Primary Retroperitoneal Solitary Fibrous Tumour: A Case Report

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Introduction

Solitary Fibrous tumours (SFTs) were first described in 1931 by Klemperer et al. as pleural tumours. Since then, it has been reported in many extra-pleural sites but is found to be exceedingly rare. Most of them are reported to arise from the pleura, and only 30% are of extra-pleural origins. Less than a hundred cases of Primary Retroperitoneal Solitary Fibrous Tumours have been described till now. These are rare soft-tissue sarcomas, with mesenchymal origins. The symptoms of these tumours depend on the location. The diagnosis is done mainly by imaging such as ultrasonography, Computed tomography or Magnetic resonance imaging. The standard of treatment for these tumours is by surgical excision with clear margins. The role of adjuvant chemotherapy is controversial.

Case Presentation

A 20-year-old lady presented with complaints of abdominal pain for 6 months, which was of mild intensity at first but gradually increased in intensity over the last few days before the presentation. It was present in the lower abdomen initially however progressed to generalized abdominal pain with an increase in intensity. She did not have any urinary complaints, fever, nausea or vomiting. She presented on the 3rd day of her menstrual cycle, which was of normal duration and flow in regular intervals of 28+/- 2 days. She did not give any significant medical or surgical history in the past. No significant history was found in the family either.

On examinations, she was a healthy-looking female, with no pallor, or lymphadenopathies. She had a soft, scaphoid abdomen, with a vague mass palpable in the left iliac fossa, ˜6x6cm in diameter, with ill-defined margins, non-tender, and not attached to the overlying skin. Bowel sounds were present on auscultation. Other systemic examinations were normal.

On ultrasonography, a complex heterogenous solid-cystic lesion was seen in the left adnexa measuring ˜9.9 x 6.0x 6.4 cm. MRI was then done to confirm the diagnosis, which showed an 11.7 x 8.2x4.8cm (CCxAPxT) size complex heterogenous signal intensity mass in the retroperitoneum just medial to the left psoas muscle and lateral to iliac vessels. (Figure 1) Anteriorly the mass was extending up to the anterior abdominal wall, displacing the psoas muscle laterally and iliac vessels medially. Multiple variable size irregular shape cystic areas were seen within it. Variable thickness septa and solid components were present. Multiple flow void areas were noted in the mass, suggesting marked vascularity. Which gave a differential diagnosis of retroperitoneal soft tissue sarcoma or neurogenic tumour.

So, with the provisional diagnosis of Primary Retroperitoneal Mass, the patient underwent “Laparoscopy Assisted Transperitoneal Excision of Retroperitoneal Mass”. During surgery, a large mass measuring ˜15x10cm

solid mass with 2 lobes with an irregular surface was seen with the larger lobe having cystic areas. (Figure 2) There was dense adhesion of the mass posteriorly with the psoas muscle. The visualized retroperitoneal organs were normal. There was blood loss of ˜1000ml from the part of the mass adhered to the psoas muscle. An intra-abdominal drain was placed, which was removed on the 4^{th} postoperative day. The rest of her stay in the hospital was uneventful and was sent home from the hospital on the 6^{th} postoperative day.

In her histopathology report, gross examination showed 2 large nodular bosselated encapsulated soft tissue measuring 9x6x2cm and 4.5x3x3cm were seen with a cut section showing a grey-white area with a cystic area within it. (Figure 3 (A) and (B)) On microscopic examination, mitotic figures or necrosis were not present, Tumour was composed of compact cellular and loose myxoid areas with spindle-like cells and ovoid cells coursed by round to slit-like and occasionally ramifying capillary seized vessels, (Figure 4 (A)) punctuated by variously sized hemangiopericytomatous vessels, many with discernible fibromuscular walls and cystic spaces. The morphological features which were consistent with Solitary Fibrous Tumour and margins, however, were positive for tumour.

The Immunohistochemistry showed Tumour cells positive for CD34, SMA and STAT-6 and negative for CK, S100 and desmin. (Figure 4 (B) and (C)) The Ki67 proliferation index was 10%.

The risk of metastasis according to Demicco et al: overall risk class: low (2/7). Age <50 years (Score 0); Tumour size 10-15cm (Score:2); Mitotic count 0/10HPF (Score 0); Tumour Necrosis <10% (Score 0)

The patient was followed up at 3 months and 6 months with Contrast-enhanced computed tomography, which did not show any recurrences.

Discussion

Klemperer and Rabin first described SFTs in 1931. These are soft-tissue spindle-cell neoplasms. SFTs are classified by the World Health Organization (WHO) as intermediate fibroblastic or myofibroblastic tumours, which means that SFTs are considered tumours that rarely metastasize. These tumours usually affect the pleura. 30% of these tumours are reported to be extra-pleural which includes the salivary glands, nasal cavity, orbit, the upper respiratory tract, thyroid, genitourinary system, peritoneum, retroperitoneum and pelvis.

As the case described above, SFTs in the retroperitoneum, are rarely found and less than 100 cases have been described so far. The main features of SFTs are the large size they can reach as they do not have any specific symptoms. This leads to the need for major surgery for resections of the primary.

The tumour on microscopic examination has a “patternless pattern” which makes histopathological diagnosis challenging. This pattern is a storiform arrangement of spindle cells combined with a “hemangiopericytoma-like appearance” and increased vascularity of the lesion. Other spindle cell tumours such as leiomyoma, angiomyolipoma, inflammatory myofibroblastic tumours, and gastrointestinal stromal tumours are some of the differential diagnoses.

Immunohistochemistry is very helpful. Solitary Fibrous Tumours are positive for Bcl-2, vimentin, CD99, and CD34 and negative for expression of S100, cytokeratin, EMA, SMA, CD117, CD31, and desmin normally. Around 75% of extrapleural SFTs express a positive for a combination of Bcl-2 and CD34, which guides histopathologically towards the diagnosis of SFT.

If SFTs show high mitotic activity (that is more than 4 mitoses in 10 HPF), high cellularity, necrosis, pleomorphism and hemorrhagic activity in histopathological examination, they are considered to be malignant.

Sometimes, paraneoplastic syndromes may be present in SFTs, mainly hypoglycemia. It is thought to arise due to tumour producing Insulin-like growth factor – 2 (IGF-2). These paraneoplastic symptoms may sometimes be the presenting symptoms for these tumours. Normally when complete resection is achieved by surgery, these symptoms subside.

Computed Tomography imaging is unable to differentiate primary retroperitoneal SFTs from other solid retroperitoneal tumours; however, for the surgeons, it is invaluable as it gives an anatomical overview and provides information required to plan the right approach and strategy for complete resection of the tumour with clear margins.

Surgery is the mainstay of treatment and the only effective treatment available in most cases. When there is a clear negative margin, the recurrence rates appear to be low and positive resection margins affect the recurrence rates.

As SFTs are quite rare, especially more so in retroperitoneum, there is a lack of studies that define the best management guidelines. For adjuvant treatment, only case reports and observational studies are available which are also dependent upon individual cases. The tumour has high vascularity therefore, antiangiogenic drugs, such as bevacizumab, interestingly, are used initially. An important study on the matter proposed a strategy to use conventional chemotherapeutic agents to keep the disease stable and for treating advanced disease. Local and distant recurrences and distant seen even in benign cases, which show unpredictable behaviour, with the potential for malignant transformation.

The potential of SFT for malignant transformation is the basis of performing computed tomography during follow-up.

Conclusion

Primary retroperitoneal soft tissue tumours should be managed aggressively. Surgery is the primary treatment option. A laparoscopic approach is a viable option for resection, but a margin free of tumours is mandatory for decreasing recurrence rates. Solitary fibrous tumours are diagnosed only with histopathological examination of excised specimens, and IHC can be used for confirmatory purposes. The rarity of the disease and lack of clinical guidelines tend to confuse the clinicians and a multidisciplinary team approach is mandatory for proper management.

Disclaimer The authors declare no conflicts of interests

Ethical Approval

Ethical approval of case report is not needed in accordance with the local ethical guideline

Consent

Written informed consent was obtained from the patient to include the clinical details

Reference

Figure 1: MRI of the abdomen and pelvis showing a complex heterogenous signal intensity mass in the retroperitoneum (A)

Figure 2: The excised specimen, shows a large soft tissue tumour with 2 lobes with the larger lobe having cystic areas.

Figure 3: (A) Gross section of the specimen, showing 2 large nodular bosselated encapsulated soft tissue and the same speci

Figure 4: (A) Microscopic H&E stain shows no mitotic figures, with no necrosis. Tumour composed of compact cellular loos