CASE REPORT

Alveolar soft part sarcoma of the tongue

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Summary The clinical, histopathological, immunohistochemical and histochemical features of an alveolar soft part sarcoma involving dorsum surface of the tongue of a 17-year-old girl are described. This is a rare tumour of uncertain histogenesis. There has been no evidence of recurrence or metastasis after one year of follow-up.
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Introduction

Alveolar soft part sarcoma (ASPS) is a rare malignant neoplasm of the head and neck region of uncertain histogenesis. ASPS commonly originates in the head and the neck region with 25% of the cases occurring in the tongue.1,2 ASPS presents as a slow-growing, painless mass that occurs in female patients between 15 and 35 years old.2,3 Metastasis occurs in the lungs, brain or bones and the prognosis is unfavourable.2 The aim of this paper is to report an additional case of ASPS on the tongue in a 17-year-old girl.

Case report

A 17-year-old girl was referred to the Oral Diagnosis Section, Odilon Berhens Hospital with a swelling on the tongue of three months duration (Fig. 1(A)). Extraoral examination revealed neither facial alterations nor cervical node enlargement. Intraoral examination showed a nodule at the dorsum surface of the tongue, asymptomatic, elastic consistency, measuring 2 cm in diameter and covered by a mucosa of normal colour and texture. The fine needle aspiration was negative for liquid. Clinical diagnosis was benign mesenchymal neoplasm (neurofibroma, schwannoma and lipoma). An excisional biopsy was performed under local anesthesia. The surgical specimen was fixed in 10% neutral phosphate-buffered formalin.

Histological findings showed nests of large granular cells separated by fibrovascular stroma (Fig. 1(B) and (C)). The
cells were large rounded or polygonal with abundant granular eosinophilic cytoplasm and vesicular nuclei with small nucleoli. Mitotic figures and nuclear pleomorphism were not present. Periodic acid-Schiff (PAS) with and without diastase digestion demonstrated deposits of intracellular glycogen and rhomboid shaped crystals (Fig. 1(D)). Cross-striations were not identified with the phosphotungstic acid-hematoxylin or Masson tricrome staining, respectively.

Immunohistochemical performed on formalin-fixed, paraffin-embedded tumour tissue revealed few tumour cells positive for the antibodies against muscle specific actin (Clone HHF-35, diluted 1:100, DAKO Corporation), cytoplasmic (but not nuclear) reactivity for MyoD1 (5-BA, diluted 1:100, DAKO Corporation) and myogenin (Clone F5D, diluted 1:100, DAKO Corporation). Strong staining was observed for desmin (Clone A0611, diluted 1:50, DAKO Corporation), neuron-specific enolase protein (NSE) (Clone A0587, diluted 1:100, Dako Corporation) (Fig. 1(E)) and smooth muscle actin (Clone 1A4, diluted 1:50, DAKO Corporation) (Fig. 1(F)). No immunoreactivity was seen for vimentin (Clone Vim 3B4, diluted 1:100, DAKO Corporation), S-100 (Z0311, diluted 1:75, DAKO Corporation), cytokeratin AE1/AE (Clone AE1 & AE3, diluted 1:50, DAKO Corporation). Antigen retrieval with citrate buffer (0.01 M, pH 6.0, 95°C, 30 min) was performed to all the antibodies, except to S-100. Diagnosis of the ASPS, solid pattern, was made. The patient was referred to the Oncology Service for further evaluation and no metastatic lesion was detected.

**Discussion**

ASPS is a rare malignant tumour corresponding to less than 1% of all soft tissues sarcomas. The lower extremities are the most common location, but it also occurs in the head and neck region (27%), with 25% of the cases affecting the
tongue. The age for lingual ASPS is much younger than that for ASPS in other anatomical locations. Lingual ASPS are reported in patients less than 21 years of age with a female predilection of 2:1.

Microscopic features, histochemical and immunohistochemical are important to the definitive diagnosis of ASPS. The cells reveal varying amounts of intracellular glycogen and characteristically PAS-positive, diastase-resistant rhomboid or rod-shaped crystals. In our case, the cells were PAS-positive and showed diastase-resistant crystals in the cytoplasm. This aspect has been observed in 80% of cases in others studies, and is helpful to differentiate this tumour from rhabdomyoma. Rhabdomyoma present cells PAS-positive, but the intracitoplasmatic glycogen granules disappeared after pretreatment with diastase. Moreover, rhabdomyoma shows cross-satiations in phosphotungstic acid-hematoxylin and Masson tricrome staining.

Since its first description, ASPS has been object of debate and controversy. Despite earlier attempts to prove a neuroectodermal, endocrine or myogenic lineage of differentiation in this neoplasm, its histogenesis has not been established. Although the immunohistochemical profile indicates a myogenic origin, it does not present consistent findings. The cells of the tumour do not stain with antibodies against cytokeratin, epithelial membrane antigen, neurofilaments, glial fibrillary acidic protein, serotonin, synaptophysin, met-enkephalin and leu-enkephalin, but occasionally express S-100 protein and NSE. In our case the neoplastic cells were positive to NSE and negative to S-100. Muscle markers differ somewhat, but most investigators were able to demonstrate immunoreactivity for vimentin, muscle-specific actin and desmin. Negative staining for vimentin has also been reported. Among the muscle markers that we investigated, desmin was strongly positive. Among specific markers of myogenous differentiation, negative and positive labeling is described for myoglobin and myogenin, respectively. While MyoD1 shows nuclear staining in muscle cells, positive cytoplasmic labeling is observed on the tumoural cells in ASPS and is considered negative.

Histologic parameters such as atypia, cellularity, necrosis, and number of mitosis per high power field seem to represent the most reliable criteria of malignancy of soft tissue sarcoma. However, these parameters are imprecise and subjective for predicting clinical behaviour of ASPS and may cause diagnostic confusion with other diseases.

ASPS is characterized by relatively slow grow and seldom recurs locally after complete resection; however, it presents a high rate of metastasis. The main sites of metastasis are lung, bone, and brain. While recurrences or metastasis were reported in 30% of lingual ASPS in one study, only one out of 10 patients presented lung metastasis in another one. Therefore patients with lingual ASPS may be follow-up for long time.

Treatment of lingual ASPS includes surgical excision, sometimes combined with adjuvant radiotherapy or chemotherapy. According to a series of cases reported, adjuvant therapy may not be necessary if the small primary lingual ASPS can be completely resected and the patient does not experience clinical recurrence or metastasis. The survival for patients with no evidence of metastasis at the time of diagnosis is 60.0% at 5 years, 38.0% at 10 years, and 15.0% at 20 years. Prognostic factors are patient’s age, tumour size, and the presence of metastasis at diagnosis. In contrast to ASPS in other parts, lingual ASPS have a good prognosis. In the present case no evidence of metastasis or recurrence was observed after one year.

References