Solitary fibrous tumour: a case series showing the radiological appearances and spectrum of disease

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Learning Objectives

After viewing this presentation, the reader should be able to:

1. Describe the typical radiological appearance of solitary fibrous tumour, so that in clinical practice the diagnosis can be suggested.
2. Understand the behaviour of solitary fibrous tumours.
Background

Solitary fibrous tumours (SFTs) are uncommon mesenchymal tumours with no known associations or causative factors, that have been reported to occur in almost all tissues of the body, although are most commonly found to arise from the subcutaneous and deep soft tissues, pleura and dura [1]. Extra pleural SFTs typically present as lumps, which cause pain or other symptoms when compressing surrounding structures. [2]. Many SFTs are also found incidentally, particularly in the case of pleural tumours found on chest-x-rays.

Solitary fibrous tumours and haemangiopericytomases (HPCs) were originally described as distinct tumour entities [3,4]. Subsequently, it was found histologically that these two tumours are both of mesenchymal cell origin, leading to reclassification of many HPCs as SFTs [5]. The current consensus is that the term HPC is reserved for tumours arising from the dura and sinonasal cavity with the remainder termed SFTs [6].

Macroscopically, SFTs are usually well circumscribed, often partially encapsulated lesions, which when sectioned are often multinodular, white and firm. They rarely contain calcification. The lipomatous subtype may contain macroscopic fat. They may contain haemorrhagic foci, and 10% may have areas of necrosis and infiltrative margins. Microscopically they are characterised by the "patternless pattern" of spindle cells separated by a variable amount of collagen. Also present is the haemangiopericytomatous 'stag horn' branching pattern of vessels [7]. CD34 immunostaining is important to distinguish the tumour from several differential diagnoses including synovial sarcoma [6].

Surgical excision is the mainstay of treatment for SFTs, however the surgical technique may be difficult due to tumour vascularity, and as such, there is a role for pre-operative endovascular tumour embolisation. Whilst there are relatively low rates of recurrence and metastasis if the surgical resection margins are clear, 10-15% will however recur locally or metastasise [6,8,9]. Whilst poor histological prognostic factors have been proposed such as tumour size >10 cm, presence of necrosis, cellular atypia and high mitotic rates, aggressive behaviours cannot reliably be predicted by either histological grade or findings, or radiological findings, and as such, long term follow-up is recommended for all tumours [6,7,10]. Metastases occur haematogenously with preference for the lungs and liver and lymphatic spread is uncommon [7,9].
Imaging Findings OR Procedure Details

This case series illustrates imaging of six cases of histologically proven SFT from patients who presented to a single tertiary sarcoma referral centre in Sydney, Australia. Ethics approval was obtained from the Hospital Ethics Board for access to, and use of patient records, imaging and pathology results. The series will focus on soft tissue SFT (three cases), but also show cases arising in the dura and pleura to show typical features. An unusual case of SFT of bone will also be shown.

SFTs present on imaging as discrete, well circumscribed, soft tissue masses Fig. 1 on page 6, Fig. 2 on page 6.

They generally cause mass effect with displacement rather than infiltration of the surrounding structures, although uncommonly can be infiltrative Fig. 3 on page 7, Fig. 4 on page 8.

SFTs are highly vascular tumours, and in many cases, prominent peri- and intralesional vessels can be identified on ultrasound, CT, MRI and angiography Fig. 5 on page 9, Fig. 6 on page 10. Flow voids can be seen within the lesions on MRI Fig. 7 on page 11. Given their vascularity, preoperative endovascular tumour embolisation is often performed Fig. 8 on page 12. In the case of the T1 vertebral body tumour, embolisation was not performed due to risk of embolising the spinal cord supply. The resection procedure was abandoned intra-operatively due to multiple litres of blood loss requiring activation of the mass transfusion protocol.

On MRI, lesions are generally T2 hyperintense to muscle, with little oedema in adjacent soft tissues seen on fat suppressed T2 weighted sequences. T2 signal of the mass can be homogenous or heterogeneous Fig. 9 on page 13, Fig. 10 on page 13.

SFTs demonstrate T1 hypointense signal Fig. 11 on page 14.

Enhancement on MRI or CT is usually homogeneous but lesions can demonstrate peripheral enhancement if large and/or if there is central necrosis Fig. 11 on page 14, Fig. 12 on page 15, Fig. 13 on page 16.

On CT SFTs can be seen to rarely contain calcification, and the lipomatous subtypes contain macroscopic areas of fat. Our cases did not show either calcification or fat. Cystic spaces are not commonly described. However, of our cases, the chest wall tumour demonstrated this feature Fig. 14 on page 17.
On PET-CT only mild increased glucose metabolism is usually seen in SFTs, suggestive of a low grade tumour Fig. 15 on page 18, Fig. 16 on page 18.

Most tumours behave in a benign way and resection is curative. Prognosis is not always predicted by the radiological or histological appearance. Complete resection is the best predictor of cure. While some tumours may show histological features of malignancy such as infiltrative margin, high mitotic rate, hypercellularity and cytological atypia, the absence of these features does not always preclude local recurrence or metastasis. Typical histological features are shown Fig. 17 on page 19.

Complications from solitary fibrous tumours are usually based on the tumour site. For example, the T1 vertebral body tumour demonstrated epidural extension, with spinal cord compression and cord oedema Fig. 18 on page 19. Despite this, the patient presented with few symptoms, and only a semi-urgent decompression laminectomy was undertaken.

When lesions act malignant they can present with local recurrence or distant spread Fig. 19 on page 20, Fig. 20 on page 21, Fig. 21 on page 22.

These lesions have a differential diagnosis. The differential diagnosis includes lymphoma, vascular malformation and sarcoma. In the chest wall case, schwannoma could be suggested and in the dural case, meningioma.

A summary of the findings of our six cases is provided Table 1 on page 23.
Fig. 1: Right buttock tumour. Axial proton density sequence shows a well circumscribed tumour with a smooth outline.

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**Fig. 2:** Left chest wall extra pleural tumour. Axial contrast enhanced CT scan shows a bilobed tumour with smooth contours.

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Fig. 3: Left ischiorectal fossa tumour. Axial proton density sequence shows displacement without infiltration of adjacent organs: the rectum is displaced to the right, prostate anteriorly and left obturator internus muscle to the left.

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**Fig. 4:** T1 vertebral body tumour. A) Sagittal T1 weighted sequence shows the inferior extension of the epidural spread and associated infiltration of the posterior T2 vertebral body B) Axial T2 weighted sequence shows a mixed pattern of growth. There is epidural tumour spread and displacement of the cord to the left. Anteriorly there is breach of the vertebral body cortex and infiltration of the longus colli muscles. C) Axial CT scan shows smooth scalloping of the bony margin and sclerosis suggestive of a lesion which has grown slowly and been present for some time, consistent with an intermediate aggressive lesion.

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Fig. 5: Right buttock tumour. A) Axial proton density sequence immediately superior to the tumour shows large associated vessels. B) Doppler ultrasound shows internal vascularity.

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**Fig. 6:** Left chest wall tumour. Diagnostic selective left subscapular artery angiography shows internal vascularity.

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Fig. 7: Left ischiorectal fossa tumour. Coronal T2 weighted sequence shows prominent intra- and perilesional vessels.

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Fig. 8: Left ischiorectal fossa tumour. A) Arterial and B) Venous phase diagnostic angiography showing large feeding and draining vessels. C) Angiography post coil embolisation of the three main feeding vessels to the tumour.

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Fig. 9: Left ischiorectal fossa tumour. A) Axial fat suppressed T2 weighted sequence shows a heterogenous T2 hyperintense mass and lack of surrounding reactive tissue change or oedema. B) Coronal T2 weighted sequence shows heterogenous T2 hyperintense mass. The rectum is displaced to the right.

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Fig. 10: Left chest wall tumour. Axial fat suppressed T2 weighted sequence shows hyperintense, homogenous signal tumour without surrounding oedema.

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**Fig. 11:** Right buttock tumour. A) Axial T1 weighted sequence shows a T1 hypointense mass. B) Axial post contrast fat suppressed T1 weighted sequence shows homogenous enhancement.

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Fig. 12: T1 vertebral body tumour. (A) Sagittal T1 weighted and (B) Sagittal post contrast T1 weighted sequences show homogeneous contrast enhancement.

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**Fig. 13:** Left ischiorectal fossa tumour. A) Sagittal T1 weighted sequence and B) Sagittal post contrast fat suppressed T1 weighted sequence shows heterogenous enhancement with low enhancement centrally, with a histological correlation of central necrosis. Note also the prominent vessels anterior and inferior to the mass. C) Macroscopic photograph of the resected tumour specimen. The yellow areas correspond to necrosis, which may in part be secondary to embolisation, although necrosis was present on the pre-embolisation biopsy.

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**Fig. 14:** Left chest wall tumour. A) Axial contrast enhanced CT chest shows relatively homogenous enhancement with a few small non-enhancing foci. B) Axial fat suppressed T2 weighted sequence shows these foci to be T2 hyperintense and cystic, and not due to fat.

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**Fig. 15:** Left ischiorectal fossa tumour. Coronal PET-CT shows peripheral mild glucose uptake. Histological correlation was of central necrosis.

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**Fig. 16:** Multiple right pleural tumours. A) PA chest radiograph shows a spiculated right upper zone opacity and subtle opacities medially at the right base. B) and C) Coronal PET-CT chest series with marked uptake of a RUL lesion (post excision histology: primary lung cancer - adenocarcinoma), and very mild uptake of several right pleural lesions (post excision histology: all SFTs). This patient had a background of previous removal of a pleural solitary fibrous tumour.

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**Fig. 17:** Left ischiorectal fossa tumour. A) Histology on biopsy shows a patternless arrangement of uniform spindle cells with intervening collagen and a branching (haemangiopericytomatous) vascular pattern. B) The tumour demonstrates strong diffuse positive staining in a CD34 immunostain.

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**Fig. 18:** T1 vertebral body tumour. A) Sagittal fat suppressed T2 weighted sequence demonstrates cord oedema. B) Coronal post contrast T1 weighted sequence shows spinal cord compression and cord displacement to the left by the tumour.

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**Fig. 19:** Haemangiopericytoma of the left anterior cranial fossa. A) Axial and B) Coronal post contrast fat suppressed T1 weighted sequences show dural recurrence 6 years following primary tumour resection. There is homogenous enhancement and lobular tumour outline, with adjacent encephalomalacia due to surgery.

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**Fig. 20:** Haemangiopericytoma of the left anterior cranial fossa. A) Sagittal T1 weighted and B) Sagittal post contrast T1 weighted sequences show homogenous enhancement of a dural deposit 12 years post primary tumour resection.

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Fig. 21: Haemangiopericytoma of the left anterior cranial fossa. Coronal contrast enhanced CT chest and abdomen 14 years post primary tumour resection, showing typical appearance of pulmonary and hepatic metastases: enhancing lesions which may contain central necrosis.

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Table 1: Summary of imaging findings of the six cases presented.

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<table>
<thead>
<tr>
<th>Location</th>
<th>Size (cm)</th>
<th>CT Appearance</th>
<th>MR Appearance</th>
<th>Margin</th>
<th>Surrounding Oedema</th>
<th>Prominent Vessels</th>
<th>PET CT Glucose uptake</th>
<th>Pre-operative angiography findings</th>
<th>Metastases/Recurrence</th>
<th>Surgical Margin</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chest wall, extra pleural</td>
<td>4.6 x 4.4 x 5.2cm</td>
<td>Heterogeneous enhancement</td>
<td>T1: hypointense, T2: hyperintense, T1+: Moderate enhancement, less enhancement at its deep aspect</td>
<td>Well circumscribed</td>
<td>Nil</td>
<td>Per-lesional</td>
<td>Mild-moderate</td>
<td>Hypervascular mass, completely devascularised by embolisation procedure</td>
<td>No</td>
<td>Clear</td>
</tr>
<tr>
<td>T1 vertebra</td>
<td>Length of epidural component 5.5cm</td>
<td>Homogeneous density, arises from and replaces T1 with spread to epidural space and spine T3.</td>
<td>T1: Low T2: Mildly high T1+: Homogeneous</td>
<td>Mixed pattern with well circumscribed and infiltrative areas</td>
<td>Nil</td>
<td>Nil</td>
<td>Mild</td>
<td>Not performed due to location and relationship to cord</td>
<td>No</td>
<td>Unable to be resected due to heavy intraoperative blood loss</td>
</tr>
<tr>
<td>Ischiatic fossa</td>
<td>8.6 x 6.2 x 7.9cm</td>
<td>-</td>
<td>T1: heterogeneous T2: hypointense, heterogeneous T1+C: Peripheral enhancement</td>
<td>Well circumscribed</td>
<td>Nil</td>
<td>Intra- and peri-lesional</td>
<td>Moderate peripheral uptake, low uptake centrally</td>
<td>Hypervascular tumour</td>
<td>Left ilium bone lesion - indeterminant</td>
<td>Clear</td>
</tr>
<tr>
<td>Bullock</td>
<td>8.9 x 6.7 x 8.6cm</td>
<td>-</td>
<td>T1: hypointense T2: hyperintense T1+: Homogeneous enhancement</td>
<td>Well circumscribed</td>
<td>Nil</td>
<td>Small peri- and intra-lesion vessels</td>
<td>Mild-moderate</td>
<td>Not performed</td>
<td>No</td>
<td></td>
</tr>
<tr>
<td>Intracranial dural HPC: Local Recurrence x2</td>
<td>-</td>
<td>-</td>
<td>T1: hypointense, T2: hypointense, heterogeneous enhancement</td>
<td>Well circumscribed</td>
<td>Minimal, Background exceptional acia from surgery</td>
<td>Nil</td>
<td>2nd recurrence hypometabolic</td>
<td>Not performed</td>
<td>Local recurrence, metastasis (lung, liver, intradural space)</td>
<td>Dural infiltration at 1st recurrence. Margin or primary tumour not described in report</td>
</tr>
<tr>
<td>Intracranial dural HPC: Metastases – 1,2,3 intradural, liver, lung</td>
<td>1.2/3 intradural: 1.3 x 2.0 x 2.1cm L/size: up to 8.6 x 6.8 x 6.7cm L/size: up to 9.6 x 5.6 x 6.7cm</td>
<td>Lung and liver: Heterogeneous, Peripheral enhancement, central low attenuation</td>
<td>Intracranial T1: Hypointense T2: Hypointense T1+: Heterogeneous</td>
<td>All well circumscribed</td>
<td>Nil</td>
<td>Lung and liver: mild to moderate</td>
<td>Not performed</td>
<td>As above</td>
<td>Dura: Extends to margin. Lung and liver: not resected</td>
<td></td>
</tr>
<tr>
<td>Pons x 5 (RUL, RM, x 2; RUL, x 2)</td>
<td>Largest lesion measures 1.3cm diameter</td>
<td>-</td>
<td>All well circumscribed</td>
<td>Nil</td>
<td>Mild</td>
<td>Moderate uptake in medial primary lung cancer</td>
<td>Not performed</td>
<td>Pleural recurrence</td>
<td>1 lesion abuts margin. 4 lesions clear</td>
<td></td>
</tr>
</tbody>
</table>
Conclusion

Solitary fibrous tumours are uncommon mesenchymal tumours occurring in almost any tissue of the body, with a predisposition for the pleura, soft tissues and dura. They are well circumscribed, vascular, enhancing soft tissue lesions with large associated vessels, and on MRI have characteristic intra and perilesional flow voids. They have mild uptake on PET CT in keeping with the usual histological low to intermediate grade.

Management of these cases may be assisted by including SFT within the differential diagnosis should the typical features be seen. This can assist appropriate referral to specialist surgeons, surgical planning where there is a role for pre-operative tumour embolization, and decisions regarding surveillance and follow up.

Whilst usually benign, SFTs can recur locally or metastasise. Behaviour cannot be reliably predicted on imaging or histological features, and as such long term follow-up is recommended.
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