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CASE REPORT

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Primary retroperitoneal solitary fibrous tumor: A case report

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Key Clinical Message

Primary retroperitoneal masses have numerous differential diagnoses, many of which are rare entities. These can be neoplastic or nonneoplastic. Among the rare conditions are solitary fibrous tumors, which can either be benign or malignant. It is a mesenchymal, spindle-cell tumor, reported first in 1931 as a pleural tumor by Klemperer et al. A 20-year-old lady, with abdominal pain for 6 months, was diagnosed with a retroperitoneal mass on the left lower abdomen on USG which was confirmed by an MRI scan of the abdomen. The patient underwent laparoscopy-assisted excision of the mass. The final histopathological reports and immunohistochemistry reports revealed a solitary fibrous tumor. Solitary fibrous tumors (SFTs) are rare tumors in the retroperitoneum. In our search, fewer than a hundred cases have been reported. It has a characteristic "patternless pattern" in a microscopic study. Adverse outcomes of SFTs are associated with atypical features in histology, such as nuclear pleomorphism, necrosis, increased cellularity, and mitoses >4/10 HPF and size more than 10 cm. The standard of care is surgical excision with clear margins. Open surgeries have been done traditionally; we present a case where we performed the excision laparoscopically.

KEYWORDS

laparoscopic surgery, oncology, solitary fibrous tumour, surgery, urology

1 | INTRODUCTION

Solitary fibrous tumors (SFTs) were first described in 1931 by Klemperer et al.[1] as pleural tumors. Since then, it has been reported in many extrapleural sites but is found to be exceedingly rare. Most of them are reported to arise from the pleura, and only 30% are of extrapleural origins.[2] Less than a hundred cases of primary retroperitoneal solitary fibrous tumors have been described till now. These are rare soft tissue sarcomas, with mesenchymal origins. The symptoms of these tumors 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 tumors is by surgical excision with clear margins. The role of adjuvant chemotherapy is controversial.[3]

We encountered one such patient with primary retroperitoneal SFT and have tried to present the case with the best resources available at our center.

2 | 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 third 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, ~6 × 6 cm in diameter, with ill-defined margins, non-tender, and not attached to the overlying skin. Bowel sounds were present on auscultation and other systemic examinations were normal.

On ultrasonography, a complex heterogenous solid-cystic lesion was seen in the left adnexa measuring ~9.9 × 6.0 × 6.4 cm. MRI was then done to confirm the diagnosis, which showed an 11.7 × 8.2 × 4.8 cm (CC × AP × T) 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-sized irregular-shaped 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 tumor.

So, with the provisional diagnosis of primary retroperitoneal mass, the patient underwent "Laparoscopy Assisted Transperitoneal Excision of Retroperitoneal Mass" (Figure 2). During the surgery, a large solid mass measuring ~15 × 10 cm with 2 lobes with an irregular surface was seen with the larger lobe having cystic areas. (Figure 3) There was dense adhesion of the mass posteriorly with the psoas muscle. The visualized retroperitoneal organs were normal. There was blood loss of ~1000 mL from the part of the mass adhered to the psoas muscle. An intra-abdominal drain was placed, which was removed on the fourth postoperative day. The rest of her stay in the hospital was uneventful and was sent home from the hospital on the sixth postoperative day.

In her histopathology report, gross examination showed two large nodular bosselated encapsulated soft tissue measuring 9 × 6 × 2 cm and 4.5 × 3 × 3 cm were seen with a cut section showing a gray-white area with a cystic area within it. (Figure 4A,B) On microscopic examination, mitotic figures or necrosis were not present, tumor 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 5A) punctuated by variously sized hemangiopericytomatous vessels, many with discernible fibromuscular walls and cystic spaces. The morphological features which were consistent with solitary fibrous tumors and margins, however, were positive for tumor.

The immunohistochemistry showed tumor cells positive for CD34, SMA, and STAT-6 and negative for CK, S100, and desmin. (Figure 5B,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); tumor size 10–15 cm (Score 2); mitotic count 0/10HPF (Score 0); tumor necrosis <10% (Score 0).[4]

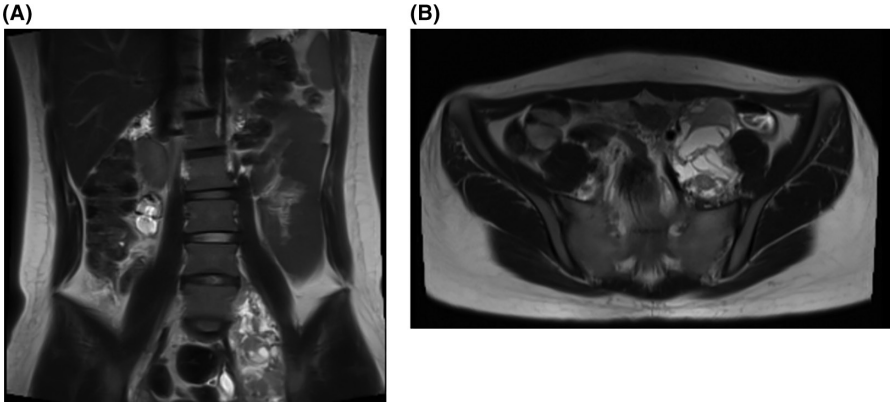

FIGURE 1 MRI of the abdomen and pelvis showing a complex heterogenous signal intensity mass in the retroperitoneum (A) coronal view (B) transverse view.

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

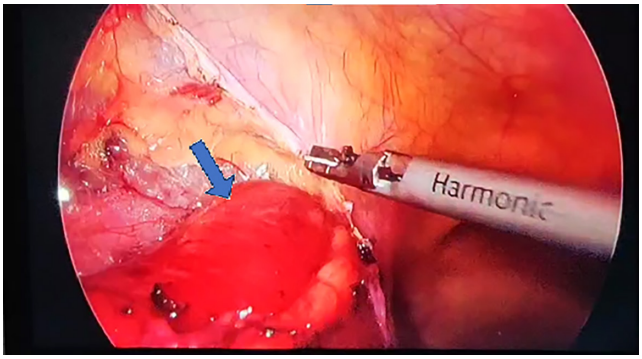

FIGURE 2 Laparoscopic image of the mass (arrow) seen during dissection.

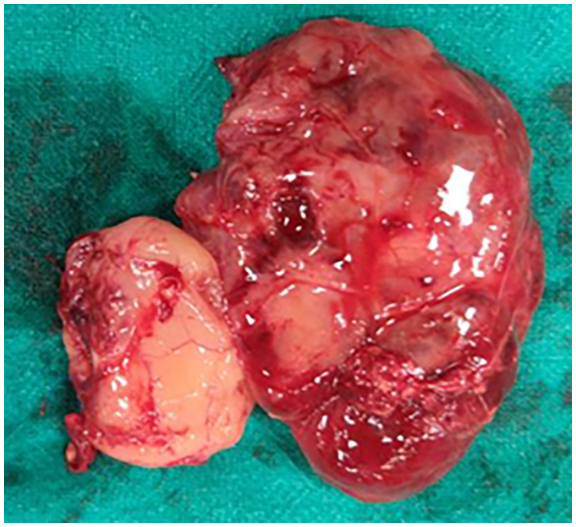

FIGURE 3 The excised specimen, shows a large soft tissue tumor with two lobes with the larger lobe having cystic areas.

3 | 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 tumors, which means that SFTs are considered tumors that rarely metastasize.[5] These tumors usually affect the pleura. Only 30% of these tumors are reported to be extrapleural which includes the salivary glands, nasal cavity, orbit, upper respiratory tract, thyroid, genitourinary system, peritoneum, retroperitoneum, and pelvis.[6]

The differential diagnosis of primary retroperitoneal masses is given in Table 1.[7] As the case described above, SFTs in the retroperitoneum are rarely found and less than 100 cases have been described so far.[2] The main feature of SFTs is 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.

Computed tomography imaging is unable to differentiate primary retroperitoneal SFTs from other solid retroperitoneal tumors[8,9]; 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 tumor 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.[10]

The tumor on microscopic examination has a "pattern-less 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.[11] Other differential diagnoses include spindle cell

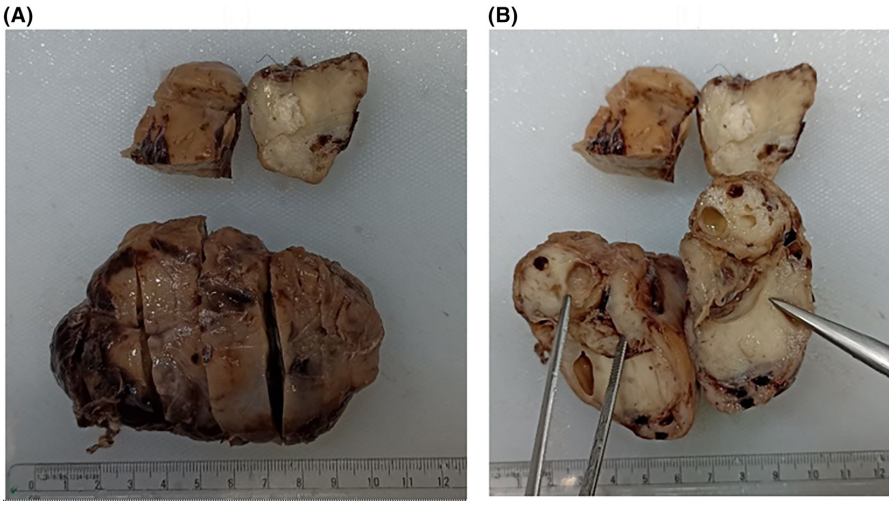

FIGURE 4 (A) Gross section of the specimen, showing two large nodular bosselated encapsulated soft tissue and the same specimen on cut section (B) showing gray white areas with cystic areas within it.

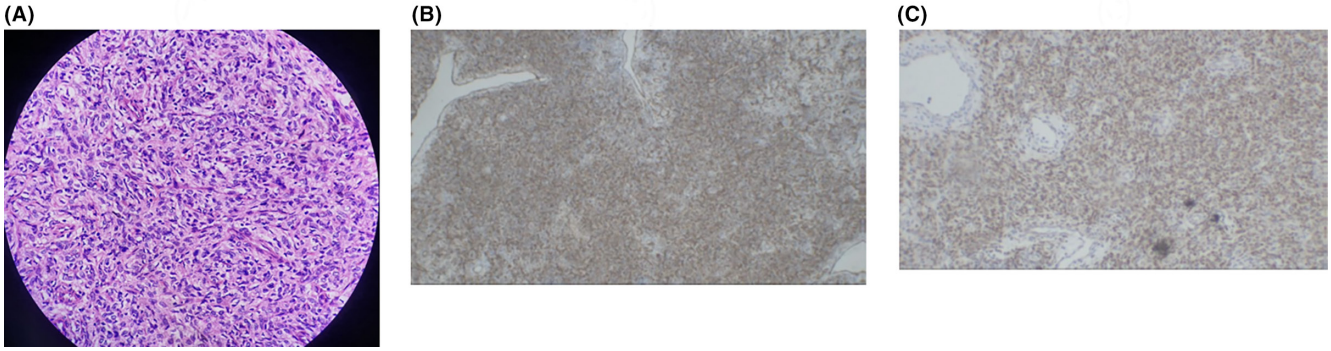

FIGURE 5 (A) Microscopic H&E stain shows no mitotic figures, with no necrosis. Tumor composed of compact cellular looser myxoid areas with spindle-like cells and ovoid cells. (B) Immunohistochemistry CD34 (Qbend10) with positive straining. (C) Immunohistochemistry STAT (EP325) with positive staining.

TABLE 1 Differential diagnosis of primary retroperitoneal masses.[7]

Solid: Neoplastic
Lymphoid tumors
Lymphoma
Sarcoma
Malignant fibrous histiocytoma
Liposarcoma
Leiomyosarcoma
Neurogenic tumors
Paraganglioma
Schwannoma
Neurofibroma
Ganglioneuroma
Immature teratoma
Solid: Nonneoplastic
Extramedullary hematopoiesis
Retroperitoneal fibrosis
Erdheim–Chester disease
Cystic: Neoplastic
Mucinous cystadenoma
Mature teratoma
Cystic mesothelioma
Cystic: Nonneoplastic
Mullerian cyst
Lymphangioma
Pancreatic pseudocyst
Epidermoid cyst
Urinoma
Lymphocele
Hematoma

tumors such as leiomyoma, angiomyolipoma, inflammatory myofibroblastic tumors, and gastrointestinal stromal tumors.

Immunohistochemistry is very helpful in diagnosing these tumors. Solitary fibrous tumors are positive for Bcl-2, vimentin, CD99, and CD34 and negative for expression of S100, cytokeratin, EMA, SMA, CD117, CD31, and desmin normally.[12] Around 75% of extrapleural SFTs express a positive for a combination of Bcl-2 and CD34, which guides histopathologically toward the diagnosis of SFT.[6]

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.[13]

Sometimes, paraneoplastic syndromes may be present in SFTs, mainly hypoglycemia. It is thought to arise due to tumor producing insulin-like growth factor-2 (IGF-2). These paraneoplastic symptoms may sometimes be the presenting symptoms for these tumors.[14] Normally when complete resection is achieved by surgery, these symptoms subside.

Demicco et al. have provided a risk stratification model to assess the risk of solitary fibrous tumors for the development of metastasis. This model is given in Table 2.[4] However, it is important to note that, this study included intrathoracic, head and neck, trunk, extremity, and intra-abdominal tumors but not primary retroperitoneal SFTs most likely due to its rarity, hence the validity of this risk assessment for stratification of primary retroperitoneal SFTs are questionable.

As SFTs are quite rare, especially in retroperitoneum, there is a lack of studies defining the best management guidelines. For adjuvant treatment, only case reports and observational studies are available which are also

TABLE 2 Modified 4-variable risk stratification model for development of metastasis in SFTs.[4]

Risk factor	Score
Age	
<55	0
>55	1
Tumor size (cm)	
<5	0
5–10	1
10–15	2
<15	3
Mitotic count (/10 hpf)	
0	0
1–3	1
>3	2
Tumor necrosis	
<10%	0
>10%	1
Risk class	**Total score**
Low	0–3
Intermediate	4–5
High	6–7

Abbreviations: hpf: high power fields; SFT, solitary fibrous tumors.

dependent upon individual cases. The tumor 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 to treat advanced disease.[15] Local and distant recurrences and distant seen even in benign cases, which show unpredictable behavior, with the potential for malignant transformation.

The potential of SFT for malignant transformation is the basis of performing computed tomography during follow-up.[16]

4 | CONCLUSION

Primary retroperitoneal soft tissue tumors should be managed aggressively. Surgery is the primary treatment option. Though our case had a positive surgical margin, improving the techniques will make the laparoscopic approach a viable option for resection. Having a margin free of tumors is mandatory for decreasing recurrence rates. Solitary fibrous tumors 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. We hope this case report adds to the few available data on primary retroperitoneal solitary fibrous tumors.

AUTHOR CONTRIBUTIONS

Pramesh Prasad Shrestha: Conceptualization; data curation; formal analysis; investigation; validation; visualization; writing – original draft. **Mahesh Bahadur Adhikari:** Supervision; writing – review and editing. **Bipin Maharjan:** Supervision; writing – review and editing. **Ravi Kiran Gautam:** Writing – review and editing. **Birodh Basnet:** Writing – review and editing. **Deepak Kumar Yadav:** Writing – review and editing.

CONFLICT OF INTEREST STATEMENT

The authors declare no conflicts of interests.

DATA AVAILABILITY STATEMENT

The data that supports the findings of this study are available in the supplementary material of this article.

ETHICS STATEMENT

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 publish this report in accordance with the journal's patient consent policy.

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