The Journal of Laryngology & Otology (2007), ***121***, *72–75*.

doi:10.1017/S0022215106003604
Printed in the United Kingdom
First published online 1 November 2006

Clinical Record

Ameloblastoma: a rare nasal polyp

D Bray, A Michael, D T Falconer*, H S Kaddour

Abstract

Ameloblastoma is an odontogenic neoplasm of enamel organ type tissue which does not undergo transformation to the point of enamel formation. We present the second case in the English literature of maxillary ameloblastoma that presented with nasal obstruction and rhinorrhoea, and the first to be excised using a combined maxillotomy and endoscopic ethmoidectomy. The patient had no previous dental history. The unusual presenting symptoms, as well as the highly destructive nature of these lesions when arising in the maxilla, make them worthy of consideration in the differential diagnosis of nasal and maxillary masses. We discuss the clinical features, pathology and management of these lesions and review the literature.

Key words: Ameloblastoma; Maxillary Neoplasms; Nasal Polyp

Introduction

Ameloblastoma is an odontogenic neoplasm of enamel organ type tissue which does not undergo transformation to the point of enamel formation. We present the second case in the English literature of maxillary ameloblastoma that presented with nasal obstruction and rhinorrhoea, and the first to be excised using a combined maxillotomy and endoscopic ethmoidectomy. The patient had no previous dental history. The unusual presenting symptoms, as well as the highly destructive nature of these lesions when arising in the maxilla, make them worthy of consideration in the differential diagnosis of nasal and maxillary masses. We discuss the clinical features, pathology and management of these lesions and review the literature.

Case report

An 81-year-old man presented to the ENT clinic with a four month history of progressively worsening, right-sided nasal obstruction and clear rhinorrhoea. There was no history of dental or concurrent medical disease. He denied allergies and was a non- smoker.

Anterior rhinoscopy revealed a smooth, polypoidal mass in the right nostril, with moderate left nasal septal deviation. The mass arose from the right maxillary antrum and could be seen at the middle meatus. The post-nasal space was clear, there was no facial swelling, deformity or crepitus, and the infraorbital nerve was unaffected. The rest of the ENT examination was unremarkable. Haematological and biochemical tests were normal.

A computed tomography (CT) scan of the paranasal sinuses showed a homogenous, polypoidal mass obliterating the nasal vestibule and involving the right maxillary, ethmoid and frontal sinuses (Figure 1). There was bony destruction of the anterior and medial walls of the right maxillary antrum and leftward deviation of the cartilaginous nasal septum. A magnetic resonance imaging (MRI) scan showed an extensive, mixed density tissue mass filling the right maxilla and extending to the right nasal cavity and ethmoid air cells (Figure 2). The orbit and cribriform plate were not breached. A biopsy of the nasal component was taken under general anaesthesia. Histologically, the lesion was a classical plexiform ameloblastoma with no evidence of malignancy.

Surgery was as follows. A Weber Ferguson incision was made through the upper lip and round the right side of the nose to the medial canthus (Figure 3). The maxilla was then downfractured bilaterally at the Le Fort one level. This provided excellent exposure of the tumour mass sitting in the right maxilla, which was enucleated from the maxillary antrum, nasal cavity and ethmoids en bloc. The tumour was easily separated from the surrounding structures and complete clearance of the tumour was possible via an endoscopic right ethmoidectomy using a 0° Hopkins rod endoscope. The tumour mass measured $60 \times 55 \times 30$ mm and weighed 40 g (Figure 4). Histological analysis confirmed ameloblastoma (Figure 5). The biopsies taken from the normal tissues surrounding the tumour showed no evidence of neoplasia.

Post-operative recovery was uneventful, and the patient ate breakfast the next day without complication. He was discharged two days post-operatively.

There were no complications during eight post-operative months' follow up. The patient had an excellent bilateral nasal airway and no further rhinorrhoea. He remained under surveillance in the out-patient clinic.

Discussion

Ameloblastoma has been described under various terms in the literature and is classified with the odontogenic neoplasms of the jaws arising from epithelial tissue. Other terms that have been used are adamantinoma, multilocular cyst, adamantinoblastoma and carcinoma of the tooth germ residue.

Cusack was the first to recognize this tumour, in 1827,[1] and it was later described by Falkson in 1879[2] (quoted in

From the Departments of Otolaryngology and *Maxillofacial Surgery, Harold Wood Hospital, Romford, UK.
Accepted for publication: 8 February 2006.

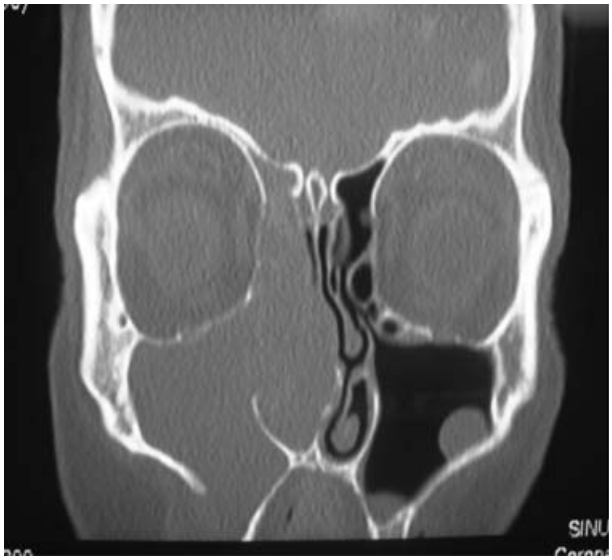

FIG. 1
Computed tomography scan of the paranasal sinuses.

Lucas).[3] The term 'adamantinoma' was cited by Malassez in 1885[4] to describe an odontogenic tumour supported by fibrous stroma. The term, however, was misleading because the lesions contained no enamel. This anomaly was addressed by Ivey and Churchill in 1933; they referring to the lesion as an 'ameloblastoma', thereby eliminating any suggestion of enamel production.[5] The

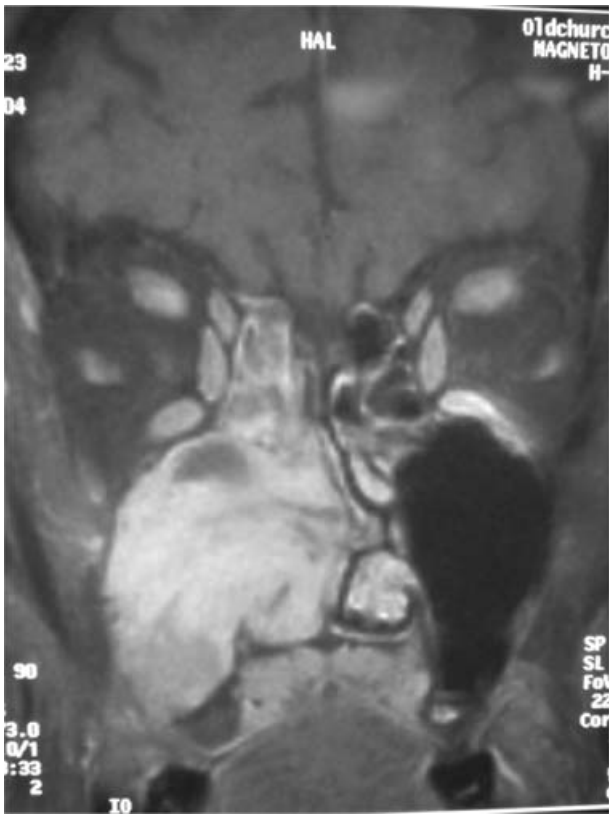

FIG. 2
Coronal T2-weighted magnetic resonance imaging scan.

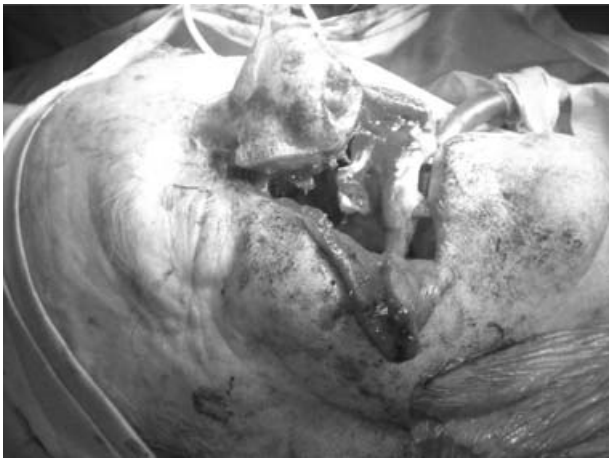

FIG. 3
Maxillotomy via Weber Ferguson incision.

World Health Organization defines ameloblastoma as 'an invasive, potentially malignant neoplasm that consists of proliferating odontogenic epithelium supported by fibrous stroma'.[6]

In 1952, Hertz reviewed the theories of pathogenesis of the ameloblastoma.[7] Subsequently, Hinds *et al*.[8] and then Gorlin *et al*.[9] collected and summarized the four possible sources of cells that could give rise to these lesions, as follows: (a) the epithelial lining of an odontogenic cyst; (b) dental lamina or enamel organ; (c) stratified squamous epithelium of the oral cavity; and (d) displaced dental epithelial remnants (accounting for tumours arising in soft tissues without bone involvement).

Ameloblastomas are exceedingly rare tumours and their incidence constitutes only 0.03 per cent of all neoplasms. Only 1 per cent of odontogenic tumours and cysts are estimated to be ameloblastomas, and origination in the mandible is more common than the maxilla by a factor of four to one.[9,10] A small minority may be found in other regions of the body, such as the pituitary gland, the tibiae, the pharynx, the ovaries, the sacrum and the mastoid process.[11] Kegel, in 1932, described a predominance in negroes of 11:1 when compared with the Caucasian population,[12] but this has been disputed by subsequent authors.[13]

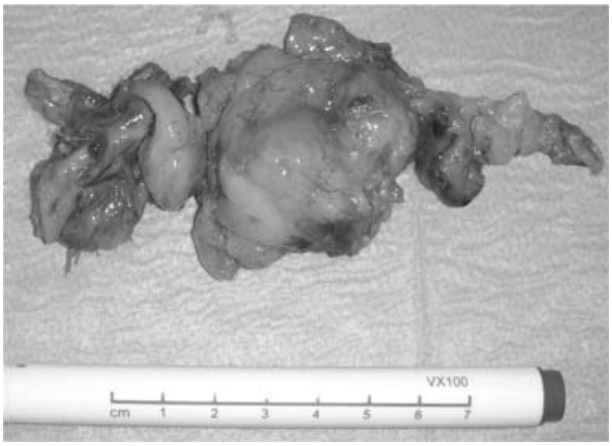

FIG. 4
En bloc resection specimen.

https://doi.org/10.1017/S0022215106003604 Published online by Cambridge University Press

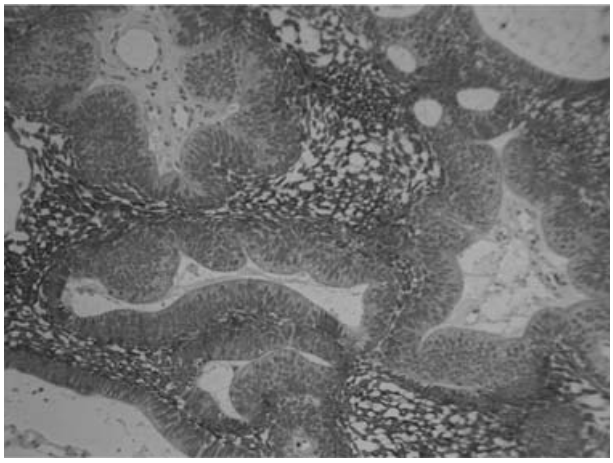

FIG. 5
Ameloblastoma (CK8 stain; ×50).

Among the histological types of ameloblastoma, follicular and plexiform patterns are the most common. Less common cellular variants are the desmoblastic ameloblastoma, basal cell ameloblastoma, keratoameloblastoma, papilliferous keratoameloblastoma, clear cell ameloblastoma and unicystic ameloblastoma.[14] Except for the unicystic variant, which has a low recurrence rate, no significant differences in the behaviour of these variants have been observed.[15]

Histologically, ameloblastomas are considerably locally invasive but are benign and therefore do not metastasize. It is theoretically possible for ameloblastoma to undergo transformation to a higher grade of malignancy, such as adenocarcinoma. Repeated stimulation by radiotherapy, cautery and surgery may be decisive in this transformation.[16]

Classically, the average age of presentation is in the fifth decade; however, ameloblastoma may occur in all age groups. Young and Robinson reported a series of 31 cases in children.[17] Their conclusion, however, was that ameloblastoma in children may be a different type of lesion to that seen in adults, probably an odontoma or mixed odontogenic tumour.

As in our reported case, the patient may be asymptomatic for a long period. In one series, the mean length of time the tumour was considered present before diagnosis was five years.[10] With the passage of time and with slow growth of the tumour, gingival and/or facial deformity may arise due to asymmetrical swelling of the jaw or maxilla. In more advanced stages, difficulty in mastication, deglutition and articulation may arise. Other symptoms arise from dental involvement, with dental pain, loose teeth and non-eruption of teeth. Recent dental extraction is often reported, which is almost certainly a consequence of the tumour rather than of aetiological significance. In cases (such as the present one) in which the tumour is located in the upper jaw or maxilla, the patient may complain of epistaxis, nasal obstruction and/or persistent rhinorrhoea.[16]

Radiographic examinations, including plain radiography, panoramic radiography and conventional tomography, have played an important role in the diagnosis and management of ameloblastoma. The radiologic findings of expansion of cortical plates with scalloped margins, multiloculation (with a 'soap bubble' appearance), resorption of tooth roots and predilection in location are key to correct diagnosis. However, these findings are not pathognomic for ameloblastoma and may indicate odontogenic keratocyst, odontogenic myxoma, ameloblastic fibroma, giant cell granuloma, immature ossifying fibroma or aneurysmal bone cyst. In addition, ameloblastoma frequently recurs after inappropriate surgery, and accurate pre-operative evaluation of the boundaries of the tumour is essential. Computed tomography can delineate soft tissue masses, destruction of cortical bone and extension of tumour into adjacent structures more clearly than can conventional radiography, but, again, this is not pathognomic for ameloblastoma. In addition, CT images are susceptible to streaking artefacts caused by dental materials. Minami *et al.* assessed MRI results in cases of maxillomandibular ameloblastoma and concluded that several common findings were present, as follows: multilocularity, mixed solid and cystic components, irregularly thick walls, papillary projections, and marked enhancement of the walls and septa.[18] These authors found that MRI was superior to conventional radiography and CT in demonstrating components of the tumour, features of the walls of cystic components and the nature of cystic fluids but not in delineating cortical margins and soft tissue invasion.

Once the histological diagnosis has been established, partial maxillectomy with a 1cm resection margin is the treatment of choice for adult maxillary ameloblastoma. Regular follow-up surveillance of the maxillary antrum is mandatory. Conservative management and curettage were once in vogue; however, the tumour's capacity for continued growth and local invasion led to recurrence rates approaching 100 per cent; more aggressive treatment strategies were thus adopted.[19]

Curiously, in Young and Robinson's study of ameloblastoma in the zero to nine year age group, the results of conservative treatment with curettage and local excision were favourable and recurrence-free.[17] Ameloblastomas are resistant to radiotherapy, with a 72 per cent recurrence rate in one study.[11] Radiotherapy may however have a palliative role in the management of patients unsuitable for surgery, and temporary resolution has been reported.[16]

The advent of endoscopic technology has afforded access to the sino-nasal spaces, previously thought unreachable. Invasion of ethmoid air cells, pterygomaxillary fossa, temporal fossa and skull base was thought irresectable as recently as the 1980s.[20]

- **This paper describes a rare ameloblastoma originating from the maxillary sinus and presenting as a nasal polyp**
- **These tumours tend to be benign and slow-growing but with local tissue invasion. Treatment is surgical, as the tumour is resistant to chemoradiation therapy**
- **In this case, surgical treatment was by a combined endoscopic ethmoidectomy and external maxillotomy. This approach avoided the need for a dental obturator**

In our case, the tumour had extended beyond the maxillary antrum to the ethmoid air cells. Previously, these patients would have undergone partial maxillectomy for tumour clearance and to enable access to the maxillary antrum for post-operative surveillance. In our case, a maxillotomy was performed by downfracturing the maxilla at the Le Fort one level. The medial wall of the right maxilla was excised, and this was combined with an endoscopic ethmoidectomy to facilitate full ethmoid clearance. The maxillotomy was then reconstructed with titanium

https://doi.org/10.1017/S0022215106003604 Published online by Cambridge University Press

plates. This is the first report of this combined approach, which not only enabled en bloc resection of the tumour, previously thought impossible, but also spared the patient the significant morbidity of a dental obturator. Out-patient surveillance of the maxillary antrum was possible with a 30° Hopkins rod endoscope.

Conclusion

We reported a case of maxillary ameloblastoma with ethmoid involvement, presenting as nasal obstruction and rhinorrhoea. Treatment with a combined approach maxillotomy and endoscopic ethmoidectomy was successful and avoided the associated morbidity of a dental obturator.

References

1 Cusack JW. Report of the amputations of portions of the lower jaw. *Dublin Hosp Rec* 1827;**4**:1–38
2 Falkson R. *Virchows Archiv fur pathologische Anatomie und Physiologie und fur Klinische Medizin* [in German] 1879;**76**:504
3 Lucas RB. *Pathology of Tumours of the Oral Tissues*. London: Churchill, 1964;30
4 Malassez L. *Archives de Physiologie Normale et Pathologique* [in French] 1885;**6**:379
5 Ivey RH, Churchill HR. The need of a standardised surgical and pathological classification of tumours and anomalies of dental origin. *Am Assoc Dent Sch Trans* 1933;**7**:240–5
6 *International Histological Classification of Tumors. Histological Typing of Odontogenic Tumours, Jaw Cysts and Allied Lesions*, monograph no 5. Geneva: World Health Organization, 1971
7 Hertz J. Adamantinoma: histopathologic and prognostic studies. *Acta Chir Scand* 1952;**102**:405–32
8 Hinds EC, Pleasants JE, Snyder PL. Management of ameloblastoma. *Oral Surg* 1954;**7**:1169–77
9 Gorlin RJ, Chaudry AP, Pinborg JJ. Odontogenic tumors. *Cancer* 1961;**14**:73–101
10 Small IA, Waldron CA. Ameloblastoma of the jaws. *Oral Surg Oral Med Oral Pathol* 1955;**8**:281–97
11 Pantazopoulos PE, Nomikos N. *Ann Otol Rhinol Laryngol* 1966;**75**:1160–7
12 Kegel RFC. *Arch Surg* 1932;**25**:498
13 Lucas RB. *Pathology of Tumours of the Oral Tissues*. London: Churchill, 1964;30
14 Pillai RS, Ongole R, Ahsan A, Radhakrishnan RA, Pai KM. Recurrent desmoplastic ameloblastoma of the maxilla: a case report. *J Can Dent Assoc* 2004;**70**:100–4
15 Thompson IO, Van Rensburg LJ, Phillips VMJ. Desmoblastic ameloblastoma: correlative histopathology, radiology and CT-MR imaging. *J Oral Pathol Med* 1996; **25**:405–10
16 Shaw HJ, Katsakis DK. Ameloblastoma of the maxilla: a clinical study with 4 cases. *J Laryngol Otol* 1972;**87**: 873–84
17 Young DR, Robinson M. Ameloblastomas in children: report of a case. *Oral Surg* 1962;**15**:1155–62
18 Minami M, Kaneda T, Yamamoto H, Fukimura H, Nishizawa Y, Fujitani K *et al*. Ameloblastoma in the maxillomandibular region: MR imaging. *Radiology* 1992; **184**:389–93
19 Sedhev MK, Huvos AG, Strong EW, Gerold FP, Willis GW. Ameloblastoma of maxilla and mandible. *Cancer* 1974;**33**:324–33
20 Wenig BL, Sciubba JJ, Cohen A, Goldstein A, Abramson AL. An unusual cause of nasal obstruction: ameloblastoma. *Otolaryngol Head Neck Surg* 1985;**95**: 426–32

Address for correspondence:
Mr D Bray,
6 Bedford Terrace,
Royal Tunbridge Wells TN1 1YJ,
UK.

E-mail: dbray@doctors.org.uk

Mr D Bray takes responsibility for the integrity of the content of the paper.
Competing interests: None declared