

Squamous Odontogenic Tumour: Clinicopathologic Analysis of a Rare Entity

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Abstract

Objectives: To report and describe 4 cases of squamous odontogenic tumor (S.O.T) that were retrieved from the biopsy files of Obafemi Awolowo University Teaching Hospital Complexes Ile-Ife, Nigeria(OAUTHC) during a period of 2007 - 2016, review the literature and to discuss the clinico-pathologic features of this rare tumor.

Methods: Hematoxylin and eosin stained sections of each of the tumors histologically diagnosed as S.O.T were reviewed with the objective of reconfirming the diagnosis in the biopsy report file.

Results: 4 cases of S.O.T. were diagnosed. The mean age of occurrence was 33.5 years (range 21 to 54 years). There was equal sex predilection, 3 (75%) occurred in the mandible and 1 (25%) occurred in the maxilla. All the mandibular lesions occurred in the posterior molar region of the mandible while the single maxillary lesion occurred in the cuspid-premolar.

Discussion: There was equal gender preponderance which was consistent with reported literature and a third decade prevalence which is however in contradiction to the prevalent 4th decade in documented reports. The posterior mandible and cuspid-premolar maxillary location was consistent with literature. There was 0% concordance for SOTs due to its uncommon nature. Ameloblastin (AMBN) and heparinase are proteins implicated in its pathogenesis

Conclusion: Squamous odontogenic tumor is a very rare distinct indolent benign odontogenic tumor commonly misdiagnosed. It presents with well described radiographic and unequivocal histopathologic features which enables its diagnosis. Conservative treatment is indicated in most cases with rare occurrence of recurrences following adequate treatment.

Keywords: Squamous Odontogenic Tumor; Concordance; Cuspid-premolar; AMBN; Heparinase

Introduction

Squamous odontogenic tumor (SOT) is a locally infiltrative neoplasm consisting of islands of well-differentiated squamous epithelium in a fibrous stroma [1]. First described in the 20th century by Pullon in 1975 [2] in his case series of 6 cases, it is a rare entity in which both intraosseous and extraosseous components have been reported in documented literature [3-6]. It exhibits a relatively slow growth that might however invade bone and adjacent structures [2]; and in some cases, have only been discovered incidentally in routine dental radiographs. A related entity, the squamous odontogenic tumor-like proliferations in odontogenic cysts (SOT-LPOC) has also been described in documented literature wherein there are multiple islands of squamous odontogenic epithelium present in the wall of odontogenic cyst [7]. Both are however rare entities with less than 170 cases of both cases reported globally.

SOT occurs between 8-74 years with a mean of 38.7 years [1], with a slight male predilection of 1.4:1. Even though it presents both intraosseously and peripherally, the localization is usually centrally and no mandibular - maxilla predilection with a molar propensity in the mandible and a canine- premolar susceptibility in the maxilla [5,8,9]. Multi-centric presentations as well as familiar presentations of SOTs have also been well documented [10], however it presents usually unitarily: a racial predilection however has not been elucidated [4,9]. It's etiology is still unknown and histogenesis from the rest cells of Malassez in the periodontal ligament has been proposed [11].

The clinical features range from being largely asymptomatic to a painful bony expansion, which may include mobility of affected teeth [4], maxillary lesions have been suggested to be more aggressive in clinical presentation [4]. A characteristic triangular-shaped unilocular radiolucency between the diverging apices of the adjacent roots is consistent with SOT with occasional scalloping and saucerization of the underlying bone [1,8]. Extensive SOTs may also exhibit multilocularity [8] and saucerization may also be found in their peripheral variants due to mainly a pressure effect on the underlying alveolar bone [5]. Histologically, SOTs comprise solid islands, oval or round, of squamous odontogenic epithelium without peripheral palisading with central microcystic degeneration interspersed on a mature fibrous connective tissue stroma [1,3]; other variable histopathological findings include lamellar calcification and globular eosinophilic structures within the epithelial islands. Histochemical and immunohistochemical studies have excluded the presence of amyloid in the epithelial islands [9] and also demonstrated immunorexpression of Notch 1, 3, 4 proteins in conjunction with Jagged1 and delta1 proteins [11]. The purpose of the present article is to report the clinico-pathologic analysis of four diagnosed cases of SOTs from our institution and review the literature of this rare entity.

Materials and Methods

Biopsy records of all histopathologically diagnosed SOTs seen at the Oral Maxillofacial Pathology unit of Obafemi Awolowo University Teaching Hospital Complexes, Ile-Ife (OAUTHCI) between January 2007 and December 2017 were retrieved. Slides from ancillary hospitals were also reviewed. Hematoxylin and eosin stained section of each of the tumours were reviewed with the objective of reconfirming the diagnosis. Information on demographic data and clinical parameters including age, gender, site, and duration of lesion prior to presentation and associated symptoms were recorded. Information on radiographic features and histologic features were also noted.

Results

Demographic distribution

A total of 4 cases were diagnosed as squamous odontogenic tumour between 2007 - 2016, their demographics is represented in table 1. The age range was between 21 - 54 years with a mean age of 33.5 years, the 3rd decade was the more common decade (n = 2, 50%) and there was no gender predilection as the male: female ratio was 2:2. Swelling was the predominant symptom with all the patients presenting with swelling, two of the patients (50%) also complained of associated pus discharge while one patient complained of associated pain.

S/No	Age	Decade	Sex	Symptom	Duration	Location	Clinical impression	Radiographs
1	23	3	Male	Painful swelling	5 years	Mandible	Dentigerous cyst	Corticated unilocular
2	54	6	Female	Swelling with pus discharge	10 years	Mandible	Ameloblastoma	Non-corticated multilocular
3	21	3	Female	Swelling with pus discharge	4 years	Maxilla with involvement of sinus	Periapical cyst	Unilocular radiolucency with flecks of radiopacity
4	36	4	Male	Swelling	6 years	Mandible	Ameloblastoma	Corticated unilocular

Table 1: Demographic distribution of cases.

There was a mandibular predilection (n = 3, 75%), all mandibular lesions were in the molar region while the maxilla lesion was between the canine - cuspid region. there was also extension of the maxillary lesions into the adjoining maxillary sinus. Radiographic presentation was principally unilocular radiolucency (n = 3, 75%) while the most long-standing of the lesions had a multilocular appearance. Radiopaque flecks were also observed in a case of the unilocular lesions.

The duration of the lesions was variable, from 4 years to 10 years with a mean duration of 6.3 years to underlie their slow growing status. All the initial clinical impressions of the lesions were wrong with 0% concordance as two of the lesions were mistakenly diagnosed as ameloblastomas whilst the remaining two were mistakenly diagnosed as odontogenic cysts.

Histological features

All the cases of SOT exhibited solid islands of well differentiated squamous epithelium in oval or round multi-sized shapes, some of these also exhibited irregular shapes of solid islands in areas. All these were interspersed in a fibrous connective tissue stroma which were bland and devoid of chronic inflammatory cells in most cases but infiltrated by aggregates of chronic inflammatory cells predominantly lymphocytes and plasma cells in few areas. The squamoid odontogenic cells exhibited uniformity in size with prominent intercellular bridges and microcystic degeneration without peripheral palisading.

The cells at the periphery of the tumor islands appear flattened with absence of nuclear reverse polarization, and the central cells do not exhibit stellate reticulum-like differentiation. No mitotic figures were seen in any of the cases and calcific- like areas were seen within some epithelial islands in parts of cystic degeneration. Few vascular channels were also seen with areas of diffuse fresh hemorrhage, in some cases, small nests resembling rests of Malassez or remnants of the dental lamina were evident in the peripheral area of the tumor. No cellular atypia was evident in all the cases.

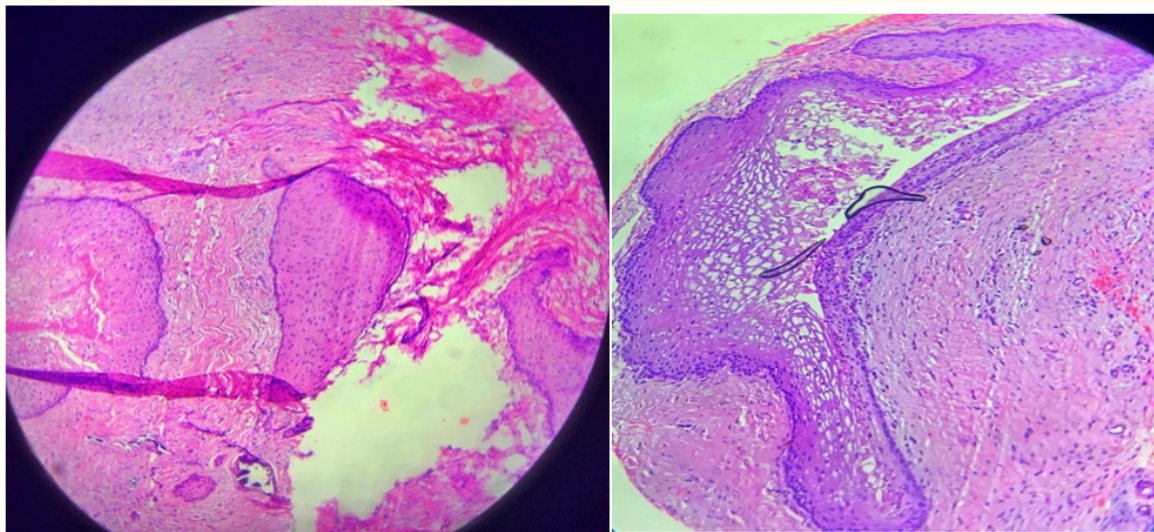


Figure 1: A) oval and round solid islands of squamous epithelial odontogenic cells in a background of mature fibrous connective cells. B) An irregularly shaped solid island with central microcystic degeneration (x100).

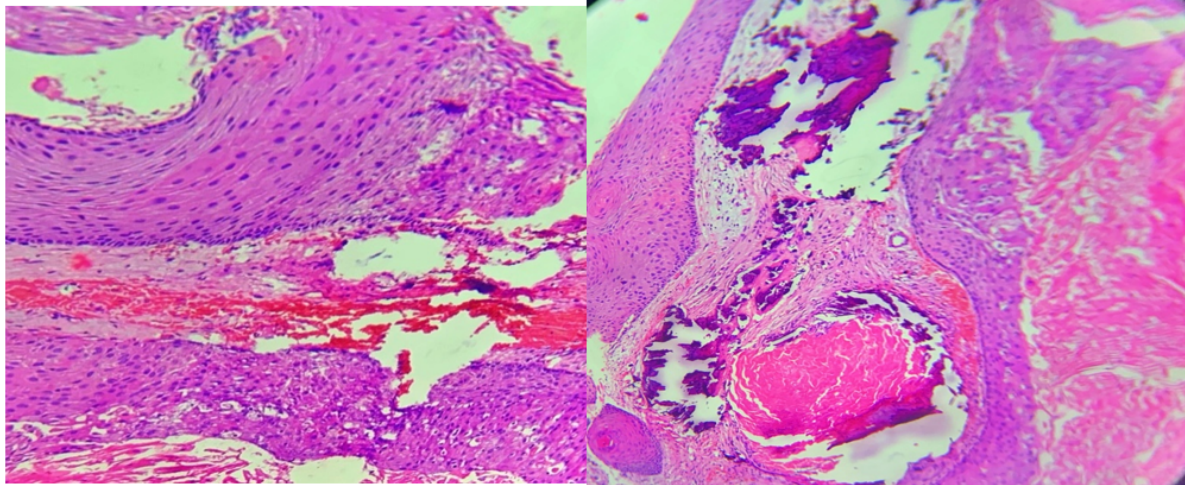


Figure 2: A) Tumor cells showing the characteristics of well- differentiated squamous epithelium. B) Calcific like areas seen within the cystic degeneration.

Discussion

Following description of the 6 cases by Pullon in 1975 [2] numerous publications have reported case reports and case series of SOT with its diagnosis and management reported by other authors [12,13]. The World Health Organization in its 4th classification of Head and Neck tumors defined it as a benign epithelial odontogenic tumour in which the tumour cells show terminal squamous differentiation. The epithelial islands occasionally show foci of central cystic degeneration'. Different authors have documented that only around 50 - 55 cases have been reported globally [1,11,14], Chrcanovic., *et al.* [7] in their review of SOTs reported about 110 cases globally (108 central and 2 peripheral). Due to its rarity, the incidence is thus unknown, however studies on odontogenic tumours as a whole have reported a relative frequency of squamous odontogenic tumor among odontogenic tumors to range from approximately 0 - 2% [15]. In the African continent, the case series of Adebisi., *et al.* [9] and the case report of Moussa., *et al.* [6] have documented the occurrence of SOTs in sub-Saharan Africa. Occurrences of cases have however been documented in large case studies on odontogenic tumors [16,17].

Although odontogenic epithelial origins have been described for SOT in the literature, the pathogenesis of this tumor is still unclear at best [8,10] with the intraosseous SOT thought to originate histogenetically from epithelial rests of Malassez, while dental lamina and gingival epithelium are suspected to be the origins of peripheral variant [11]. Of the numerous studies focused on the aetiopathogenesis and mechanisms of development, the mutation of the ameloblastin (AMBN) has been extensively documented in SOT as well as in adenomatoid odontogenic tumor [18]. The heparanase gene, which codes the heparanase enzyme has also been implicated in the cytodifferentiation of SOT and its mutation also linked to SOT development [19]. The immunohistochemical studies of Siar., *et al.* [11] revealed the immuno-expression of Notch receptors 1, 3, 4 and their ligands, Jagged1 and Delta1; these factors and proteins may play a role in the cellular transformation events that take place within epithelial rests of Malassez leading to the development of SOT. Immunohistochemical studies have further confirmed the proliferative activity of the odontogenic epithelium by its heavy staining for keratin 13/16; also, the squamous differentiating cells in the center of the tumor islands expresses a strong positive reaction for involucrin staining [5].

SOTs present a wide age range distribution with reports of occurrence between the ages of 8 and 74 [6] and a mean age of 36 - 38 years [9,15] documented and mainly reported in the 4th decade of life [1]. However, some authors also reported a predilection for 3rd decade [7].

In our present study, the mean age was 32.5 years and the 3rd decade was the most common reported decade. This is in contrast with previous documented reports of 4th decade prevalence [6,10] and even also with the previous Nigerian study of Adebisi, *et al.* [9] but in tandem with Malathi, *et al.* [5] and Neville, *et al.* [5]. The wide age range of SOTs might have led to this discrepancy, earlier decade presentations have also been observed in the multicentric variety of SOTs [10]. Discrepancies also exist in gender predilection as some authors have observed nil gender predilections [3-5] while others have documented a slight male predilection [7,8,10]; Goldblatt, *et al.* [9] even documented a female preponderance. Even though there is no gender predilection in our study with a male: female ratio of 1:1, our sample size is too small to be able to reinforce or support earlier theories of gender preponderance. Nil racial predilection however has been agreed on by all authors [4,9] with Elmuradi, *et al.* however stating an African-American predilection for multi-centric presentation of SOTs [10].

A nil maxilla - mandibular predilection has also been reported [1,7] with equal occurrence in both jaws; the cuspid - premolar region the most favored site in the maxilla and the posterior region the most common location in the mandible [7]. However, all the cases of SOTs reported in both Odukoya, *et al.* and Adebisi, *et al.* [9] were all in the mandible. Our present study also presents a mandibular predilection (75%, n = 3). The paucity of the respondents in these cases may not be enough to make assertions but there seems to be a mandibular predilection for SOTs in our part of the world; this however needs to be confirmed by more comprehensive case studies/series. Some authors have also reported mandibular predilection [6,11].

Haghighat, *et al.* [4] highlighted common clinical presentation in SOT to include occasional local pain, gingival swelling, and mobility of adjacent teeth, it is however reported to be usually asymptomatic in its indolent growth pattern. The patients in our present study also presented with associated local pain and pus discharge observed in others. None of the cases was discovered on routine investigation. Maxillary lesions have also been suggested to undertake a more aggressive course than their mandibular counterparts [20]; the sole maxillary lesion in our study presented with recurrence following treatment but this was due mainly to inappropriate treatment and not necessarily as a result of its aggressiveness.

The classical radiographic presentation is that of a triangular or semi lunar shaped radiolucency between or along roots of adjacent teeth, with or without corticated margins [1,15]. Mardones, *et al.* [8] further reported other non-specific radiographic patterns that resembles other odontogenic and non-odontogenic cysts and tumours; a multilocular appearance has also been described for extensive lesions. This thus makes SOT a radiological differential diagnosis in a variety of odontogenic lesions and bone lesions. Presence of radiopacities in radiographic cases of SOTs have also been documented [10]. Root resorption of affected teeth are usually rare, both unilocular and multilocular radiolucencies were observed with both corticated and non-corticated margins observed likewise. Furthermore, a case involving flecks of radio-opacities was also observed which incidentally was the sole maxillary case.

All cases of SOTs were misdiagnosed with 0% concordance rate and in the audit of histopathologically diagnosed lesions in our center, no clinician ever made an initial clinical impression of SOT. This corroborates the rarity of this entity and also the non-specificity of its clinico-radiographic features. Ameloblastoma was the most common lesion it was misdiagnosed as. Despite its rarity, SOT should however be on the list of differential diagnosis of slow growing lesions with unilocular radiolucencies especially in cuspid - premolar regions.

The histological features in our present study was in tandem with histo-morphological features reported in documented literature [9]. Multiple multi-sized solid islands were in oval, round and a times irregular shapes of well differentiated squamous epithelium in a background of fibrous connective tissue stroma. Central microcystic degeneration, calcific like bodies and loss of palisading peripheral cells are all in tandem with reported literature as well as rests resembling rests of dental lamina. Other histopathological features that have been reported in studies include presence of clear cells, papillary hyperplasia or papillomatous proliferation [21] and presence of eosinophilic coagulum within cystic areas [9]. None of these adjunct features were evident in our present study. In the diagnosis of SOT, care should be taken to mistake the features of SOT with that of acanthomatous ameloblastoma which it is usually misdiagnosed as and also squamous cell carcinoma. Absence of nuclear palisading by the peripheral epithelial layer of the islands should assist the pathologist in excluding the

diagnosis of ameloblastoma while SOT's bland histologic features of the epithelial nests should exclude consideration of carcinoma [21]. Absence of nuclear hyper chromaticity and pleomorphism can also serve to exclude ameloblastoma.

The main modality of treatment of SOT is conservative surgical removal via local excision or thorough curettage usually with preservation of affected tooth or teeth [3]. More radical treatment has however been prescribed for maxillary lesions as they are more aggressive; such aggressive lesions may be treated by en bloc resection and hemi maxillectomy especially when there is infiltration of maxillary sinus and nasal cavities [20]. All the cases in our study were treated by excision and only the maxillary lesion presented with recurrence. Recurrence in SOTs is rare and attributed to incomplete removal of the initial tumours [3]; this was the exact case in our maxillary lesion.

Conclusion

Squamous odontogenic tumor is a very rare distinct indolent benign odontogenic tumor commonly misdiagnosed. It presents with well described radiographic and unequivocal histopathologic features which enables its diagnosis. Conservative treatment is indicated in most cases with rare occurrence of recurrences following adequate treatment.

Conflict of Interest

The authors confirm that this article content has no conflict of interest.

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