Epithelioid sarcoma of the tongue

X Leroy, A Delobelle, J L Lefebvre, V Cabaret, F Bloget, M O Vilain

Abstract
A case of epithelioid sarcoma in the tongue is reported. The patient, a 35 year old woman, presented with a non-ulcerated painful lesion of the tongue. Microscopically, the tumour was characterised by multiple coalescent nodules with central geographic necrosis infiltrating the lingual muscle. The tumour cells were epithelioid with abundant eosinophilic cytoplasm and atypical nuclei. Immunohistochemically, the tumour cells stained for vimentin, keratin, and epithelial membrane antigen. These morphological and immunohistochemical appearances led to the diagnosis of epithelioid sarcoma of the tongue. Seven years later, the patient died with metastatic dissemination to the scalp, lungs, and brain. No case of epithelioid sarcoma arising in the tongue has been described previously.

(J Clin Pathol 1997;50:869–870)

Keywords: epithelioid sarcoma; tongue; prognosis; immunohistochemistry

Epithelioid sarcoma is a rare malignant neoplasm occurring usually in the extremities of young adults. We present the first case in the literature of a primary epithelioid sarcoma located in the tongue, and demonstrate the use of immunohistochemistry in the differential diagnosis.

Case report
A 35 year old woman without a history of cancer presented with a painful non-ulcerated nodular lesion in the left half of the tongue of several months duration. A biopsy showed a sarcomatous proliferation. A hemiglossectomy was performed, followed by postoperative brachytherapy despite tumour free margins. Three years later, bilateral cervical lymph node metastases were removed. The patient died four years later with metastatic dissemination to the scalp, lungs, and brain in spite of radiation therapy.

Pathological findings
A 2.5 cm firm mass of the tongue was noted. Histologically, the tumour was poorly defined in the lingual muscle, typified by multiple nodules sometimes coalescent with central necrosis, which often appeared geographic (fig 1). The tumour cells were either fusiform or epithelioid, polygonal with a plump eosinophilic cytoplasm (fig 2). The nuclei were vesicular and sometimes had prominent nucleoli. Mitotic activity was high (25 mitotic figures per 10 high power fields). No vascular invasion was seen. The lymph node metastases were similar to the primary tumour.

Immunohistochemically the tumour cells stained for cytokeratin (KL1 diluted 1/120; Immunotech, Marseille, France) (fig 3), epithelial membrane antigen diluted 1/50 (Dako, Trappe, France), and vimentin diluted 1/200 (Dako). No staining was seen with antibodies to S-100 protein diluted 1/400 (Dako), cytokeratin 7 diluted 1/100 (Dako) or CD34 diluted 1/250 (Immunotech). Therefore, this lesion was diagnosed as an epithelioid sarcoma of the tongue.

Discussion
Since the first description in 1970 by Enzinger,1 epithelioid sarcoma has been known to involve the superficial or deep soft tissues in
eral nerve sheath tumours are S-100 protein positive. Ultrastructurally, the tumour cells of epithelioid sarcoma show intercellular junctions and abundant intermediate filaments. Several morphological variants of epithelioid sarcoma have recently been described: a fibroma-like variant, a chondroid variant, an angiomatoid form mimicking angiosarcoma, and a proximal-type with large cells and rhabdoid features that represents a more aggressive variant of epithelioid sarcoma.

The histogenesis of this tumour is uncertain: a pluripotential mesoderm origin is proposed by some authors. A relation with extrarenal malignant rhabdoid tumours was also suggested.

Epithelioid sarcoma is a malignant neoplasm with frequent local recurrences and often metastatic dissemination in regional lymph nodes, in the scalp, lungs, and brain.

Halling et al in a recent review of 55 cases found three histological variables associated with a poor prognosis: vascular invasion, tumour size of more than 5 cm, and more than 30% necrosis.

The treatment is radical surgery with radiation therapy; however, 50% of patients develop metastatic lesions and the five year survival rate is 70%.

We thank Dr J M Coindre and the French sarcoma group for reviewing the histological slides.

References

Epithelioid sarcoma of the tongue.

X Leroy, A Delobelle, J L Lefebvre, V Cabaret, F Bloget and M O Vilain

*J Clin Pathol* 1997 50: 869-870
doi: 10.1136/jcp.50.10.869

Updated information and services can be found at:
[http://jcp.bmj.com/content/50/10/869](http://jcp.bmj.com/content/50/10/869)

**Email alerting service**

Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

**Notes**

To request permissions go to:
[http://group.bmj.com/group/rights-licensing/permissions](http://group.bmj.com/group/rights-licensing/permissions)

To order reprints go to:
[http://journals.bmj.com/cgi/reprintform](http://journals.bmj.com/cgi/reprintform)

To subscribe to BMJ go to:
[http://group.bmj.com/subscribe/](http://group.bmj.com/subscribe/)