An unusual abdominal mass in a young male

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Abstract

Solitary fibrous tumour is a rare spindle cell neoplasm of uncertain histogenesis and unpredictable biological behaviour, which was first described in the pleura and subsequently in many extrapleural locations. The clinical, radiologic and histologic features of a case of pelvic extraperitoneal solitary fibrous tumour in a young male are presented.

Keywords

Solitary fibrous tumour; extraperitoneal pelvic tumour.

Introduction

Solitary fibrous tumours (SFTs) are mesenchymal and arise from primitive fibroblast-like cells in connective tissue[1]. Klemperer and Rabin first described SFTs as a distinctive entity in 1931 [2,3]. They have been reported in a wide range of sites outside the thoracic cavity including abdominal and retroperitoneal sites[4–6], head and neck[7,8] and soft tissues[1,9,10]. Extrathoracic solitary fibrous tumours (ESFTs) are indolent and less frequently found than intrathoracic SFT tumours[11]. Among the extrathoracic sites, the orbits and the soft tissues of the extremities (muscles and subcutaneous tissues) are the most commonly reported sites[1,10]. We present a case of pelvic SFT in a young male.

Case report

A 28-year-old man presented with a 2-week history of flu-like symptoms with a high temperature and swollen cervical lymph nodes. There was also a few days history of lower abdominal pain which became continuous, in contrast to his other symptoms which resolved later. This patient was otherwise fit and well with no significant past medical or surgical problems. There was no family history of malignant disease. On abdominal examination, a solid suprapubic mass was revealed, extending upwards as far as the umbilicus and measuring around 15 cm in size. The rest of the abdomen was clear to palpation.

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A computed tomography (CT) scan was ordered and this revealed a large anterior heterogeneously enhancing soft tissue mass that extended superiorly to above the umbilicus and inferiorly down to lie in front of the bladder. The mass measured a maximum diameter of $15 \times 8 \text{ cm}$ with a blood supply arising from the right femoral and iliac vessels. The tumour appeared separate from the bladder and bowel. There was no evidence of metastatic disease. It was thought that the primary tumour might be a sarcoma and percutaneous biopsy might therefore be accompanied by the risk of seeding, so none was performed. A positron emission tomography (PET) scan showed that the pelvic mass exhibited mild fluorodeoxyglucose (FDG) uptake with no definite metastatic disease.

**Surgery**

The patient underwent elective excision of the mass. On inspection the tumour appeared to be completely encapsulated and extraperitoneal. It was arising from the right side of the pelvis, deep to the inguinal ring. It was vascular, with an extensive blood supply from the iliac vessels as noted in the pre-operative CT scan. There was no invasion of any pelvic or abdominal organ and it was only attached by the vascular pedicle, which made excision relatively straightforward. He made a very good post-operative recovery with no complications. Follow-up was arranged at 3-month intervals for the first year, 6-monthly intervals for the second year and then once a year subsequently. Periodic CT scans to confirm the absence of any recurrent disease were arranged.

**Pathology**

Macroscopically there was an encapsulated well-circumscribed mass measuring $160 \times 150 \times 80 \text{ mm}$ and weighing 1.3 kg. Microscopic sections revealed a partially encapsulated large tumour mass composed of small spindle- and stellate-shaped cells with hyperchromatic...
nuclei predominantly arranged haphazardly in a variably fibrous/collagenous stroma. There was marked variation in cellularity ranging from solid sheets of tumour cells to hypocellular myxoid areas. The tumour was highly vascular. Focally there were collections of large pleomorphic multinucleate tumour cells. There was no evidence of necrosis or proliferative activity.

Immunohistochemistry showed that the tumour cells were diffusely positive for CD34, Bcl-2 and CD99, and negative for epithelial membrane antigen, desmin, Calretinin, WT1 and cytokeratin. The combined morphological and immunohistochemical features were those of an SFT.

Discussion

SFT is a clearly outlined mass ranging in size from 1 to 30 cm. They are found with similar frequency in both genders, mostly in the 5th to 7th decades of life, with an age range of 5 to 92 years\textsuperscript{[12,13]}. When extrapleural, the tumour is most commonly found in the orbits and the extremities\textsuperscript{[1]}.

SFTs are a subtype of spindle cell lesions, which can be difficult to diagnose due to their rarity and also their homogeneity with other spindle cell subgroups. The pathologist can utilize histology to make an accurate diagnosis despite the wide histological variability. A large number of published papers suggest that SFTs do not have any particular symptoms and that these are dependent on local mass effects. Presentation of the mass can be from 1 month to 10 years after onset of symptoms\textsuperscript{[1,14]}. Some cases are diagnosed when the patient presents with hypoglycaemia as the tumour cells of SFT secrete an insulin-like growth factor II (IGF-II). The hypoglycaemia resolves after resection of the tumour\textsuperscript{[1,15,16]}.

Most SFTs are well-defined, yellow to tan coloured, regularly lobulated masses with an elastic feel on examination and surrounded by a thin clear membrane. A common histological feature of any SFT is a combination of 3 components: fibrous tissue, cellular structures, and highly vascularized sections that contain an abundance of dilated small- to medium-sized vessels. These are organized around large or small quantities of tumour cells, creating the commonly known patternless configuration\textsuperscript{[17]}.

SFTs can be categorized into fibrous and cellular variants based on the quantity and distribution of the organized fibrous stroma. The most common variant of SFT is fibrous SFT with abundant fibrous stroma. The less common cellular SFT variant has a more uniform appearance on histology with a moderate to high cellularity, numerous intermixed haemangio-pericytoma-like blood vessels, and only lightly organized fibrosis\textsuperscript{[1]}.

SFTs can also be grouped into 2 groups histologically: benign or malignant. Malignancy can be identified on histology by increased cellularity, nuclear pleomorphism, cellular atypia, necrosis, haemorrhage and 4 or more mitoses per 10 high-power microscopic fields. Larger tumours are more likely to be malignant, especially when larger than 10 cm in diameter. However, malignancy cannot be excluded by a size less than 10 cm alone\textsuperscript{[1]}. Extrapleural SFT has
unpredictable characteristics, with the above-mentioned malignant histological behaviour not being fully reliable. Some SFTs with a benign microscopic appearance behave aggressively while others with a malignant appearance do not. Therefore, all patients with extrapleural SFT require complete surgical resection and long-term follow-up\(^1\)

The study by Cranshaw et al.\(^{11}\) suggested that SFTs should be treated in the same way as other high-grade soft tissue tumours. He reported that extrathoracic SFTs with atypical or malignant characteristics on histology have a higher potential for malignancy and therefore a worse prognosis than previously reported. Vallat-Decouvelaere et al.\(^{19}\) described a subgroup of atypical and malignant tumours that behaved in a more aggressive fashion or had atypical histology and represented 11% of their series. The study by Gold et al.\(^{10}\) showed that ESFTs had a more aggressive clinical behaviour with a higher rate of recurrence compared with intrathoracic SFTs.

Pelvic SFTs show variable and non-specific findings on radiology with internal calcification on some radiographs. How the tumour appears on imaging depends on the histologic components in the tumour. This includes fibrous tissue, cellular components and highly vascularized areas consisting of numerous closely packed small- to medium-sized blood vessels\(^1\).

The available data indicate that SFTs have a variable and non-specific appearance on ultrasonography, although the internal vascularity is usually evident, particularly on colour-coded duplex scanning. CT and magnetic resonance (MR) imaging of pelvic SFTs often show stand-alone, well-defined, ovoid or rounded masses. SFTs grow slowly, developing into large masses and often displacing or compressing adjacent tissues and organs, thus giving rise to the clinical symptoms. This, with variable enhancement on CT and MR images may suggest the diagnosis of SFT\(^{1,17}\).

MR imaging returns hypointense or isointense T1 signals compared with muscle in most tumours (80%). The low T2 signal intensity in the dense mature fibrous tissue helps to identify the fibrous content of tumours, which are typical and helpful in T2-weighted imaging diagnosis. Fuksbrumer et al. found that the signal intensity in T2-weighted images was variable, and this is mainly due to the differences in the quantity of collagen and fibroblasts, and on the presence of degeneration\(^{1,16–18}\). It has also been shown that haemorrhage, cystic degeneration and necrosis can be seen in the larger tumours and the malignant tumours. However, this is uncommon and occurs in less than 5% of cases. Other investigations used to identify the origin of the low T2 signal intensity foci (such as haemosiderin and calcification) in the tumour include gradient echo MR images, T1-weighted MR images and CT imaging. Intermediate-to-high signal intensity on T2-weighted images due to increased oedema and vascularity can indicate malignant areas within the tumours\(^{1,17}\). Lesions with a high fibrous content and a poor vascular network may demonstrate progressive contrast enhancement during the arterial and portal phases that become marked on delayed images\(^{18}\).

There is only a limited literature available on PET/CT findings in both malignant and benign SFTs. Increased cellularity in both benign and malignant SFTs is the reason for the increased FDG uptake, regardless of the benign or malignant nature of the disease. However radiotracer may not be taken up in some lesions\(^{1,11}\). The most important differential diagnoses of SFT are fibromatosis, fibrous histiocytoma, fibrosarcoma, synovial sarcoma, and scirrhous metastatic carcinoma\(^{1,20}\).

A useful non-specific but differentiating imaging indicator of SFTs that can guide the radiologist is the presence of a large collateral feeding vessel. The feeding vessels can be seen with CT, MR imaging, ultrasonography, and angiography. Alveolar soft tissue sarcoma may contain similar vessels. A percutaneous core biopsy is required to determine the correct diagnosis. SFTs should be dealt with in specialist tertiary referral centres. This is because of the vascularity of the tumour and the presence of collateral vessels, which could make excision of the tumour challenging. Tumour excision may be preceded by embolization if necessary, to limit the vascularity\(^{20}\).

Immunohistochemical studies can also be used to differentiate between SFTs and other spindle cell neoplasms. SFT tumour expresses CD34, with descriptions of positivity to Bcl-2, type HI insulin-like growth factor, and neural crest and muscle markers\(^{1,14,18,21}\).

Surgery is the most effective treatment for localized disease, sometimes combined with radiotherapy in the treatment of those tumours graduating towards soft tissue sarcomas\(^{1,15}\). Metastases are usually blood borne, and are found in the lungs, liver, adrenal glands, bones, brain, muscles, and the gastrointestinal tract. Chemotherapy is used as palliation in advanced disease, and sometimes also preoperatively to decrease tumour size and eradicate micro-metastases, making meaningful surgery possible. The role of chemotherapy as adjuvant treatment in localized disease is not agreed\(^{1,22}\).
Complete wide local excision with negative margins is the most important indicator of a good prognosis. Most extrapleural SFTs, regardless of histology, will behave benignly as long as wide surgical excision is performed with a margin of 1–2 cm. Positive margins contribute to up to 40% of local recurrences and 75% of metastatic disease.

Disease progression does not correspond with the histopathologic findings in some patients. This necessitates the need for long-term follow-up for all patients[20]. Most recurrences occur within the first 2 years, with a few still occurring up 17 years after resection. Relapse is more likely if there is deterioration in the histologic indictors of malignancy and, of course, with haematogenous metastasis. Hence, 6-month radiologic follow-up imaging during the initial 2 years after the resection and yearly thereafter seems prudent[1,23]. Our patient will have long-term follow-up, mainly because of the large size of his tumour. He will be seen at 3-monthly intervals in the first year, 6-monthly intervals in the second year and then once a year thereafter.

Modern molecular diagnostic tools are making it possible to improve differentiation between the different tumour entities and the identification of particular molecular abnormalities may make it possible to introduce focussed treatment modalities in the future[1,22].

**Conclusion**

SFTs are found in pleura and many different extrapleural locations. SFT should be considered in the differential diagnosis of extrapleural lesions in which MR imaging suggests a fibrous content. SFT is diagnosed on microscopic examination of the tissue by means of immunohistochemistry. CT and MR imaging are useful diagnostic tools that may show a well outlined ovoid or rounded mass with variable enhancement on images. CT and other radiology investigations are used for identifying the location of tumour, and guiding pre-operative planning, staging, spread of disease and follow-up. Surgery is the only cure if the tumour has not spread and patients require long-term follow-up due to the unpredictability of the disease[1,17,19,20].

**Teaching points**

1. SFTs are spindle cell neoplasms of uncertain histogenesis and unpredictable biological behaviour.
2. SFTs have no gender or age predilection.
3. Microscopic examination of the tissue is the only reliable diagnostic modality for SFT.
4. Radiological findings of pelvic SFTs are variable and nonspecific. However, CT and other diagnostic tools are essential for pre-operative planning, staging of disease, excluding distant metastasis and follow-up.
5. Well-defined ovoid or rounded mass with hypointense signals on MR T2-weighted images and variable enhancement on CT and MR images may suggest the diagnosis of SFTs. PET can be used to rule out metastatic disease.
6. Surgery is the only cure and only if the tumour has not spread.
7. The most important factor influencing clinical outcome is complete wide local excision with negative margins.
8. Histologic signs of malignancy include increased cellularity, nuclear pleomorphism, cellular atypia, high mitotic counts per power field, haemorrhage and necrosis. The rate of malignancy increases with size.
9. Long-term follow-up is essential, because the outcome is unpredictable.

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**References**