CASE REPORT

A solitary fibrous tumour mimicking an aggressive angiomyxoma/liposarcoma

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SUMMARY

We present a case of a solitary fibrous tumour (SFT) resembling an aggressive angiomyxoma/liposarcoma on radiological imaging, causing significant diagnostic challenges preoperatively. A 76-year-old male was incidentally found to have a large pelvic mass on a CT scan. Further evaluation with an MRI scan confirmed a presacral mass containing fat and soft tissue components. It was inseparable from the sacrococcygeal spine, rectal serosa and the posterior wall of the urinary bladder, but no evidence of invasion was seen. A prominent vascular pedicle arising from the epidural vasculature was also noted. Differentials discussed at the multidisciplinary tumour board were an aggressive angiomyxoma versus a liposarcoma. The patient underwent wide resection of the pelvic tumour, anterior resection and end colostomy. Intraoperatively, a large 20 cm pelvic mass involving the sigmoid mesocolon and presacral fascia was found. Final histology reported an SFT with extensive adipocytic metaplasia.

BACKGROUND

Solitary fibrous tumours (SFTs) are rare spindle cell neoplasms with fibroblastic differentiation. First described to originate from the pleura, this tumour has now been reported to occur throughout the body.^{1,2} Histological analysis with immunohistochemistry is usually necessary for accurate diagnosis. A characteristic inversion of the long arm of chromosome-12 producing the NAB2-STAT6 fusion protein has been identified to occur within SFTs.³ This tumour generally has an indolent course with low risks of recurrence and metastasis, and first-line treatment is surgical resection of the tumour with clear margins.

Aggressive angiomyxomas are rare, slow-growing tumours of mesenchymal origin and frequently arises from the connective tissues of the perineum and pelvis.⁴ Due to its rarity, it is often misdiagnosed, and patients are often treated surgically as for another pathology. Surgical resection with clear margins has been advocated as the treatment of choice. Disease recurrences can range from 28% to 72% of patients.^{5,6}

Liposarcoma is the most common soft tissue sarcoma in adults and can be classified into the well-differentiated, myxoid, round cell, pleomorphic and dedifferentiated types.⁷ As small liposarcomas are generally asymptomatic, they can grow to become fairly sizeable before presenting as palpable masses or with compressive symptoms.

Treatment of choice is surgical resection with clear margins.

Differentiating between these three tumours on imaging scans can be challenging because these tumours share several similar imaging characteristics—all three tumours tend to appear as discrete masses that displace adjacent structures, and they can demonstrate high signal intensity on T2-weighted MRI.

We present a case of an SFT mimicking an aggressive angiomyxoma/liposarcoma in a 76-year-old male patient, and discuss the diagnostic challenges involved, review the pathological and radiological features of each tumour and provide a review of the literature.

CASE PRESENTATION

A 76-year-old male with past medical conditions of diabetes mellitus, hypertension, hyperlipidaemia and coronary artery bypass surgery more than 10 years ago presented to our institution with acute abdominal pain and vomiting of 2 days duration, and was diagnosed with acute gangrenous cholecystitis. He underwent emergency open cholecystectomy and made good recovery from his surgery.

INVESTIGATIONS

During the admission, an incidental 13.7×9.4×13.5 cm presacral mass with lobulated margins and mixed fat and soft tissue components was noted on the preoperative CT scan of his abdomen and pelvis (figure 1). Further evaluation with an MRI was performed (figure 2) that showed a large presacral mass measuring 13.5×9.7×21.7 cm containing mostly fat with soft tissue component.

Figure 1 CT scan images showing tumour arising from the pelvis.

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Figure 2 MRI scan images.

The mass was inseparable from the sacrococcygeal spine, rectal serosa and the posterior wall of the urinary bladder, but there was no evidence of invasion. Following administration of intravenous gadopentetate dimeglumine contrast, there was enhancement of the soft tissue component of the large mass. A vascular pedicle communicating with the epidural vasculature was also noted. No enlarged pelvic lymph nodes were detected.

A CT scan of his thorax revealed two subcentimetre right upper lobe pulmonary nodules that were indeterminate for metastases. Serum carcinoembryonic antigen level measured 2.1 µg/L, which was within normal limits.

The patient did not have symptoms arising from the mass except for increased urinary frequency, needing to pass urine almost hourly. He did not notice any constitutional symptoms such as loss of appetite or weight, and his bowel habits remained normal.

DIFFERENTIAL DIAGNOSIS

The case was reviewed at the multidisciplinary tumour board (MDTB) and the differential diagnoses discussed included a liposarcoma and aggressive angiomyxoma. Recommendation was for en-bloc resection with clear margins. Percutaneous or transenteric biopsy prior to surgery carried the risks of tumour seeding and procedural complications such as bleeding, infection and bowel perforation. It would have made minimal change to the surgical management in this case, as the mass was deemed to be resectable on imaging.

TREATMENT

The patient was agreeable for surgery, and he underwent an elective laparotomy. Intraoperatively, a large 20 cm pelvic mass involving the sigmoid mesocolon and presacral fascia was found. The mass filled the entire pelvis, and caused bilateral ureters to be displaced bilaterally. However, both ureters and the bladder were not involved. No peritoneal nodules were found. A wide resection of the pelvic tumour, with en-bloc low anterior resection was performed. The estimated blood loss was 9 200 mL, and 11 units of pack cells, 5 units of fresh frozen plasma, 3 units of platelets and 2 units of cryoprecipitate were transfused during the surgery. The entire procedure took 6 hours and 55 min. The decision for an end colostomy

was made in view of the prolonged surgery, significant blood loss and intraoperative blood transfusion. He was transferred to the intermediate care area postoperatively.

The patient was monitored closely with hourly vitals and strict intake and output measurements. His nasogastric tube was put on low intermittent suction with 4-hourly aspiration. He received chest physiotherapy and incentive spirometry, and calf compressors were applied. His postoperative haemoglobin level was 11.8 g/dL, and no further blood transfusion was required immediately post-operation. His stoma remained healthy and started functioning on postoperative day (POD) 4, and he was started on clear feeds on POD 5, full feeds on POD 6 and diet on POD 7. He was transferred to general ward on POD 7.

On POD 9, routine blood checks revealed elevated inflammatory markers, and a purulent discharge was noted from his laparotomy wound. The patient also started developing symptoms suggestive of ileus. A CT scan of his abdomen and pelvis was performed, and this revealed a 6.4 cm collection in the presacral region containing fluid and gas locules. A second rim-enhancing collection was noted in the anterior abdomen underneath the midline laparotomy scar, extending towards the left flank, measuring 16×1 cm. An image-guided percutaneous drainage of the collection was performed, and the fluid was sent for microscopy and culture. This grew *Enterococcus hirae*, *Klebsiella pneumonia* and *Escherichia coli*. Infectious diseases physicians were consulted, and the patient was started on a course of intravenous imipenem for a total of 4 weeks. The patient was also started on total parenteral nutrition for 3 days until his ileus resolved. He remained clinically well and afebrile thereafter, and was discharged home on POD 43 after completing the course of antibiotics. His wound had healed by that time.

Unfortunately, the patient was readmitted 1 week later due to lower limb swelling and caregiver stress. Blood investigations revealed that hypoalbuminaemia was the likely cause of his lower limb swelling, and his nutrition was optimised. He was subsequently transferred to a community hospital for rehabilitation.

OUTCOME AND FOLLOW-UP

The final histology on the resected specimen reported an SFT with extensive adipocytic metaplasia, measuring up to 21.0 cm. No invasion into the adherent colon, cytological atypia or necrosis was seen. The mitotic count was 1/30 high power fields (HPFs) (figures 3 and 4). The spindle cells showed immunopositivity for CD34 and bcl2. Fluorescent in-situ hybridization (FISH) test for MDM2 amplification was negative, effectively ruling out a dedifferentiated liposarcoma. The histology was discussed at the MDTB, and close clinicoradiological monitoring was advised. Radiotherapy was considered but the risk:benefit ratio was deemed to be unfavourable, due to the tumour's location in the pelvis.

The patient was last reviewed in the outpatient clinic 12 months after surgery. He remained well and was putting on weight and had a good appetite. A surveillance CT scan that was done 2 weeks prior to the clinic visit revealed no radiological evidence of recurrence and the lung nodules remained stable in size. Reversal of his colostomy was discussed with the patient but he was not keen to undergo surgery at the present.

DISCUSSION

While SFTs are thought to be generally benign, about 15% of tumours recur or metastasise. Large tumours ≥15 cm, ≥4

Figure 3 Spindle cell proliferation admixed with adipocytes (low power).

mitoses per 10 HPFs, hypercellularity, nuclear pleomorphism and areas of tumour necrosis have been associated with higher risk of malignant behaviour.¹ Histologically, SFTs are characterised by a 'pattern-less' growth pattern—uniform elongated spindle cells arranged in a haphazard pattern, with prominent branching vasculature.⁸ Immunoreactivity for CD34 has been well documented, with some tumours showing reactivity for bcl-2 protein as well.⁹ However, the behaviour of SFTs do not always correlate with histological factors, and all patients are best followed up on a long-term basis. Surgical resection with clear margins offers the best oncological outcome. Preoperative angioembolisation has been advocated by some authors to increase the chances of a successful surgery.^{10 11}

On the other hand, aggressive angiomyxomas are characterised by their high local recurrence rate. This tumour does not typically demonstrate aggressive behaviour such as local invasion or metastasis. When detected, aggressive angiomyxomas are often noted to span across the pelvic diaphragm,¹² posing significant challenges to obtaining clear surgical margins. Histologically, aggressive angiomyxomas are composed of spindle cells and abundant vascularity embedded in myxoid matrix. The tumour cells are positive for oestrogen and progesterone receptors, implying a hormonal role in the growth of these tumours.¹³ While resection with clear margins is generally advocated, several sources have reported successful application of hormonal therapy, mainly in a recurrent setting.^{14 15}

Figure 4 Spindle cell proliferation admixed with adipocytes (high power).

Liposarcomas typically arise de novo. Malignant transformation from existing benign lipoma is extremely rare. Local recurrence is common. Distant metastasis is rare except in cases of dedifferentiated liposarcoma, where distant recurrences can occur in up to 30% of cases.¹⁶ The presence of lipoblasts is critical to the histopathological diagnosis of a liposarcoma. FISH analysis for the MDM2 gene amplification is often helpful in clinching the diagnosis. Surgical resection with clear margins is the mainstay of curative treatment, and radiotherapy may be used as a possible adjunct in certain cases.¹⁷

On imaging scans, these three tumours share several similar characteristics, making it hard to differentiate between them. All three tumours tend to appear as discrete masses that displace adjacent structures. All can demonstrate high signal intensity on T2-weighted MRI—SFTs and liposarcomas due to presence of fat¹⁸ while aggressive angiomyxoma due to their myxoid matrix.¹²

Several factors could have helped to discern between the three tumours. A case series by Wignall *et al* suggested that the presence of a prominent feeding vessel could be used as a distinguishing feature for SFTs.¹⁸ In this patient, a feeding vessel arising from the epidural vasculature was reported (figure 2). This could have tilted the diagnosis more towards an SFT. On the other hand, aggressive angiomyxoma have been described to display a characteristic 'swirled' internal pattern on T2-weighted MRI after gadolinium contrast,¹⁹ which is normally absent in the other two tumours. Although liposarcomas can usually be recognised by their internal septations, the heterogeneity in appearance often increases with tumour grade.

In this patient when the scan images were reviewed preoperatively, both aggressive angiomyxoma and liposarcoma could not be ruled out. The tumour contained significant fatty component, making liposarcoma a distinct possibility. Intriguingly, the T2-weighted MRI actually showed a 'swirled' pattern (figure 2), suggestive of an aggressive angiomyxoma. With those observations, it was difficult to make a definitive call based on imaging alone. It was only after a histopathological analysis of the surgically resected specimen that the definitive diagnosis of SFT was made which, in retrospect, made the imaging features highly unusual for an SFT.

Another interesting feature of this case lies in its histological finding. In addition to the usual histological and immunohistochemistry features suggestive of an SFT, the tumour was found to have extensive adipocytic metaplasia which explained the significant fat component seen on the scan images. A fat-forming variant

Learning points

- ▶ Making an accurate diagnosis of solitary fibrous tumour based on imaging scan alone continues to be a significant challenge, especially with the wide array of possible differentials and the occasional rare variant posing added difficulties. Cases like these should be discussed at a multidisciplinary tumour board and the imaging scans reviewed by experienced radiologists.
- ▶ Preoperative biopsy for histopathological correlation may be worthwhile in selected cases, especially if the diagnosis may lead to different surgical plans.
- ▶ Good surgical margins are mandatory for the best possible oncological outcome, and all patients should be on close surveillance postoperatively, given the risk of recurrence and metastasis.

of SFT has been described in literature. Previously known as lipomatous haemangiopericytoma, few cases have been reported of this variant. This variant tends to affect middle-aged males, and the clinical course tends to be benign.⁸ The fatty components can be seen as poorly enhancing foci on CT scans while on T2-weighted MRI they appear with high signal intensity. Histologically, they resemble SFTs with the additional finding of mature, non-atypical adipocytes.¹⁸

Contributors GHCT: conception or design of the paper, acquisition, analysis and interpretation of data for the paper, drafting the manuscript. DN: conception or design of the paper, acquisition, analysis and interpretation of data for the paper, drafting the manuscript. TH: acquisition, analysis and interpretation of data for the paper, revising manuscript for content. MT: conception or design of the paper, revising manuscript for content.

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