Cervical Synovial Sarcoma in a Young Boy

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SUMMARY

Synovial sarcomas comprise about 8% of all tumours of somatic soft-tissues, and are the most common sarcomas of the hands and feet. Occasionally they may occur in the trunk, but they have rarely been reported in the neck.

We present a case of cervical soft-tissue mass producing symptoms in a 12-year-old-boy. This mass could be defined partially by radiological techniques, but its nature was not suspected because of its rarity in children and particularly in view of its unusual site.

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CASE REPORT

A 12-year-old Cape Coloured boy presented with a 3-week history of dysphagia for solids and a change of voice. There was no history of dyspnoea, weight loss, trauma or previous disease.

On general examination he appeared healthy. The ears, nose and nasopharynx were normal. The vocal chords and epiglottis were not demonstrated. At the level of the hyoid bone was a firm mass which moved on tongue movement. No other abnormality was found. Blood and biochemistry tests were normal.

On screening, a large midline mass was seen, situated in the hypopharynx at the hyoid level, with a smooth convex upper border (Fig. 1). At barium swallow the upper convexity of the mass was outlined and barium passed behind it and along the lateral food channels to the oesophagus (Figs 2 and 3). At this stage the diagnosis included a cyst, ectopic thyroid tissue or a tumour arising from any structure in the anterior wall of the hypopharynx, etc. The thyroid isotope scan was normal.

Laryngoscopy revealed a fleshy white tumour, with intact overlying mucosa, arising from the left anterolateral wall of the hypopharynx in the vallecula. The vocal chords and epiglottis were not seen.

At operation the tumour was removed piecemeal and appeared necrotic and avascular. The pedicle of the tumour was at the level of the arytenoid mounds.

Histology revealed a synovial sarcoma which did not appear to arise from any synovia and was, in fact, fairly far removed from any bone, tendon, cartilage or joint.

Lung tomography was negative. The boy recovered well from the operation and was symptom-free a month later.

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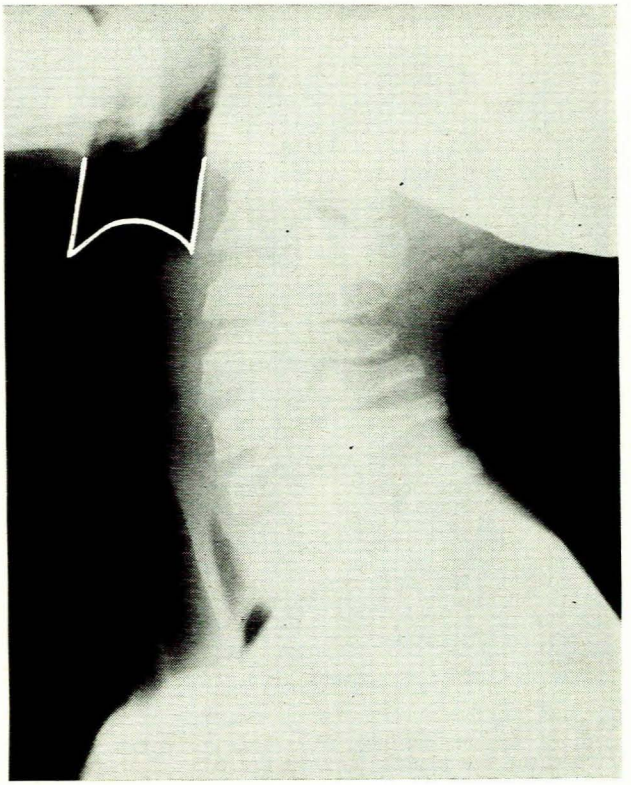

Fig. 1. Soft-tissue mass (outlined) at the level of the hyoid.

DISCUSSION

The term synovial sarcoma implies an origin from synovial cells lining joints, tendon sheaths or bursae, but the tumour is rarely demonstrated as arising from any of these structures, although it may be in close approximation.[1,3] It has a predilection for the extremities and limb girdles,[1-5] and in adults the commonest site is the knee, followed by the ankle and foot. A review of the literature reveals that other sites have been reported in the gluteal region, the bursae of the abdominal wall,[2] and the temporal region of the scalp.

Twelve cases of sarcomas in the neck have been reported[6-10] arising variously from the sublingual area, the hypopharynx, and the larynx, attached to the hyoid bone and to the thyrohyoid membrane.

The tumour is rare in children. Crocker and Stout[4] found 9,5% of soft-tissue tumours in children under the age of 15 years, and only 1,8% of these were synovial sarcomas. There is a slightly higher male sex incidence. Most series report the average age at discovery as 32 - 38 years, although it has been seen at 72 years of age and in the foot of a newborn baby.[11]

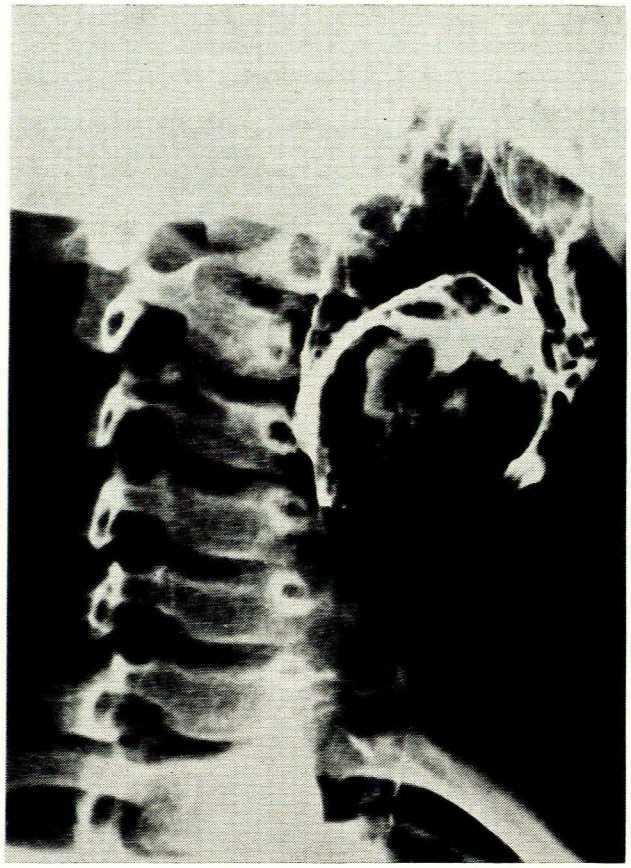

Fig. 2. Barium outlining the mass and passing behind it down the oesophagus (oblique aspect).

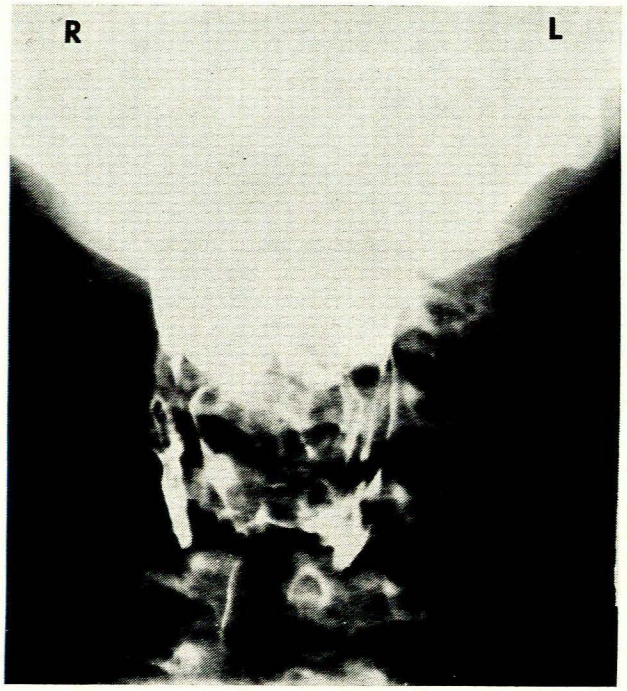

Fig. 3. Barium outlining the sides of the mass (frontal aspect).

The symptoms are very variable and may range from a painless mass (usually near a joint or bursa) to occasional prolonged unexplained pain. Paucity of symptoms and insidious growth may cause a delay in diagnosis of as long as $2\frac{1}{2}$ years. In our patient, who had a short history, the symptoms were explained by the mechanical effect due to the site of the tumour.

In their series of 134 cases Cadman *et al.*[1] found lung metastases in 81,1%, spread to regional lymph nodes in 23% and to the bones in 20%. Death followed in about $6\frac{1}{2}$ years. Metastases have also been seen in the skin, liver, paravertebral regions, bladder and intestines.[11]

In plain films of the local lesion, many demonstrate a fairly well-defined soft-tissue mass, which may be lobulated. If calcification is seen it is usually discrete. When the tumour is contiguous to bone there may be erosive changes due to pressure by the mass. Actual bony erosion may occur, appearing as cystic areas of rarefaction.[1,5]

The tumour is a lesion of somatic soft tissues and may have a biphasic histological pattern with a spectrum ranging from an adenocarcinoma to a small cell fibrosarcoma.[3] Jernstrom[12] suggests these tumours are a neoplastic expression of supporting mesenchymal body tissues to form synovial-lined spaces under certain conditions.

Histology is typical and shows two definite cell forms, spindle and epithelioid. There is an absence of reticulin fibres in the epithelial-like areas, which are surrounded by an abundance of reticulin fibres in the fibrosarcoma-like parts. Special stains show Alcian blue and mucicarmine positive material, but the latter may be negative in poorly-differentiated tumours.

Treatment varies from local to wide excision, with or without radiation and/or amputation. Whatever the treatment, there is a high incidence of local recurrence, and 60% of cases may have metastases within 16 months. This was usually caused by blood spread, but lymphatic spread also occurred.[3] However, the prognosis appears better in children, with 22,5% apparently cured after 5 or more years.[4]

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