PARAPHARYNGEAL SYNOVIAL SARCOMA: MR IMAGING

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SUMMARY

Synovial sarcoma is a rare, malignant soft-tissue tumour, which usually arises in the extremities of a young adult. They are rare in other areas of the body including the neck. We present a case of synovial sarcoma occurring in the neck with MR findings. A heterogeneous multiloculated tumour with fluid-fluid levels, calcification and haemorrhage occuring in a young adult, should raise the suspicion of a synovial sarcoma.

Key words: magnetic resonance, synovial sarcoma, neck

CASE REPORT

A 11- year old female presented with gradual enlargement of a lump in the left side of the neck over a period of several months. The patient's general health was good. On examination the patient was afebrile with a firm mass measuring approximately 4 cm in diameter palpable in the left side of the neck beneath the angle of the mandible. There was no lymphadenopathy. The spleen and liver were not palpable and the skin and mucosa of the head and neck region were normal.

Magnetic resonance imaging of the neck was performed with axial T1-weighted and T2-weighted spin-echo, and coronal short TI-inversion recovery sequences (STIR). Gadolinium-enhanced (Gd-DTPA) images in the axial and coronal planes were also obtained. T1- weighted axial spin-echo images (Fig1) demonstrated a lobulated mass with intermediate signal intensity and focal region of higher signal intensity suggesting the presence of subacute haemorrhage.

Coronal STIR (Fig2) and axial T2-weighted (Fig3) images revealed a heterogeneous but predominantly high signal intensity mass with internal septation and multiple fluid-fluid levels. There were a few foci of signal void within the mass suggesting the presence of haemosiderin or calcification. Gadolinium-enhanced images (Fig4) demonstrated heterogeneous nodular and septal enhancement. The tumour measured 4.5x3.5 x2.5cm. The main bulk of the tumour was located in the left parapharyngeal space, with extension inferiorly to the paralaryngeal space. Tumour extended inferiorly to the level of the superior pole of the left thyroid gland with displacement of the larynx and upper airways to the right. Superiorly the tumour extended to the level of the palatine tonsil. The posterior tonsillar pillar (palatopharyngeus muscle) was displaced anteriorly and the posterior belly of the digastric muscle laterally. On retrospective review the margins of the posterior belly of the digastric muscle and longus capitis muscle were ill-defined. The tumour appeared inseparable from the pharyngeal constrictor muscle. There was distortion, rotation and displacement of the oropharynx and hypopharynx which was displaced to the right. The left pyriform sinus was obliterated. The left

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submandibular gland was displaced antero-inferiorly, the carotid sheath and sternocleido-mastoid muscle were displaced laterally.

At surgery an encapsulated mass extending to the base of the skull was excised. This contained liquefied haemorrhage and some soft, pale gelatinous tissue in part of the mass.

Histology of the mass revealed a monophasic synovial sarcoma with poorly differentiated areas. Immunostains demonstrated spotty keratin-reactivity, with little or no labelling for desmin or S100 protein. There was diffuse strong CD99 staining of the tumour consistent with the diagnosis of synovial sarcoma.

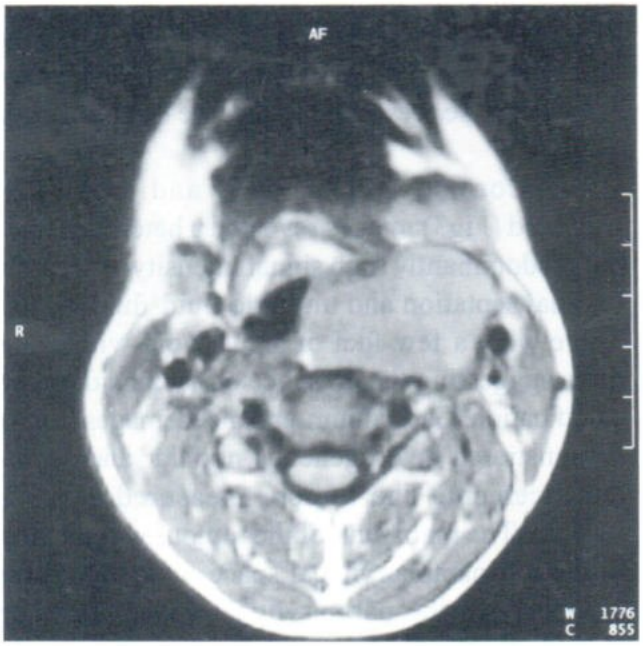


Fig 1. T1-weighted spin-echo axial image shows a well-defined lobulated mass with intermediate signal intensity. On the lateral aspect the mass demonstrated higher signal intensity suggesting the presence of haemorrhage.

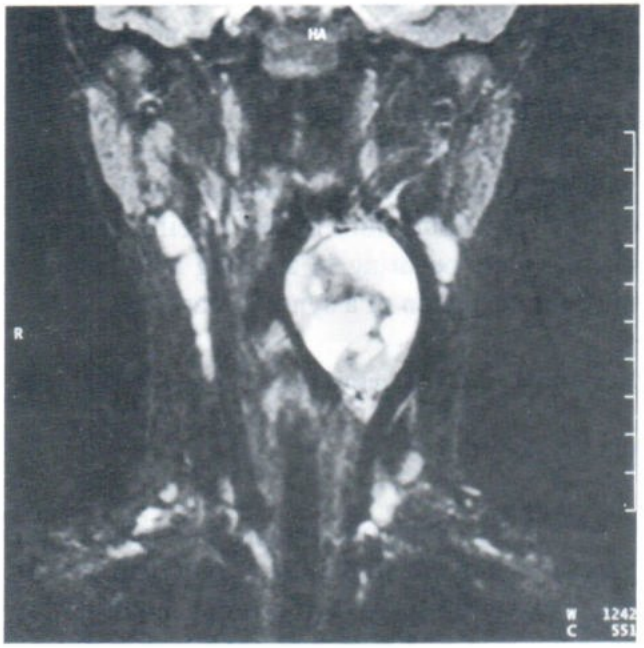


Fig 2. Coronal STIR MR image demonstrated heterogeneous but predominantly high signal intensity mass with several foci of signal void suggesting the presence of calcification or haemosiderin.

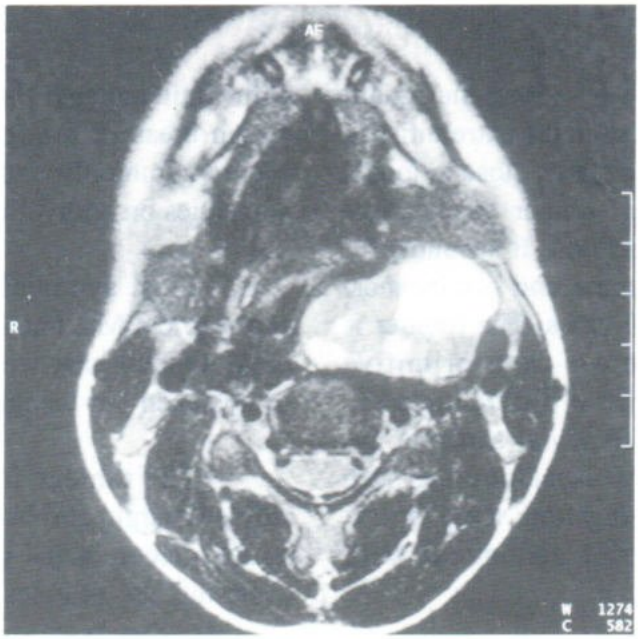
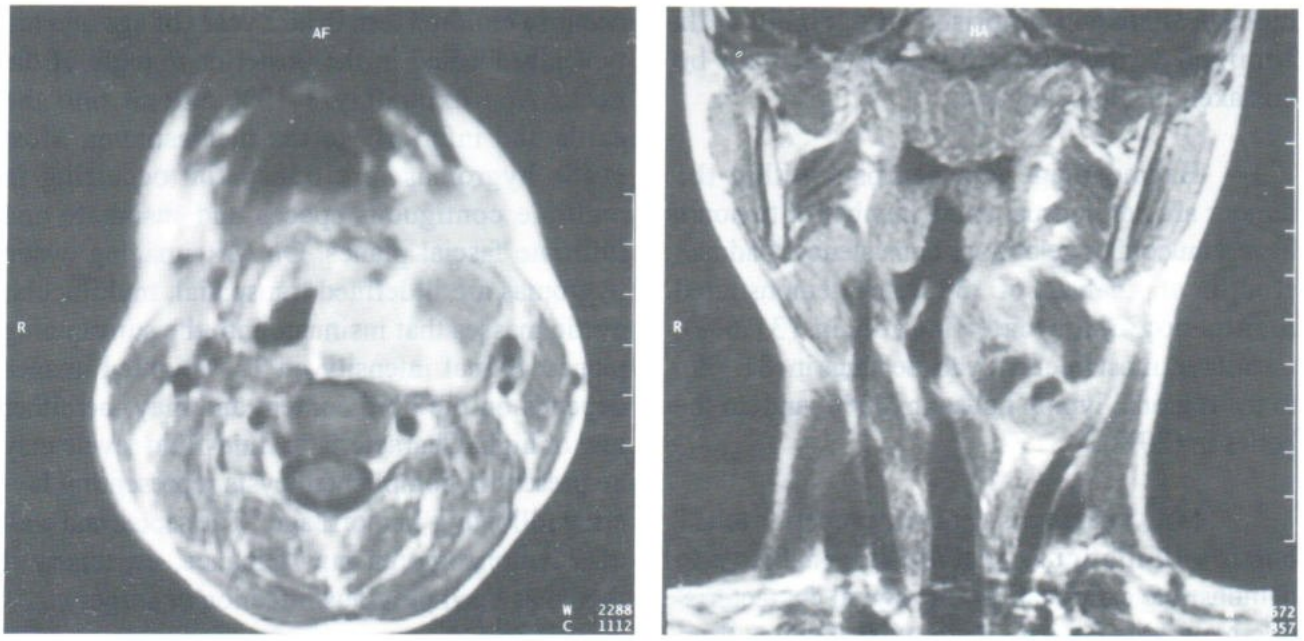


Fig 3. T2-weighted spin-echo axial image reveals a multilocular mass with fluid-fluid levels.



4A. **4B.**

Fig 4. A) Axial and B) coronal gadolinium-enhanced MR image displays nodular and septal enhancement.

DISCUSSION

Synovial sarcoma is a rare malignant tumour and frequently arises in the extremities in young adults more often distally, close to joints, tendon sheaths and bursae. Tumour rarely arises from an intra-articular location. It accounts for 8-10% of soft tissue sarcoma and occurs most commonly between 20-40 years. The tumour is considered to arise denovo from primitive mesenchymal cells that have differentiated to resemble synovial cells. Synovial sarcoma has been reported to occur in unusual locations such as the cervical region arising from the pharynx, larynx, chest wall, anterior abdominal wall, gluteal region and vagina.[1,2,3]

Tumours rarely arise in the cervical region. The first case of hypopharyngeal and laryngeal synovial sarcoma was reported by Jernstrom in 1954.[1] The tumour is usually characterised by a biphasic pattern comprised of spindle cells admixed with epitheloid cells and may be mistaken for fibrosarcoma.[4]

Haemorrhage and necrosis within the tumour are not unusual and a number of tumours have a mucoid or gelatinous consistency. Cadman et al, in an analysis of 134 cases demonstrated roentgenographic calcification in (31.6%), metastases most frequently to the lungs in (81.1%), to the regional lymph nodes in (23%) and to the bones in (20%).[4]

MR imaging is generally superior to CT but it's limitations include it's inability to demonstrate calcification and signal characteristics generally do not distinguish benign from malignant tumours.[5]

On MR imaging synovial sarcomas have predominantly heterogeneous signal intensity multiloculated masses with internal septation and multipile fluid-fluid levels.[6] The signal intensities of the fluid indicate the presence of haemorrhage within areas of cyst or necrosis.[7] Calcification occurs commonly and manifests as low signal intensities on both T1 and T2-weighted images.

Most synovial sarcomas enhance with contrast. On MR imaging approximately a third of the tumours have well defined margins and can be mistaken for benign tumours.[8]

The tumour in our patient had an ill-defined margin and had enlarged cervical lymph nodes. Secondaries to regional lymph nodes occur in 23% of cases.[4]

The differential diagnosis of multiloculated cystic masses in the neck includes thymic cyst, cystic hygroma/lymphangioma and cervical teratomas. The majority of cystic hygromas, occur in children less than 2 years of age and are typically located in the posterior triangle of the neck. Approximately 3-10% extend into the axilla and inferiorly to the mediastinum. Most cystic hygromas are transpatial occurring in multiple contiguous spaces and insinuate and infiltrate fascial planes. On MR imaging cystic hygromas are illdefined transpatial, multilocular cystic masses that insinuate into the surrounding planes. Signal intensity on T1-weighted images depends on the lipid content and may be either low or high. Characteristic fluid-fluid levels may be present in complicated cysts representing layering following hemorrhage.[9] The septae and wall of a lymphangioma may enhance, especially if there has been previous infection or surgery.

Thymic cysts are rare and the majority occur between 2 and 13 years of age and are located in the lower half of the neck and mediastinum. On imaging, thymic cysts are generally thin-walled unilocular or multilocular masses closely associated with the carotid sheath.

The mass is frequently elongated at one or both ends, tapering into a tract or cord.[10]

Three percent of teratomas are found in the neck. Cervical teratomas are multilocular, heterogeneous masses containing both solid and cystic components with scattered calcification.[11]

Other parapharangeal non-mucosal neck masses include neurogenic tumours and other soft -tissue sarcomas. Fluid-fluid levels within a cystic mass have been described in a schwannoma and a neurofibroma.[12] Extracranial schwannomas are most commonly found in the mid-neck. On MR imaging, schwannomas are of intermediate signal intensity on T1-weighted images and hyperintense on T2-weighted images depending on the cellularity. Twenty percent of schwannomas have cystic degeneration and may mimic other cystic masses. Some older schwannomas (ancient schwannomas) have degenerative changes with haemorrhage, haemosiderin and fibrosis.

CONCLUSION

Although signal characteristicS of synovial sarcomas are non specific, MRI is useful to demonstrate the extent of the tumour and pressure effects on adjacent airway. The MRI findings of a heterogeneous, multiloculated tumour in a young adult with fluid-fluid levels, calcification, and haemorrhage suggest synovial sarcoma

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