Recurrent Desmoplastic Ameloblastoma of the Maxilla: A Case Report

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Abstract

A case of desmoplastic ameloblastoma recurring within 2 months of curettage is presented. This tumour appeared in the premolar region of the left maxilla with involvement of the antrum. The 24-year-old female patient was initially treated by curettage with wide surgical margins. Later, partial maxillectomy was carried out followed immediately by iliac bone graft. The case was followed with periodic plain radiography and computed tomography. The presence of a pulpally infected premolar and the atypical radiographic appearance obscured the disease. The biologic profile of this tumour is not fully understood because of the limited number of reported cases, coupled with inadequate long-term follow-up. A review of the lesion with emphasis on the pathogenesis of recurrence is discussed.

MeSH Key Words: *ameloblastoma/pathology; maxillary neoplasms; neoplasm recurrent, local*

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This article has been peer reviewed.

Ameloblastomas are benign, locally aggressive, polymorphic neoplasms of proliferating odontogenic epithelial origin.[1] They are the most common clinically significant odontogenic neoplasms affecting the jaws.[2] Among the histologic types of ameloblastoma, follicular and plexiform patterns are the most common; variants include acanthomatous and granular cell types. Less common cellular variants are the desmoplastic ameloblastoma, basal cell ameloblastoma, keratoameloblastoma, papilliferous keratoameloblastoma, clear cell ameloblastoma and unicystic ameloblastoma. Except for the unicystic type, which has a low recurrence rate, no significant differences in the behaviour of these variants have been observed.[1] A desmoplastic variant with features of other histologic types is termed a "hybrid."[3]

In 1984, Eversole and others[4] described 3 cases of a variant of ameloblastoma. The characteristic feature was extensive stromal desmoplasia with small nests, cords and strands of odontogenic epithelium. This variant was included in the World Health Organization's *Histopathological Typing of Odontogenic Tumors*.[5] It occurs mainly in the anterior region of the jaws and appears radiographically as a diffuse, mixed radiolucent–radiopaque lesion with a honeycomb or soap bubble pattern and indistinct borders.[6]

The desmoplastic variant differs from other histologic types of ameloblastoma in that it is located in the anterior or premolar regions of the maxilla or mandible and its radiographic appearance is often more typical of a fibro-osseous lesion. Because only a few cases have been reported, the true biologic profile is yet to be fully understood. The purpose of this paper is to report a case of recurrent as well as residual desmoplastic ameloblastoma examined periodically by radiography before surgery.

Case Report

A 24-year-old woman reported to the department of oral medicine and radiology complaining of a swelling in the left anterior region of her upper jaw that had been present for 2 weeks. It was not associated with pain and had enlarged slowly to its current size. One month previously, the woman had undergone endodontic treatment of the upper left second premolar. She had no history of displacement of teeth. There was no other relevant medical history.

Figure 1: *Intraoral photograph showing the swelling between the canine and premolar teeth.*

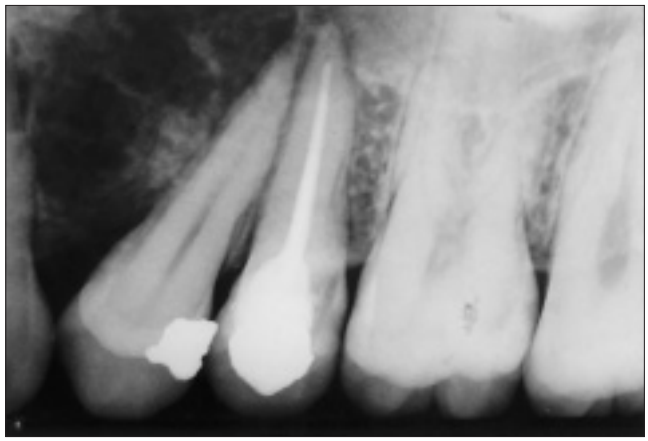

Figure 2: *Radiograph showing a honeycomb pattern of radiolucency, displacement of the roots and involvement of the floor of the antrum.*

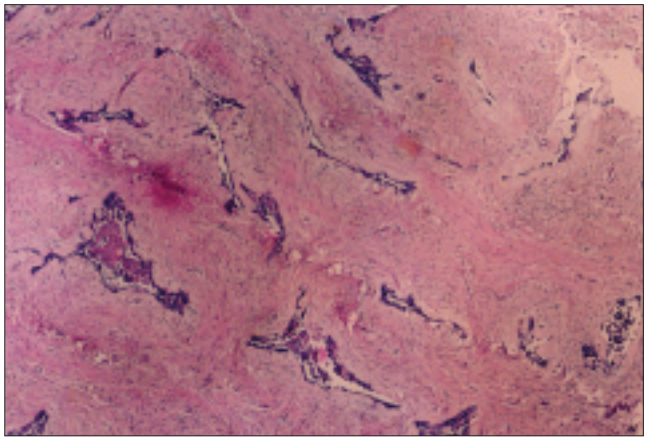

Figure 3a: *Photomicrograph (magnification ×5) showing extensive stromal desmoplasia.*

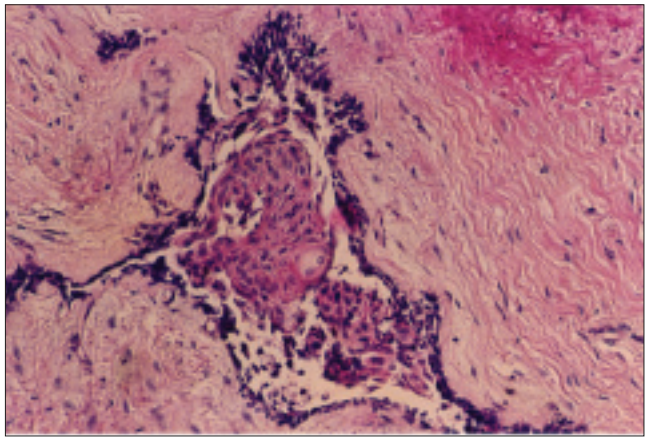

Figure 3b: *Photomicrograph (magnification ×20) showing nest of ameloblast cells with squamous metaplasia.*

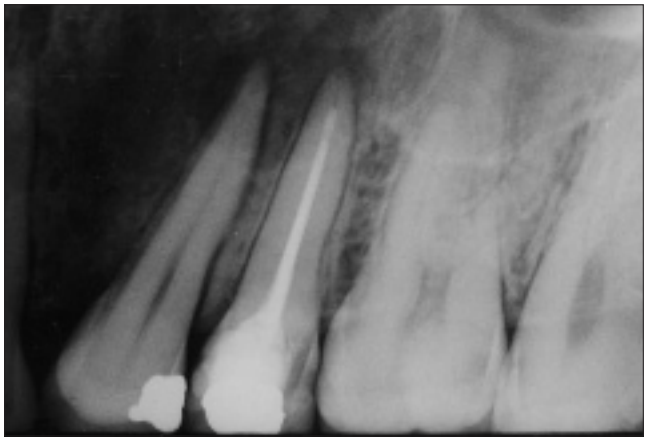

Figure 4: *Radiograph showing a new radiolucency at the periapical region of the premolars and separation of roots.*

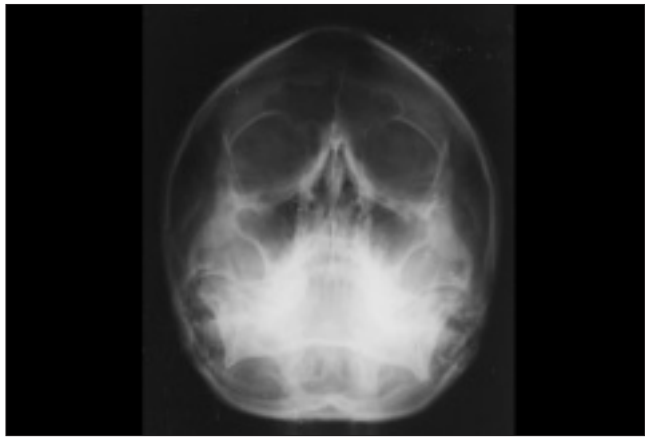

Figure 5: *Water's view showing haziness over the floor of the left maxillary sinus.*

Clinical examination revealed obliteration of the left nasolabial fold and none of the cervical lymph nodes was palpable. A firm, nontender, well-circumscribed, spherical swelling measuring 1 cm in diameter was observed between the canine and the premolar teeth, involving the labial sulcus (**Fig. 1**). The overlying mucosa was normal and no sinus tracts were present. None of the teeth in the region was carious, mobile or tender on percussion. Electrical and thermal vitality tests were positive for the canine and first premolar and negative for the second premolar.

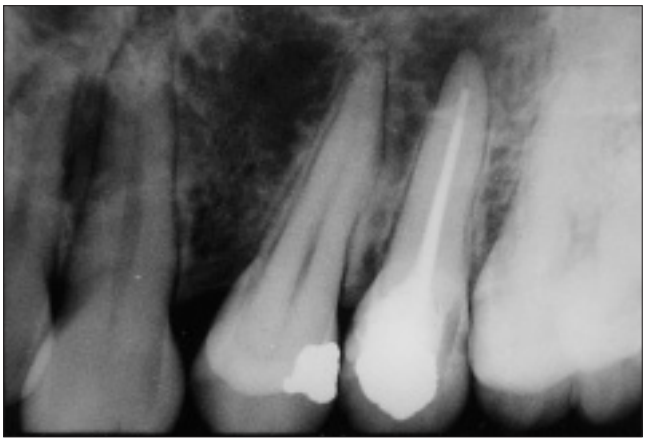

Figure 6: *Radiograph showing horizontal spread of the tumour to involve the mesial aspect of the canine.*

Figure 8: *Coronal computed tomography section showing involvement of the floor of the sinus.*

A radiograph of the region showed a triangular, honeycomb radiolucency with ill-defined margins, which was causing distal displacement of the root of the first premolar (**Fig. 2**). The floor of the maxillary sinus appeared to be involved by the lesion. The apical regions of the premolars were in close proximity. The second premolar showed a well-obturated root canal with diffuse apical radiolucency and loss of lamina dura. There was no evidence of root resorption. Evaluation of the radiograph taken before endodontic treatment of the second premolar also showed the presence of the lesion, which had been initially diagnosed as a periapical abscess by the endodontist.

A provisional diagnosis of a fibro-osseus lesion involving the left maxilla was made. Biopsy of the lesion revealed a nonencapsulated mass with columnar ameloblastic cells in a peripheral palisade pattern, a central stellate reticulum with extensive squamous metaplasia and foci of cystic degeneration in a dense cellular, fibrous stroma of collagen fibres (**Figs. 3a, 3b**). No dysplastic features were seen.

The biopsy was diagnostic for acanthotic desmoplastic ameloblastoma. The patient preferred to undergo surgery in her hometown, where the tumour was curettaged with wide

Figure 7: *Axial computed tomography section showing involvement of the anterior and medial walls of the left maxillary sinus.*

margins. Involvement of the maxillary sinus was not documented in the discharge summary.

Two months after the curettage, she reported back complaining of occasional dull pain of insidious onset in the same region. She had no history of sinusitis, nasal blockage, epistaxis or paresthesia of the face. Clinical examination showed no abnormalities. A radiograph of the region revealed a new diffuse radiolucency at the apical region of the premolars and separation of the root apices (**Fig. 4**). Water's view showed haziness over the left maxillary sinus (**Fig. 5**). A recurrence was suspected and surgery advised.

For personal reasons, the woman was unable to undergo surgery for 2 months. During this period, she was kept under observation. Periodic radiographic examination showed horizontal spread of the tumour to involve the canine and the first molar (**Fig. 6**).

Computed tomography with a contrast medium carried out before the surgery showed an osteolytic lesion arising from the alveolar ridge and involving the anterior, medial walls and the floor of the left maxillary sinus (**Figs. 7** and **8**). The woman underwent partial maxillectomy with immediate placement of a nonvascularized iliac bone graft. Histologic examination revealed normal bone in the margins of the excised mass.

Discussion

Desmoplastic ameloblastoma is most likely to occur in the anterior or premolar region of the jaws; there is no difference in prevalence between the maxilla and mandible. Cases have been reported in patients aged 18 to 70 years with a mean of 41.2 years.

No difference between sexes has been reported.[7] The incidence of desmoplastic ameloblastoma is low; rates of 0.9% to 12.1% of all ameloblastomas have been reported.[8–12] This type of tumour has been reported mainly in Chinese (in Malaysia and Hong Kong), Malaysians, Afro-Caribbeans and Japanese.[8]

No typical radiographic features are associated with this variant of ameloblastoma, although a mixed radiolucent–radiopaque appearance with ill-defined borders has been observed in many cases.[7,13] The current case lacked the typical features of ameloblastoma, which was, therefore, not considered in the differential diagnosis. The presence of pulpally involved teeth in the region and the diffuse radiolucency masked the presence of the tumour. Displacement of the roots was probably overlooked and considered normal by the endodontist. The age of the patient, location and radiographic features led us to the provisional diagnosis of a fibro-osseous lesion.

Histopathologically, desmoplastic ameloblastomas are nonencapsulated tumours with extensive collagenous stroma or desmoplasia containing small islands and nests of ameloblast cells. They have little tendency to mimic ameloblasts and the typical palisade pattern may be absent.[7] The follicles tend to be morphologically irregular or compressed.[14] Desmoplastic ameloblastoma must be histologically differentiated from ameloblastic fibroma, odontogenic fibroma and squamous odontogenic tumour.[8]

As the tumour is nonencapsulated, the cells infiltrate between the trabeculae of the cancellous bone leaving them intact for some time. Thus, the tumour actually extends beyond the radiographic margin,[15] which could be the reason for the ill-defined radiographic borders and the high recurrence rate after curettage. Marx and others[16] reported unpublished data from an analysis of 34 mandibular ameloblastomas, which showed that the tumour extended 2.3–8.0 mm beyond the radiographic margin. As a result, they have recommended resection of 1 cm of normal-appearing bone beyond the radiographic margin. It has been suggested that an altered intracellular distribution of collagen XVII and consequent loss of critical cellular attachments may contribute to the infiltrative and progressive growth characteristics of ameloblastoma.[17]

Only 5 cases of recurrent desmoplastic ameloblastoma have been previously reported.[8,9,11] Three of them were treated by enucleation or curettage. Desmoplastic ameloblastoma may have a propensity to recur with a frequency equal to that of other types of ameloblastoma.[9] Recurrence rate of conventional mandibular ameloblastomas treated by curettage ranges from 33.3% to 90%, whereas for those affecting the posterior maxilla it appears to be 100%.[18,19] Curettage is an inappropriate treatment for ameloblastomas of the posterior maxilla because recurrence is inevitable and difficult to treat. Such tumours should be excised with an extensive margin of apparently unaffected bone on the first attempt.[15]

Ameloblastoma of the posterior maxilla is dangerous because of its close proximity to the orbit, pterygomaxillary fossa and cranium and due to the difficulty in achieving an adequate surgical margin. Intracranial extension can be lethal. The entire maxilla possesses a thin cortical plate that offers little resistance to the tumour, thereby enhancing its rapid spread into the adjacent vital structures.[15,20] In our case, this was evident from the rapid horizontal spread of the tumour to involve the mesial aspect of the canine and first molar.

Some investigators have demonstrated oxytalan fibres in the tumour stroma and have suggested that the stroma originated in the periodontal membrane.[21] Immunohistochemical studies suggest that the desmoplasia originates from *de novo* synthesis of extracellular matrix proteins.[7] Because ameloblastomas are known to recur years after initial treatment, the need for long-term periodic follow-up (preferably lifelong) for early detection of recurrence should be emphasized.[22,23]

Conclusion

In the current case, the tumour of the alveolar region was recurrent, whereas that of the antrum was residual. The involvement of the antrum was missed by the previous surgeon who probably relied mainly on plain radiographs. Computed tomography is especially important in determining the borders of lesions, particularly of the maxilla. In this case, the nonencapsulation and trabecular infiltration of the tumour coupled with incomplete removal by curettage was responsible for the recurrence. Curettage for ameloblastoma of the posterior maxilla should never be considered as a treatment option. Recurrence is still possible after resection if the borders are not free of the tumour. Desmoplastic ameloblastoma should be included in the differential diagnosis of any mixed radiolucent–radiopaque lesion with diffuse borders involving the anterior–premolar region of the jaws. ◆

Acknowledgement: *We would like to thank Dr. Anuradha C. K. Rao, department of pathology, Kasturba Medical College, Manipal, for her professional help in preparing and interpreting the slide, and providing the photomicrographs.*

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The authors have no declared financial interests.

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