

REVIEW ARTICLE

THE UBIQUITOUS SYNOVIAL SARCOMA

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Although named synovial sarcoma by early pathologists, a synovial origin for this common tumour has not been proven to date. The histological resemblance to developing synovium, encouraged the name synovial sarcoma, which is still in use, simply because a cell of origin has not been definitely indentified.¹ The term, carcinosarcoma of soft tissue is suggested as an alternative and more appropriate name.¹

Synovial sarcoma commonly occurs around the knee and ankle joints.² However tumours have been reported in practically every site in the body, except in the brain, meninges, and intraosseous locations. Some unusual sites reported are orbit, heart, superior vena cava, lung, oesophagus and fallopian tube.³⁻⁸

We report for the first time, a mesenteric synovial sarcoma, of the small intestine, in a 55 year old man, who presented with an abdominal mass of 19cm diameter (Figure 1). Histology revealed a classic biphasic synovial sarcoma, confirmed with immunostains.

Omental and abdominal wall tumours have been reported previously.

Intra articular synovial sarcomas are rare. Figure 2 shows an intra articular synovial sarcoma of the knee joint in a 35 year old man who presented with pain in the joint. The x ray showed appearances of 'loose bodies'. Histologically the characteristic features of a monophasic synovial sarcoma were seen. The intra articular location and the pain caused, facilitated an early diagnosis in this case, and would carry a good prognosis. (Courtesy Dr. R. Waduge).

Synovial sarcomas are classically biphasic tumours, comprising histologically easily recognizable epithelial and sarcomatous components. This descriptive, (rather than histogenetic) pattern can be easily seen on a reticulin stain. In fact, this is a useful stain to bring out the biphasic nature of more monophasic appearing synovial sarcomas (Figure 3).

Incidentally, reticulin staining in synovial sarcoma remains one of the few uses for this silver staining technique, in contemporary histopathological practice. When the epithelial component shows glandular spaces / structures, diagnosis on haematoxylin and eosin stained sections is easy.

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When the epithelial component is of columnar type cells or solid nests of pale cells, the reticulin stain is of immense use to bring out the epithelial configuration. Squamous differentiation in this component is exceptional, but can mimic a squamous cell carcinoma when it does occur.^{1,2}

The sarcomatous component is classically of spindle cells, and characteristically monotonous in appearance. However, focally, a whorled pattern, fasciculation or lobulations and haemangiopericytomatous pattern can be seen. The stroma contains characteristic wiry collagen. Foci of hyalinization, and myxoid change can also be present. Rarely, extensive osseous metaplasia can mimic a bone forming tumour, or myositis ossificans.²

The monophasic synovial sarcoma comprises predominantly of the spindle cell component, and can mimic a fibrosarcoma, haemangiopericytoma, or other spindle cell neoplasms histologically. In this variety too, the reticulin stain can bring out subtle epithelial elements, and can be very useful, especially when immunohistochemistry is not readily available. The existence of a purely epithelial synovial sarcoma is still questionable.

A poorly differentiated variety is being increasingly diagnosed. Figure 4 shows a poorly differentiated synovial sarcoma, arising in the limb of a 35 year old man. Note the lack of any clue to the origin of the tumour. In this type, the tumour cells can appear spindly, small, large or clear, and can mimic a round blue cell tumour. Figure 5 shows a synovial sarcoma with pseudovascular structures with unusual stromal sclerosis. Classic appearance in other areas of the tumour, and immunohistochemistry, helped in the diagnosis of a synovial sarcoma in this case. Some tumours can have extreme cystic change, which could mislead the cursory observer (Figure 6).

Routine special stains, except for the reticulin stain, are of little value in the diagnosis of this tumour. Mast cells are conspicuous in synovial sarcomas, and staining for these cells with a metachromatic stain such as crystal violet etc. can give a useful clue to the pathologist, especially in the Sri Lankan context.

The mainstay of diagnosis of synovial sarcoma rests on immunohistochemistry. Cytokeratin is consistently demonstrable in the epithelial components, and to some extent in the spindle cell component. Of the cytokeratins, 7, 14 and 19, are useful, as 8 and 18, can be seen in synovial sarcoma as well as other soft tissue sarcomas.² Epithelial Membrane Antigen, is another epithelial marker that is present in synovial sarcoma.

Some synovial sarcomas stain positively with CD99 and BCL2.^{2,7} These are potential pitfalls for pathologists in Sri Lanka, who are at times compelled to use limited panels of antibodies to cut costs. Focal reactivity with calretinin is also seen, and is important when differentiating from a mesothelioma.² Immunoreactivity for S100 protein can pose a problem when differentiating from a malignant peripheral nerve sheath tumour. With reduction of the epithelial component, the staining for epithelial markers becomes focal and necessitates immunostaining of multiple sections, which can be an expensive exercise in the Sri Lankan setting.



Figure 1. Mesenteric synovial sarcoma. The red arrow points to the intestine, cut open to show the lumen

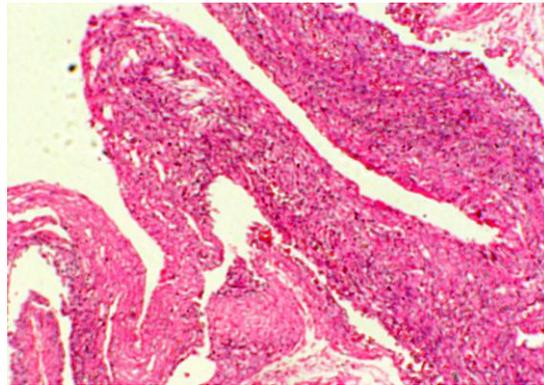


Figure 2. Intra-articular synovial sarcoma, protruding into the joint space (Haematoxylin and eosin x4)

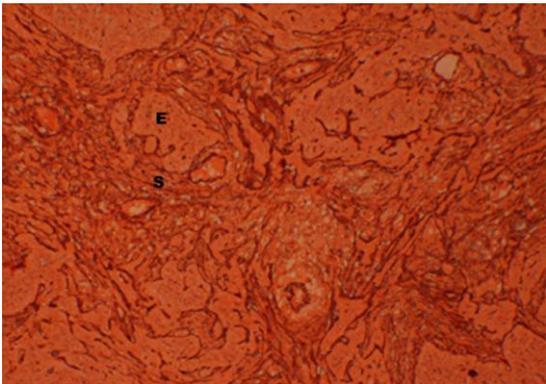


Figure 3. Biphasic reticulin fibre staining (S – Sarcoma pattern, E – Epithelial pattern) (Gordon & Sweet's silver stain for reticulin fibres x10)

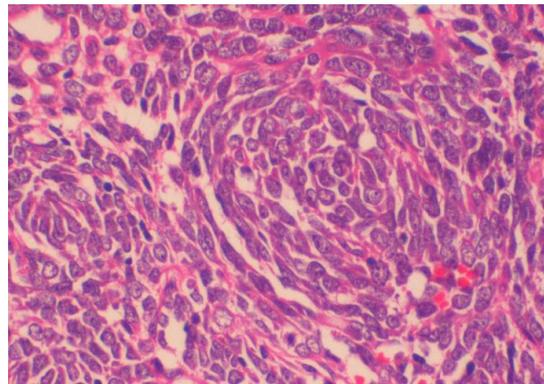


Figure 4. Poorly differentiated synovial sarcoma, resembling a blue cell tumour (Haematoxylin and eosin x40)

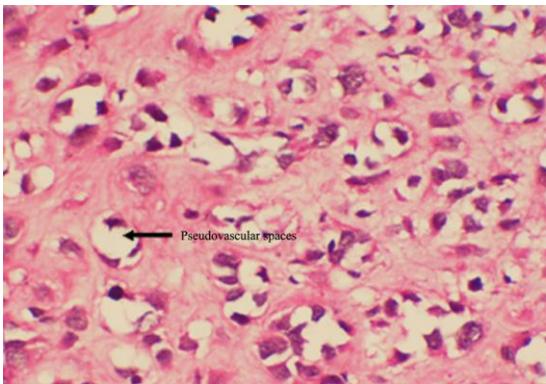


Figure 5. Pseudovascular spaces in a hyalinized stroma (Haematoxylin and eosin x40)

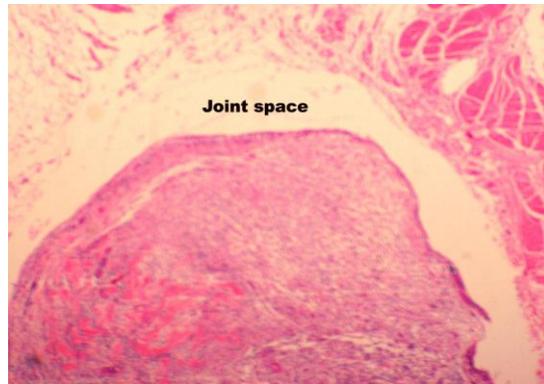


Figure 6. Cystic spaces rimmed by tumour tissue (Haematoxylin and eosin x10)

Cytogenetics play a major role in the accurate diagnosis of synovial sarcoma. Commonly a balanced reciprocal translocation t(X;18) (p11.2;q11.2) is consistently found in over 90% of synovial sarcomas.⁸ The SYT/SSX fusion messenger RNA can be detected by reverse transcriptase polymerase chain reaction, or FISH and frozen or paraffin embedded tissue may be used.¹ This is the method of choice for diagnosis of difficult cases, especially when there is equivocal immunostaining.

Local recurrence and metastases to lung and lymph nodes occur. The tendency for nodal metastases is higher than for other sarcomas.² Small tumours (<5 cm), young age at diagnosis, and distally located tumours bear a better prognosis.

Apart from diagnosing and typing the tumour, the histopathologist should also comment on the mitotic activity, presence of necrosis, the presence of cellular features such rhabdoid change, the status of the excision margins, and DNA ploidy, all of which affect the prognosis.² Recent studies suggest that expression of the cancer associated cell membrane glycoprotein 'dysadherin' may be a significant prognostic factor.⁹

References

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