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Case Report

Management and Outcome of Lateral Cleft Lip in Goldenhar Syndrome: A Rare Case Report

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Abstract

Goldenhar syndrome, or Oculo-Auriculo-Vertebral syndrome, is a poly-malformative disorder, that associating most often an epi-bulbar dermoid cyst, pretragial diverticula and vertebral anomalies. That may vary greatly in range and severity from case to case.

We report a rare observation of Goldenhar syndrome, associating lateral labial cleft, dermoids of the occular limbus and pre-tragic excretion.

The child had a normal auditory meatus without any evidence of skin abnormalities.

Radiography of the spine, echocardiography did not show any associated anomaly.

Goldenhar Syndrome is a rare malformation, related with an anomaly of development of the first gill arches. Although most cases were sporadic. The modalities of autosomal recessive inheritance and the dominant have been described. Also referred to as oculo-atrial dysplasia, it is most often unilateral affecting the soft tissues and less the bone component. It usually includes peribulbar dermoids or dermolipomas, pre-auricular cuticular diverticula, abnormalities of the shape of the auricle, conductive hypo-acoustics, abnormalities of the facial area and sometimes vertebral anomalies. It can be added to these malformative alterations a coloboma of the eyelid, a Duane syndrome and very rarely microphthalmia or anophthalmia.

Through this observation, we will discuss the pathogenic, clinics aspects of this rare entity and the therapeutic measures taken.

Keywords: *Goldenhar Syndrome; Lateral Facial Cleft; Infants; Surgery*

Introduction

Goldenhar syndrome (SG) or facio-auricular-vertebral dysplasia is a complex syndrome characterized by a combination of maxillomandibular hypoplasia, ear deformity, ocular dermoid and vertebral abnormalities. It is associated in the most severe forms of hemifacial microsomy [1-4].

Rare pathology; whose prevalence is estimated between 1/5,600 to 1/20,000 births [2]. Most cases are not familial; occur sporadically. The incidence of lateral cleft lip is 1 in 100,000 - 300,000 and constitutes about 0.3 - 1% of all facial clefts [2].

The genetic origin of the disease is complex and heterogeneous, several genes and chromosomes would be involved [4,7].

Case Report

We describe the case of a 2-year-old patient with a form considered minor of the disease.

Observation

2 years-old patient reported to the Department of pediatric surgery, Constantine Algeria, with a transverse cleft present in the left angle of mouth, with incompetent lips epibulbar dermoid cyst, pretragial diverticula (bilateral preauricular tags). There was no history of head and neck trauma or maternal exposure to teratogenic agents.

The remainder of the clinical examination, moreover, was without any abnormality.

Echocardiography as well as the other paraclinical investigations, CT-scan; abdominopelvic ultrasound, were without abnormalities.

Correlating the history and clinical findings a final diagnosis of Goldenhar Syndrome was given (Figure 1-4).

Figure 1: *Clinical photo showing unilateral cleft.*

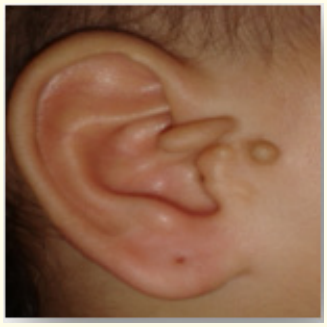

Figure 2: *Multiple accessory tags.*

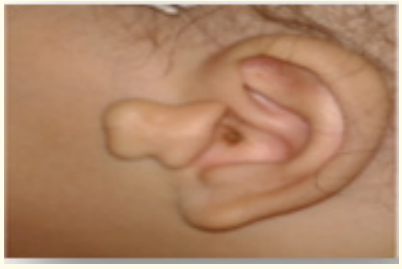

Figure 3: *Preauricular tags.*

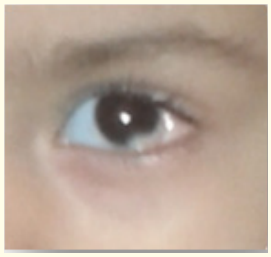

Figure 4: *Eyelid coloboma of the left eye.*

The intervention under general anesthesia was done, in which the orbicularis muscle was reconstructed by reconstruction of the angle of the lip on its mucous and cutaneous surface. Excision of preauricular tags. The patient was referred to the ophthalmic department, where she underwent a simple excision of limbic tumors (Figure 5-7).

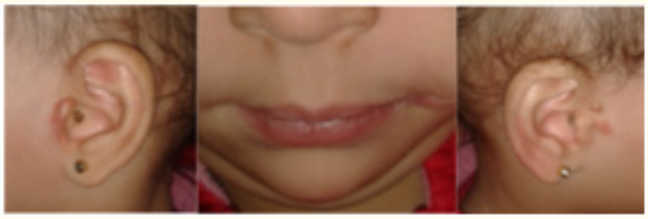

Figure 5-7: *Postoperative aspect.*

During the 2-years follow-up, the parents were satisfied with the aesthetic post-surgical result.

Discussion

Goldenhar syndrome (GS), or oculo-auriculo-vertebral syndrome, affects the soft tissues more often than bone structures [1]. Goldenhar syndrome usually includes cleft lip and especially lateral labial cleft, dermoid or dermolipoma of limb, pre-auricular cutaneous diverticula, fistulas in the same area, malformations of the external ear [1-4].

It can be added to these malformative changes: colobomas of the upper eyelid, conductive hypoacusis, bilateral asymmetry of face and macrostomia, abnormalities of the lower part of the facial mass (macrostomy, micrognathia, partial in occlusion of the dental arches), costal or vertebral abnormalities and finally visceral [4,7].

In rare cases, lipomas of the corpus callosum are described [1,2].

Von Arlt was the first to describe GS, in 1881; then in 1952, Dr. Maurice Goldenhar, named the malformation complex as Goldenhar [2].

Several other diagnosis can also be invoked as Collins and Wildervanck Syndrome (Syndroma cervicoocuioacusticum); Treachers Collin's Syndrome; Miller Syndrome characterized by distinctive craniofacial malformations [3].

In our case, relating to the clinical presentation, para clinic investigation and the congenital history of this malformation, a final diagnosis of Goldenhar Syndrome was given, in its minor form.

GS syndrome is made up of anomalies, mainly of first and second branchial arch derivatives. Characteristic features include structural malformations of the external and middle ears, face, and jaw [4]. Many theories have been proposed to explain the pathogenesis of this disorder; it has been suggested in first that hemorrhage involving the first and second branchial arches causes hypoplasia and malformation of the face and auricle [6,7].

This theory cannot explain the multisystemic manifestations of this disease.

The most accepted theory, it explains that GS would imply a molecular mechanism involving cell adhesion molecules that unify the two disease processes and explain the multisystem anomalies of the GS [4,6].

The management of this disease is multidisciplinary and depending to the association and the severity [5,7].

Dr. Trisser established a classification system for craniofacial clefts that the most widely used of those available today.is notated 0 to 14, with clefts 0 to 7 describing facial clefts and clefts 8 to 14.

The transverse or lateral cleft (Tessier number 7, is the most common form without skeletal abnormalities which corresponding to our case report [9].

LK Makhija., *et al.* attempt to classify lateral cleft lip 7, in severity grade

- Grade I: Slight widening of angle of mouth.
- Grade II: Cleft extending till the anterior border of the masseter.
- Grade III: Cleft extending beyond the anterior border of the masseter.

The main objective of lateral cleft surgery is to create symmetrical commissure [9].

There are various techniques, like simple linear flap, triangular flap, Z plasty, W plasty, more important factors Correction are to correct different function as, speech problems and compromised chewing ability. In our patient, We have used the triangular flap technique The well-repaired orbicularis or is counters the lateral contractile force of the skin scar [8].

Conclusion

Goldenhar syndrome is a rare multifactorial disease. Clinically stands as a combination of several abnormalities such as limbal tumors, malformation of the ears of different degrees, as well as cardiac and vertebral malformations, cleft lip or palate of different stages. Lateral clefts in Goldenhar syndrome with minor manifestations requires perfect correction because all the challenges are focused on the aesthetic aspect.

Conflict of Interest

The authors have no conflict of interest.

Bibliography

1. Mutanabbi M., *et al*. "Goldenhar syndrome - a case report". *Mymensingh Medical Journal* 23.3 (2014): 586-589.
2. Saccomanno S., *et al*. "Role of 3D-CT for orthodontic and ENT evaluation in Goldenhar syndrome". *Acta Otorhinolaryngologica Italica* 34.4 (2014): 283-287.
3. Ruchi Bhuyan., *et al*. "Goldenhar Syndrome: A rare case report". *Journal of Oral and Maxillofacial Pathology* 20.2 (2016): 328.

4. Lam CH. "A theory on the embryogenesis of oculo-auriculo-vertebral (Goldenhar) syndrome". *Journal of Craniofacial Surgery* 11.6 (2000): 547-552.

5. Bogusiak K., *et al*. "Treatment strategy in Goldenhar syndrome". *Journal of Craniofacial Surgery* 25.1 (2014): 177-183.

6. Spinelli-Silva S., *et al*. "Distal deletion at 22q11.2 as differential diagnosis in Craniofacial Microsomia: Case report and literature review". *European Journal of Medical Genetics* 61.5 (2018): 262-268.

7. LK Makhija., *et al*. "Transverse facial cleft: A series of 17 cases". *Indian Journal of Plastic Surgery* 44.3 (2011): 439-443.

8. Milind Joshi., *et al*. "Lateral cleft lip and macrostomia: Case report and review of the literature". *Journal of Indian Association of Pediatric Surgeons* 19.4 (2014): 242-243.

9. Kuriyama M., *et al*. "Tessier number 7 cleft with oblique clefts of bilateral soft palates and rare symmetric structure of zygomatic arch". *Journal of Plastic, Reconstructive and Aesthetic Surgery* 61.4 (2008): 447-450.

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