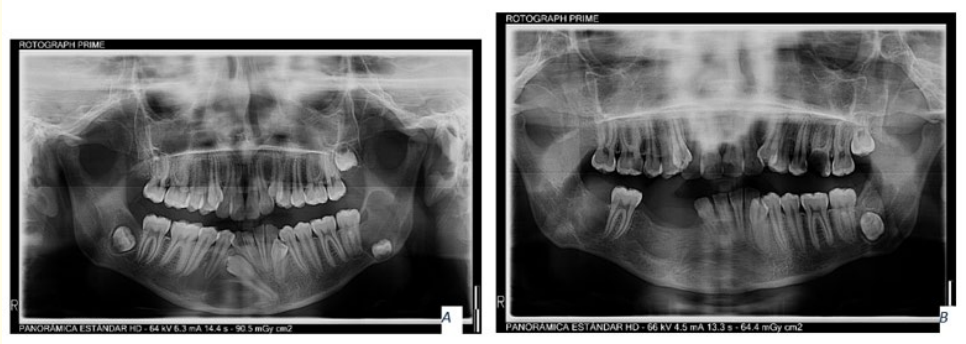


Figure 42A and 42B: *Initial radiographic image 06/14/2021 (Figure A); control radiographic image 06/06/2024 (Figure B).*

Patient mother

Under local anesthesia, a sample was taken from the maxillary branch.

Procedure

Field antisepsis with 10% povidone iodine; placement of surgical fields; anesthesia of the inferior dental nerve trunk and lingual nerve and infiltration of the buccal nerve with 4% carticaine hydrochloride - adrenaline 1=100,000; incision in the anterior edge of the branch, mid-crestal incision and distal discharge to piece 45 with a Parker No. 5 Bard type scalpel, blade 15; full-thickness flap stripping; root remnant extraction; puncture and aspiration with obtaining whitish content with a soft and granular texture (ricotta); access ostectomy with a blunt instrument due to the externalization of the lesion; taking of 2 fragments with a cold scalpel (Figure 43A-43C); fixation in 10% formalin and sent to FOUBA UBA with its corresponding protocol; toilette and hemostasis of the wound; repositioning and suturing with a simple 3/0 nylon stitch.

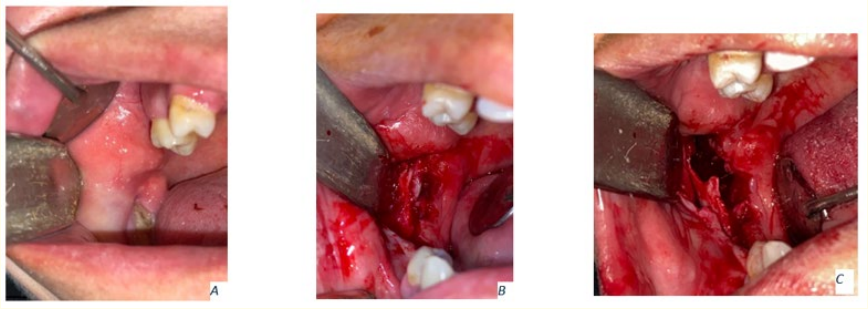
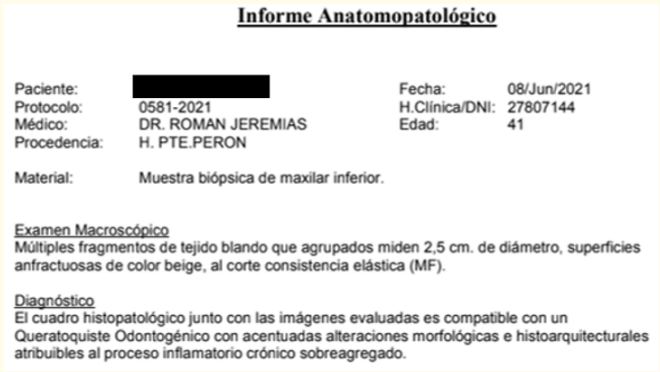


Figure 43A-43C: *Preoperative surgical site (Figure A); full-thickness flap detachment and access to the lesion (Figure B); surgical site after ostectomy and biopsy sample collection (Figure C).*

Post-surgical instructions were given in written and verbal form. Amoxicillin 875 mg + clavulanic acid 125 mg was indicated; dexamethasone 8 mg intramuscularly; chlorhexidine gel 2%; placement of extraoral cold in the surgical area; 24-hour control; 72 hours and 10 days later for suture removal.

Histopathological report (Figure 44)

- Macroscopic examination: Multiple soft tissue fragments that grouped together measure 2.5 cm in diameter, beige-colored anfractuous surfaces, elastic consistency (MF) when cut (Figure 45).
- Diagnosis: The histopathological picture together with the images evaluated is compatible with odontogenic keratocyst, with marked morphological and histoarchitectural alterations attributable to the chronic inflammatory process.



Informe Anatomopatológico

Paciente: [redacted] Fecha: 08/Jun/2021
Protocolo: 0581-2021 H.Clínica/DNI: 27807144
Médico: DR. ROMAN JEREMIAS Edad: 41
Procedencia: H. PTE.PERON

Material: Muestra biópsica de maxilar inferior.

Examen Macroscópico
Múltiples fragmentos de tejido blando que agrupados miden 2,5 cm. de diámetro, superficies anfractuosas de color beige, al corte consistencia elástica (MF).

Diagnóstico
El cuadro histopatológico junto con las imágenes evaluadas es compatible con un Queratoquiste Odontogénico con acentuadas alteraciones morfológicas e histoarquitecturales atribuibles al proceso inflamatorio crónico sobreagregado.

Figure 44: *Right lower maxilla biopsy report.*

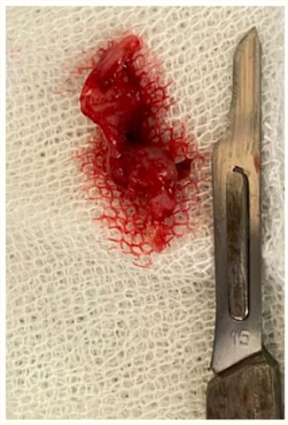


Figure 45: *Biopsy sample of the left maxillary branch.*

Control (Figure 46A and 46B)

In the radiographic control performed on the patient 2 months after the sample was taken, the new bone formation of the ramus and coronoid process and the absence of lesions were observed. The patient was not reoperated on and radiographic controls are currently continuing.

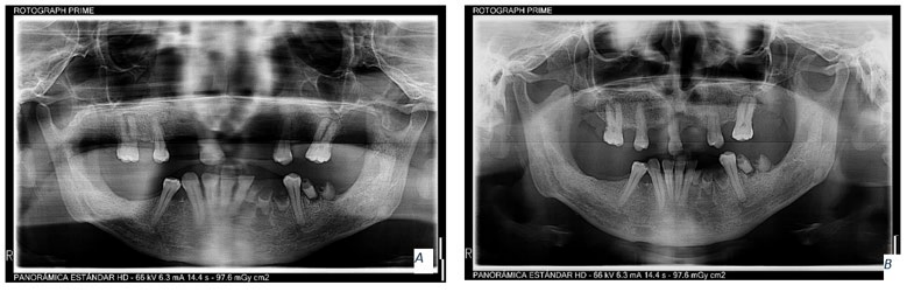


Figure 46A and 46B: *Comparison of radiographs between the initial situation 03/17/2021 (Figure A) and the control radiograph 12/10/2021 (Figure B).*

Patient age 22 (Figure 47A-47C)

Lower maxillary lesion

A sample was taken from the right maxillary body, associated with teeth 46, 47 and 48, under local anesthesia.

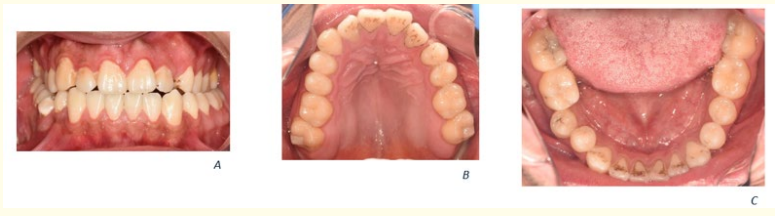


Figure 47A-47C: *Clinical images of the initial pre-surgical situation. Frontal photograph (Figure A); upper occlusal (Figure B) and lower occlusal (Figure C).*

Procedure

Field antisepsis with 10% povidone iodine; placement of surgical fields; anesthesia of the inferior dental and lingual nerve trunk and infiltration of the buccal nerve with 4% carticaine hydrochloride - adrenaline 1=100,000; incision in the anterior edge of the branch with mesial discharge to tooth 45; full-thickness flap curettage; puncture and aspiration with obtaining citrine-colored liquid; access ostectomy with a No. 8 round bur; excision of the lesion; wound cleansing with physiological solution; repositioning and simple suturing with 3/0 silk (Figure 48A-48F).

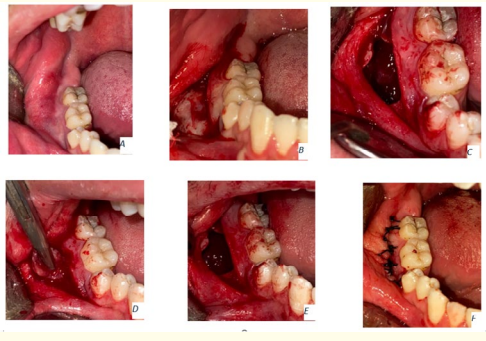


Figure 48: *Pre-surgical image (Figure A); full-thickness flap carving (Figure B); ostectomy and access to cystic cavity (Figure C); taking of excisional biopsy sample (Figure D); surgical site (Figure E); simple suture with 3/0 silk (Figure F).*

Post-surgical instructions were given in writing and verbally. Amoxicillin 875 mg + clavulanic acid 125 mg was indicated; dexamethasone 8 mg intramuscularly; chlorhexidine gel 2%; placement of extraoral cold in the surgical area; 24-hour monitoring; 72 hours and 10 days later for suture removal.

Maxillary lesion

The total excision of the lesion together with the retained tooth 23 was performed under general anesthesia.

Procedure

Under nasotracheal anesthesia, the surgical field was antisepsised with 10% povidone-iodine; drape placement; pharyngeal pack placement; infiltrative anesthesia with lidocaine + epinephrine dilution 1/10; intracrevicular incision from tooth 27 to tooth 22 with mesial discharge; mucoperiosteal flap curettage; puncture and aspiration with obtaining whitish content with a granular appearance (ricotta); ostectomy with piezosurgery and gouge; extraction of tooth 23 with a straight elevator under odontosection with piezosurgery; total excision of the lesion with blunt instruments; extraction of tooth 28 included in the lesion; cavity milling with a round bur; Carnoy's solution was applied for 5 minutes and the wound was washed thoroughly with saline solution; hemostasis and wound cleansing; flap repositioning and simple 3/0 Vicryl suture (Figure 49A-49H).

Cephalothin 1g was administered intravenously every 6 hours, Ketorolac 30 mg every 8 hours, dexamethasone 2 ml IV at 12 hours and a hydration plan of 28 drops per minute. Monitoring and discharge at 24 hours; outpatient monitoring at 24 hours, 72 hours and 7 days. Cephalexin 1g was prescribed every 12 hours for 10 days; flurbiprofen 200 mg every 12 hours for mild pain; ketorolac 10 mg sublingually for acute pain; chlorhexidine 2% antiseptic gel.

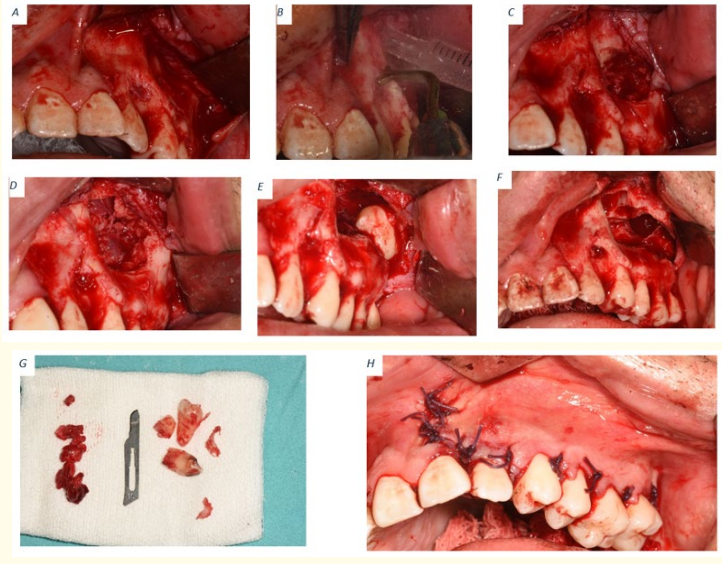


Figure 49A-49G: *Maxillary surgery. Incision and detachment of triangular flap with mesial discharge to tooth 22 (Figure A); vestibular ostectomy with piezosurgery (Figure B); surgical window of ostectomy (Figure C); cystic membrane and retained tooth 23 can be seen (Figure D); surgical site after tooth 23 extraction and removal of cystic membrane, where retained tooth 28 can be seen in close relation to the lesion (Figure E); surgical lodge after pathology extraction and tooth 28 (Figure F); biopsy sample (Figure G); repositioning and simple suture with vicryl. 2/0 (Figure H).*

Pathological report

Maxilla (Figure 50)

Macroscopic examination:

1. Multiple fragments of soft tissue with a membranous appearance that together measure 2.7 cm in diameter, beige-colored anfractuous surfaces, elastic consistency (MF) when cut.
2. Tooth fragments and multiple fragments of mineralized soft tissue that together measure 1 cm in diameter, brownish-white anfractuous surfaces.

Diagnosis

1) and 2) the histopathological picture corresponds to an odontogenic keratocyst with intramural satellite lesions and chronic inflammatory process added. It is accompanied by laminar bone tissue in sectors with the presence of mixed hematopoietic and adipose bone marrow (sample 2).

Universidad de Buenos Aires
Facultad de Odontología
Cátedra de Anatomía Patológica
Laboratorio de Patología Quirúrgica

Informe Anatomopatológico

Paciente: [redacted] **Fecha:** 26/Set/2023
Protocolo: 1216-2023 **H.Clínica/DNI:** 41723922
Médico: DR. ROMAN JEREMIAS **Edad:** 24
Procedencia: H. PTE.PERON

Material: Muestras biópsicas de maxilar superior.

Examen Macroscópico
1) Múltiples fragmentos de tejido blando de aspecto membranoso que agrupados miden 2.7 cm. de diámetro, superficies anfractuosas de color beige, al corte consistencia elástica (MF).
2) Fragmentos dentarios y múltiples fragmentos de tejido blando y mineralizado que agrupados miden 1 cm. de diámetro, superficies anfractuosas blanco prduscas (MF).

Diagnóstico
1 y 2) El cuadro histopatológico corresponde a Queratoquíste Odontogénico con lesiones satélites intramurales y proceso inflamatorio crónico sobreagregado. Acompaña tejido óseo laminar en sectores con presencia de médula ósea mixta hematopoyética y adiposa (muestra 2).

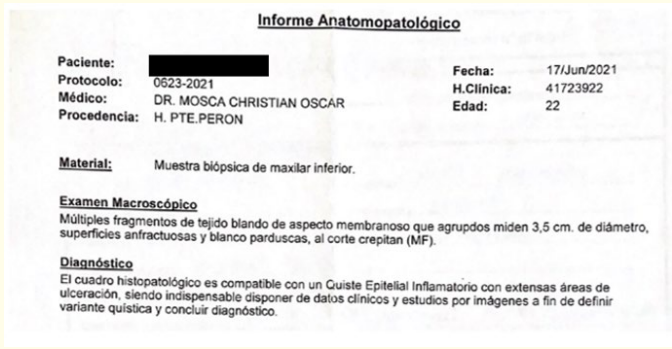
Figure 50: *Anatomopathological result of the upper jaw.*

Lower jaw (Figure 51)

Macroscopic examination: Multiple fragments of soft tissue with a membranous appearance that together measure 3.5 cm in diameter, with an irregular, brownish-white surface, and crepitate when cut (MF).

Diagnosis

The histopathological picture is compatible with an inflammatory epithelial cyst with extensive areas of ulceration. Clinical data and imaging studies are essential to define the cystic variant and conclude the diagnosis.



Informe Anatomopatológico

Paciente: [redacted]
Protocolo: 0623-2021
Médico: DR. MOSCA CHRISTIAN OSCAR
Procedencia: H. PTE.PERON

Fecha: 17/Jun/2021
H.Clínica: 41723922
Edad: 22

Material: Muestra biópsica de maxilar inferior.

Examen Macroscópico
Múltiples fragmentos de tejido blando de aspecto membranoso que agrupdos miden 3,5 cm. de diámetro, superficies anfractuosas y blanco parduscas, al corte crepitan (MF).

Diagnóstico
El cuadro histopatológico es compatible con un Quiste Epitelial Inflamatorio con extensas áreas de ulceración, siendo indispensable disponer de datos clínicos y estudios por imágenes a fin de definir variante quística y concluir diagnóstico.

Figure 51: *Lower jaw pathology report.*

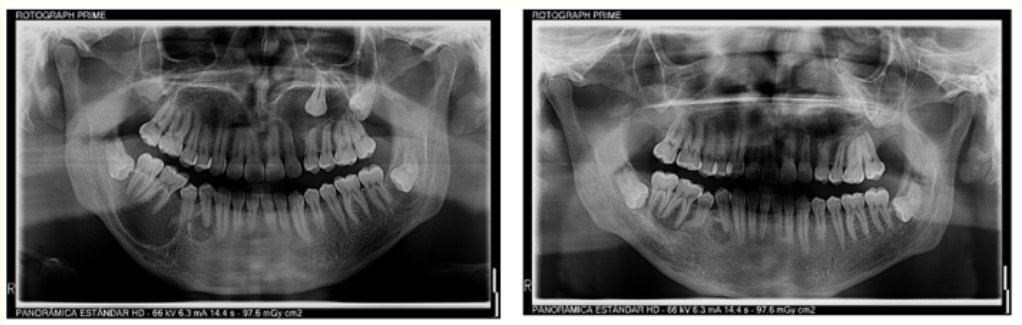


Figure 52A and 52B: *Radiographic comparison between initial image 06/09/2021 (Figure A) and control x-ray 04/24/2024 (Figure B).*

Discussion

Gorlin and Goltz described the classic triad composed of multiple basal cell carcinomas, maxillary keratocysts and bifid ribs that characterized the diagnosis of this syndrome. The diagnostic criteria were established by Evans., *et al*. and modified by Kimonis., *et al*. in 1997, according to them the diagnosis of the syndrome is established when two major criteria or one major and two minor criteria are met. These criteria have been modernized with the inclusion of molecular confirmation, the replacement of the bifid ribs of the major criterion by medulloblastoma and also some changes in the minor criteria that now includes cardiac fibroma [14]. In the case presented, the presence of major and minor diagnostic criteria mentioned above were of vital importance to confirm the presumptive diagnosis of our patients.

Multiple odontogenic keratocysts affect 75 - 100% of people with Gorlin Goltz syndrome and may be the first sign of this genetic disorder. Thus, it is essential that the dentist knows how to investigate and obtain an early diagnosis, and consequently, provide adequate treatment with a better prognosis. The appearance of malignant brain and epithelial tumors are possible complications in cases of late diagnosis [15]. We agree with this, because in our case the diagnosis was made through radiographic findings of multiple cystic-looking lesions, with subsequent confirmation of keratocyst through biopsy. The presence of basal cell carcinomas and their confirmation through biopsy reports provided by the surgery and dermatology service were also of vital importance for the diagnosis.

According to the literature, keratocyst has an incidence of 3-11% of maxillary cysts and its condition is predominantly in men, with a 2:1 ratio compared to women. This lesion occurs in an age range between the second and third decade of life, with a second peak of incidence between the fifth and sixth decade of life. Although any bone, maxilla or mandible can suffer this injury, most of the time the mandible has a predominance of 2:1 and up to 75% of the time this lesion is found in the posterior region of the mandible [36]. In our experience we agree that in daily clinical practice keratocyst is one of the most frequently diagnosed pathologies. The case report shows a predominance in men over women [3,4]. In relation to the age of appearance we find it in the first, second and fifth stage of life. It is most frequently diagnosed in the mandible in the posterior region.

Clinically, keratocyst presents as a solitary lesion except when associated with Gorlin Goltz syndrome. It is important to emphasize that, in its initial clinical stage, it does not present symptoms, it is generally discovered due to complementary tests requested by the dentist. However, in advanced stages, it can manifest itself through buccal or lingual expansion, pain, swelling or secretion, and in rare cases paresthesia of the lower lip [36]. Likewise, we agree on the association of multiple lesions with Gorlin Goltz syndrome. Our findings were radiographic in asymptomatic patients. Only in one case was it manifested by a buccal vestibular expansion associated with a superinfection of the lesion. Paresthesia in the lower lip occurred in only one case, with recovery after the surgical procedure. In our experience, this was the second case in which we found an association with the syndrome.

There is a difference between keratocysts associated with the syndrome and those not; keratocysts of odontogenic origin are generally single and are found in adult and elderly patients, with a preference for the posterior portion of the mandible with thick epithelium and a recurrence of approximately 61%, while those associated with the syndrome appear multiple, in young patients, with equal distribution in the maxillary bones, thinning of the lining epithelium and have a recurrence of 82% of relapses [15]. In our experience, we agree on the appearance of multiple keratocysts in Gorlin syndrome. The location of the lesions showed greater localization in the lower jaw compared to the upper jaw, the radiographic controls performed to date only showed one recurrence. In cases treated in patients without association with the syndrome, the recurrence rate in our practice is the same.

In relation to treatment, the inability to accurately assess the distance from the main cyst to the satellite cysts in the alveolar bone represents the greatest challenge when making the decision and planning the surgical technique because they are responsible for recurrence [39]. Other factors to take into account when planning are the extent and location of the lesion, the appearance of the lesion (unilocular or multilocular), the presence of cortical perforation, the involvement of soft tissues and the age of the patient [40].

There are several treatment modalities, which can be classified into non-conservative or radical treatments, and conservative treatments accompanied by adjuvant methods. Among the non-conservative or radical treatments we find resection, which is the most aggressive way to treat a keratocyst; however, it has been shown to be the most effective in preventing recurrence of the lesion. Conservative treatments include marsupialization, decompression, and enucleation, with or without adjuvant therapy (cryotherapy, peripheral osteotomy, or application of Carnoy's solution) [36].

Conservative treatments include simple enucleation of the cyst (without curettage) as an appropriate treatment method for keratocysts. Recurrence rates are higher with this treatment method and range from 9% to 62.5%. At a minimum, curettage is recommended after cyst removal. Peripheral osteotomy is primarily used as an adjunct to bone removal when resections can be avoided. Almost all recurrence theories involve the possibility of leaving fragments of the cyst lining or capsule within the bone cavity. As the cyst increases in size, the cyst edge may become irregular or scalloped, or surgical access to the cyst may be compromised. There is a higher risk of not removing the entire cyst. A peripheral ostectomy with rotary instruments allows the surgeon to remove as much bone as necessary to ensure that the entire lining disappears [39].

Adjuvant therapies include cryotherapy, a rarely used method, complementary to enucleation or resection of the keratocyst, in which solutions such as liquid nitrogen at 20°C induce cellular necrosis in the bone, maintaining inorganic bone fragments that, in the future, help bone regeneration in the area to be treated. It is important to emphasize its management, since, in healthy tissue, it can show devitalization. The use of Carnoy's solution aims to prevent the recurrence of the lesion by penetrating it through the bone margin of between 1 and 1.5 mm. This solution should not exceed five minutes to avoid its function as an inducer of local neurotoxicity, which depends greatly on the exposure time. It is composed of alcohol (6 ml), chloroform (3 ml), acetic acid (1 ml) and ferric hydrochloride (0.1 mg). The solution should be placed after enucleation of the QQO, curettage and cleaning of the surgical niche, and should not come into contact with surrounding tissues [36].

Decompression involves opening the cystic lesion through a small incision and inserting a catheter to maintain the opening and drainage. Decompression is often followed by a cystectomy a few months later when the lesion has been reduced in size. In this way, the second step of the surgical technique can be performed without damaging important anatomical structures such as the teeth or inferior alveolar nerve, thereby reducing the possibility of spontaneous fracture. Marsupialization focuses on externalizing the cyst through a surgical window in the buccal mucosa and cystic wall, where the cyst-mucosal margins are sutured to create an open window that communicates with the oral cavity [36].

The reason for decompression is to reduce pressure within the cystic lesion, a goal also achieved by marsupialization, to expose the lesion capsule to the oral environment for definitive resolution. Both methods, decompression and marsupialization, are particularly effective in pediatric treatments and in patients with severe pathologies and extensive lesions. This method allows a relative absence of bleeding and scarring; however, due to the difficulty in controlling the amount of liquid nitrogen applied to the cavity, the resulting necrosis and swelling can be unpredictable. In addition, an increased risk of subsequent spontaneous fracture was also observed. The advantage of these techniques is that they are minimally invasive, can be performed under local anesthesia, and avoid the risk of resection and consequently facial deformity. Both decompression and marsupialization, although considered effective techniques, have cases of relapse in long-term follow-up [15].

Recurrence has produced a change in the paradigm of keratocyst treatment. Surgical resection or en bloc resection, which consists of the surgical removal of a complete section of the maxilla or mandible, is the only approach that does not show recurrence of keratocysts in surgical follow-ups but, despite the high long-term success rate, it also produces a very high cost in terms of morbidity, such as bone loss, continuity and mandibular and facial deformation. Resection can also be only marginal, which is an advantageous technique because it allows surgical removal of lesions while maintaining an intact bone margin to ensure continuity. Resection treatment does not seem justified for asymptomatic lesions, even large ones, that affect only the spongy part of the bone, without expansion, erosion or perforation of the cortical bone. Resection should be reserved only for tumors that have perforated the cortical layer, especially those that are recurrent. Lesions limited to the medullary part, on the contrary, should generally be approached more conservatively. In the case of "en bloc" resection at the mandibular level, it is indicated, if possible, to proceed with immediate reconstruction of the mandible with autologous bone grafts or free flaps of the fibula or iliac wing. In the case of extensive, multilocular lesions, in risk areas or recurrent lesions, in the phase of malignant degeneration or invasion of deep planes, the use of a more radical therapy, such as en bloc resection, would seem more appropriate, in order to avoid a recurrence of the lesion and its further expansion [15,41-43].

Regarding treatment, we opt for conservative treatment, using decompression procedures in the case of large lesions, risk of fracture and proximity to noble structures, always evaluating the patient's cooperation and the evolution of the treatment through monthly radiographic studies. We use alternating lavages through the decompression cannula with 0.2% chlorhexidine and sterile physiological solution, 3 times a day. We prefer enucleation followed by the use of Carnoy's solution as adjuvant therapy, applying it for 5 minutes in the cavity and then with profuse lavages with sterile physiological solution. Two of the cases presented were treated with decompression,

one of these evolved favorably leading to the total disappearance of the lesion after radiographic follow-up; the other case allowed the reduction of the size of the lesion to be subsequently operated on. As a disadvantage of decompression we find the formation of septa that subsequently make correct curettage of the pathology difficult. We agree, as the articles show, on radical treatment in case of recurrences, but we do not take it as the method of choice, although this is the most effective method, the aesthetic and functional consequences are unpleasant for the patient and his environment.

Conclusion

Gorling Goltz syndrome is a pathology that requires multidisciplinary work in order to provide the patient with comprehensive treatment covering all the systems affected by it. The importance of the dentist is relevant in the early diagnosis of radiographic findings of multiple odontogenic keratocysts in the jaws; this highlights the value of requesting imaging studies in the dental consultation and the importance of evaluating the stomatognathic system covering all the tissues that compose it.

Regarding treatment, it is vital to act immediately, avoiding the growth of lesions. We prefer conservative therapies and short, medium and long-term controls, as long as the lesion does not invade deep planes or is large, preventing it and requiring radical action.

The patient with GGS must have lifelong follow-up by the health team. Annual radiographic controls are vital by the dentist to diagnose relapses or the development of new keratocysts.

Bibliography

1. RJ G. "Nevoid basal-cell carcinoma syndrome". *Medicine (Baltimore)* 66.2 (1987): 98-113.
2. I Palacios-Álvareza., *et al*. "Síndrome de Gorlin". *Actas Dermo-Sifiliograficas* 109.3 (2017).
3. Kiran NK., *et al*. "Nevoid basal cell carcinoma syndrome (Gorlin-Goltz syndrome)". *Contemporary Clinical Dentistry* 3.4 (2012): 514-518.
4. Gorlin RJ GR. "Multiple nevoid basal- cell epithelioma, jaw cysts and bifid rib: a syndrome". *New England Journal of Medicine* 262 (1960): 908-912.
5. Ganguly S., *et al*. "Gorlin-Goltz syndrome: a rare case". *Indian Journal of Dermatology* 60.2 (2015): 216.
6. Naveen N Kumar., *et al*. "Gorlin-Goltz syndrome: A rare case report". *Contemporary Clinical Dentistry* 9.3 (2018): 478-483.
7. Jindřich Pazdera., *et al*. "Gorlin-Goltz syndrome with familial manifestation". *Biomedical Papers of the Medical Faculty of the University Palacky, Olomouc, Czech Republic* 166.1 (2022): 112-116.
8. Rocío Gilabert Rodríguez., *et al*. "Síndrome de Gorlin-Goltz: manejo del carcinoma basocelular facial". *Revista Española de Cirugía Oral y Maxilofacial* 35.1 (2013).
9. Yamamoto K., *et al*. "Further delineation of 9q22 deletion syndrome associated with basal cell nevus (Gorlin) syndrome: Report of two cases and review of the literature". *Congenital Anomalies* 49.1 (2009): 8-14.
10. MM C. "Nevoid basal cell carcinoma syndrome: molecular biology and new hypotheses". *International Journal of Oral and Maxillofacial Surgery* 28.3 (1999): 216-223.
11. Andressa Bolognesi Bachesk., *et al*. "Gorlin-Goltz Syndrome: The importance of clinical investigation and a multidisciplinary approach". *International Journal of Odontostomatology* 15.1 (2021): 189-195.

12. KJLJea. MN. "Multidisciplinary oral rehabilitation of an adolescent suffering from juvenile Gorlin-Goltz syndrome- a case report". *Head and Face Medicine* 15.1 (2019): 5.

13. Evans DG., *et al*. "Complications of the nevoid basal cell carcinoma syndrome: results of a population based study". *Journal of Medical Genetics* 30.6 (1993): 460-464.

14. Anne Kristine Larsen., *et al*. "Manifestations of Gorlin-Goltz syndrome". *Danish Medical Journal* 61.5 (2014): A4829.

15. Francesco Spadari., *et al*. "Multidisciplinary approach to Gorlin-Goltz syndrome: from diagnosis to surgical treatment of jawbones". *Maxillofacial Plastic and Reconstructive Surgery* 44.1 (2022): 25.

16. Lida Velazque Rojas., *et al*. "Diagnóstico y tratamiento conservador del queratoquiste odontogénico". *Odontoestomatologia* 24.39 (2022).

17. Jackson IT., *et al*. "Penetration of the base by dissecting keratocyst". *Journal of Cranio Maxillo Facial Surgery* 21.8 (1993): 319-325.

18. Crowley Te. "Odontogenic keratocysts: A clinical and histologic comparison of the parakeratin and orthokeratin variants". *Journal of Oral and Maxillofacial Surgery* 50.1 (1992): 22-26.

19. Gorlin RJ., *et al*. "Nevoid Basal Cell Carcinoma Syndrome". *Medicine* 66.2 (1987): 98-113.

20. José Luis Cadena Anguiano., *et al*. "Queratoquiste odontogénico. Revisión de 20 años en la Unidad Médica de Alta Especialidad, Hospital de Especialidades CMN «La Raza» IMSS 1980-2000". *Revista Mexicana de Cirugía Bucal y Maxilofacial* 6.1 (2010): 4-13.

21. Wright JM., *et al*. "Update from the 4th edition of the World Health Organization Classification of Head and neck Tumours Odontogenic and maxillofacial bone tumors". *Head and Neck Pathology* 11.1 (2017): 68-77.

22. Barnes L., *et al*. "World Health Organization classification. Pathology and Genetics of Head and neck Tumors". 3rd edition. IARC Press (2005).

23. Shear M. "Cysts of the oral and maxillofacial regions". 4th edition. Oxford: Blackwell Munksgaard (2007).

24. Robert Wakolbinger and Johann Beck-Mannagetta. "Long-term results after treatment of extensive odontogenic cysts of the jaws: a review". *Clinical Oral Investigations* 20.1 (2016): 15-22.

25. M Anthony Pogrel and R C K Jordan. "Marsupialization as a definitive treatment for the odontogenic keratocyst". *Journal of Oral and Maxillofacial Surgery* 62.6 (2004): 651-655.

26. Merva Soluk-Tekkesin and John M Wright. "The World Health Organization Classification of Odontogenic Lesions: A Summary of the Changes of the 2022 (5th) Edition". *Turkish Journal of Pathology* 38.2 (2022): 168-184.

27. JG M. "The odontogenic keratocyst: A 20-year clinicopathologic review". *Laryngoscope* 108.2 (1998): 280-283.

28. Javier Sánchez Sánchez., *et al*. "Odontogenic keratocyst: diagnostic characteristics and conservative surgical treatment". *International Journal of Medical and Surgical Sciences* (2021).

29. Cazar Almache M., *et al*. "Queratoquiste odontogénico de amplia dimensión: reporte de un caso". *Research Society and Development* 9.12 (2020): 1-12.

30. Alonso MA., *et al*. "Tratamiento con descompresión de un queratoquiste odontogénico". *Revista Odontológica Mexicana* 24.2 (2020): 124-133.

31. Pampin F., *et al*. "Presentación radiológica inusual de queratoquiste odontogénico mandibular". *Revista Anuario de la Sociedad de Radiología Oral y Máxilo Facial de Chile* 24.2 (2018): 43-46.

32. Alba Forteza-López., *et al*. "Tratamiento del tumor odontogénico queratoquístico: revisión sistemática". *Revista Española de Cirugía Oral y Maxilofacial* 41.1 (2018): 26-32.

33. R B Brannon. "The odontogenic keratocyst: A Clinicopathologic study of 312 cases. Part I. Oral Pathology. Clinical Features". *Oral Surg Oral Med Oral Pathol* 42.1 (1976): 54-72.

34. S Omura., *et al*. "Odontogenic keratocyst appearing as a soap bubble or honeycomb radiolucency: Report of a case". *Journal of Oral and Maxillofacial Surgery* 55.2 (1997): 185-189.

35. AKGNMCRBO EFMO. "Effectiveness in the recurrence rate of marsupialization, enucleation, and resection as treatment alternatives against odontogenic keratocyst". *Odontología Sanmarquina* (2022).

36. Ochoa Moreira JA., *et al*. "Técnicas para el tratamiento del queratoquiste, revisión de la literatura y presentación de un caso". *Revista Científica Odontológica (Lima)* 11.2 (2023): e159.

37. Alstad V and J Abtahi. "Surgical removal of keratocystic odontogenic tumours via a Le Fort I osteotomy approach: a retrospective study of the recurrence rate". *International Journal of Oral and Maxillofacial Surgery* 46.4 (2017): 434-439.

38. Borghesi A., *et al*. "Odontogenic keratocyst?: imaging features of a benign lesion with an aggressive behaviour". *Insights into Imaging* 9.2 (2018): 883-897.

39. Ghali GE and M Scott Connor. "Surgical management of the odontogenic keratocyst". *Oral and Maxillofacial Surgery Clinics of North America* 15.3 (2023): 383-392.

40. Santana Daiana Cristina Pereira., *et al*. "Odontogenic Keratocyst: Eight-Year Follow-Up After Conservative Treatment". *International Journal of Odontostomatology* 15.2 (2021): 520-525.

41. Merva Soluk-Tekkeşin and John M Wright. "The World Health Organization Classification of Odontogenic Lesions: A Summary of the Changes of the 2017 (4th) Edition". *Turkish Journal of Pathology* 34.1 (2017).

42. Jorge Andrés Ochoa Moreira., *et al*. "Técnicas para el tratamiento del queratoquiste, revisión de la literatura y presentación de un caso". *Revista Científica Odontológica (Lima)* 11.2 (2023): e159.

43. Spiker AM., *et al*. "Gorlin Syndrome". StatPearls (2022).

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