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CASE REPORT

CD34 expressing ameloblastic fibrosarcoma arising in the maxilla: a new finding

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Ameloblastic fibrosarcoma (AFS) is a rare malignant tumor of the jaw. The malignant mesenchymal component of AFS has been described as 'fibroblast-like', although little is known about the immunophenotype, except for vimentin expression. Here, we present a case of AFS in a 62-year-old woman. The mesenchymal component displayed the features of either dermatofibrosarcoma protuberans or fibrosarcoma, and was positive for CD34. This is the first reported case of CD34 expressing AFS in the maxilla.
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A 62-year-old woman was referred to the Asan Medical Center for a further evaluation of recurrent multiple bone and joint pain following a surgery for a protruded intervertebral disc at L4-5, which had been performed 18 months earlier. During the course of evaluation, a jaw mass was identified and located in the right anterior region of the maxilla, at the root area of lateral and central incisors which had been extracted years ago (Fig. 1). The mass was firm and non-tender, and the patient claimed to have first noticed it 10 years earlier. The rest of the head and neck examination was normal. Chest X-ray and laboratory tests were unremarkable with no evidence of metastatic disease.

A biopsy was performed on the mass, but proved non-diagnostic because of inappropriate targeting. The specimen was submitted for frozen section, which disclosed cellular spindle cells with inconspicuous epithelial component. The frozen section diagnosis was a spindle cell tumor of at least intermediate malignancy, and a complete excision was recommended. The resected tumor was an ill-demarcated, firm, ovoid mass, measuring 3 cm in greatest dimension. The cut surface was solid, pinkish tan and focally fish-fleshy. Microscopically, the tumor consisted of spindle-shaped, fibroblast-like cells arranged in storiform and fascicular patterns resembling dermatofibrosarcoma protuberans or fibrosarcoma (Fig. 2a). In between the spindle cell bundles, scattered islands and strands of an epithelial component were also noted. These islands were composed of a few layers of uniform cuboidal or columnar cells that were histologically consistent with ameloblastic origin, exhibiting peripheral palisading and evidence of reverse polarization (Fig. 2b). Focal areas of ossification and osteoid like material were present at the periphery, but there was no obvious dentin or enamel. The spindle cells were cellular and hyperchromatic with mild pleomorphism, and showed three to five mitoses per 10 high power fields without atypical forms (Fig. 2c).

Immunohistochemical staining was carried out using formalin-fixed and paraffin-embedded tissue. The malignant spindle cells were positive for CD34 and vimentin, but negative for smooth muscle actin, S100 protein, MyoD1, CD68 and CD117 (Fig. 3a). In contrast, the benign looking ameloblastic epithelium showed positive immunoreactivity for pancytokeratin (AE1/AE3) (Fig. 3b). Histologic and immunohistologic examination led to the diagnosis of ameloblastic fibrosarcoma.

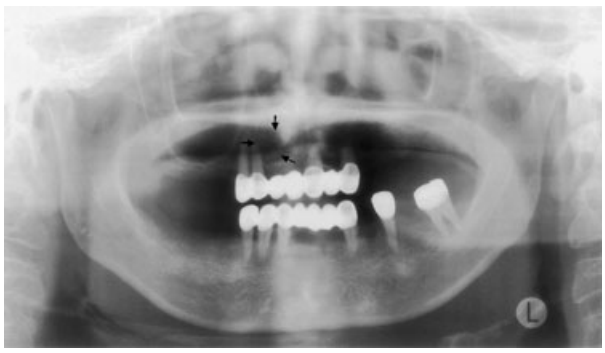

Figure 1 A panoramic X-ray view reveals a relatively well demarcated round osteolytic lesion at the right anterior maxillary region.

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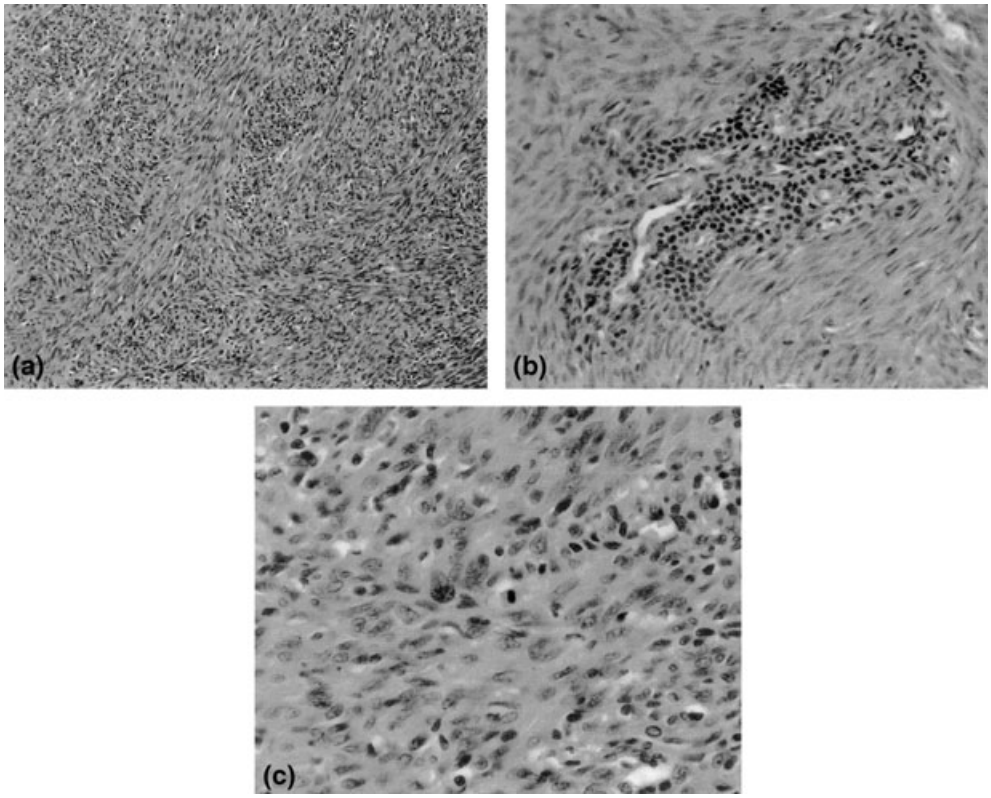

Figure 2 (a) The spindle-shaped, fibroblast-like cells are arranged in storiform pattern resembling dermatofibrosarcoma protuberans (HE 40×). (b) Scattered islands and strands of an epithelial component are present in between the spindle cell bundles (HE 200×). (c) The spindle cells are cellular and hyperchromatic with mild pleomorphism, and show a mitosis in the center (HE 400×).

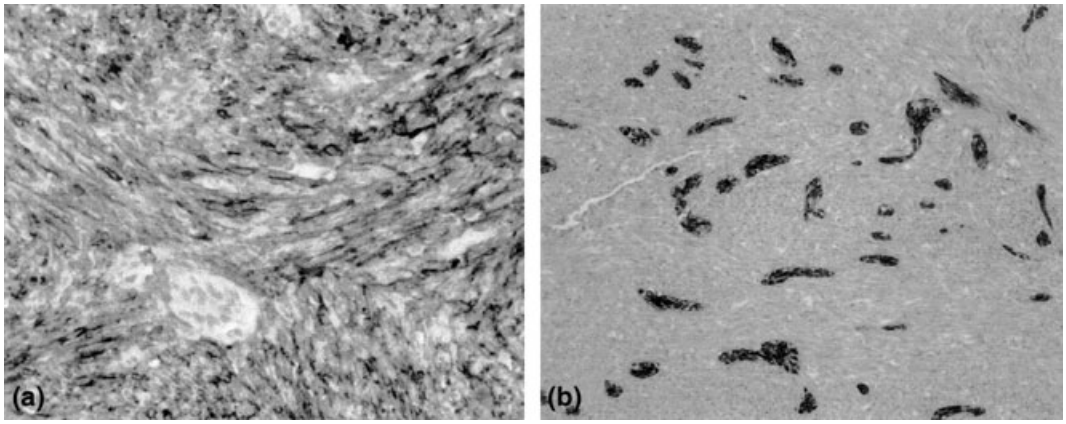

Figure 3 Immunohistochemistry for CD34 (a) and pancytokeratin (b). The malignant spindle cells are positive for CD34 (200×). The benign looking ameloblastic epithelium shows a positive immunoreactivity for pancytokeratin (AE1/AE3) (100×).

Comments

Ameloblastic fibrosarcoma (AFS) is a rare malignant tumor of the jaw and histologically characterized by a biphasic pattern of benign epithelial elements scattered throughout the malignant mesenchymal tissue. Depending on the elaboration of dentin or dentin and enamel, AFS has also been designated as ameloblastic dentinosarcoma or ameloblastic odontosarcoma, respectively. Although clinical presentations vary according to the cases, swelling and pain are the most common features. The information available about the treatment and prognosis is limited because of the rarity of the cases, although surgical wide excision and close follow-up seem to be the first choice of the treatment. AFS occurs either *de novo* or from a pre-existing or recurrent ameloblastic fibroma (1), and this low-grade sarcoma has a high rate of local recurrence; according to Bregni et al. (2), 23 (37%) of the 62 reported cases have had at least one recurrence. The male-to-female ratio was 1.6:1

in total 62 cases with 49 tumors located in the mandible (79%) and 13 in the maxilla (21%). The most commonly affected site within the jaw is the posterior part of the mandible. AFS can present at any age; the patient age at time of clinical presentation in previously reported cases was 3–83 years (mean 27.3 years) (2). The case described herein is unusual in that the tumor was located in the right anterior region of the maxilla and occurred significantly later than the mean age. There was no evidence of a pre-existing ameloblastic fibroma, but this could not be ruled out, given the age of the mass.

In the differential diagnosis of AFS, dermatofibrosarcoma protuberans, fibrosarcoma and other spindle cell sarcomas should be ruled out, because the mesenchymal cells of AFS in case of an inconspicuous epithelial component could be arranged in bundles simulating herring-bone, cartwheel, or storiform pattern. In particular, dermatofibrosarcoma protuberans is a difficult entity to distinguish from this case, because the spindle cells in the present AFS expressed CD34 and were arranged in a storiform pattern with a not very conspicuous epithelial component. Cytokeratin immunostaining is helpful in identifying the epithelial nests and thus excluding the pure sarcomas.

Until recently, immunohistochemical stain was of limited value in the diagnosis of AFS because of the rarity of cases and the relative lack of immunohistochemical data in the previous reports (3, 4). Here, we showed that an AFS was positive for CD34. This 110-kDa transmembrane glycoprotein is expressed on hematopoietic stem cells, endothelial cells, the interstitial cells of Cajal, and a group of dendritic cells present in the dermis, around blood vessels, and in the nerve sheath (5). CD34 positive fibroblasts in many organs are thought to represent uncommitted cells capable of multidirectional differentiation. A previous report showed an inverse relation between CD34 expression and myofibroblastic differentiation (6). CD34 expression is well documented in vascular tumors, dermatofibrosarcoma protuberans, solitary fibrous tumors, malignant peripheral nerve sheath tumors, gastrointestinal stromal tumors and epithelioid sarcomas. However, to our knowledge, CD34 immunostaining has not previously been performed in the case of AFS. Here, we found that the mesenchymal components of AFS showed diffuse and moderate expression of CD34. Although the function of CD34 in AFS remains unsolved, these data could provide a possible additional clue for the diagnosis and better understanding of these neoplasms.

References

1. Muller S, Parker DC, Kapadia SB, Budnick SD, Barnes EL. Ameloblastic fibrosarcoma of the jaws. A clinicopathologic and DNA analysis of five cases and review of the literature with discussion of its relationship to ameloblastic fibroma. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 1995; **79**: 469–77.
2. Bregni RC, Taylor AM, Garcia AM. Ameloblastic fibrosarcoma of the mandible: report of two cases and review of the literature. *J Oral Pathol Med* 2001; **30**: 316–20.
3. Tajima Y, Utsumi N, Suzuki S, Fujita K, Takahashi H. Ameloblastic fibrosarcoma arising de novo in the maxilla. *Pathol Int* 1997; **47**: 564–8.
4. Yamamoto H, Caselitz J, Kozawa Y. Ameloblastic fibrosarcoma of the right mandible: immunohistochemical and electron microscopical investigations on one case, and a review of the literature. *J Oral Pathol* 1987; **16**: 450–5.
5. Van de Rijn M, Rouse RV. CD34. A review. *Appl Immunohistochem* 1994; **2**: 71–80.
6. Chauhan H, Abraham A, Phillips JR, Pringle JH, Walker RA, Jones JL. There is more than one kind of myofibroblast: analysis of CD34 expression in benign, in situ, and invasive breast lesions. *J Clin Pathol* 2003; **56**: 271–6.