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Unusual presentation of ameloblastoma and a review of its biological behaviour

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Abstract

Ameloblastoma is a benign odontogenic tumour occurring primarily in the mandible with rare occurrence outside the jaw. We report a case of an ameloblastoma presenting as a polypoidal mass in the right nasal cavity of a 72-year-old woman. She presented with progressive nasal blockage symptoms and examination revealed a mass occupying the right nostril. Initial tissue biopsy confirmed conventional ameloblastoma. Subsequent imaging study showed a mass involving the right maxilla. Primary tumour from the maxilla extending into the sinonasal tract was favoured over a much rarer primary ameloblastoma of the sinonasal tract, in view of the tumour bulk in the maxilla. Unfortunately, the patient defaulted her follow-up. Ameloblastomas are known to recur locally thus excision with clear margins is paramount. Recurrence is also influenced by factors such as the presence of B-Raf proto-oncogene (BRAF) and Smoothened, frizzled class receptor (SMO) genetic mutations, with SMO mutation found to be associated with maxillary location in the older age group. Ameloblastomas can metastasise without undergoing malignant cytological changes, with mandibular tumours being far more likely to do so. Long-term follow-up and regular computed tomography (CT) monitoring is required post excision.

Keywords: *ameloblastoma, maxillary tumour, nasal mass, odontogenic tumour*

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Introduction

Ameloblastoma is a benign odontogenic tumour of the jawbone predominantly occurring at the mandible with propensity for local extension if left untreated (Masthan *et al.,* 2015). It is often asymptomatic but may present with swelling and or pain in the jaw. Although they are benign tumours, ameloblastomas have a strong inclination of recurrence after treatment, with a rate of up to 90% (Goh *et al.,* 2021). We describe an unusual case of ameloblastoma presenting as a polyp within the nasal cavity.

Case Report

A 72-year-old woman presented with right nasal block with progressing severity over 1 year. It was associated with episodes of mucopurulent discharge of the same duration and a few episodes of self-limiting trivial epistaxis. Rhinoscopy showed a pink mass occupying the entire right nostril, remnant of head of inferior turbinate seen. The mass appeared to be friable and bled easily on touch during procedure, but the operator was unable to advance the scope to visualize posteriorly. On further examination, the oral cavity was found to be

fully edentulous. A small pink soft tissue mass protruded through a focal mucosal breach over the right gingivobuccal sulcus, representing the inferior extension of the same lesion that occupied the right nasal cavity.

A biopsy of the nasal mass was taken; macroscopic examination showed multiple pieces of brownish tissue measuring 10mm in aggregate diameter. Microscopic examination showed multiple fragments of tissue focally lined by stratified squamous epithelium. Most fragments are lined by palisading basaloid columnar epithelium arranged in anastomosing strands and islands. These columnar epithelia exhibited uniform elongated hyperchromatic nuclei, inconspicuous nucleoli and reversed polarity with presence of subnuclear vacuolation. The central stroma exhibited extensive squamous metaplasia with minimal areas of typical loose stellate reticulum-like cells (Figure 1). There was no dysplasia or evidence of malignancy seen. Immunohistochemically, the columnar epithelium and stellate reticulum were positive for CK19. The interpretation was that of an ameloblastoma.

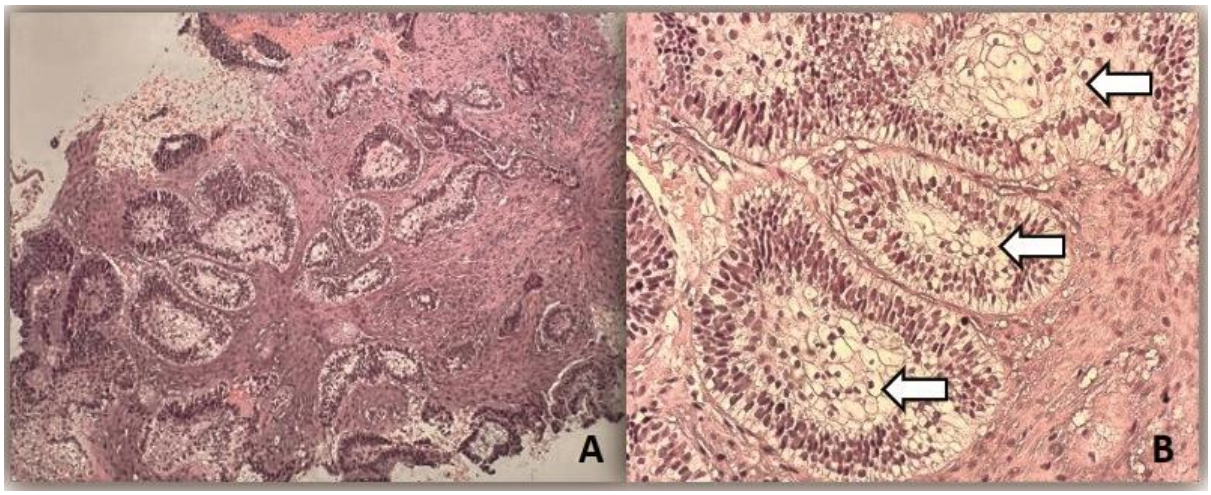

Figure 1. Histopathological examination, A (H&E, x100) and B (H&E, x400) of the mass showing fragments of tissue composed of palisading basaloid columnar epithelium with reversed polarity and loose stellate reticulum cells (white arrow).

Her subsequent CT scan 2 days later demonstrated that both the intranasal mass and the gingivobuccal lesion were contiguous extensions of a large heterogeneously enhancing tumour, centred within the right maxillary sinus. The mass measured 6.7 x 6.2 x 5.1cm (AP x W x CC) (Figure 2). The mass showed extensive destruction of the surrounding maxillary and nasal cavity walls with extension into the nasal cavity and infratemporal fossa. Areas of hypodensities within the mass seen, likely of necrotic areas. Multiple air pockets noted at the anterior and inferior aspects of the mass, which was likely from previous biopsy. Unfortunately, this patient was lost in follow-up.

Discussion

Ameloblastoma is the most common odontogenic tumour, apart from odontomas. It is a benign intraosseous progressively growing epithelial odontogenic neoplasm characterized by expansion and a tendency for local recurrence if not adequately removed. The peak incidence occurs in the 4th and 5th decades of life. Approximately 80% of all ameloblastomas are found in the mandible; they occur most often in the posterior region, followed by the anterior mandible, posterior maxilla and anterior maxilla (Hertog *et al.,* 2012; Siar *et al.,* 2012).

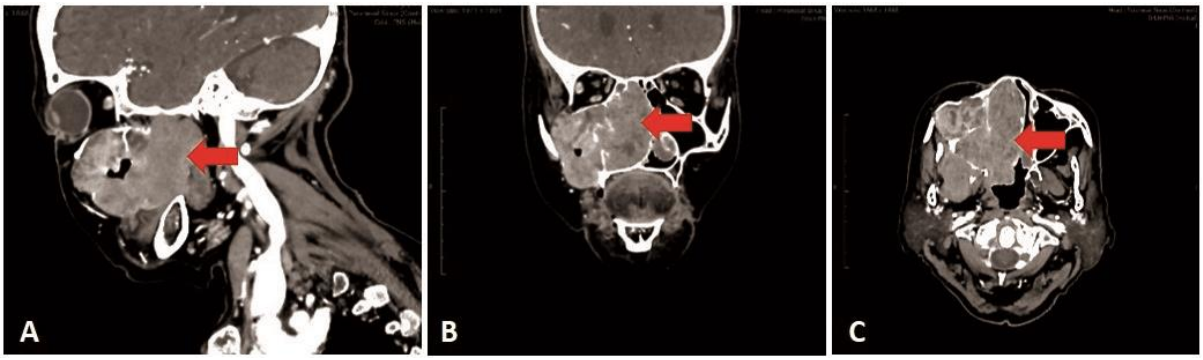

Figure 2. Sagittal (A), coronal (B) and axial (C) views showing a large heterogeneously enhancing solid mass (red arrow) occupying the right infratemporal space, with its epicentre at the right maxillary sinus. The tumour demonstrated extensive destruction of the maxillary sinus and nasal cavity walls with extension into the nasal cavity and infratemporal fossa. Areas of hypodensities within the mass seen likely of necrotic areas.

They are usually asymptomatic but may present with a painful or painless mass around the jaw region. Ameloblastomas are locally aggressive, invading from the bone to the surrounding soft tissues. They usually have a progressive growth with large local extension, bone destruction and dental resorption.

Ameloblastomas arising from sites other than normally situated odontogenic epithelium (extragnathic) is extraordinarily rare. Ameloblastoma of the sinonasal tract is included in these extragnathic ameloblastomas (Schafer *et al.,* 1998). As ameloblastomas are locally aggressive, a primary gnathic ameloblastoma with spread to the sinonasal tract should first be ruled out. These tumours usually come as painless swellings with possible concurrent loosening of teeth, malocclusion and altered sensation in the teeth and occasionally pain. Local extension of the tumour may erode and destroy the bone and subsequently spread to the adjacent soft tissues. Extensive local progression has been reported with one case seen extending up to the brain (Bettoni *et al.,* 2018). Our case could possibly be a right sinonasal ameloblastoma, metastasis from a previous tumour or a right maxillary ameloblastoma with intranasal extension. With the bulk of the tumour at the right maxillary sinus, and no prior history, the third option seems most likely.

Local recurrences of ameloblastomas show a marginal predilection to females with a ratio of 1:1.3. Reccurent ameloblastomas are seen in as many as 16.35% of cases to the mandible and 15.68% of cases to the maxilla (Goh *et al.,* 2021). Maxillary ameloblastoma is known to be more aggressive due to the cellular attributes of ameloblastoma and the character of bone in the maxilla (Darshani Gunawardhana *et al.,* 2010). The rate of local recurrence is increased 3.15 folds in cases with conservative treatment in contrast to radical treatment (Almeida Rde *et al.,* 2016). Therefore, in terms of patient management, most authors conclude that surgical excision with clear margins should remain the mainstay of treatment (Almeida Rde *et al.,* 2016; Hosalkar *et al.,* 2021; Pandiar *et al.,* 2021).

Metastasizing ameloblastoma is a distinct subtype in the WHO Classification of Head and Neck tumours and is differentiated from ameloblastic carcinoma by the absence of cytologic atypia. Metastatic deposits usually occur in the lungs, followed by the lymph nodes and bones (Tilakaratne *et al.,* 2023). Metastasis has been associated with poor patient outcome (Hosalkar *et al.,* 2021). Metastasizing ameloblastoma harbours BRAF p.V600E mutation similar to conventional ameloblastoma. Reported risk factors for metastasis include large size, rapid enlargement, prolonged clinical course, inadequate surgical excision, and multiple recurrences at the primary

site (Hosalkar *et al.,* 2021; Pandiar *et al.,* 2021).

The reason for metastasis in a benign entity such as ameloblastoma is not well-understood. It has been postulated that the basis may be similar to that seen in metastasis of other benign neoplasms such as benign metastasizing leiomyoma, giant cell tumour of bone and pleomorphic adenoma. The leading reason is due to incomplete removal with tumoural seeding during surgical removal. A higher frequency of surgical interventions due to multiple recurrences is associated with increased likelihood of metastasis (Hosalkar *et al.,* 2021). However, the authors also highlight that the flaw in this theory is that the immune mechanism is able to destroy distant seeding of benign tumour cells. Other routes of spread such as hematogenous, lymphatic and aspiration have been proposed with no exact mechanism established to date (Pandiar *et al.,* 2021).

A key factor in predicting metastatic behaviour is the tumour location. Mandibular ameloblastoma has been found to metastasise more frequently by virtue of being the primary location of most ameloblastomas (Pandiar *et al.,* 2021). Another factor is the histological subtype. Plexiform ameloblastoma has been associated with higher rates of metastasis. It has been theorized that the bud-like structures seen in this histological subtype are able to dislodge and become implanted at distant sites. In contrast, one review found that follicular subtype was the more commonly metastasizing subtype (Pandiar *et al.,* 2021). Other factors predicting higher likelihood of metastasis include male gender, large size of primary tumour, fast local spread and prolonged disease (Hosalkar *et al.,* 2021; Pandiar *et al.,* 2021).

Approximately 90% of ameloblastomas harbour mutations involving the MAPK signalling pathway, with BRAF V600E being the commonest alteration. In contrast, SMO mutations occur less frequently but have been shown to correlate with maxillary tumours arising in older patients, suggesting distinct molecular pathways according to tumour location (Sweeney *et al.*, 2014). Furthermore, tumours harbouring SMO mutations or multiple concurrent genetic alterations demonstrate a higher risk of recurrence compared with BRAF-mutated tumours (Gültekin *et al.,* 2018). Case series have demonstrated remarkable tumour regression following treatment with BRAF inhibitors such as dabrafenib, either alone or in combination with MEK inhibitors, particularly in unresectable, recurrent or metastatic BRAF-mutated ameloblastomas (Kaye *et al.*, 2014; Tilakaratne *et al.,* 2023; Grynberg *et al.*, 2024). These findings support the role of molecular profiling for prognostication and may guide targeted therapeutic strategies in selected patients (Brown *et al.*, 2014; Sweeney *et al.*, 2014; Gültekin *et al.*, 2018; Tilakaratne *et al.,* 2023; Grynberg *et al.*, 2024).

Although treatment outcome could not be assessed because the patient defaulted follow-up, this case remains educational because it illustrates an uncommon presentation of maxillary ameloblastoma masquerading as a benign nasal polyp. Recognition of this presentation is important to avoid diagnostic delay and to prompt appropriate cross-sectional imaging for assessment of the primary tumour.

Conclusion

Behaviour of ameloblastoma depends on its location, completeness of excision and presence of certain genetic mutations. Regional spread of ameloblastoma is more likely and need to be ascertained before interpreting an extragnathic primary, as this diagnosis affects prognosis. Targeted therapy for ameloblastoma may be contemplated in some cases.

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Competing Interest

The authors declare that they have no competing interests.

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