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UNICYSTIC AMELOBLASTOMA WITH UNUSUAL CLINICAL AND DIVERSE HISTOPATHOLOGICAL PRESENTATION: A CASE REPORT WITH SUGGESTED SURGICAL APPROACH.

Dental Science

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ABSTRACT

Ameloblastoma is locally invasive, second most common tumor of odontogenic origin. WHO categorized ameloblastomas clinically into solid, cystic (unicystic), desmoplastic and peripheral (extraosseous) variants, of which unicystic accounts for about 13% of all cases. Granular cell change in ameloblastoma though is a recognized phenomenon; its occurrence however is rare in unicystic entity. Here is a unique case of unicystic ameloblastoma exhibiting granular cell differentiation and diverse histopathological presentation, which has perforated the bone and presented as soft tissue growth (peripheral variant).

KEYWORDS:

Peripheral ameloblastoma, Granular variant, Follicular variant, Acanthomatous, Plexiform variant.

INTRODUCTION:

Ameloblastoma is a benign, locally aggressive neoplasm of odontogenic epithelial origin. It is the second most common odontogenic neoplasm next to odontoma. [1][2][3] Its incidence rate is around 0.5 per million population per year, with highest reported cases in Asia and African continents. [4]

Ameloblastoma can occur as four variants - unicystic, solid/multicystic, desmoplastic and peripheral type. [5] Peripheral variant is extraosseous with histological findings similar to intraosseous type with low recurrence rate. Excluding odontoma, the incidence of ameloblastoma is at least equal to the incidence of all the other odontogenic neoplasms combined. [2]

Its incidence, collective with its clinical behavior, makes ameloblastoma the most significant odontogenic neoplasm of concern to oral and maxillofacial surgeons. As seen with nearly every odontogenic neoplasm, the ameloblastoma may occur centrally within bone or peripherally, without an intraosseous component, in the soft tissues overlying the alveolar ridge. [3][4][5] Intraosseous lesions outnumber peripheral lesions in the ratio of 9:1. [1][4][5] Most of the peripheral variants however, are intraosseous neoplasms which invaded the bone & fused with the oral epithelium and therefore, true peripheral ameloblastomas, are relatively very rare.

Unlike the solid ameloblastomas, a less aggressive variant that tends to occur in younger individuals is the unicystic ameloblastoma. [5] Although plexiform, follicular & acanthomatous are frequently encountered histological variants, a granular cell change in ameloblastomas is a well recognized entity. However, it is rare in case of mural unicystic ameloblastoma.

CASE REPORT:

A 27 year old male reported with a chief complaint of swelling in left lower back tooth region since 5 months. Patient was apparently asymptomatic 5 months back, later noticed a small growth in lower left 3rd molar region associated with mild pain. He gave a history of gradual increase of growth from the past 3 months.

On examination no significant facial asymmetry was noticed. Intra-orally, a diffuse growth in the alveolar mucosa distal to 37 was noticed, measuring 2 X 1cm extending anterioposteriorly from distal surface of 37 to retromolar region and superioinferiorly from occlusal level of 37 extending into vestibule. Obliteration of buccal vestibule was noticed. Color of the growth was slightly erythematous, with indentations of left maxillary posterior teeth (Figure - 1). On palpation, the growth was tender, firm and non-pedunculated. Based on history and clinical findings, a provisional diagnosis of fibro-epithelial hyperplasia was considered and panoramic imaging was advised.

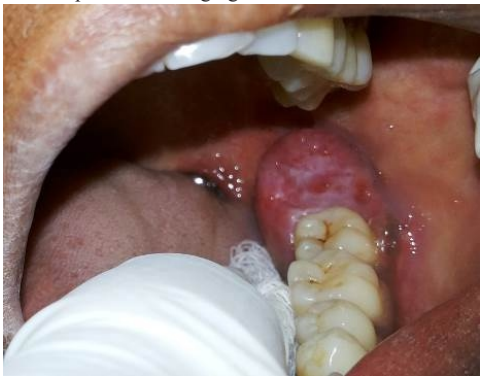

Figure - 1: Intra-oral clinical presentation of lesion proper over retromolar trigone with indentations of maxillary molars.

Orthopantomograph (OPG) revealed, a unilocular radiolucency with diffuse borders over left ramus of the mandible involving the crown and a portion of root of impacted 38 (Figure - 2). Considering these findings Computed Tomography (CT) was advised to know the extent of lesion. It showed bony destruction on the left posterior mandibular region extending from distal aspect of impacted 38, involving the ascending ramus of the mandible up to coronoid process (Figure - 3).

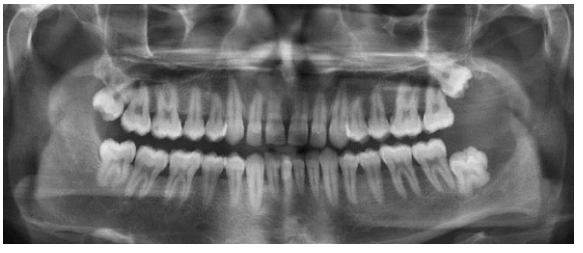

Figure – 2: OPG reveals impacted left mandibular third molar exhibiting root resorption along with associated unilocular radiolucent lesion.

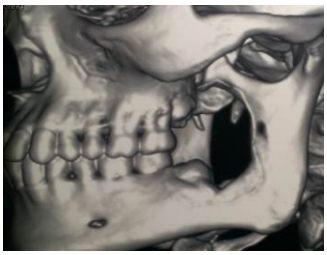

Figure – 3: 3D image shows evidence of bony destruction involving the ascending ramus & coronoid process of the mandible.

Based on the radiographic findings, a differential diagnosis of dentigerous cyst in relation to 38 was made. Prior to incisional biopsy, fine needle aspiration was performed and it was found to be negative. Incisional biopsy was planned under local anaesthesia at the lesional site and two specimens were collected, one from the alveolar mucosal growth and the other from bone for histopathological analysis.

On microscopic examination, the H&E stained section from the soft tissue specimen revealed a parakeratinized stratified squamous epithelium of even thickness overlying a fibrovascular connective tissue stroma showing odontogenic tumor epithelial cells arranged in the form of follicles which were lined peripherally by tall columnar ameloblast like cells with reversal of polarity, and superficial stellate reticulum like tissue. The stellate reticulum in few follicles exhibited cystic degeneration and acanthomatous changes in the form of squamous metaplasia. The intervening connective tissue stroma showed dilated vascular channels engorged with RBCs. A diagnosis of "peripheral ameloblastoma" was made (Figure - 4).

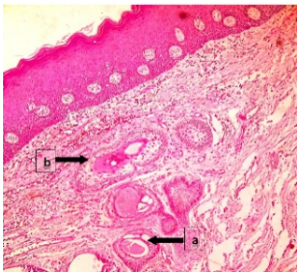

Figure – 4: H&E stained sections from the soft tissue lesion exhibiting parakeratinized stratified squamous epithelium overlying a fibrovascular connective tissue stroma with follicles of odontogenic epithelial tumour islands showing (a)cystic degeneration & (b)squamous metaplasia. (Peripheral Ameloblastoma).

On microscopic examination, the stained section from the intra bony lesion showed a well defined cystic lumen lined by ameloblastomatous epithelial lining with basal tall columnar ameloblast like cells & superficial stellate reticulum like tissue. The cystic lining showed mural proliferations into the connective tissue capsule with sub-epithelial hyalinization (Figure - 5). The connective tissue capsule also showed numerous interconnecting, anastomosing cords & strands of odontogenic tumor epithelial cells with intervening connective tissue showing multiple dilated blood vessels typical of plexiform ameloblastoma (Figure - 6). Deeper connective tissue capsule revealed solid ameloblastomatous follicles, while few exhibiting acanthomatous change with squamous metaplasia and others show granular differentiation of the central stellate reticulum like tissue (Figures - 7, 8, 9). A diagnosis of "mural unicystic ameloblastoma - granular cell variant" was made.

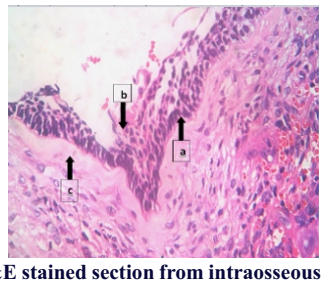

Figure - 5: H&E stained section from intraosseous lesion showing a cystic lumen lined by stratified squamous odontogenic epithelium exhibiting mural proliferation with (a)basal tall columnar ameloblasts like cells, (b)superficial stellate reticulum like tissue and (c)subepithelial hyalinization(Unicystic ameloblastoma – mural type).

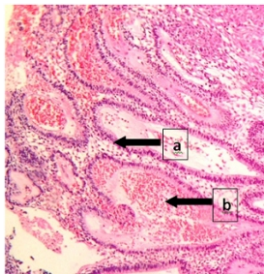

Figure – 6: H&E stained sections from deeper connective tissue stroma exhibiting odontogenic tumour epithelial islands showing Plexiform variant of ameloblastoma with (a)interconnecting strands of tumour cells & (b)dilated vascular channels within the stroma

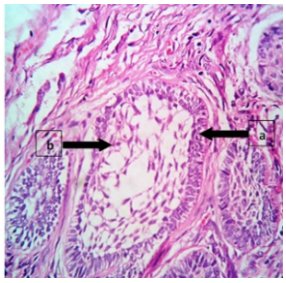

Figure – 7: H&E stained sections from deeper connective tissue stroma exhibiting odontogenic tumour epithelial islands showing Follicular variant of ameloblastoma with (a)peripheral tall columnar ameloblasts like cells and (b)central stellate reticulum like tissue

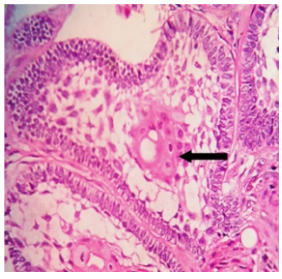

Figure – 8: H&E stained sections from deeper connective tissue stroma exhibiting odontogenic epithelial tumour islands showing Acanthomatous variant of ameloblastoma with squamous metaplasia of stellate reticulum like tissue

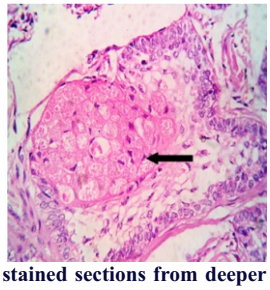

Figure - 9: H&E stained sections from deeper connective tissue stroma exhibiting odontogenic tumour epithelial islands showing Granular cell variant of ameloblastoma with eosinophillic granules in cytoplasm of stellate reticulum like tissue.

With these histopathologic findings, a final diagnosis of mural unicystic ameloblastoma- granular cell variant was made, which has perforated the bone and presented as a peripheral variant.

DISCUSSION:

Ameloblastoma is a benign, slow-growing, locally invasive tumor of odontogenic origin involving the mandible (80 %) and maxilla, the

evidence of which was first described by Cusack in 1827.[6] The 2005 WHO classification for ameloblastomas includes four subtypes. The solid/multicystic is the most common type (91 %), followed by the unicystic (6 %), the extra osseous (2 %), and the desmoplastic type (1 %).[7]

The radiological appearance is frequently similar to that of a dentigerous cyst, but the presence of root resorption should alert to the possibility of ameloblastoma.[8] In 2009, Ngwenya et al. supported the use of the terms 'unilocular' and 'multilocular' rather than 'unicystic' and 'multicystic' to describe the radiological appearances of ameloblastoma. A real distinction between unicystic and multicystic, can only be made after microscopic examination of a resected specimen.[9]

The extra-osseous variant, peripheral ameloblastoma is a rare odontogenic tumor and accounts for about 1% of all ameloblastomas. Most of the lesions described under peripheral ameloblastoma, were not truly peripheral lesions, infact were intraosseous variants that have penetrated through the alveolar bone, fused with the oral epithelium, and eventually presented themselves clinically as peripheral lesions.[10] Among all intraosseous ameloblastomas, unicystic variant accounts for 6-15%.[11] They predominantly affect the younger age group in the 2nd decade of life.[12] The term "Unicystic Ameloblastoma" was given by Robinson & Martinez.[13] Leider et al proposed three pathogenic mechanisms for the evolution of unicystic ameloblastoma: (1) the reduced enamel epithelium associated with a developing tooth undergoes ameloblastic transformation with subsequent cystic development; (2) ameloblastomas arise in dentigerous or other types of odontogenic cysts in which the neoplastic ameloblastic epithelium is preceded momentarily by a non-neoplastic stratified squamous epithelial lining; and (3) a solid ameloblastoma undergoes cystic degeneration of ameloblastic islands with subsequent fusion of multiple microcysts and develops into a unicystic lesion. But most authors strongly favored the idea that these lesions are cystic neoplasms *de novo*.[14]

The diagnosis of unicystic ameloblastoma can lone be made histologically and cannot be predicted preoperatively on clinical or radiographic grounds. Examination of the entire lesion through sectioning at many levels is mandatory for securing the final diagnosis, as mural proliferations into the cystic capsule upto various depths is always a possibility.[15]

Although granular cell change in classic ameloblastoma is a well recognized phenomenon, its occurrence in the context of unicystic ameloblastoma is infrequently appreciated.[16][17] There has been considerable interest as to the nature of granular cells in ameloblastoma ever since it was recognized.[18] It is evident from the literature, that there exist two main lines of interpretation in that some consider it to be metabolic, whilst others of the view that it represents a degenerative process as an aging phenomenon in long standing lesions. However, it may also affect young patients.[19][20][21] But recent immunohistochemical studies suggest that this phenomenon is related to increased apoptotic cell death of the lesional cells and the phagocytosis by neighboring neoplastic cells.[22]

The biological behavior of granular cell ameloblastoma does not appear to differ from the other histologic subtypes; it can be locally aggressive and has a relatively high chance of recurrence.[23] Reichart et al stated a 33.3% recurrence rate for granular cell ameloblastoma, which was higher, compared to the more common follicular, plexiform and acanthomatous subtypes.[24] In Hartman's study, 11 of 15 patients (73%) developed recurrence.[25]

Surgery is the standard treatment for ameloblastomas. Historically, the extent of resection has been controversial, embracing two surgical options: "conservative" vs "radical". Peripheral ameloblastomas should be viewed as a relatively innocuous lesion totally lacking the persistent invasiveness of intra osseous variant, so must be treated conservatively, with supraperiosteal surgical excision and adequate disease free margins. Solid multicystic ameloblastomas require radical surgical intervention, whereas unicystic ameloblastomas require only conservative surgical enucleation, unless infiltration from the epithelial cyst lining into the cyst wall has been demonstrated. In such cases, the treatment should follow that outlined for solid multicystic ameloblastoma.[10]

Gardener points out that the characteristic slow growth of ameloblastomas is significant in that it may take years before recurrence becomes evident. Therefore, it is imperative that the surgical sites be examined, along with the radiographs thoroughly for at least 10 years and preferably longer.[26] However, similar to the other types of solid or multicystic ameloblastomas, the prognosis is more dependent on the method of surgical treatment, i.e. granular cell ameloblastomas treated by enucleation or curettage exhibit a high recurrence rate, due to the fact that the border of the tumor within cancellous bone lies beyond the apparent macroscopic surface and the radiographic boundaries of the lesion. Therefore, radical surgical methods are recommended.[27] Noteworthy is that, granular cell ameloblastomas may rarely behave in a malignant fashion giving rise to metastasis. Patients should be kept under periodic observation because of reports of recurrences occured even up to 8 years after initial treatment.[28]

Conclusion:

Like most of the peripheral ameloblastomas which presented as soft tissue growths were falsely interpreted as extraosseous lesions, our reported case showed a similar presentation of a peripheral ameloblastoma, but which was a counterpart of an intra-osseous unicystic variant which has perforated the alveolar bone giving rise to a soft tissue growth. Hence, thorough radiological & histopathological investigations are indicated before committing a diagnosis of peripheral ameloblastoma. From a surgical point of view, the present lesion should be treated as a solid ameloblastoma, though it is unicystic, because of histological presentation of a mural variant with a more aggressive granular cell differentiation. Therefore, an early diagnosis and prompt surgical treatment in granular cell ameloblastoma is of prime importance.

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