CASE REPORT

Unicystic ameloblastoma of the maxillary sinus: Pitfalls of diagnosis and management

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Abstract

Background: Ameloblastoma is a common odontogenic tumor of the jaws that comprises 3 variants: conventional (solid), unicystic and peripheral ameloblastomas. Unicystic ameloblastoma (UA) in the maxillary sinus is very rare. With a secondary infection, the clinical features may lead to incorrect diagnosis and treatment.

Patients and Methods: A 19-year-old man was referred for the management of sinusitis and a mass at the right cheek. A few weeks earlier, the patient presented with acute cellulitis at the same area and underwent an incision and drainage in a primary care unit without any appropriate investigation. A radiographic examination revealed a massive lesion in the right maxillary sinus. An unerupted tooth within the lesion was found at the level of the orbital floor.

Results: The patient was successfully treated by enucleation of the tumor and curettage. The specimen was sent for histopathological examination, and the definite diagnosis was UA. The patient has been followed-up periodically for 5 years without recurrence.

Discussion: This case report suggests that primary care doctors should pay attention to differential diagnosis of orofacial lesions. It is therefore of great benefit to organize continuing education for general physicians who initially meet oral disease patients as a 'gate keeper'. Errors of clinical diagnosis and management of orofacial lesions would be minimized. Pitfalls of diagnosis and management of UA in the maxillary sinus were briefly reviewed. Hippokratia 2010; 14 (3): 217-220

Keywords: unicystic ameloblastoma; maxillary sinus; jaw tumor; orofacial infection

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Ameloblastoma is a common odontogenic epithelial tumor of the jaws. It is believed that this tumor arises from cell rests of the dental lamina, developing enamel organ, epithelial lining of an odontogenic cyst, or basal cells of the oral mucosa. The characteristic of ameloblastoma is unique because of its locally aggressive behavior and a high recurrence rate. In general, there are 3 variants of ameloblastoma: conventional (solid), unicystic and peripheral ameloblastomas. The first 2 types are intra-osseous ameloblastomas, while the other occurs in soft tissue[1-3].

Conventional ameloblastoma can be found in any age of life with no gender predilection, but it is rare in childhood and adolescence. Radiographically, the lesion is commonly seen as a multilocular radiolucency with scalloped borders. Jaw expansion and dental root resorption may be noticed. The tumor is slow growing, but locally aggressive until it becomes gigantic and destroys adjacent tissues[1-3].

Compared with the solid variant, unicystic ameloblastoma (UA) has a less aggressive nature and a lower recurrence rate. It is usually encountered in young populations.

The radiographic presentation as a unilocular radiolucency is more frequent than the multilocular pattern. The tumor can be either a tumor de novo or arising from an odontogenic cyst. Most patients are asymptomatic unless its size enlarges. UA is frequently associated with an unerupted tooth. Both solid ameloblastoma and UA typically occur in the mandible, especially the molar-ramus area. Ameloblastoma of the maxillary sinus is very rare[1-3].

The aim of this article was to report a case with UA in the maxillary sinus presenting as sinusitis and cellulitis. The clinical features misled a primary care physician into incorrect diagnosis and treatment. Pitfalls of diagnosis and management of ameloblastoma in the maxillary sinus were briefly reviewed.

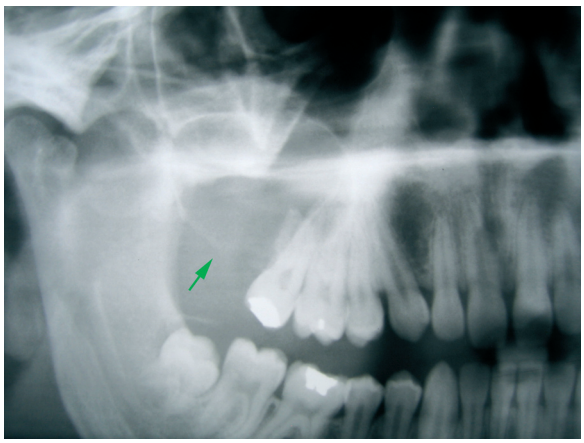


Figure 1: A part of panoramic radiograph showing a uniloc-ular radiolucent lesion involving the upper right molar teeth and the tuberosity (green arrow).

Case description

A 19-year-old white man was referred to our depart-ment with recurrent sinusitis and an enlarging mass at the right cheek. A few weeks ago, the patient was diagnosed with acute cellulitis at the same region by a family doctor. The lesion was managed with an incision and drainage 'without' radiographic and histopathological examina-tion. The patient's medical and familial histories were unremarkable.

A physical examination revealed a large, intra-osse-ous mass located at the right cheek. The lesion was pal-pable at the right maxillary vestibule and was covered by intact oral mucosa. The right upper third molar was clinically absent. There was no pain, rhinorrhea, nasal obstruction, or other antronasal abnormalities.

A panoramic film showed a radiolucent lesion with relatively well-circumscribed margins involving the right maxilla, extending from the apices of the upper right mo-lar teeth to the tuberosity. Dental root displacement was noticed, but there was no root resorption. Neighboring structures were radiographically normal (Figure 1). Com-puted tomography (CT) demonstrated a massive cystic lesion invading the entire right maxillary sinus. The lat-eral nasal wall and labial cortex of the maxilla were in-volved. The right upper third molar within the lesion was found at the level of the orbital floor (Figure 2).

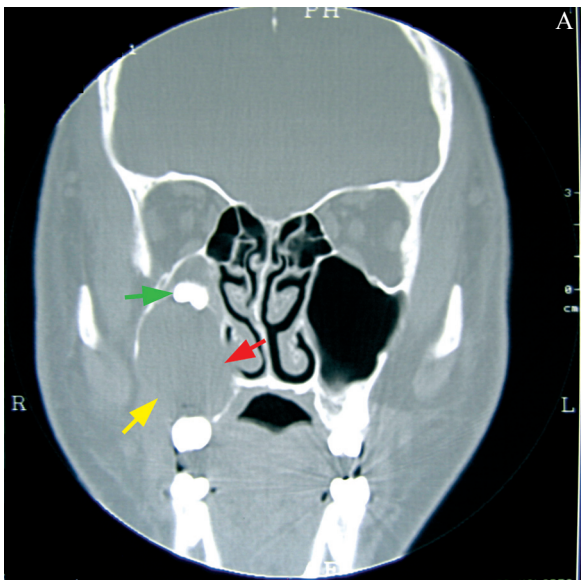
The patient elected to undergo enucleation and cu-rettage of the lesion via an intraoral vestibular approach with preservation of the maxillary sinus walls, lateral na-sal wall and orbital floor. The impacted third molar tooth within the lesion was removed at the time of surgery. A postoperative antibiotic regime was amoxicillin 500 mg 3 times a day for 1 week, together with postoperative oral care as usual. The immediate post-operative period was uneventful.

Histological sections from the surgical specimen re-vealed cystic spaces lined by palisading ameloblast-like cells demonstrating basal vacuolization, and reverse cell polarity (Figure 3). The definite diagnosis was UA. The patient has been followed-up periodically for 5 years without recurrence.

Ethical approval of this article was not required by the Committee of Human Subject Protection in Biomedi-cal Research (Comitι de Protection des Personnes: CPP) of Paris and its suburb, while we followed the National Guideline on Informatics System and Human Liberty (Dıclaration de Commission Nationale de l'Informatique et des Libertιs: CNIL) in order to protect patient confi-dentiality in a rare case report. The patient did not allow us to publish his clinical pictures.

Discussion

Diagnostic consideration for an orofacial infection in-cludes conditions of sinonasal, dental, and facial soft-tis-sue origins. Acute cellulitis of the cheek caused by dental infections is common (usually from canines or posterior teeth). However, when dental diseases are absent, diagno-



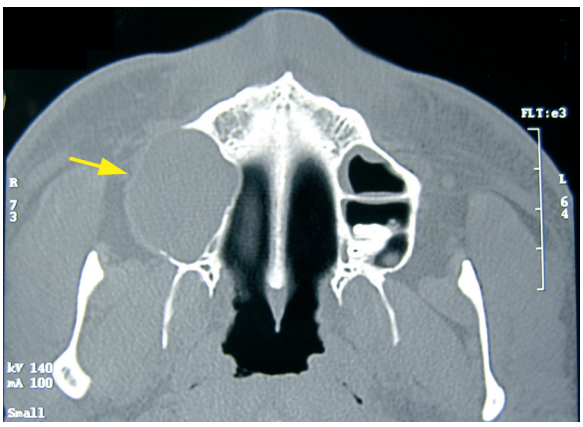


Figure 2: CT scan showing a cystic lesion in the right maxillary sinus involving the lateral nasal wall (red ar-row) and the lateral antral wall (yellow arrow). The right upper third molar was found at the orbital floor level (green arrow). (A: coronal section, B: axial section).

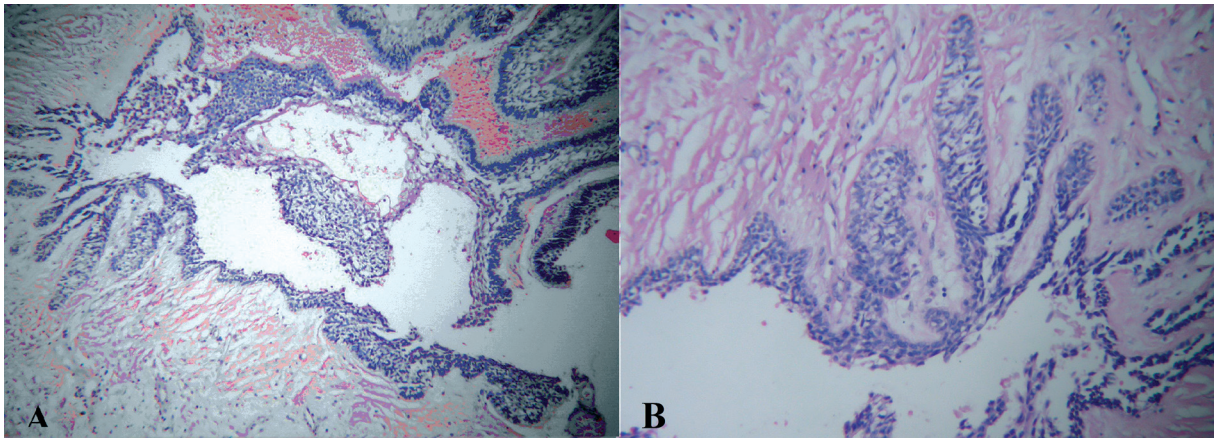


Figure 3: Photomicrograph showing a cystic space lined by ameloblastomatous epithelium (Haematoxylin and eosin stain, original magnification, A: 10x, B: 20x).

sis of the condition becomes a challenge. In this regard, an appropriate radiograph (e.g. a panoramic or Water film or CT) should be considered.

The differential diagnosis of a maxillary antral lesion includes lesions of sinonasal, odontogenic and minor salivary gland origins. Antral pseudocyst frequently occurs in the maxillary sinus. Its classical presentation is a dome-shaped radiopacity within the maxillary sinus with the density of soft tissue/fluid, whereas intrabony radiolucencies is quite rare[4].

Odontogenic cysts and tumors are high on the differential diagnosis when a lesion is encountered in the tooth-bearing area of the jaws and/or there is dental structure within the lesion. These include radicular cyst, odontogenic keratocyst, ameloblastoma, adenomatoid odontogenic tumor, ameloblastic fibroma, odontogenic myxoma, and glandular odontogenic cyst. The definite diagnosis cannot be ruled out on clinical or radiographic grounds; histopathologic confirmation is required for the diagnosis. There is general agreement that when a jaw lesion reaches a large size, a secondary infection can superimpose on the lesion.

Histologically, UA presents as a cystic lesion comprising a fibrous stroma with ameloblastomatous lining which can be classified into 3 subtypes: luminal, intraluminal (intracystic), and intramural (infiltrating) subtypes. Characteristic histopathologic features of ameloblastoma include palisading, reverse cell polarity and basal vacuolization of the ameloblast-like cells, and stellate reticulum-like tissue. A combination of 2 or 3 subtypes is possible. An intramural subtype seems more aggressive than other subtypes, requiring radical surgery and a long-term follow-up[2,5].

Treatment of UA remains controversial and greatly differs from that of conventional ameloblastoma. Algorithms for managing solid ameloblastoma and cyst-like lesions (expansile radiolucent lesions with no calcified matrix) of the jaws in our department are described in our previous publications[6,7]. Enucleation alone yields a high recurrence rate of UA, possibly because of insufficient removal of the tumor especially in regions with anatomical difficulties such as the posterior portion of the jaws. Therefore, to eliminate the tumor cells, the application of Carnoy's solution (which previously consisted of a mixture of absolute alcohol, chloroform, glacial acetic acid and ferric chloride; chloroform is no longer used because of its carcinogenicity) before curettage of surrounding bone is useful[5].

Chapelle et al[8] recommends enucleating all unilocular cystic lesions of the jaws. If a definite diagnosis is UA, a 'wait and see' protocol can be applied. However, in the retromolar trigone and ascending ramus of the mandible where odontogenic keratocyst and ameloblastoma is common, excising the overlying oral mucosa, coupled with the treatment with Carnoy's solution or liquid nitrogen after enucleation, is recommended. Aspiration or incisional biopsy is beneficial in cases involving multilocular lesions. However, it may not be representative of the entire lesion, and inflammation may lead to an erroneous diagnosis[8,9]. Some patients are diagnosed with UA after multiple recurrences[3,5,10].

To ensure that the tumor is totally removed, Tashiro[11] recommends applying gentian violet on the surrounding bone. The stained bone requires careful curettage and/or peripheral ostectomy. If the inferior dental nerve of the mandible is involved, nerve transposition is of benefit prior to the curettage and/or ostectomy[2]. Decompression or marsupialization is suitable for large cysts, irrespective of the histological diagnosis. These techniques reduce the lesion size, minimize the extent of subsequent treatment (enucleation with/without curettage, or resection), and allow the possibility for an incisional biopsy[2,3,8].

Resection is reserved only for cases with multiple recurrences and/or expansive or destructive lesions[2,12]. A systematic review showed weak evidence supporting the lower recurrence rate following the resection of UA[3]. It may therefore be unethical if an aggressive approach is applied without considerable benefits over drawbacks.

In our department, all cyst-like lesions of the jaws are treated by simple enucleation[7]. Despite a relatively low

recurrence rate, continuation of close follow-up is recommended once a year during the first 5 years and then every 2 years[8,12]. CT scan is considered useful for both the treatment planning (e.g. identification of the extension of the lesion, cortical perforation, and relationship to other important structures) and the surveillance of recurrence when the tumor involves the maxillary sinus[12].

Conclusion

Large UA of the maxillary sinus is rare. A large-sized tumor with a secondary infection may mislead a clinician as seen in the present case. In our patient, we hypothesized that the expanding lesion with the perforation of labial maxillary cortex (anterior wall of the maxillary sinus) made the antrum predispose to infection as sinusitis and cellulitis. Orofacial problems are common in general practice[13]. A recent British survey revealed that oral disease patients often present to their general doctors before specialists[14]. However, the primary care doctors seemed to be unfamiliar with oral lesions. The oral cavity is not routinely examined by general medical practitioners[14]. It is therefore of great benefit to organize continuing education for general physicians who may initially meet oral disease patients as a 'gate keeper' in the primary care setting[13-16]. Errors of clinical diagnosis and management of orofacial lesions would be minimized.

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Disclosure of potential conflicts of interest

The authors had full freedom of manuscript preparation and there were no potential conflicts of interest.

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