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CASE REPORT

Ameloblastic fibrosarcoma: A rare malignant odontogenic tumor

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KEYWORDS
Odontogenic tumor;
Ameloblastic fibrosarcoma;
Malignant;
Differential diagnosis;
Surgical resection

Summary
Introduction: Ameloblastic fibrosarcoma (AFS) is a rare malignant odontogenic tumor. It can arise de novo, however one-third of cases may arise from a recurrent ameloblastic fibroma, in which case they appear to present at an older age.
Case report: A 16-year-old female presented with one month history of right mandibular mass. Computerized tomography (CT) scan showed a large destructive mass. A biopsy of the mass was performed. Histologically, it consisted of a mixed epithelial-mesenchymal odontogenic neoplasm composed of benign islands of well-differentiated ameloblastic epithelium within a malignant fibrous stroma consisting of spindle cells or fibroblasts with a brisk mitotic activity. The malignant spindle cell proliferation showed positive staining with p-53 and a high proliferation index with ki-67. A diagnosis of AFS was rendered.
Conclusion: The differential diagnosis includes other odontogenic sarcomas, ameloblastic carcinosarcoma and spindle cell carcinoma. Treatment of choice is wide surgical excision, with long-term follow-up. Postoperative chemotherapy and radiotherapy has been used successfully in a few reported cases. AFS is a locally aggressive malignant tumor, with regional and distant metastases being uncommon.

Introduction

Ameloblastic fibrosarcoma (AFS) is an extremely rare malignant odontogenic tumor that was first described in 1887 [1]. To the best of our knowledge only 71 cases of AFS have been reported in the literature with most cases occurring in the mandible within the third decade of life.

Case Report

We reported a rare case of AFS in a 16-year-old female, who presented with a month history of right mandibular mass, resulting in difficulty in swallowing. The patient was worked up and a maxillofacial CT-scan was performed. CT scan showed a destructive 5-cm mass on the right molars. Additionally, it showed cortical expansion and perforation with invasion into pterygopalatine space causing destruction of facial bones (Fig. 1). An incisional biopsy was done. Microscopically, there was a biphasic pattern composed of bland appearing epithelium that resembled ameloblastic fibroma, but quantitatively less, along with a malignant mesenchymal component (Fig. 2A). The mesenchymal cells showed

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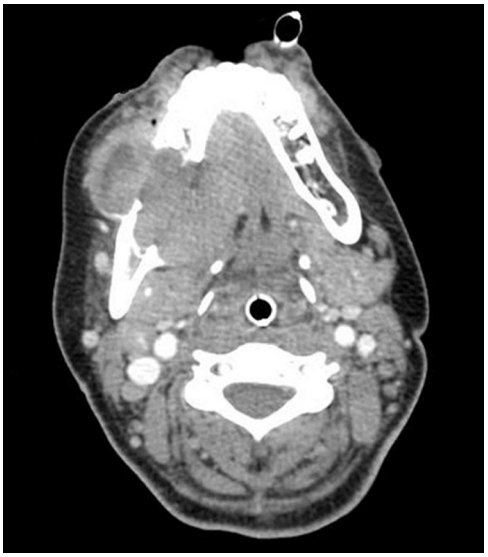

Figure 1 Computed Tomography (CT) scan showing a mass lesion straddles the right mandibular ramus and extends laterally and medially from the right mandibular ramus causing bony destruction.

marked cellularity, nuclear atypia and occasional mitoses (Fig. 2B). The benign epithelial component showed uniform positivity for pan-cytokeratin (Fig. 3A), while the malignant mesenchymal component was positive for p53 (Fig. 3B) with a high proliferation index for Ki67 (Fig. 3C), while negative for c-KIT (CD-117) (Fig. 3D). Based on the morphology and immunohistochemical staining pattern, a diagnosis of AFS was rendered.

Discussion

AFS is a rare neoplasm, in which the clinical and pathological distinction from other neoplasms is essential for appropriate care. The terms ameloblastic dentinosarcoma and ameloblastic odontosarcoma have been used in the past for these types of neoplasms depending on the presence of dentin or enamel as some authors consider these lesions as histological variants of the same neoplasm. However, in the recent World Health Organization (WHO) ''classification'' of odontogenic tumors, ameloblastic odontosarcoma and dentinosarcoma are listed separately from AFS. AFS occurs within a wide age range from 3 to 89 years [2].

The mean age at time of presentation for all reported cases is 27.3 years [3]. Of the 62 cases analyzed by Huguet et al., 20 arose in previously benign AFS [4].

The usual clinical presentation consists of a patient who complains of a painful but occasionally painless, facial mass with accompanying paresthesia or dysesthesia. The duration of symptoms varies widely from a few weeks up to 2 years.

Radiologically, AFS presents as destructive expansile radiolucent mass with irregular and ill-defined borders. Grossly the tumor may be cystic or solid with a fleshy whitish to yellow consistency that usually causes destruction the bone. The epithelial component is present in the form of nests and branching cords with anastomosing strands of odontogenic epithelium exhibiting peripheral palisading that resembles the developing enamel organ. The mesenchymal cells vary from hyperchromatic spindle to stellate that exhibit moderate to marked nuclear pleomorphism with a high number of mitotic figures. Dentin matrix material may be present within the intercellular areas. Ultrastructually, these tumors exhibit features of fibroblasts.

The sarcomatous mesenchymal component of AFS is positive for p53 and proliferating cell nuclear antigen (PCNA) as compared to negativity for these stains in AF [4]. The mesenchymal component of recurrent AF and AFS usually show higher labeling indices for Ki-67 as compared with non-recurrent AF [5]. Williams et al. identified diffuse nuclear positivity for p53 in the sarcomatous component along with positivity for c-KIT (CD 117) [6]. However, expression of CD-117 is variable in AFS and can show a negative staining pattern. Pontes and coworkers demonstrated a positive

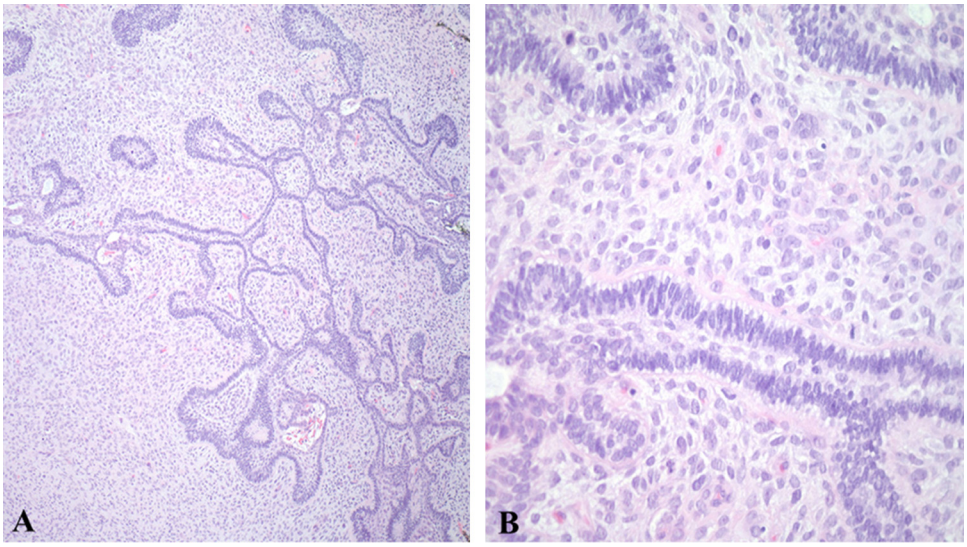

Figure 2 A. Biphasic pattern with benign odontogenic epithelium surrounded by hypercellular mesenchymal component (H & E × 100). B. Malignant proliferation of mesenchymal component with mitosis (H & E × 400).