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Epithelioid sarcoma with metastatic spread to the tongue

Stuart C. A. Winter, M.R.C.S., Nicholas Steventon, F.R.C.S., Ketan A. Shah, F.R.C.P.*, Graham J. Cox, F.R.C.S.

Abstract
Epithelioid sarcoma (ES) is a rare tumour that seldom presents to the otolaryngologist. It typically occurs in the extremities of young adolescents; however, it has the capability of metastasizing, often to the lungs or skin. The diagnosis is by histopathological examination and immunohistochemistry. We present a case of metastatic ES occurring in the tongue, a tumour not reported previously in the English literature.

Key words: Sarcoma; Tongue; Neoplasm; Metastasis

Introduction

Epithelioid sarcoma (ES) was first described by Enzinger in 1970.[1] It is a morphologically distinct neoplasm of uncertain histogenesis, which can be mistaken for a variety of benign and malignant conditions. It is most prevalent in young adolescents, although no age group is exempt. It tends to affect males more commonly. The primary site is often the upper extremities and requires aggressive resection to prevent local recurrence. Metastasis occurs in up to 50 per cent of cases up to five years from treatment of the primary tumour.[2] Commonly, the tumour metastasizes to the lung and, although rarely primary tumours arise in the head and neck, there has been no previous documented evidence of ES metastasizing to the tongue.[3] ES should be considered by the otolaryngologist as part of the differential diagnosis, particularly when there is a history of a primary lesion elsewhere.

Case report

A 64-year-old man presented with a firm nodule on the lateral border of the tongue which was causing increasing dysphagia and pain. Two years previously he had been diagnosed with ES of the right hand, for which he had undergone amputation. However, several months later, he had represented with recurrence in the forearm, and a subsequent pleural biopsy had confirmed pleural metastasis.

The tongue lesion was excised with a wide margin, as part of a palliative procedure. The histopathological examination of the specimen showed an infiltrate of tumour with nodular architecture and central necrosis. The cells had an 'epithelioid' appearance with pleomorphic nuclei and a moderate amount of eosinophilic cytoplasm. Focal spindle cell morphology was noted. A few mitoses were in evidence (Figure 1). Immunohistochemical staining supported the diagnosis of ES. The tumour cells were shown to express cytokeratin and epithelial membrane antigen, vimentin and actin. S-100 and CD-31 were negative.

The patient's dysphagia improved, however, he died eight months following the palliative resection.

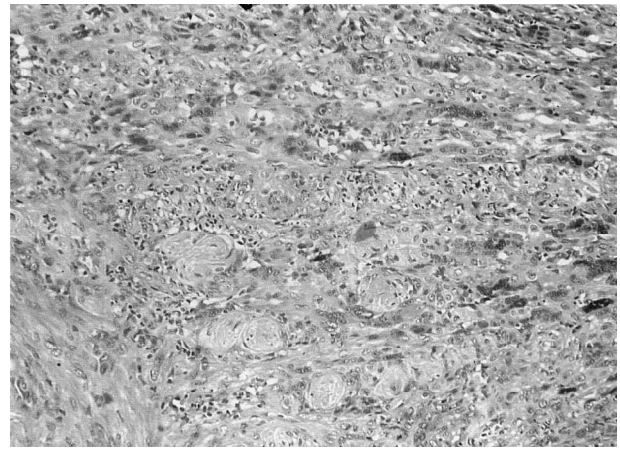

Fig. 1
Histological 'epithelioid'-shaped cells. (H&E; ×10)

Discussion

ES most often presents as a primary lesion in the extremities of young adults. Rare published cases detail intra-oral primary lesions, including the tongue.[4,5] Although the tumour is slow-growing the clinical behaviour is unpredictable, with local recurrences occurring and distant metastatic spread. The lung is the most commonly reported site of tumour spread, but lymph node involvement as well as, brain, scalp, bone and liver are reported.[6] However, there are no reports of the metastatic disease in the tongue. The tumour may be mistaken for a benign inflammatory process, or another sarcoma such as an epithelioid angiosarcoma. In the head and neck, the main differential diagnosis is squamous cell carcinoma.[7] The diagnosis is on histological examination; common features include nodular architecture of the tumour cells with areas of necrosis, epithelioid appearance and cytoplasmic eosinophilia.

From the Department of Otolaryngology, Radcliffe Infirmary, Oxford and the Department of Cellular Pathology*, The John Radcliffe Hospital, Oxford, UK.
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Immunohistochemically, most tumours stain positive for cytokeratins, epithelial membrane antigen and vimentin. Up to 70 per cent are positive for CD-34. Typically, antibodies directed against S-100 and CD-31 are negative. Histological predictors of poor outcome include vascular invasion, tumour size >5 cm, more than 30 per cent necrosis and positive resection margins.[2]

The treatment of the primary lesion is with wide resection; in cases affecting the extremities this may involve amputation. The local recurrence rate appears to be decreased by aggressive initial resection, however, this does not appear to influence the metastatic rate. Rates up to 50 per cent are reported, occurring five years or more from the initial presentation.[6] While primary intra-oral lesions have rarely been described, there are no reports of metastatic lesions occurring in the tongue. The diagnosis of ES needs to be considered by the otolaryngologist, particularly when an epithelioid sarcoma has been diagnosed elsewhere.

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Address for correspondence:
Mr G. J. Cox, F.R.C.S.,
Department of Otolaryngology,
Radcliffe Infirmary,
Woodstock Road,
Oxford OX2 6HE, UK.

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