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Case Report

Undifferentiated Spindle Cell Sarcoma in the Retroperitoneum- Case Report

Agil Selvam*, Kumaresh Pandian, Kothe Pallavi Baban, Vinoth M, Arcot Rekha

Saveetha Medical College, Saveetha Nagar, Thandalam, Chennai, India.

***Corresponding author:** Selvam A, Saveetha Medical College, Saveetha Nagar, Thandalam, Chennai, India.

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Abstract

Retroperitoneal Sarcoma (RPS) remains a rare malignancy, occurring in just 0.5 to 1.0 per 100,000 population [1]. The diagnosis and management of this tumor requires a multidisciplinary approach [2]. An important part of the diagnosis is the identification of the histological subtype since this can affect the prognosis and treatment options. A 39-year-old patient presented to the emergency with complains of pain in the left lower limb and left lumbar and iliac region for the past 15 days. The pain aggravated in the last 3 days. Swelling in the left abdominal quadrant was present for the past 10 days. Imaging done showed a retroperitoneal tumor which at excision was revealed as undifferentiated spindle cell sarcoma. Undifferentiated high-grade spindle cell sarcoma is one of the rare forms of malignancy occurring in just less than 10% of the word population. The diagnosis is often missed and hence following up with the HPE report is very essential to improve the outcome and prognosis of this disease.

Keywords: CD 34, S-100; Retroperitoneal sarcoma; Undifferentiated High-Grade spindle cell sarcoma

Introduction

Retroperitoneal sarcoma presents with vague symptoms and is difficult to diagnose or is discovered incidentally on imaging. Retroperitoneal sarcoma can expand massively in the retroperitoneum prior to detection and diagnosis, making these resections challenging. Contrast-enhanced CT of the abdomen and pelvis is often sufficient for the diagnosis of retroperitoneal sarcoma.

Case Report

A39-year-old male patient presented to the emergency with complains of pain in the left lower limb and left iliac and lumbar regions for the past 15 days. The pain has aggravated over the past 3 days. Tingling sensation was present over the left posterior quadrant radiating to the left lower limb. The pain aggravated on walking and relieved on taking rest. The patient gave no history of trauma or injury. There was no history of tuberculosis.

Per-abdomen Examination

Abdomen was soft, Mass was present over the left lumbar quadrant; 10x10 cm fixed, mixed in consistency.

Straight Leg Raising Test

Left lower limb (restricted) and the patient had a flexion deformity of 15° which could be straightened albeit with pai. Patient was evaluated further in the hospital and MRI imaging showed retroperitoneal sarcoma. CT- guided biopsy revealed malignant spindle cells and the patient was planned for explorative laparotomy.

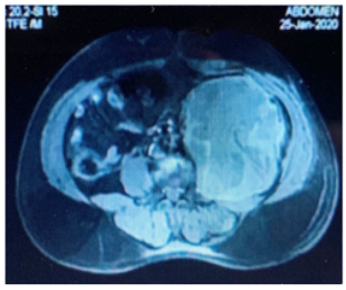

Figure 1a

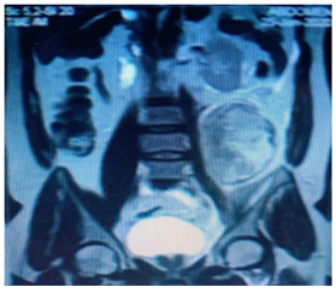

Figure 1b

Figure 1a and **1b:** showing MRI images of retroperitoneal sarcoma.

Patient underwent explorative laparotomy with intraoperative finding of 15x12cm retroperitoneal mass overlying the psoas muscle on the left side without injuring the nerves and

overlying vasculature. Specimen was removed in toto. Marking clips were placed on the tumour bed. The post-operative period was uneventful.

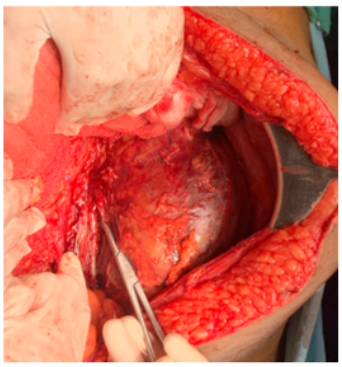

Figure 2a

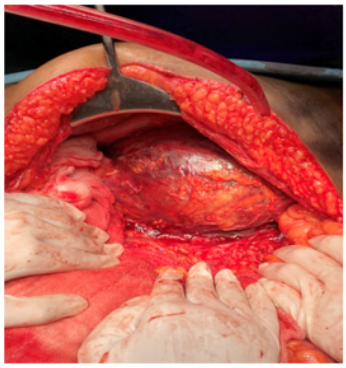

Figure 2b

Figure 2a and 2b: Showing intraoperative finding of 15x12cm retroperitoneal mass overlying the psoas muscle.

Histopathological examination revealed undifferentiated spindle cell sarcoma. Immunohistochemistry sent showed, S-100, SMC, and CD 34- Negative. The patient was referred to oncology for adjuvant therapy.

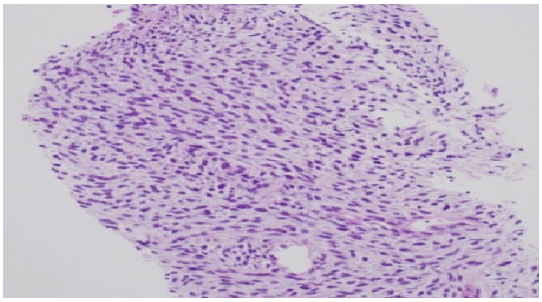

Figure 3: Depicting histopathological examination, revealing undifferentiated spindle cell sarcoma.

Discussion

Patients with sarcomas often present late, because these tumours arise in the capacious potential spaces of the retroperitoneum and can grow very large without producing symptoms [3,4]. Moreover, even when symptoms do occur, they are nonspecific, such as abdominal pain and fullness, and are easily dismissed as being caused by other less serious processes [5]. Imaging plays a major role in the diagnostic workup of these patients, being required not only for tumour detection, staging, and operative planning, but also for guiding percutaneous or surgical biopsy of these tumours. Therefore, image-guided, and surgical biopsies have a relatively greater role to play in the diagnosis of retroperitoneal sarcomas than is the case for sarcomas elsewhere in the body [6,7]. After the diagnosis is made, the surgical team needs to determine if the retroperitoneal sarcoma can be resected. Therefore, one of the first determinations to be made is whether the tumour is localized, it's local extent, and also if there is evidence of intra- or extra-abdominal metastatic spread of tumour. The location and size of the tumour, its relationship to adjacent organs, presence or absence of local extension, relationship to and/or involvement of major vascular structures, as well as the presence of normal anatomic variants and anomalies of major abdominal arteries and veins, are all crucial pieces of information that need to be provided. Accurate staging is important as it facilitates determination of appropriate surgery, establishes prognosis, and provides a guide for adjunctive therapy [8]. This staging system takes into consideration histological grade, tumour size and depth relative to the superficial muscular fascia, presence or absence of lymph node involvement, and the presence or absence of distant metastases. The AJCSS of soft tissue sarcoma is shown in table 1.

Table 1: CLASSIFICATIONS

HISTOLOGICAL GRADE (G)

- G1- Well differentiated
- G2- Moderately differentiated
- G3- Poorly differentiated

PRIMARY SITE (T)

- Tx- Primary tumour cannot be assessed
- T0- No evidence of primary tumour
- T1- Tumour less than 5 cm in diameter

T1a- Superficial tumour

T1b- deep tumour

- T2- Tumour 5 cm or more in diameter

T2a- Superficial tumour

T2b- Deep tumour

REGIONAL LYMPH NODES (N)

- Nx- Regional lymph nodes cannot be assessed
- N0- No regional lymph node metastasis
- N1- Regional lymph node metastasis

DISTANT METASTASIS (M)

- Mx- Distant metastasis cannot be assessed
- M0- No distant metastasis
- M1- Distant metastasis

Neoadjuvant therapies (chemotherapy, external be am radiation, or combination radiation and chemotherapy) are safe in well-selected patients and may be considered only after a careful review. The preferred modality is chosen based on the pattern of

recurrence. Chemotherapy is being employed in histologies with the highest systemic risk (e.g. leiomyosarcoma and high-grade undifferentiated sarcoma), and radiotherapy in those with the highest loco regional risk (e.g. well-differentiated sarcoma and low/intermediate undifferentiated sarcoma). None of the neoadjuvant/adjuvant modalities, however, has proven its value in terms of improved survival rates. Retroperitoneal sarcomas comprise a spectrum of histologic types/subtypes; the most common are liposarcoma and leiomyosarcoma. For many of these RPSs, the histologic type/subtype defines the grade of the tumour. They were classified into 6 histologic groups: liposarcoma, leiomyosarcoma, malignant peripheral nerve sheath tumour, sarcoma not otherwise specified (NOS), solitary fibrous tumour, translocation-associated (synovial sarcoma, rhabdomyosarcoma, Ewing sarcoma, fibro sarcoma, inflammatory myofibroblastic tumour), or other (malignant granular cell tumour, malignant mesenchymoma, myosarcoma). Patient evaluation for metastatic RPS should aim to give the patient access to clinical trials and comprehensive multidisciplinary care. A proper treatment plan should be designed, taking into consideration histopathology, previous therapy, disease-free interval, symptomatology, comorbidities, and functional status [9]. Complete surgical excision is the only chance at a cure and should be performed at specialized centers by an experienced team. Recurrent and metastatic disease should be addressed on a case-by-case basis by a multidisciplinary team and possibly in prospective studies.

Conclusion

It is essential that in these types of cases a thorough evaluation through surgery is required. If reported as in our case immunohistochemistry would be helpful in follow up management like requirement of radiotherapy and chemotherapy. If improperly treated, findings may worsen and risk of losing the patient is on the higher side.

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