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CASE REPORT

Odontogenic sarcoma with smooth muscle differentiation: Report of a case and review of the literature

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Summary A case of odontogenic sarcoma with smooth muscle differentiation arising in a 70-year-old woman is described. The lesion grew out of an extraction socket and enlarged rapidly. Radiographically, a large radiolucent lesion with ill-defined margins was observed in the left posterior mandible. Histopathologically, islands of odontogenic epithelium with a surrounding malignant mesenchymal proliferation were noted. The latter exhibited hyperchromatic oval and spindle-shaped cells with 3–4 mitoses in some high-power fields and foci of necrosis. Lesional cells demonstrated immunoreactivity with vimentin and alpha smooth muscle actin, but were negative for antibodies directed against S100, CD34 and CD31. Cytokeratins highlighted the epithelial islands, but did not react with the mesenchymal proliferation. To date, fewer than 70 cases of odontogenic sarcoma have been described in the literature. To our knowledge, this is the first report of an odontogenic malignancy showing smooth muscle differentiation.

Introduction

Malignant mixed odontogenic tumours are rare gnathic entities with malignant mesenchymal and benign epithelial

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components, or with malignant epithelial and mesenchymal components. An odontogenic sarcoma is a mixed odontogenic tumour with benign epithelial and malignant mesenchymal components.[1] The epithelial component resembles the odontogenic epithelium of normal tooth development or that observed in pathologic lesions such as ameloblastoma or ameloblastic fibroma.[1] The mesenchymal component usually exhibits features of a fibrosarcoma. If the lesion contains dentin or enamel and dentin, the most appropriate diagnoses would be ameloblastic dentinosarcoma or ameloblastic odontosarcoma, respectively.[1,2] The differences exist in nomenclature rather than in prognosis.[1] The term odontogenic carcinosarcoma is reserved for lesions with both malignant mesenchymal and odontogenic epithelial components.

Fewer than 70 cases of odontogenic sarcoma have been described in the literature, all of which were diagnosed as odontogenic fibrosarcomas. To our knowledge this is the first case report of an odontogenic sarcoma with smooth muscle differentiation.

Case report

A 70-year-old woman underwent an extraction of the mandibular left first molar. Two weeks later, she presented to the oral surgeon with complaints of pain at the extraction site. Upon examination, a large, lobulated, pink, fungating mass with foci of ulceration was observed growing out of the extraction site (Fig. 1). A panoramic radiograph showed a large radiolucent lesion with poorly defined margins in the posterior mandible (Fig. 2). An incisional biopsy was performed and submitted for histopathologic evaluation. Microscopic examination revealed a malignant neoplasm consisting of oval and spindle-shaped cells with hyperchromatic nuclei. Interspersed among the neoplastic cells were islands of epithelium, some of which exhibited a basaloid appearance, consistent with odontogenic epithelium. Large areas of necrosis and numerous mitotic figures, some of which were atypical, were observed. Some high power fields contained 3—4 mitoses (Fig. 3). Immunohistochemical studies using formalin-fixed, paraffin-embedded tissue showed positive immunoreactivity of the spindle and oval-shaped cells for antibodies directed against vimentin and alpha smooth muscle actin, but these cells were negative for antibodies directed against S100, CD34, CD31 and cytokeratin. In contrast, the epithelial islands were highlighted by cytokeratin antibodies. Approximately, 20—30% of the neoplastic cells were Ki-67 positive, a measure of proliferative activity. Chest and abdomen scans were negative for any lesions. The patient underwent a left hemi-mandibulectomy with removal of the left submandibular gland. The patient elected not to have reconstructive surgery. At an eight-month follow-up visit, there was no evidence of recurrent disease.

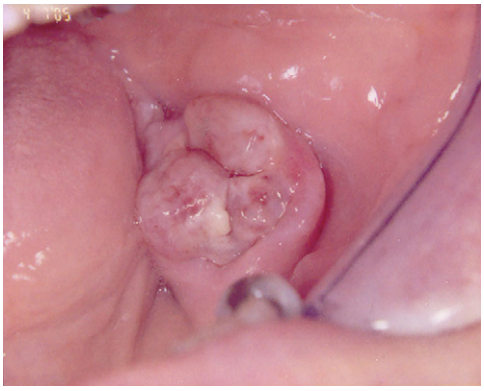

Figure 1 Clinical appearance prior to incisional biopsy, showing a fungating mass on the posterior mandibular ridge.

Discussion

An ameloblastic fibrosarcoma is a rare mixed odontogenic tumour with a malignant mesenchymal component.[1] In this report, the term odontogenic sarcoma with smooth muscle differentiation is used in preference to odontogenic leiomyosarcoma because the latter implies smooth muscle origin rather than smooth muscle differentiation of the ectomesenchymal tissue surrounding the odontogenic epithelium. Furthermore, the epithelial component resembled odontogenic epithelial islands that were not ''ameloblastic'' in appearance.

The first extensive review of the odontogenic sarcoma was by Muller et al.[3] in 1993. Subsequently, the lesion was reviewed by Bregni et al.[4] who identified 60 cases in the literature and described two new cases. Four other reports describing one case each and the present case brings the total to 67 cases.[2,5—7] The odontogenic sarcoma exhibits a male predilection with 39 cases occurring in males (58.2%) and 26 in females (38.8%). The gender was not stated for two cases (3.0%). Of these, 37 cases were in the mandible and 11 were in the maxilla.

Odontogenic sarcomas may arise from an ameloblastic fibroma or de novo.[6] The short duration and age of the patient in this case suggest that the lesion arose de novo. Typically, lesions present in the 2nd and 3rd decades, consistent with the younger age presentation of the ameloblastic fibroma.[1,4] Previous studies[4] reported an average age of 27.3 years with a range of 3—83 years. The five subsequently reported cases, including the one described here, raise the mean age to 28.9 years. Thirty-one of 58 (53.4%) well documented cases appeared to arise from a pre-existing ameloblastic fibroma.[2—11]

Based on less than 70 reported cases, ameloblastic fibrosarcomas are considered low-grade lesions because they rarely metastasize but are locally aggressive lesions.[2,4,6,12] However, about 20% of patients die of locally aggressive disease in 3 months to 19 years.[12] Due to the high recurrence potential, wide-surgical excision with close clinical-follow-up are recommended.[4]

The current case is the first report, to our knowledge, of an odontogenic sarcoma with documented smooth muscle differentiation. A recently published case study reported positive immunohistochemical reaction with CD34, but not with smooth muscle actin antibodies.[2] In contrast, the present case was CD34-negative but SMA-positive. Due to the limited descriptions of immunohistochemical studies in earlier reports, it is possible that some of these previously documented lesions represent odontogenic sarcomas with

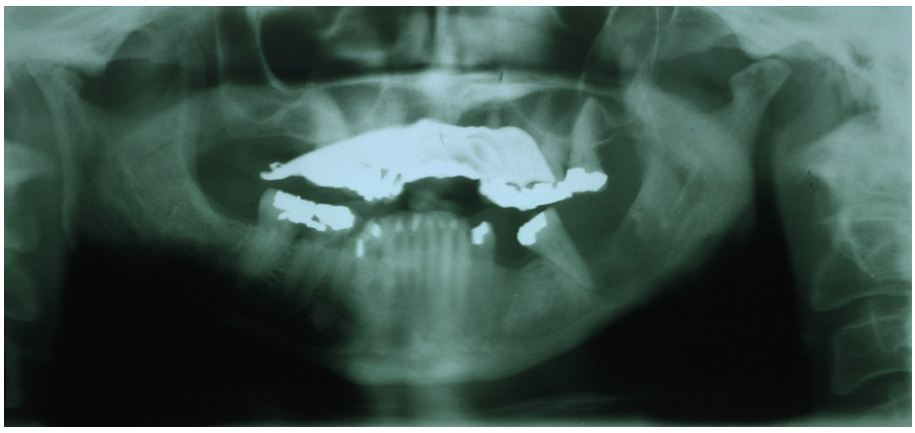

Figure 2 A panoramic radiograph reveals a poorly circumscribed radiolucent lesion in the left posterior mandible.

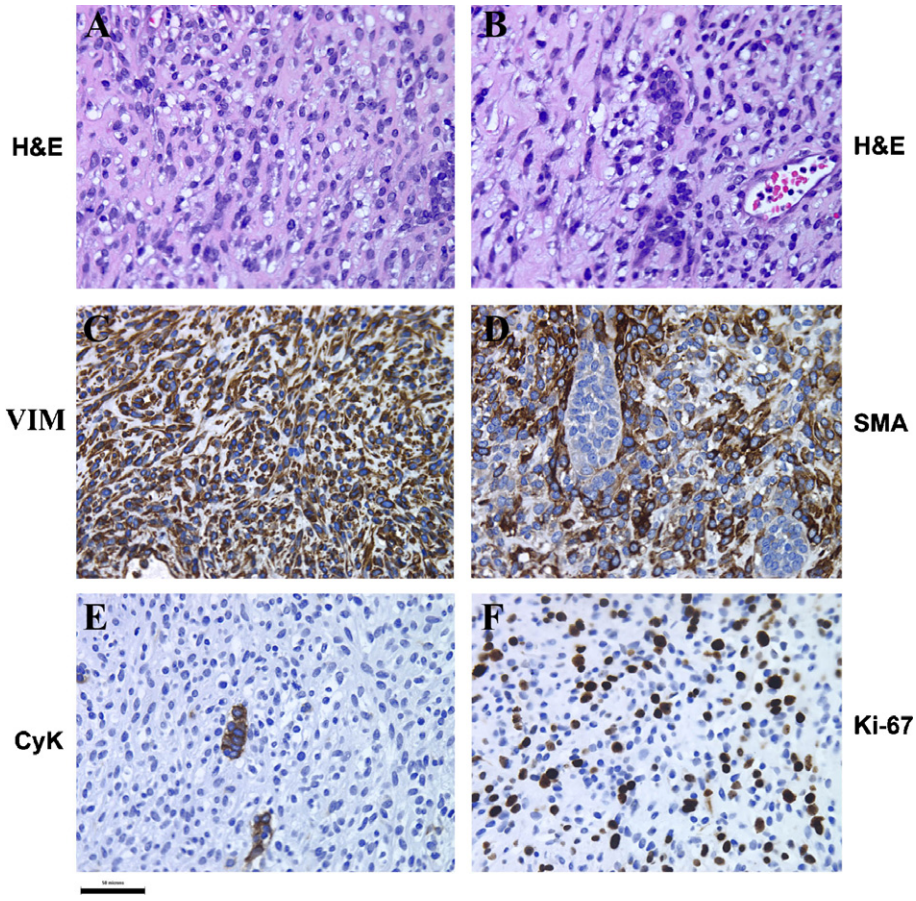

Figure 3 (A, B) Histopathologic appearance of the lesion showing hyperchromatic nuclei, numerous mitoses, and odontogenic epithelial islands (hematoxylin and eosin stain, H & E). (C—F) Immunohistochemistry for vimentin, smooth muscle actin, pancytokeratin and Ki-67. The spindle cell proliferation was immunoreactive with vimentin (VIM) and smooth muscle actin (SMA) whereas the epithelial component was immunoreactive with pancytokeratin (cyk). Approximately 20—30% of the lesional cells stained positively with Ki-67 (bar = 50 μm).

varying types of differentiation rather than exclusively fibrosarcomas.

In summary, odontogenic sarcomas are rare lesions that exhibit a male predilection and are seen more frequently in the mandible than in the maxilla. These lesions usually develop in pre-existing ameloblastic fibromas, but de novo lesions have also been described. The present case appears to represent the first report of an odontogenic sarcoma with smooth muscle differentiation.

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