A rare case of oral epithelioid sarcoma of the gingiva

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Epithelioid sarcoma (ES) is a rare malignant soft tissue tumor. ES can be classified into proximal, distal, and fibroma-like subtypes. These tumors show both mesenchymal and epithelial immunophenotypes. Microscopically, the proximal type ES is usually characterized by nodules of spindle and epithelioid cells growing in granuloma-like pattern often presenting with central necrosis. Immunohistochemically these tumors are vimentin, pancytokeratin, and usually EMA (80%) positive. CD34 (50%) and CD99 (25%) may be positive, and occasionally SMA and S-100 immunopositivity has been reported. No specific genetic alterations have been found in ES. As far as we know, this is the first case in the literature to present ES in gingival mucosa. (Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2011;111:e25-e28)

In February 2010 a 33-year-old man with no medical history presented to the Department of Oral and Maxillofacial Surgery with a gingival mass and some tenderness and loosening of the teeth in the area (Fig. 1).

In the clinical examination, teeth were somewhat loose but vital, and sensation in the lip area was normal. Panoramic radiography and a computerized tomographic (CT) examination were performed, and then an open biopsy. Radiologic examination revealed a bone-destructive tumor in the right premolar area of the mandible with no radiologically pathologic lymph nodes (Fig. 2). Initial biopsy diagnosis was malignant mesenchymal neoplasm.

The tumor was excised with 20-mm margins, and the patient underwent an elective neck dissection. When the tumor was measured grossly, its width was 20 mm with an invasion depth of 18.6 mm. A small ulceration was visible on the mucosa. The epithelium around the ulcer did not show dysplastic features, and the tumor did not come into contact with the epithelium in any place. The growth pattern was mainly diffuse with only a few fascicular and sheet-like formations. The tumor cells showed some nuclear pleomorphism with prominent nucleoli (Fig. 3). Mitotic activity was high: In “hot spot” areas up to 13 mitoses/10 high-power fields were observed. The neck dissection specimen contained 20 lymph nodes. In routine hematoxylin-eosin staining, no metastatic growth was detectable; however, pancytokeratin (pan-CK) staining showed scattered metastatic cells in 2 lymph nodes.

Immunohistochemistry

The tumor was strongly positive for pan-CK, vimentin, CD99, and desmin. Some positivity was also evident for EMA, p53, CK19, Myf-4, myoglobin, and INI, but CD31, CD34, CD68, CK8, MART, S-100, and CD45 were all negative (Figs. 4 and 5).

DISCUSSION AND DIFFERENTIAL DIAGNOSIS

Epithelioid sarcoma (ES) is a rare soft tissue tumor that presents usually in adolescent and young adults as a deep dermal or subcutaneous mass in the extremities. As far as we know, this is the first reported case of gingival ES. Considering the rarity of ES and the unusual location of the tumor we very thoroughly considered the diagnosis.

Because the tumor was poorly differentiated, the diagnosis was verified immunohistochemically to rule out the possibility of other malignancies.

Primarily, we out ruled melanoma with negative S-100 and MART-1 immunostainings and lymphoma with negative CD45 staining.

Squamous cell carcinoma is the most common oral pan-CK–positive malignancy. However, in a typical squamous cell carcinoma, vimentin and desmin immunopositivity should not occur. A positive reaction to these antigens is typical for oral spindle cell carcinoma,
another rare tumor, but that tumor type should comprise an epithelial in situ or invasive carcinoma with a sarcomatoid component. In our case, the epithelium was benign with no dysplastic features and with no contact to the malignant cells. In the sarcomatoid portion of spindle cell carcinoma, various keratin immunostain-
Fig. 4. Positive markers in immunohistochemistry.

Fig. 5. Negative markers in immunohistochemistry.
ings are often only partly positive.\textsuperscript{3,4} This is in contrast to the present case, which showed a consistent pan-CK positivity. Among possible sarcomatoid tumors in this location, a potential diagnosis is synovial sarcoma, which is usually CK, CD99,\textsuperscript{5} and EMA positive. However, desmin positivity is not among the typical characteristics of synovial sarcoma.\textsuperscript{5} CD99 is one of the markers for Ewing sarcoma, but keratins are typically absent from this tumor group. The strong CK positivity also excluded rhabdomyosarcoma and angiomatoid histiocytoma. Some positivity for Myf-4 and myoglobin was also detectable but not to the same extent as in typical rhabdomyosarcoma. Other epithelioid-type tumors might also be mistaken for ES. Epithelioid angiosarcoma is typically positive for CD31 and CD34.\textsuperscript{6} Epithelioid malignant peripheral nerve sheath tumor may have similar morphologic features but is usually S-100 immunopositive.\textsuperscript{7}

ES is expected always to be CK and vimentin and often EMA positive. This is in accordance with the present case. In addition, Hasegawa et al. studied 20 ES, and these showed focal membranous CD99 positivity in 25%; desmin positivity was also evident for 15%.\textsuperscript{8}

The main prognostic factors for ES are tumor size and the presence of metastases.\textsuperscript{8} The critical tumor diameter is considered to be 7.8 cm, but the present tumor, though significantly smaller, had already metastasized. Studies have in fact shown that tumor size is not associated with metastatic potential. Early metastasis is a predictor of poor survival.\textsuperscript{8} Because the operation was so recent, we lack follow-up data on the patient.

As far as we know, this is the first report of ES localized intraorally in gingival tissue.

REFERENCES


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