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Case Report

Recurrent unicystic maxillary ameloblastoma presenting as unilateral proptosis

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Abstract

Unicystic Ameloblastoma (UA) is a rare variant of ameloblastoma which is an odontogenic epithelial neoplasm, typically affecting mandibular ramus. Maxillary ameloblastoma is a rare entity with a more disastrous consequence. Although extremely rare, their highly recurrent and locally aggressive behavior can lead to invasion of vital structures surrounding maxilla (orbit, cranium) even after several years of conservative surgical management (limited resection, curettage). We report a case of 16-year-old girl presenting with proptosis of left eye, UA left maxilla, who was treated initially with limited resection (enucleation) and curettage and the lesion recurred after two years with a more aggressive behavior, causing destruction floor of orbit. To this date there are only 23 documented cases of orbital invasion and only three of the reports are in ophthalmic literature. The ophthalmologists need to be aware of this type of rare lesion presenting as proptosis.

Keywords: Proptosis, Maxillary ameloblastoma, Unicystic ameloblastoma, Orbit

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Introduction

Ameloblastoma is a rare, benign tumor of odontogenic epithelium (ameloblasts, or outside portion, of the teeth during development) commonly appearing in the mandible. Four variants of ameloblastoma have been described by to the World Health Organization: conventional (solid/multicystic type), unicystic, peripheral (extraosseous) and desmoplastic ameloblastoma. Maxilla is an atypical and rare location for unicystic ameloblastoma (UA).[1,2] Unicystic ameloblastoma is defined as a cystic lesion presenting with clinical, radiographic, or gross features of a mandibular cyst, but histopathologically part of the cyst cavity appears to be lined by typical ameloblastomatous epithelium.[1] It was recognized in 1827 by Cusack.[3]

These tumors are rarely malignant or metastatic and slow progressing.[4] Nevertheless the resulting lesions can cause severe deformities of the face due to the high recurrence rate (upto 70%) and a very aggressive invasion of surrounding structures, which may reach upto 45% even after treatment, depending on the mode of surgery.[4] Maxillary ameloblastoma and its orbital involvement are extremely rare in literature but attribute to high recurrence, morbidity and mortality.[4] This is the first case of maxillary ameloblastoma

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invading orbit reported from the Middle East region. There are only 23 well documented cases of orbital involvement in the world literature and only three publications in ophthalmic literature.[4–6]

Case presentation

A 16-year-old girl was referred from local hospital to a tertiary hospital department of otolaryngology head and neck surgery for management of recurrent left sided maxillary swelling. She presented with history of left side cheek swelling of seven months duration, increasing in size and associated with left eye proptosis, left sided nasal obstruction and blood stained nasal discharge. She denied any history of headache, decrease or double vision. Four years prior to her presentation in this tertiary hospital she was diagnosed with maxillary ameloblstoma in a local hospital. She was treated by limited resection (enucleation) of the tumor under general anesthesia. For the next two years she was symptom free then swelling reappeared gradually increasing in size.

Physical examination revealed large intraosseous mass located at the left cheek which was palpable at left maxillary vestibule and covered by intact mucosa. The left eye had proptosis but eye movement was normal in all direction and visual acuity was also normal. Hard palate was pushed downward by the mass and lateral nasal wall was pushed medially. The left upper third molar not yet erupted.

Computed tomography scan revealed left massive maxillary unicystic mass causing destruction of the floor of the orbit and proptosis of the orbit invading, left maxillary sinus (Fig. 1). Magnetic resonance image (MRI) was performed to rule out any soft tissue or ocular invasion. The scan came negative for invasion and showed huge mass occupying left maxillary sinus and orbit.

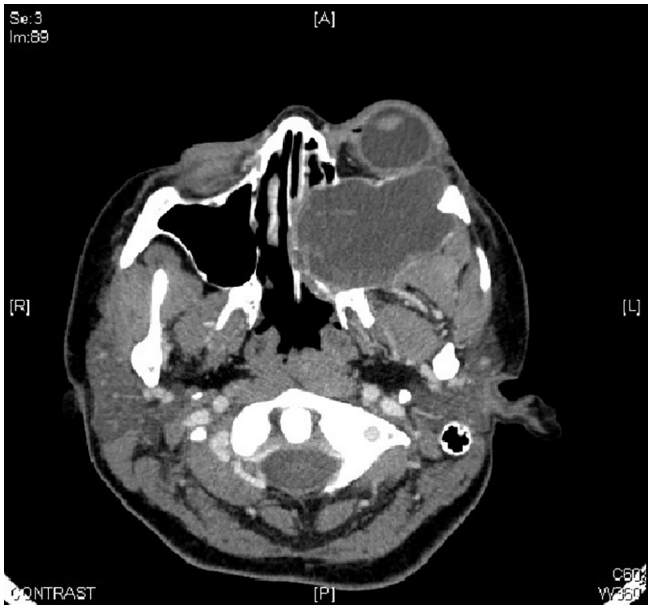

Fig. 1. Axial CT scan showing left massive maxillary unicystic mass causing destruction of the floor of the orbit and proptosis of the orbit.

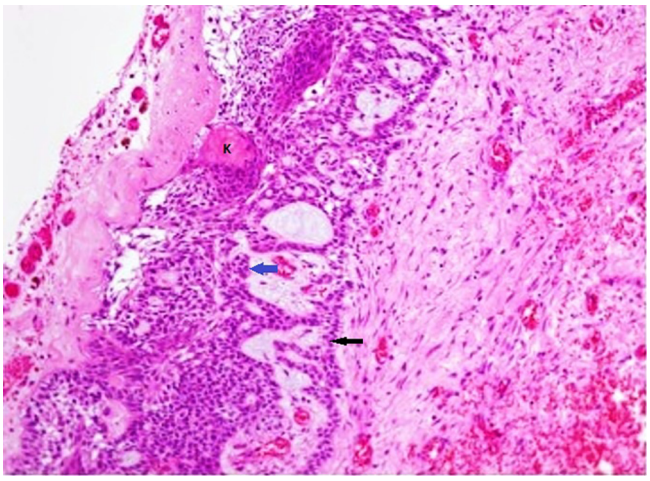

Fig. 3. Microscopic picture of the specimen showing unicystic plexiform ameloblastoma: cystic lesion lined by irregular strands odontogenic epithelium, bordered by palisading columnar cells (black thin long arrow), that surround an islands of cells resembling stellate reticulum (plexiform pattern) (blue broad short arrow), with wet keratin (K). (H & E original magnification ×40).

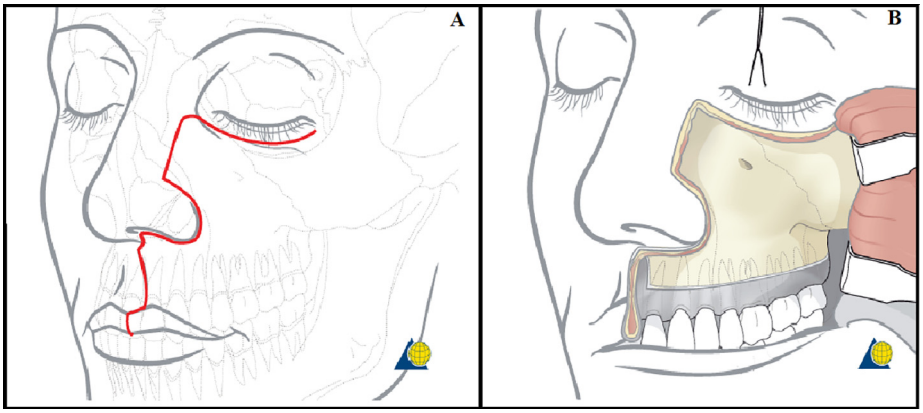

Fig. 2. Weber Ferguson approach, to access maxillary tumor extending into orbit; A: Skin incision line. B: View after elevation of cheek flap exposing anterolateral wall of maxilla. Weber Ferguson approach. AO Surgery Reference. AO Foundation Retrieved from: https://www2.aofoundation.org/wps/portal/!ut/p/a0/04_Sj9CPykssy0xPLMnMz0vMAfGjzOKN_A0M3D2DDbz9_UMMDRyDXQ3dw9wMDAzMjfULsh0VAbWjLW0!/?approach=Weber%20Ferguson%20%20&bone=&classification=&implantstype=&method=&redfix_url=&segment=&showPage=approach&treatment=&contentUrl=/srg/96/04-Approaches/A500_WeberFerguson_Midface.jsp.

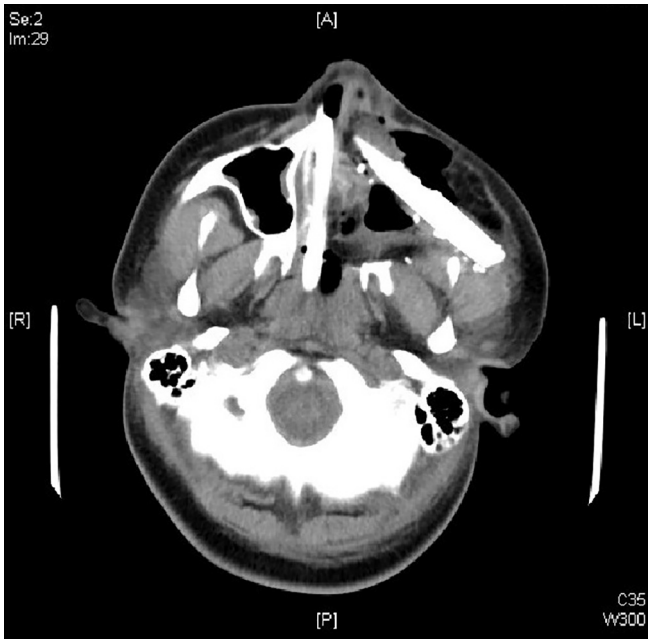

Fig. 4. Post operative axial CT scan of the patient after total left maxillectomy and fibula free flap for reconstruction.

Since the patient developed a recurrence after limited resection (enucleation), she was elected for a more aggressive treatment. Surgical resection of the mass was carried out with left total maxillectomy. The orbit was preserved via Weber Fergusson approach (Fig. 2) and reconstructed using fibula free flap. Histological sections from the surgical specimen revealed a cystic lesion lined by irregular strands of epithelium, bordered by columnar cells (palisading) that surround an islands of cells resembling stellate reticulum (plexiform pattern) (Fig. 3). The lesion was diagnosed as unicystic plexiform ameloblastoma. There were also islands of tumors within its fibrous capsule. The immediate postoperative period was uneventful. The post-operative CT scan is presented in Fig. 4. The patient remains well without tumor recurrence after 6 years.

Discussion

In this case report, we are discussing the surgical management of recurrent unicystic ameloblastoma (UA) of the maxilla involving the orbit. Maxillary unicystic ameloblastoma is a rare neoplasm, that demonstrates aggressive local behavior with invasion without distant metastasis. The thin and fragile bone of the maxilla with its spongy architecture facilitates easy infiltration of surrounding vital structures like orbit and cranium.[7]

Usually oral and maxillofacial ameloblastoma is more prevalent in the third decade or later in life[2] and men represent the majority (Table 1). Sixteen years is a very rare age for this kind of neoplasm.

The histopathological types of ameloblastoma are depicted in Fig. 5. Microscopically the unicystic variant is diagnosed by the presence of a well-defined single cystic cavity lined with odontogenic ameloblastomatous epithelium and a fibrous stroma. Sometimes it displays intraluminal plexiform patterns. Ackerman classified UA into 3 groups according to the microscopic appearance of the lesion:

Group 1. Luminal—Fibrous cystic wall is not infiltrated by tumor;
Group 2. Intraluminal – Intraluminal epithelial proliferation without any infiltration of connective tissue wall; and
Group 3. Mural – invasive islands of ameloblastomatous epithelium in the connective tissue wall not involving the entire epithelium).[1] The mural ameloblastoma is considered the most aggressive (locally) with a very high recurrence rate owing to it's invasive nature.[8] According to histopathological analysis, our patient belonged to the second subtype.

Surgery is the mainstay of treatment of UA of the maxilla. However, the ideal surgical approach for this tumor remains controversial especially in a young patient and also during the initial diagnosis. Opinions differ between conservative management like limited resection (enucleation),[9] curettage versus extensive surgical interventions like radical resection, maxillectomy (Table 1).

Although Ramesh et al. did not mention about the disease recurrence of their patient or long term follow up status; they suggested that late recurrence following treatment is common, with average being seven years.[10] The recurrence can be attributed to different factors including surgical approach during the initial diagnosis. Conservative management has better oral function and facial cosmesis but has higher incidence of recurrence than block resection and radical excision.[11] On the other hand Some reports suggests that patients can be treated conservatively by limited resection (enucleation), curettage, partial maxillectomy without recurrence[8,12] (Table 1). The vigorous curettage post limited resection (enucleation) may play a role in the implantation of foci of ameloblastoma deep into the bone.[10] Lau et al.[13]; reported recurrence rate of 3.6% of resection, 30.5% for limited resection (enucleation) alone, 16% limited resection (enucleation) with Carnoy's solution and 18% marsupialization followed by limited resection (enucleation).

Late recurrence also can be attributed to the histopathological subtypes of UA, with those invading the fibrous wall having a rate of 35.7% but others only 6.7%.[14]

Thompson et al.[15]; recommended that surgical specimens should be examined thoroughly for signs of epithelial infiltration. Their patient presented with a recurrence, after 5 years after conservative treatment, limited resection (enucleation). The histopathology specimens had revealed connective tissue infiltrated by islands and strands of odontogcnic epithelium- which is a potential risk factor for recurrence. Also the follicular variant of multicystic ameloblastoma appears to recur more frequently than the plexiform ones.

In our case, this patient presented with more aggressive recurrence than the primary disease and resulted in proptosis and destruction of the floor of the orbit. This may be attributed to both the initial conservative method of treatment and the invasion of connective tissue by the ameloblastomatous epithelium. Therefore, we preferred to use more aggressive treatment (radical resection) with reconstruction using fibula free flap and reconstructing orbital floor with titanium mesh.

Although extremely rare, maxillary ameloblastoma can present with ophthalmologic sign symptoms. Multidisciplinary approach might be needed achieve the best possible result both functionally and cosmetically. Both otolaryngologists and ophthalmologists need to keep maxillary ameloblastoma in mind when a patient presents with oph-

Table 1. Maxillary ameloblastoma case reports.

Serial no.	Year (chronological)	Author	No. of cases	Case report/ retrospective/ prospective	Age (average)	Sex	Extra maxillary extension	Histopathology	Malignancy/ metastasis During follow up	Treatment	Follow up (years)	Recurrence	Remarks (if any)
1	2016	Milman et al.[4]	23	Review	15–81 (56)	M:19	Orbit Nasal cavity	Follicular:10 Plexiform: 9	4	Conservative Surgery	0.5–31 (9.9)	16	
2	2010	Pitak-Arnnop et al.[8]	1	Case report	19	M	Nasal wall	Unicystic ameloblastoma	No	Limited resection (enucleation) with curettage	5	No	Histopathlogical subtype not mentioned
3	2010	Ramesh et al.[10]	1	Case report	30	F	Mandible	Unilocular ameloblastoma	No	Segmental mandibulectomy	...	No	
4	2006	Leibovitch et al.[5]	2	Case report	60	M	Orbit	Follicular	No	Total orbital exenteration;	1.5	No	Past history of recurrence 3 years
					73	M	Intracranial, orbit	Plexiform		Extensive resection by intracranial approach	0.5		Past history of multiple recurrence 30 years
5	1999	Iordanidis et al.[12]	1	Case report	63	F	None	Basal cell type a	No	Partial maxillectomy	4	No	
6	1997	Kieserman et al.[11]	3	Case Series	24	F	None	Ameloblastoma	Spine, humerus, clavicles, Pelvis	Caldwell-Luc procedure and removal of maxillary antrum. Palliative radiation	7	Yes	Past history of multiple recurrence 10 years, died of disease at age 31
					54	M	Nasal cavity	Cystic type ameloblastoma	No	Radical maxillectomy	4	No	
					75	M	Pterygoid	–	Eustachian tubes, infratemporal fossa. Bones lungs	Local limited resection and a partial maxillectomy	14	Yes	Died of metastaic lesion 14 years after the diagnosis
7	1993	Thompson et al.[15]	1	Case report	21	F	None	Mural	No	Limited resection (enucleation)	...	Yes (6 yrs)	Hemimaxillectomy peformed after recurrence
8	1985	Weiss et al.[6]	1	Clinicopathologic Study	72	M	Intracranial, orbit	–		Maxillectomy, local radiotherapy and exenteration		No	Past history of recurrence 6 years

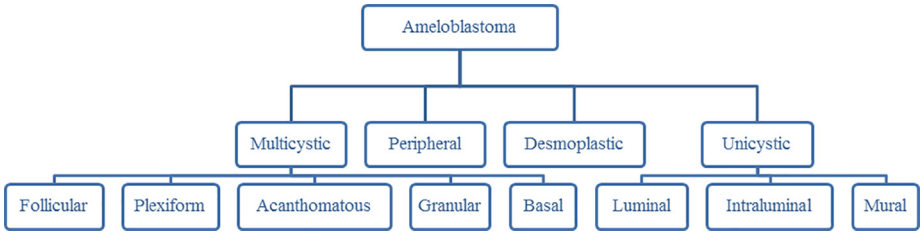

Fig. 5. Histopathological types of ameloblastoma.

thalmologic sign symptoms with a history of this rare tumor. Patients treated with conservative surgery should be evaluated by long term follow-up. For recurrence of UA of the maxilla, aggressive treatment like radical resection is a better option.

Conflict of interest

The authors declared that there is no conflict of interest.

Consent

Informed consent was obtained from the patient presented in this article.

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