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Pathology in Focus

Primary synovial sarcoma of the middle ear

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Abstract
Synovial sarcoma is a soft tissue malignancy which most commonly affects the lower limbs of young adults and rarely occurs in the head and neck region. The term synovial sarcoma may be a misnomer as most of these tumours occur in tissues not known to contain synovial tissue. There has only been one previously reported case affecting the middle ear, which was metastatic, and we report the first case of primary synovial sarcoma of the middle ear.

Key words: Sarcoma, primary synovial; Ear, middle

Introduction

Synovial sarcomas comprise eight to ten per cent of all sarcomas with approximately 90 per cent occurring in the lower extremities of young adults (Pack and Ariel, 1950; Cadman *et al.*, 1965). The head and neck region is rarely affected by this tumour with fewer than eighty reported cases (Chew *et al.*, 1992). The most common site to be involved in this region is the hypopharynx, followed by the neck, cheek, nasopharynx, tongue and larynx (Chew *et al.*, 1992).

One case of metastatic middle ear synovial sarcoma, secondary to a primary site in the left thigh, has been reported (Parker-Cross and Ladaga, 1986). To our knowledge, the case presented here is the first synovial sarcoma occurring primarily in the middle ear.

Case report

A 20-year-old female presented with a four-month history of right sided pulsatile tinnitus and increasing ipsilateral facial weakness. She had also developed right otalgia for several weeks before presentation.

Four years previously, while on holiday in the United States, the patient was treated for Lyme disease and was noted to have a very erythematous right tympanic membrane. On returning to the United Kingdom, an exploratory right tympanotomy and biopsy were performed. The biopsy specimen was reported to show granulation tissue only. Those specimens have since been independently reviewed by two pathologists who could find no sinister histological features.

Clinical examination on this occasion demonstrated no lymphadenopathy. Right facial nerve function was estimated at grade 2 with approximately 90 per cent function in all three divisions. The right tympanic membrane had been completely replaced by an erythematous pulsatile mass which appeared to be typical of a glomus tumour. The remaining cranial nerves were intact.

Pure tone audiometry demonstrated normal hearing on the left with a wide conductive loss on the affected side. High resolution CT and MRI scanning suggested a diagnosis of a glomus tumour (Figure 1). Angiography demonstrated a prominent tumour blush projected over the petrous bone which appeared to be filling almost exclusively from branches of the posterior auricular artery.

After pre-operative embolization the tumour was approached *via* a post-auricular incision. A large vascular tumour was found to have destroyed the tympanic membrane, completely filled the middle ear and extended from the tegmen tympani virtually to the isthmus of the eustachian tube. The tumour extended into the mastoid bone, comparable to a Fisch type C glomus tumour. The whole of the horizontal portion of the facial nerve, extending from the lateral half of the labyrinthine portion to midway along the descending portion of the nerve, was totally replaced by tumour. The vestibule had also been widely eroded by the tumour but there was no involvement of the jugular bulb. The cochlea and labyrinth were drilled out as far as the lateral end of the internal auditory meatus. The tumour was excised completely and a greater auricular nerve cable graft used to repair the facial nerve. The resulting cavity was packed using a combination of temporalis fascia, muscle and tissue glue. On the third post-operative day the wound was re-explored and repacked to seal a cerebrospinal fluid leak from the lateral end of the internal auditory meatus. There were no further post-operative complications.

Histological analysis of the tumour revealed it to be very cellular, composed predominantly of elongated cells with spindle-shaped nuclei. There was little cytoplasm and some cells showed a positive reaction to PAS stains. The reticulin pattern was heavy with prominent vascularity and some dilated vascular channels. There were several foci of distinct cartilaginous differentiation within the tumour. The spindle cells showed scattered mitoses and, on immunostaining, were positive for cytokeratin and faintly positive for desmin. They tested negative for CAM 5.2, smooth muscle actin, Factor VIII R Ag, S100 and chromogranin. Despite the fact that there were no typical clefts lined by epithelial cells present, their appearances were consistent with a monophasic synovial sarcoma (Figure 2 a and b).

The patient remains well 18 months after her surgery with no evidence either clinically or radiologically (as seen on repeat CT scanning) of tumour recurrence. Her right facial nerve function has recovered to grade 3 level.

Discussion

Synovial sarcomas often arise in areas remote from structures

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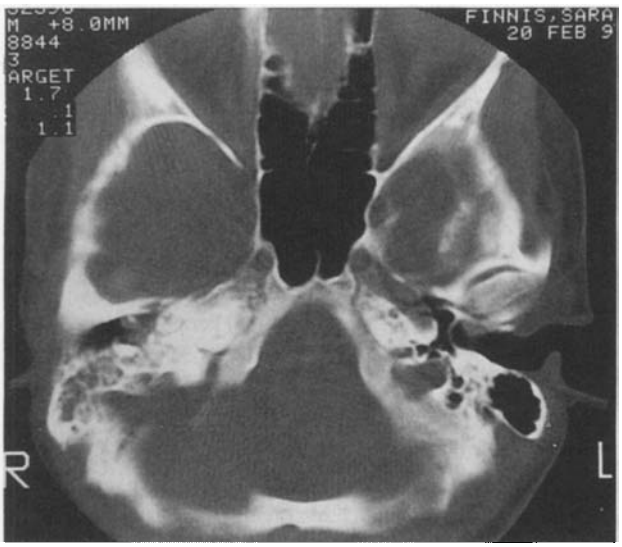

FIG. 1

CT scan demonstrating a soft tissue mass in the right middle ear, possibly with some early cochlear erosion, suggestive of a glomus tumour.

containing synovial membrane such as joints or bursae. This is particularly true in the head and neck where these lesions have been reported to occur in the hypopharynx, cheek, tonsil and palate, larynx and infra-orbital region (Chew *et al.*, 1992). There was no suggestion that the tumour in question here originated from the incudostapedial or malleo-incudal joints. It has been suggested that synovial sarcomas arise from undifferentiated mesenchymal tissue which retains the potential for synovioblastic differentiation (Jernstrom, 1954; Mackenzie, 1966). Immunohistochemical studies by Abenoza *et al.* (1986) support the theory of mesenchymal origin. Mackenzie (1966) stated that the term 'synovial sarcoma' derives from the particular pattern of mesenchymal differentiation observed.

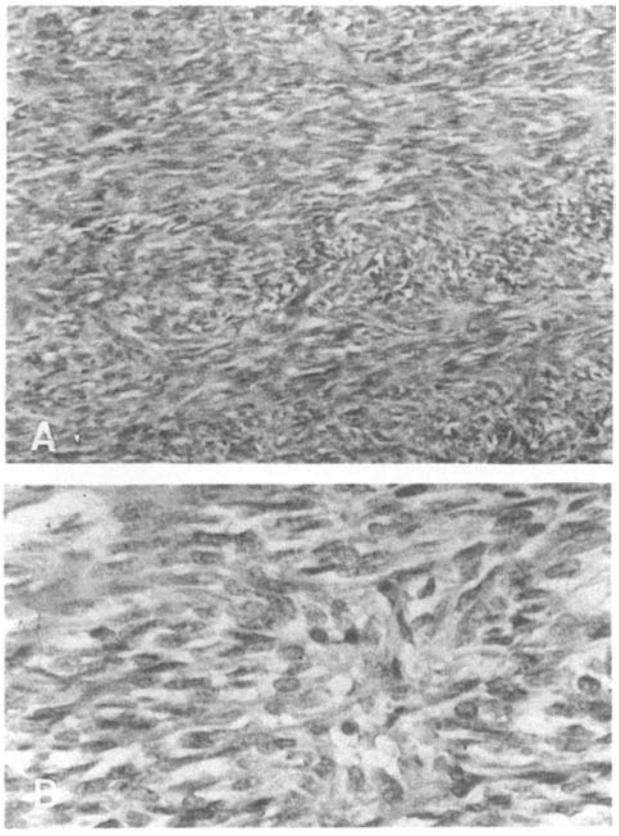

FIG. 2

Most of the tumour consisted of monotonous aggregates of spindle cells. (H & E; ×100(A) and ×200 (B)).

The tumour is classically biphasic, containing both epithelial and spindle cells. The biphasic nature of the tumour is the only diagnostic histological criterion although a monophasic variety where either cell type predominates is recognized (Mackenzie, 1966) as in this case. The different histological patterns seen make diagnosis difficult and tumours may be mistaken for other forms of undifferentiated mesenchymal tumours. Immunohistochemical studies, demonstrating positive immunoreaction for the epithelial markers, epithelial membrane antigen [EMA] and cytokeratin [CK], have been shown to increase diagnostic accuracy (Abenoza *et al.*, 1986).

With fewer than 80 reported cases, the most effective method of treating synovial sarcomas of the head and neck is uncertain. Studies have shown that the prognosis is, to some extent, dependent on the site of origin of the tumour. Cadman *et al.* (1965) have reported a 25 per cent five-year survival rate in 134 cases from all sites while Roth *et al.* (1975) reported 47 per cent five-year survival in 22 patients with synovial sarcoma of the neck. Shmookler *et al.* (1982) reported 66 per cent survival in nine patients with orofacial lesions at 2.9 years follow-up. Mamelle *et al.* (1986) have predicted a five-year survival rate of 62.7 per cent based on their series of four cases in the head and neck. Regional lymph node metastasis occurs in 12.5 per cent of cases in the head and neck region and 23 per cent of cases in the extremities (Pack and Ariel, 1950; Cadman *et al.*, 1965; Roth *et al.*, 1975). Pulmonary metastases were fatal in 10 out of 22 cases of synovial sarcoma of the neck reported by Roth *et al.* (1975). Tumours of the extremities metastasize to the lungs in 33 to 81 per cent of cases (Pack and Ariel, 1950; Cadman *et al.*, 1965; Roth *et al.*, 1975).

It would appear that synovial sarcoma occurring in the head and neck region carries a better prognosis than cases arising primarily in the extremities, based on the evidence referred to above. All authors agree on the necessity for aggressive surgical tumour clearance but some would also advocate adjuvant radiotherapy (Moore and Berke, 1987; Chew *et al.*, 1992). However, there are reports that this tumour may be unresponsive to radiotherapy (Gapany-Gapanavicius *et al.*, 1978; Quinn, 1984). Chemotherapy has not yet been shown to have any definite effect although it may help prevent growth of micrometastases (Mamelle *et al.*, 1986; Moore and Berke, 1987).

The case reported here was treated solely by wide surgical excision. Radiotherapy was not prescribed because: (i) tumour excision was thought to be complete; (ii) its benefit is not proven;

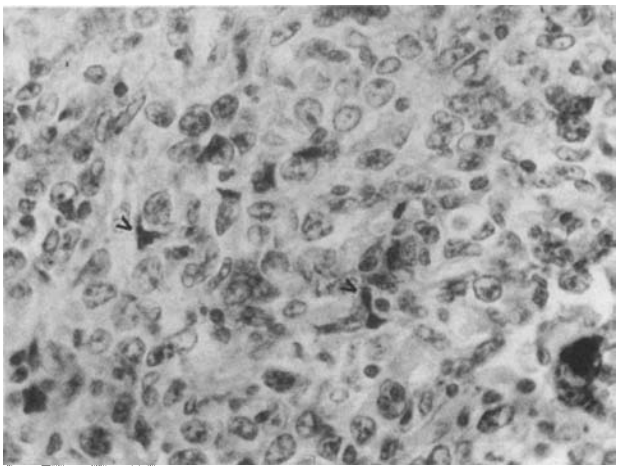

FIG. 3

Rarer areas consisted largely of rounded cells some of which reacted for epithelial membrane antigen (arrowed). (Immunoperoxidase; ×160).

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and (iii) the risk of radionecrosis of the temporal bone. Furthermore, it was felt that radiotherapy would most likely inhibit successful regeneration of the facial nerve.

Although the lymphatic drainage of the middle ear is poor and we have been advised that the tumour is histologically low grade, the prognosis in this case must remain guarded. This patient will obviously require close observation in the long-term to determine the success of her treatment.

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