

Extreme Ameloblastoma

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ABSTRACT

Ameloblastoma is a benign neoplasm arising from epithelial components of odontogenic apparatus. It is known for its aggressive biologic behavior and tendency for recurrence. Uncommonly, it may present as a massive swelling of the jaws, which has been described as 'extreme' form of ameloblastoma. We report a case of extreme ameloblastoma involving the mandible in an Indian male that measured 10.9 × 8.5 cm. The tumor was neglected by the patient for a period of 13 years. He reported to the hospital after the tumor has grown up to grotesque size. Thus, ameloblastoma may show huge dimensions with functional impairments and complications, if neglected.

Keywords: Ameloblastoma, Complications, Computed tomography, Extreme, Giant.

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INTRODUCTION

Ameloblastoma is a well-recognized prototype of benign odontogenic tumors. Molecular alterations in ameloblastoma are such that it shows relentless growth with infiltration of marrow spaces.¹⁻⁴ It can exhibit voluminous size if neglected and left untreated. Complications, like starvation and hypoproteinemia, eventually lead to death in such cases.⁵ In today's time, due to easy availability of medical care and increased health awareness, it is uncommon to find such 'extreme' ameloblastoma. Only 10 such cases are reported in literature.⁶ We report a rare case of extreme ameloblastoma which was difficult to treat because of its monstrous size and systemic complications.

CASE REPORT

A 55-year-old male reported with complaint of difficulty in food intake due to swelling on the right side of the face

and mobility of teeth. The patient appeared malnourished and weak. He was a farmer and from poor socioeconomic background.

History revealed that patient noticed a swelling involving the lower right side of the face before 12 to 13 years. The swelling was painless and caused no discomfort to the patient at onset. According to the patient, initially the swelling was small which slowly increased in size. Since past 2 to 3 years, the patient noticed looseness of the lower teeth along with reduced mouth opening. Lesion was the matter of concern to patient because of its increasing size and associated trismus, which along with mobility of teeth, caused difficulty in eating. Patient also complained of dull pain associated with swelling. Patient was a bidi smoker since approximately 20 years. He was suffering from nonproductive cough since 4 years. After consultation with a physician, it was found that the patient was suffering from chronic bronchitis.

On extraoral examination, facial asymmetry was obvious due to a huge swelling of approximately 10 × 8 cm size involving right side of face (Fig. 1). The lesion was extending anteroposteriorly from the point 1 cm anterior from the corner of mouth to just behind the lobule of ear on right side. Superoinferiorly, it extended from the point below the corner of eye to 2 cm below the inferior border of mandible on same side (Fig. 2). Overlying skin was smooth and stretched. Trismus was apparent. On palpation, surface of the lesion was nodular. Consistency was firm. The swelling was nontender.

Intraorally, narrowing of the palatal arch was noticed, as the right maxillary posterior teeth were pushed



Fig. 1: Extraoral image showing a huge swelling involving right side of face

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toward midline (Fig. 3). A pus discharging sinus was also noticed in right posterior gingivobuccal sulcus. The lesion was extending on buccal side up to the mandibular gingivobuccal sulcus on same side (Fig. 4). Lower gingivobuccal sulcus was completely obliterated. Right mandibular molars were absent. Remaining mandibular teeth exhibited mobility. Interestingly, maxillary arch showed full complement of maxillary teeth. With the above findings, the provisional diagnosis of ameloblastoma was made.

Computed tomography showed a heterogeneously enhancing expansile lesion with extensive solid and cystic components measuring about 10.9 × 8.5 cm on the right side of the face and neck, completely invading the right half of mandible and condylar process, extending inferiorly along the right side of the neck. Medially, it compressed the right submandibular gland and the tongue. Laterally, the lesion was extending into the subcutaneous tissues. Superiorly, it extended till the level of maxillary

sinus, pushing the posterolateral wall of maxilla toward midline. Inferiorly, lesion extended till the infraglottic region (Figs 5 and 6).

Diagnostic biopsy was performed under local anesthesia by an intraoral incision. The patient was kept under observation after biopsy.

Microscopic examination of hematoxylin and eosin (H&E) stained sections from the specimen showed multiple islands and follicles of odontogenic epithelium in the background of fibrous connective tissue. The epithelial islands were made up of tall columnar cells resembling ameloblasts, arranged in peripheral layers. These cells revealed round to oval hyperchromatic nuclei, oriented away from the basement membrane with subnuclear vacuolization. The cells in the centers of other follicles resembled stellate reticulum. Many follicles showed central cystic degeneration (Figs 7 and 8). Supporting stroma was fibrocollagenous connective tissue with areas of hemorrhage. Thus, the microscopic features were



Fig. 2: Profile view displaying the extent of lesion in superoinferior and anteroposterior directions



Fig. 3: Intraoral photograph showing narrowing of the palatal arch due to medial displacement and palatal tipping of right maxillary premolars and molars



Fig. 4: Intraoral photograph showing intact buccal mucosa covering the lesion. Prominent vessels are noticeable on the surface

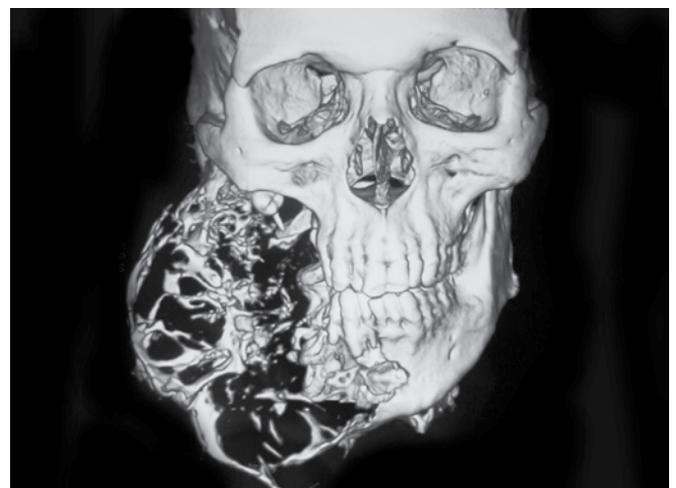


Fig. 5: Three-dimensional computed tomography showing large expansile lesion completely invading the right half of the body of the mandible

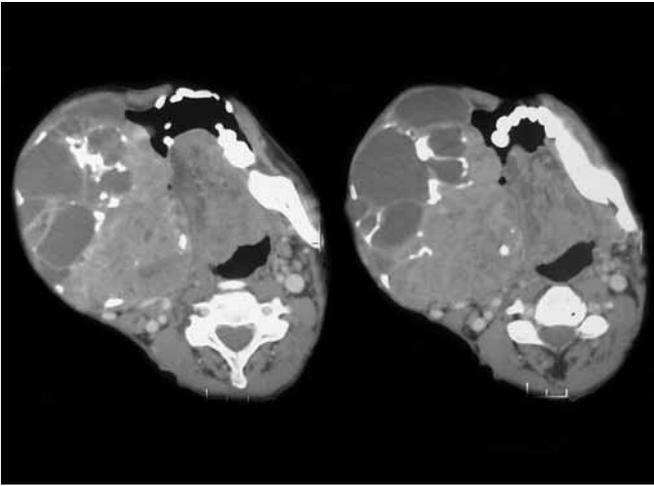


Fig. 6: Two-dimensional computed tomography showing the lesion with extensive cystic and solid areas

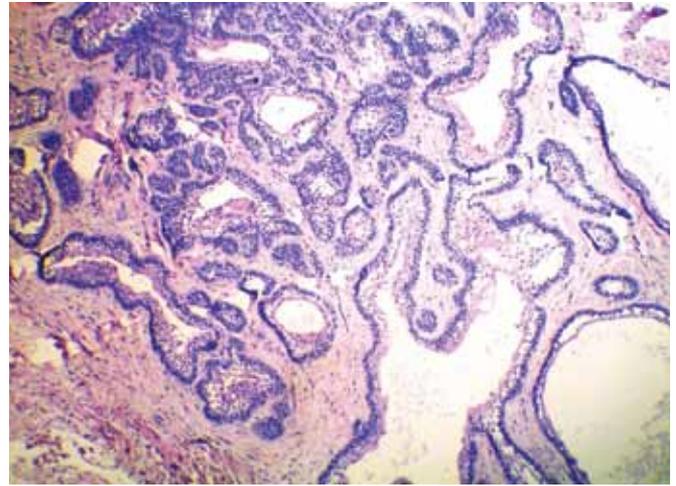


Fig. 7: Photomicrograph showing multiple follicles of varying sizes against the background of collagenous stroma (hematoxylin and eosin stain, magnification: 100x)

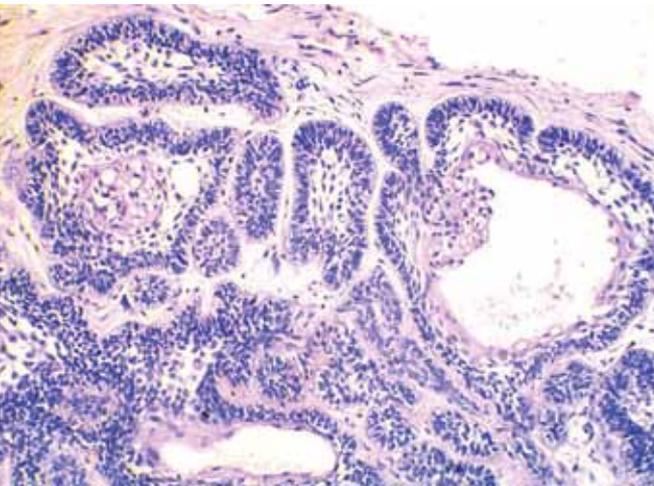


Fig. 8: Follicles of odontogenic epithelium with prominent peripheral cells and central stellate reticulum-like cells. Cystic degeneration and squamous metaplasia of central cells are noticeable (hematoxylin and eosin stain, magnification: 100x)

consistent with the diagnosis of follicular ameloblastoma. After diagnosis, again the patient underwent systemic examination. The patient was not a good candidate for major surgery due to his debilitated physical condition. Surgical removal of the lesion under general anesthesia was not advisable until significant improvement in his health. He was hospitalized once again. Patient was unable to take solid food. He was put on high protein liquid diet and nasogastric tube feeding. Unfortunately, during course in hospital, the patient died.

DISCUSSION

Ameloblastoma is a benign neoplasm originating from epithelial components of odontogenic apparatus. The entity is known for its propensity of growth up to large dimensions and proclivity for recurrence. ‘Giant’ or ‘extreme’ ameloblastomas are the lesions which show huge dimensions, causing gross facial disfigurement

and functional disturbances. Although benign, such extreme ameloblastomas can become life threatening due to associated problems. Lethal complications related to such voluminous size of tumor are starvation due to restriction from feeding, airway obstruction, and hypoproteinemia due to leakage of proteins into large cystic spaces of tumor.⁵

Studies have shown that ameloblastoma accounts for 40 to 60% of all received specimens of odontogenic tumors in various Institutes in India.⁷⁻⁹ In spite of availability and easy access to the medical care in developing countries, sometimes ameloblastomas of enormous sizes are confronted in practice. At least 10 cases of extreme ameloblastomas are reported till date. Complications resulting from regional dysfunctions are described with most of these reports.⁶ Regional dysfunctions, like trismus and difficulty in mastication and deglutition, were seen in the present case also. Similar to most of the reported cases, in our case, the patient had poor nourishment as a result of inability to take food. Thus, it becomes apparent that neglected ameloblastomas can enlarge up to extreme dimensions, causing gross facial deformities that pose considerable problems in management. The present case serves as an excellent example of ameloblastoma in terms of its relentless growth, and capacity to attain huge size which may become culprit for death.

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