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Synovial sarcoma in the retropharyngeal space

S. K. Nadig, F.R.C.S.(Ed.), M. H. S. Love, M.R.C.P., F.R.C.R.[*], D. S. Brooker, F.R.C.S.[†], S. J. Hall, F.R.C.S.

Abstract
Synovial sarcoma is an aggressive mesenchymal tumour, rarely occurring in the head and neck. Management guidelines are by extrapolation of management of sarcomas in the extremities. We present a case involving the retropharyngeal space in a 20-year-old male. Analysis of more data on head and neck synovial sarcoma is necessary to make meaningful management recommendations.

Key words: Sarcoma, Synovial; Neck

Introduction

Synovial sarcoma is a malignant mesenchymal tumour that originates from pluripotent mesenchymal cells[1] and accounts for up to 14 per cent of all soft tissue sarcomas.[2] Eighty-five per cent of these tumours occur in the extremities, especially in the knee joint. Only nine per cent occur in the head and neck.[3] We present a case of synovial sarcoma in the retropharyngeal space.

Case report

A 20-year-old male presented with a four-week history of dysphagia and hoarseness. The patient had been seen three years previously at a district general hospital with a mass in the right neck. Ultrasound at the time indicated a mass lesion posterolateral to the thyroid lobe. A right thyroid lobectomy, isthmectomy with removal of the 5 cm tumour was performed. Histopathology confirmed a synovial sarcoma but the upper margin was positive. He, therefore, underwent radical radiotherapy remaining symptom free until re-presentation.

On this occasion examination revealed a mass in the posterior pharyngeal wall obscuring the larynx. The regional lymph nodes were not enlarged. Laboratory values were within normal limits. Computed tomography (CT) showed an extensive retropharyngeal mass from C2/3 down to C6/7 posterior to the pharynx and the right pyriform fossa (Figures 1 and 2).

Endoscopic examination and biopsy confirmed the pathology. It was felt that the tumour could be removed by an external approach and partial pharyngectomy, but it was found that the tumour infiltrated the post-cricoid larynx. The patient declined a formal pharyngolaryngectomy or chemotherapy and received palliative care. He died six months later.

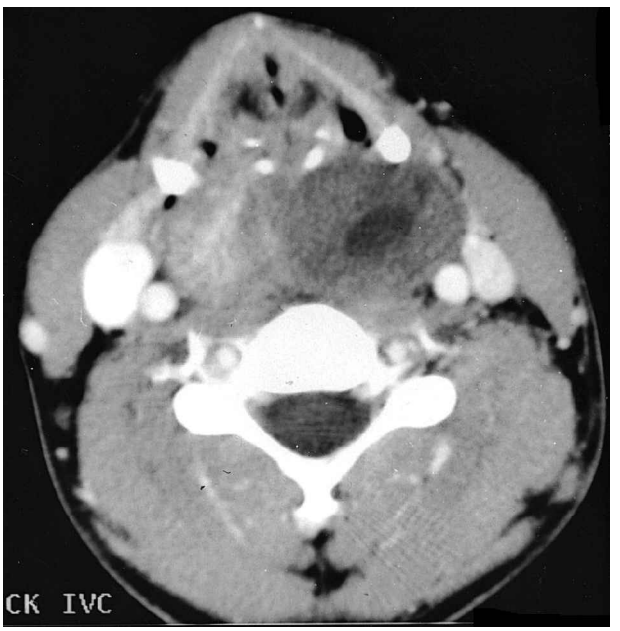

Fig. 1

Enhanced CT showing a nonhomogeneous mass in the retropharyngeal space. The supraglottic larynx is narrowed to a slit at the level of the false folds.

Discussion

Synovial sarcomas are aggressive high-grade neoplasms, occurring most commonly in young adults. Synovial sarcomas originate from undifferentiated or pluripotent mesenchymal cells,[1] near or remote from articular surfaces; however they are not derived from synovial cells. Two types, monophasic and biphasic have been identified. The biphasic type consists of two distinct cell types, epithelial cells positive for cytokeratin and epithelial membrane antigen (EMA) and spindle-shaped cells, which are present in various proportions and patterns. Histological features easily identify a biphasic synovial sarcoma, but monophasic synovial sarcoma is more difficult to distinguish from other sarcomas with fibrosarcomas, malignant schwannomas, haemangiopericytomas and leiomyosarcomas being the main differential diagnosis.

From the Departments of Otolaryngology and Radiology[*], Craigavon Area Hospital, Portadown and the Department of Otolaryngology, Royal Victoria Hospital[†], Belfast, UK.
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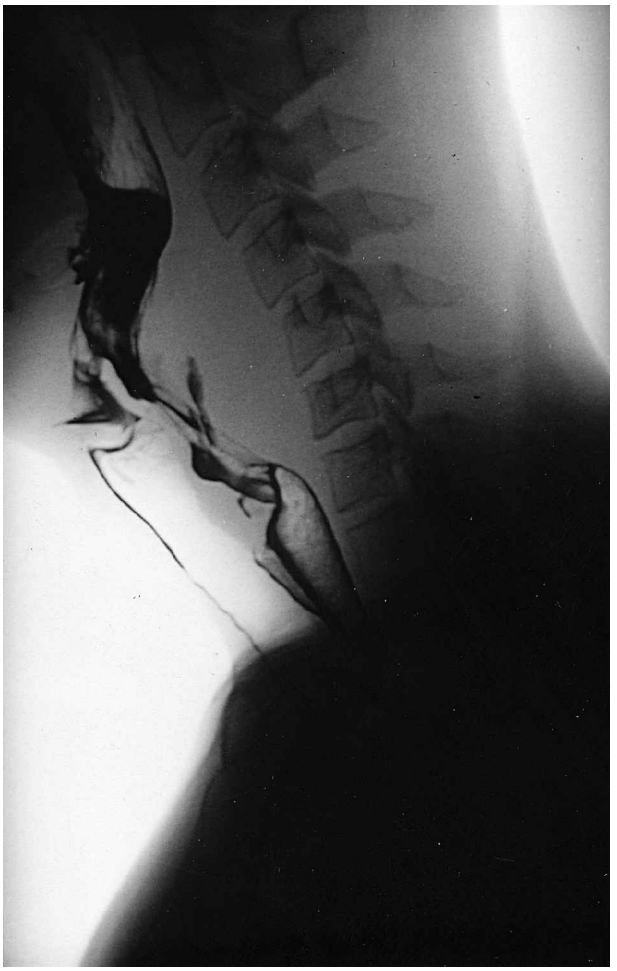

FIG. 2
Barium swallow showing a soft tissue mass in the retropharyngeal space.

CT and magnetic (MR) findings are non-specific and resemble those associated with other tumours of soft tissue.[4] Calcification, a feature on CT in this case (Figure 3), may be found in up to 30 per cent of adults with extensive calcification possibly indicating a more favourable prognosis.[4] Imaging, particularly MR, has a major role in planning surgery, radiation and determining prognosis.[5]

Rarity makes pre-operative diagnosis and treatment planning difficult. Diagnosis by FNA alone is also difficult, although there is now also a cell culture technique available. A complete wide surgical excision is the treatment of choice with regional nodal dissection only with evidence of involvement.[6] The five-year and 10-year survival rates following complete resection are higher than those in patients who undergo a non-curative operation.[7] The role of radiotherapy in the treatment of synovial sarcoma has not been clearly established. Mamelle *et al.*[8] have suggested that post-operative radiotherapy reduces the risk of local recurrence but does not improve the long-term survival. Recurrence with excision alone is as high as 90 per cent[1] but if combined with adjuvant therapy this reduces to 28–49 per cent.[2] A minimal total post-operative dose of 65 Gy is recommended for local control of this tumour.[7] Recurrent or metastatic tumours occur from four months to 62 months later; thus, long-term follow-up is important.[7] The five-year survival rate ranges from 23.5 to 45 per cent[4,5,9] with 11.2 to 30 per cent alive at 10 years.[2]

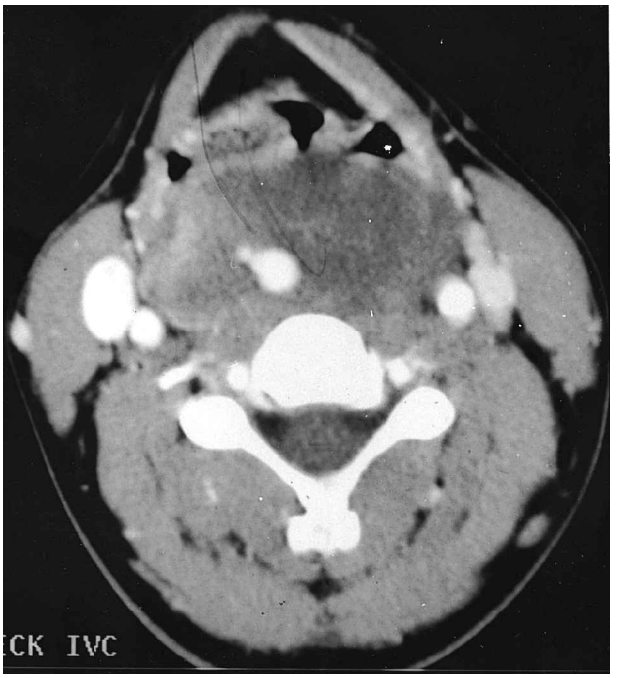

FIG. 3
Enhanced CT showing calcific concretions in the tumour.

Tumour size is the most important prognostic factor. In multivariate analysis, independent risk factors for local recurrence included larger tumour size and primary surgical resection outside the referral centres. Independent risk factors for metastasis are increasing patient age, tumour with poor histological differentiation, and tumour necrosis.[10] Head and neck tumours may have better prognosis by earlier detection, hence smaller tumours, but may be confounded by difficulty in achieving adequate margins. Death is usually due to metastatic relapse, with favoured sites being lungs 94 per cent, lymph nodes 21 per cent and bone marrow 17 per cent.[2]

In summary, this case illustrates that synovial sarcoma, a high-grade sarcoma, needs a wide primary excision with risk factor assessment to plan adjuvant therapy. Head and neck tumours may behave differently from tumours elsewhere but treatment guidelines have been extrapolated from synovial sarcomas elsewhere. There is a need to report every case so that meaningful management decisions for head and neck tumours can be made. In retrospect, perhaps the patient could have been better treated with more aggressive surgery after the initial operation. Furthermore, we would emphasize the importance of treatment based in a head and neck centre with long-term follow-up to increase local control, preserve function, and improve final outcome.

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Address for correspondence:
Mr S. J. Hall, F.R.C.S.,
Department of Otolaryngology,
Craigavon Area Hospital,
Portadown BT63 5QQ, UK.

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