



MAXILLARY ACANTHOMATOUS AMELOBLASTOMA WITH ORBITAL INVOLVEMENT - A RARE CLINICAL VARIANT

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ABSTRACT

Ameloblastoma is a group of rare benign odontogenic tumour, accounting for around 1% of all the cysts and tumours in the jaw. Predominantly this tumour occurs in the mandible and rarely crosses the midline. The typical clinical presentation is that of facial dysmorphism and functional dysfunction due to the expansile and compressive nature of the tumour. Of the six histological subtypes described, Acanthomatous variant is relatively very rare. A Large ameloblastoma with acanthomatous variant of maxilla is also unusual. This case discusses the diagnostic and therapeutic challenges in the management of a large maxillary acanthomatous ameloblastoma that had extended into the orbit, ethmoids and sphenoid sinus.

KEYWORDS

Ameloblastoma, Acanthomatous variant, Odontogenic tumour

INTRODUCTION-

Yoithaprabhuth et al have discussed in detail the Onomastics (the study on proper names and its origin) and meta-terminology of 'Ameloblastoma'. Many terminologies have been used to describe this rare odontogenic tumour of the jaw.[1] The term "Ameloblastoma" was coined by Churchill and Ivy. "Amel" is an earlier terminology in English used for enamel and "blastoma" in Greek means germ. This terminology is widely accepted and used all over the world. [1,2] In 1991, WHO defined Ameloblastoma as a 'benign but locally aggressive tumor with high tendency to recur, consisting of proliferating odontogenic tumor epithelium lying in fibrous stroma', in 2017 WHO has refined its classification and updated its types based on genetic studies. [1,2]

Ameloblastoma accounts for about 1% of all tumors of the jaw. The most common site of predilection for occurrence of Ameloblastoma is the mandible and followed by the upper jaw in the maxilla. [2] This case report discusses the diagnostic and therapeutic challenges faced during the management of a large maxillary Acanthomatous Ameloblastoma.

Case Report-

A 32-year male with no comorbidities presented to our OPD with history of nasal block for 6 months, more in the right nostril which was gradually progressive. He had no other complaints. On examination a smooth regular swelling, with well-defined margins of about 4x3 cm was seen in the right side of the hard palate which was non tender, firm in consistency and crossing the midline. [Fig.1a]. On anterior rhinoscopy, a smooth mass was seen obliterating the right nasal cavity, pushing the septum to left. Examination of eyes revealed proptosis of right eye with chemosis. Hard ill-defined mass was palpable in the inferomedial part of right orbit [Fig.1b]. Extraocular movements of right eye showed restricted adduction, abduction and depression. Distant vision of right eye was 6/9 with no improvement on pin hole vision.

Contrast Enhanced computed tomography of paranasal sinus showed an expansile, multiloculated homogenous soft tissue density lesion with multiple internal septations with epicentre located in the right maxilla. Superiorly the expansile lesion extended upward into the orbit with a cortical breach and the expansile lesion was seen abutting the medial and inferior recti and was in close proximity to optic nerve. Posterosuperiorly the lesion was extending into the posterior ethmoid and the sphenoid bone. Anteriorly there was thinning and scalloping of the anterior wall of maxilla and alveolar process. Posteriorly it was limited to body of pterygoid. The margin's were well defined and distinct [Fig.2a&2b]

Clinically in view of hard, non-tender, non-compressible, non-fluctuant swelling in right side of maxilla obliterating right nasal cavity a provisional diagnosis of bony tumor of maxilla was considered. Radiologically differential diagnosis of keratocystic odontogenic tumour or a dentigerous cyst was considered. However, diagnosis of fibrous dysplasia and ossifying fibroma could not be ruled out.

The diagnosis of Acanthomatous ameloblastoma was confirmed by endoscopic trans-nasal biopsy of the lesion. Microscopic examination showed bone with a solid and cystic lesion composed of islands of squamous epithelium, squamous metaplasia, and clusters of clear cells surrounded by stellate reticulum cells. [Fig.3]

Considering the size of the lesion, a combined external and endoscopic approach was used to completely excise the lesion. Right extended total maxillectomy was done via a Weber Fergusons incision [Fig.4] the entire tumour in the maxilla along with a part of hard palate, alveolus and soft palate was excised. Periorbita in the floor was carefully dissected keeping in mind the close proximity to the optic nerve. Endoscopy was used to dissect the tumour from the roof of the posterior ethmoid and from the greater wing of sphenoid. Optic nerve was visualised and preserved, the ethmoid and sphenoid sinus extension was cleared and the tumour removed in toto. [FIG.5a & 5b]

On seventh postoperative day trial oral feeds were started and Ryle's tube was removed. Vision had improved to 6/6 and proptosis reduced. Extra Ocular Movements were full, free and painless. [FIG.6a] Patient is on regular follow-up for the last one year and no features of recurrence or residual tumour was noted. [FIG.6b]

DISCUSSION-

Ameloblastoma is a slow growing, locally invasive benign tumour. Usually asymptomatic, unless the size of the tumour is so big that it is noticed by the patient or his relatives. Generally, causes facial asymmetry. Depending on the site of tumour it may compress the adjacent structures causing functional impairment. If the tumour originates in the upper jaw involving the maxilla, it can expand upwards towards the orbit causing visual symptoms as was seen in this case.

Maxillary Ameloblastoma with orbital extension is very rare. Milman et al in 2015, retrospectively analysed the clinicopathological database of the university of Pennsylvania and reported 8 case of maxillary ameloblastoma that had orbital involvement. [3] Abtahi et al in 2017 reviewed the existing literature for orbital involvement of ameloblastoma and have identified only around thirty-seven such cases reported.

The mean age of occurrence of orbital involvement was 52.79+/-20.62 years with the age range from 7 to 81. [4] WHO has classified ameloblastoma into three types, namely conventional, unicystic and peripheral. Of these the conventional type is the most common, this is further subtyped based on histological variants into follicular, plexiform, acanthomatous, granular cell, desmoplastic and clear cell. [5] The most common histological variant that has been associated with orbital involvement is the follicular type (48.7%), followed by the plexiform type (37%). [4] None of cases described to have orbital involvement where of acanthomatous subtype. This case describes a rare occurrence of acanthomatous ameloblastoma that presented with orbital symptoms and orbital involvement.

Acanthomatous ameloblastoma variant is histologically characterised by presence of squamous cell differentiation along with keratin formation. This squamous differentiation is thought to be due to chronic irritation as a result of infection. [6] Bhuyan et al have reported an acanthomatous ameloblastoma that recurred in a previously treated plexiform ameloblastoma. [7] Lakshmi et al have reported a rare variety of hybrid ameloblastoma that occurred in the maxilla. [8]

As the understanding of histopathogenesis of ameloblastoma has evolved and so has the classification of ameloblastoma. At present, the molecular and genetic basis for predicting the site predilection, the histological variant, treatment option and the prognosis have been the focus of research. [9,10]

Many therapeutic options have been described in literature. Conservative local excision, enucleation, marsupialisation has been described by many but has shown to have a high chance of recurrence. Wide surgical excision has been the treatment of choice. The close proximity or required margins following complete resection have not been defined. However, the safe margins of 10mm has been recommended. [11] However, Since the tumour is mostly benign the operating surgeon should be judicious in the extent of clearance, respecting the vital structures in close vicinity to the tumour. This is particularly very important in maxillary tumour extending into the orbit, ethmoid and sphenoid sinus where the tumour is in close proximity to the optic nerve as seen in the present case, use of endoscopy help in adequate clearance of the tumour near this vital structure without any morbidity.

Other modalities of therapy such as radiotherapy, chemotherapy and targeted drug therapy based on the molecular and genetic basis have been explored. The goal of management of ameloblastoma is to reduce morbidity and to prevent recurrence of tumour. [11]

CONCLUSION-

This case highlights the rare occurrence of a large maxillary acanthomatous ameloblastoma that was expanding into the orbit and was in close proximity to the optic nerve causing visual symptoms. Wide excision of the tumour was successfully achieved by a combined endoscopic as well as external approach.

The use of endoscope assisted in visualising the orbital structures at close proximity and aided complete removal of lesion with preservation of vital structures thus avoiding morbidity.

The author's emphasis the need for long term regular follow up for early identification of recurrence and provide early intervention especially in cases that are in close proximity to vital structures.



Figure 1a and 1b

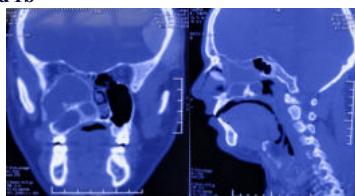


Figure 2a and 2b

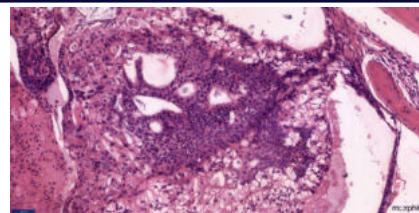


Figure 3



Figure 4



Figure 5a and 5b



Figure 6a and 6b

FIGURE CAPTIONS-

Figure 1a-Clinical picture of the patient showing a swelling over hard palate crossing midline

Figure 1b-Clinical picture showing proptosis of right eye with chemosis

Figure 2a-Coronal section of contrast enhanced CT scan of paranasal sinuses showing the multiloculated lesion with epicentre in right maxilla

Figure 2b- Sagittal section of contrast enhanced CT scan of paranasal sinuses showing extension of the lesion

Figure 3- Histopathological examination (HPE) picture (magnification 40x) Section shows bone with a solid and cystic lesion composed of islands of squamous epithelium, squamous metaplasia, clusters of clear cells surrounded by stellate reticulum cells.

Figure 4- Intra operative picture showing excision of the tumour

Figure 5a & 5b – dissected specimen showing hard palate, alveolus and soft palate

Figure 6a- Immediate post-operative picture

Figure 6b- Postoperative picture after 3 months follow up

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