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Synovial sarcoma of the pharynx

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Abstract
Synovial sarcoma is rarely seen in the head and neck region. A case of synovial sarcoma of the pharynx in a child is presented.

Key words: Pharyngeal neoplasms; Sarcoma, synovial

Introduction

Synovial sarcoma is a malignant soft tissue sarcoma occurring primarily in the extremeties. It accounts for seven to 10 per cent of all soft tissue sarcomas. It is rarely seen in the head and neck region and some 100 cases have been reported in this region since the first case reported by Jernstrom in 1954. A review of the literature has shown only nine pharyngeal synovial sarcomas in the paediatric age group and is summarised in Table I (Cachin *et al.*, 1966; Attie *et al.*, 1970; Roth *et al.*, 1975; Cutchavaree and Jimakorn, 1985; Mammelle *et al.*, 1986; Lenoir *et al.*, 1991). One further case of synovial sarcoma of the pharynx in the paediatric age group is reported here.

Case report

A 12-year-old boy was admitted with a two-week history of shortness of breath, productive cough and a sore throat. He had also noticed increasing difficulty in swallowing and change of voice in recent weeks and had also lost 8–10 lbs in weight over the past two months. Indirect laryngoscopy revealed a smooth vascular mass about 3 cm × 3 cm occluding the laryngeal inlet and the patient was breathing around the mass. It was thought to arise from the left lateral pharyngeal wall. X-ray of lateral view of the neck revealed a globular mass occluding the laryngeal inlet (Figure 1). As the airway was compromised, the same evening he underwent an emergency tracheostomy. Direct laryngoscopy revealed a smooth well-encapsulated vascular tumour occluding the laryngeal inlet arising from the lateral wall of the left pyriform sinus. The neck and larynx were uninvolved.

The tumour was biopsied and histology showed a tumour composed of two components (Figures 2 and 3) a prominent spindle cell component with dark staining nuclei forming fascicles, which stained positive with vimentin, and an epithelial component forming pseudoglandular spaces containing eosinophilic secretion. The latter areas were well delineated by CAM 5.2 and AE 1/3 and a diagnosis of biphasic synovial sarcoma was made. CT scan, X-ray chest, routine haematology and biochemical tests were normal.

He underwent partial pharyngectomy, the base of the tumour was excised with a wide margin, aryepiglottic fold and left half of the epiglottis were removed en-bloc. The defect was closed using a radial forearm flap and a distal cricopharyngeal myotomy was performed. The post-operative period was uneventful and he was having oral fluids and jelly by the seventh post-operative day and was decannulated a week later.

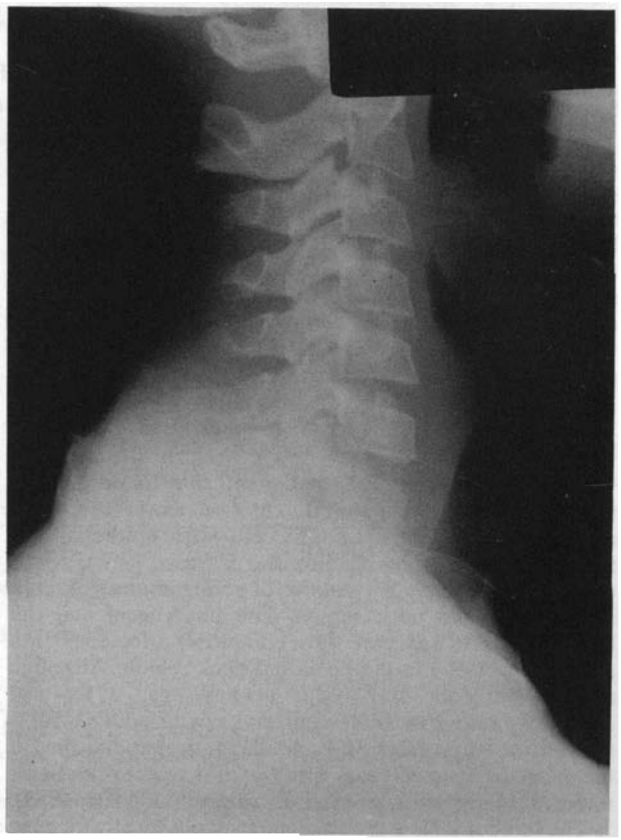

FIG. 1
X-ray soft tissue neck lateral view showing a globular mass occluding the laryngeal inlet.

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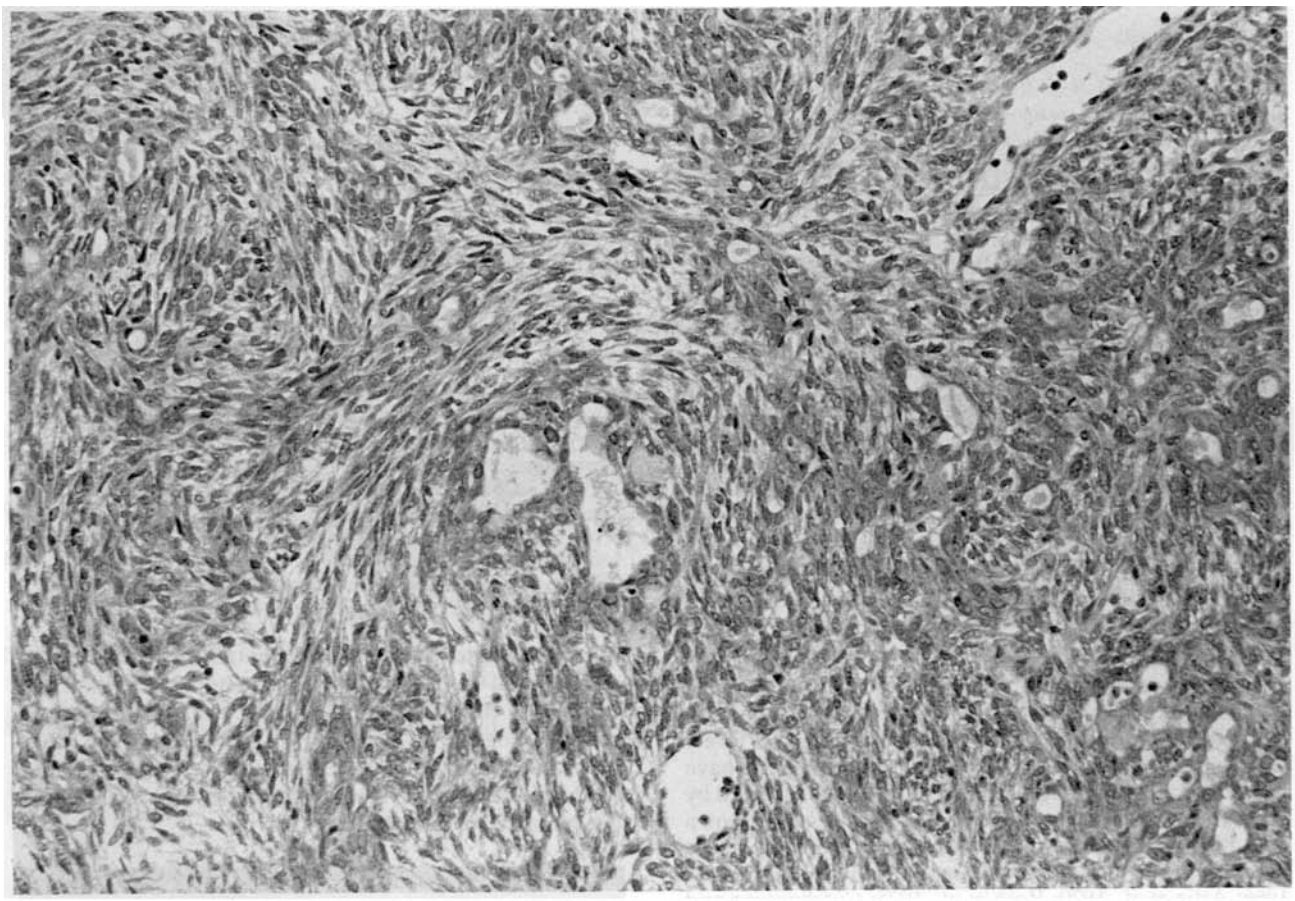

FIG. 2

Synovial sarcoma with biphasic growth pattern. There are insular areas lined by cuboidal cells surrounded by spindle cell areas. (H & E × 125).

He had a post-operative MRI scan of the head and neck region and an isotope bone scan which were normal. He had post-operative megavoltage, beam-directed radiotherapy to a dose of 45 Gy in 20 fractions over four weeks and six courses of systemic chemotherapy with vincristine, cyclophosphamide and adriamycin at three-weekly intervals. The patient is well and without any evidence of recurrence, a year after his operation.

Discussion

Synovial sarcoma is a malignant soft tissue tumour thought to arise from a pluri potent mesenchymal stem cell capable of differentiating into cells with epithelial and fibroblast-like features (Felito and Caruso, 1991), which usually occurs at the extremities of young adults. It is rare in the head and neck region. The pharyngeal and the hypopharyngeal areas are most commonly affected (Felito and Caruso 1991) in the head and neck region. Although synovial sarcoma may occur at any age, it is most commonly found in adolescent and young adults with a sex ratio of about 1.2: 1 M: F. At diagnosis, patients have a mean age of 31 to 36 years with 60–70 per cent of patients under 40 (Cadman *et al.*, 1965; Enzinger and Weiss, 1983).

The tumour is slow growing and expands locally forming a pseudo-capsule by compression of the surrounding normal tissue. The most typical presentation of synovial sarcoma is of a painless mass but some authors report that up to 40 per cent are painful (Pack and Ariel, 1950; Enzinger and Weiss, 1983).

Histogenesis of the tumour is still debated. Since it rarely involves the synovial membrane it is no longer considered as a neoplasm derived from it but rather as a lesion developing from a pluri potent mesenchymal stem cell capable of differentiation into epithelial and fibroblast-like elements. Differential diagnosis must be made from fibrosarcoma, leiomyosarcoma, spindle cell sarcoma, malignant schwannoma and adenocarcinoma.

Histologically the classic form shows a biphasic pattern with two neoplastic elements: (1) a spindle celled sarcomatous stroma, and (2) gland-like clefts lined by epithelial cells. There are two monophasic forms of which either the epithelial or mesenchymal components is difficult, or impossible, to find (Enzinger and Weiss, 1983; Leader *et al.*, 1985; Oppendal *et al.*, 1985; Moore and Berke, 1987). The monophasic epithelial form without an identifiable mesenchymal component is rare and is difficult to differentiate from a carcinoma. Dystrophic calcification and calcospherites are characteristic findings in synovial sarcoma (Michaels, 1987). The present case is a biphasic form without calcification.

The prognosis of synovial sarcoma is difficult to formulate. Enzinger and Weiss believed that size influenced prognosis as patients whose tumours were less than 4 cm in diameter seemed to do better than those with larger tumours. The influence of histological differentiation on the prognosis is unclear. Schmookler *et al.* (1982) found no association between the outcome and pathological features of the synovial sarcoma on head and neck. However, Enzinger and Weiss have correlated improved prognosis with degree of differentiation of the tumours and calcification within the tumour.

The treatment guidelines for synovial sarcoma of the head and neck have been extrapolated from those tumours

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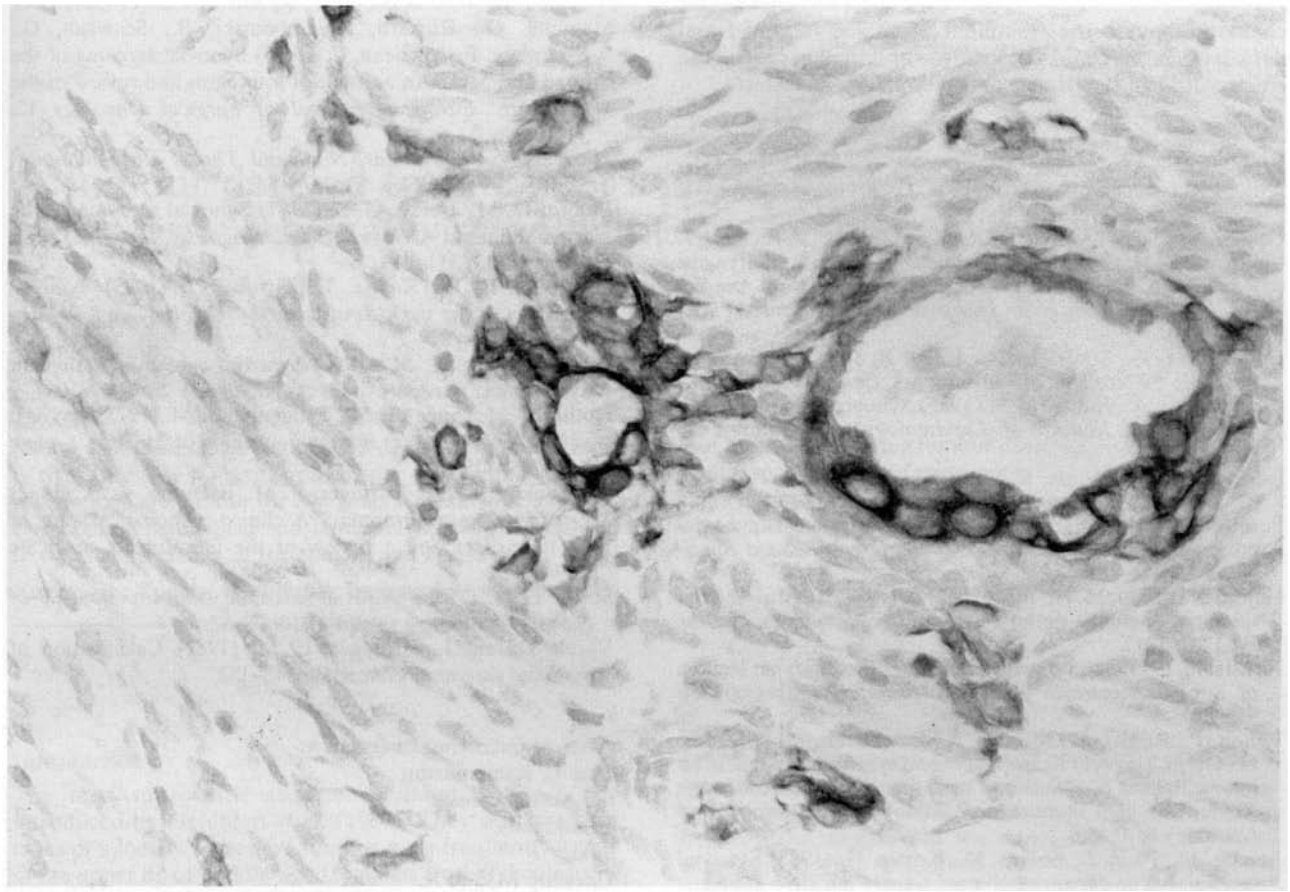

FIG. 3
Shows acinar areas positive with cytokeratin (CAM 5.2) and the spindle cells are negative for cytokeratin. (Immunoperoxidase × 313).

arising in the extremeties. Most authors advocate either radical surgery or wide excision as the treatment of choice. The role of radiotherapy is not clearly defined.

Radiation therapy has been reported to decrease the usually high rate of local recurrences (Moore and Berke, 1987). Chemotherapy is also thought to have a potential role for control of distant metastasis especially late pulmonary metastasis which can occur in up to 45 per cent of cases (Roth *et al.*, 1975; Moore and Berke, 1987).

Overall, synovial sarcoma is a grave disease with poor prognosis. Hajdu *et al.* (1977) reported five years survival of 40 per cent. The prognosis for synovial sarcoma in the head and neck region appears to be better than for synovial sarcoma in the extremities with five year survival ranging from 40 per cent 80 per cent. (Roth *et al.*, 1975; Varela and Enzinger, 1982; Soule, 1986).

In conclusion, it appears that multi-modal treatment with wide excision, post-operative radiotherapy and

TABLE I
REPORTED CASES OF PHARYNGEAL SYNOVIAL SARCOMA IN CHILDREN

Ref	Age/Sex	Location	Therapy	Follow-up
1	15/F	Retropharynx & second cervical vertebra	Neck dissection & excision + RT 60 Gy	No evidence of disease at 10 years
2	14/M	Hypopharynx & Retropharynx	Wide local excision	No evidence of disease at seven years
4	9/M	Hypopharynx	Microlaryngeal excision seven months later wide excision one year later total laryngectomy	No evidence of disease at two years
10	15/12/M	Retropharynx & first and second cervical vertebra	Chemotherapy (not completed)	Died 26 months of age
12	8/M	Retropharynx & post auricular	Wide local excision RT 50 Gy & chemotherapy	No evidence of disease at 15 months
17	13/F	Hypopharynx & Retropharynx	Excision 1959, 1962, 1963 RT 1964 & chemotherapy	Died (lung metastasis) 1966
17	15/F	Retropharynx	Excision Radiotherapy	Alive and well
17	12/M	Hypopharynx	Excision	Died of lung metastasis
17	12/F	Retropharynx	Excision X2 (1967, 1968)	Alive and well at six years (1973)
Present case	12/M	Hypopharynx	Wide excison RT 50 Gy Chemotherapy	No evidence of disease at one year

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chemotherapy, is the treatment of choice for head and neck synovial sarcoma, especially in children, to reduce the chance of local recurrence and pulmonary metastasis.

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