CASE REPORT

LEIOMYOSARCOMA OF THE INFERIOR VENA CAVA MASQUERADING AS A DUODENAL STROMAL TUMOUR

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Leiomyosarcomas originating in the inferior vena cava (IVC) are rare vascular tumours characterized by gradual growth and subtle onset. The challenges associated with these tumours stem from their unfavourable prognosis and the absence of established treatment protocols. We present a case of a 52-year-old woman who presented with abdominal pain and was subsequently diagnosed with leiomyosarcoma in the second segment of the IVC. This case highlights the significance of adopting a multidisciplinary approach and emphasizes the necessity for timely identification and intervention in IVC leiomyosarcomas to improve patient outcomes. To our knowledge, this is the first reported case of leiomyosarcoma in the IVC from Pakistan. We believe this case will contribute valuable insights to the existing knowledge on the subject.

Keywords: Leiomyosarcomas; Masquerading; Tumour; Abdominal pain

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INTRODUCTION

Leiomyosarcoma (LM), a malignancy arising from smooth muscle tissue, possesses a complex aetiology and typically carries an unfavourable prognosis. It's recognized as the most common tumour in the venous system and the second most prevalent retroperitoneal neoplasm in the elderly. Emerging in any vein, the inferior vena cava (IVC) is most affected, constituting 5–10% of soft tissue sarcomas.[1,2] LM may display intraluminal growth in 5% of cases, extraluminal growth in 62%, or a combined pattern in 33%. Affecting both genders, it shows a higher incidence in individuals in their fifth and sixth decades of life.[3] The slow growth of leiomyosarcomas often leads to nonspecific symptoms like abdominal pain, weight loss, a palpable abdominal mass, weakness, fever, anorexia, vomiting, and night sweats due to their retroperitoneal location.[4]

CASE

A 52-year-old female with no known comorbidities presented with complaints of abdominal pain and vomiting for 7 days. She was admitted to a local hospital where abdominal imaging revealed a well-defined heterogeneous mass at porta hepatis, displacing the head of the pancreas, infra-hepatic part of the IVC, caudate lobe of the liver, and right renal vein as shown in Figure 1. The baseline workup, including complete blood count, liver function tests, renal function tests, coagulation profile, viral markers, and electrolyte panel was unremarkable. Endoscopic ultrasound showed an ovoid hypo-echoic paraduodenal mass, measuring 6.2×4.2 cm, which was separable from the pancreas. A 22 G ACQUIRE fine needle biopsy was done, exhibiting atypical smooth muscle cells. Immunohistochemistry was performed which was positive for AMSA and desmin stains. Findings were consistent for a spindle cell neoplasm. Intraoperative, the right hepatic lobe was mobilized and a 4 cm mass was noticed arising from the anteromedial wall of the retrohepatic segment of the IVC, which is one of the four segments of the IVC.

The other being suprahepatic, renal and suprarenal. The mass was dissected and separated from the aorta, portal vein, and resected en bloc with the anteromedial wall of the inferior vena cava as shown in Figure 2 (a and d). Subsequently, the anteromedial wall of the IVC was repaired with a Polytetrafluoroethylene (PTFE) 6mm patch as shown in Figure 2 (b and c). An additional margin of the IVC was biopsied, which was negative for malignancy. The resected mass was sent for histopathology, showing a well-circumscribed malignant neoplasm comprising fascicles of spindle cells with patchy areas of necrosis and moderate nuclear pleomorphism (2–3 mitosis/HPF). Cigar-shaped nuclei with areas of multinucleation and Focal hypocellular hyalinized areas were also seen. Staining was positive for ASMA, Desmin, and H-Caldesmon. Post-surgical imaging showed no discrete enhancing masses and nodules.

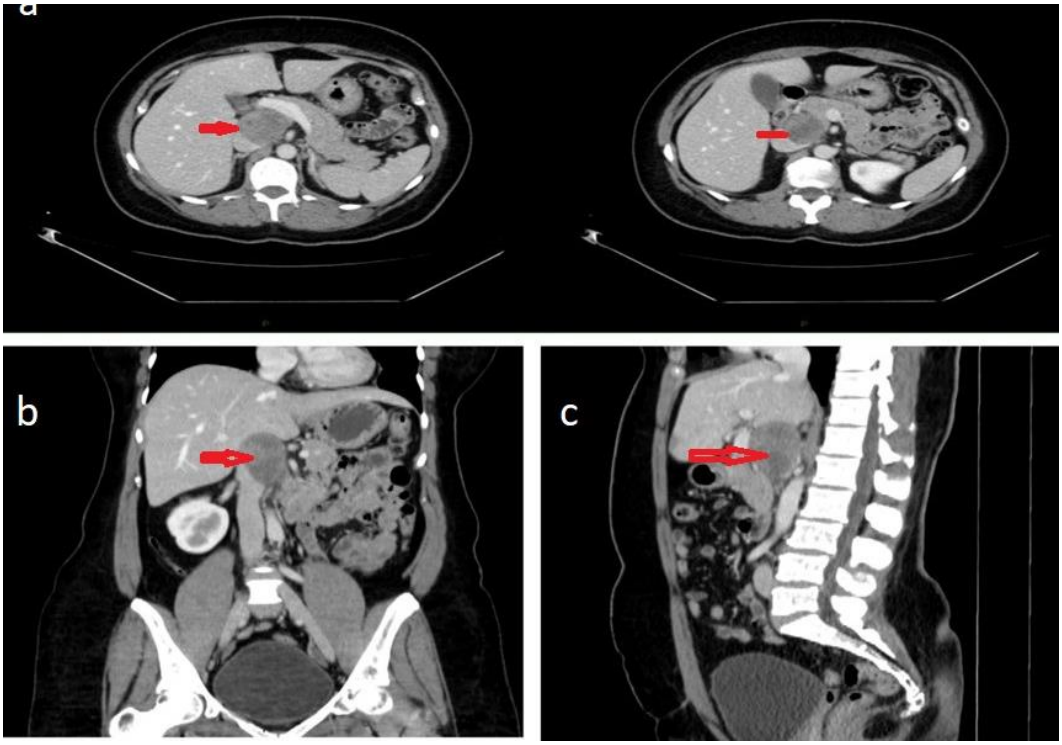

Figure-1:

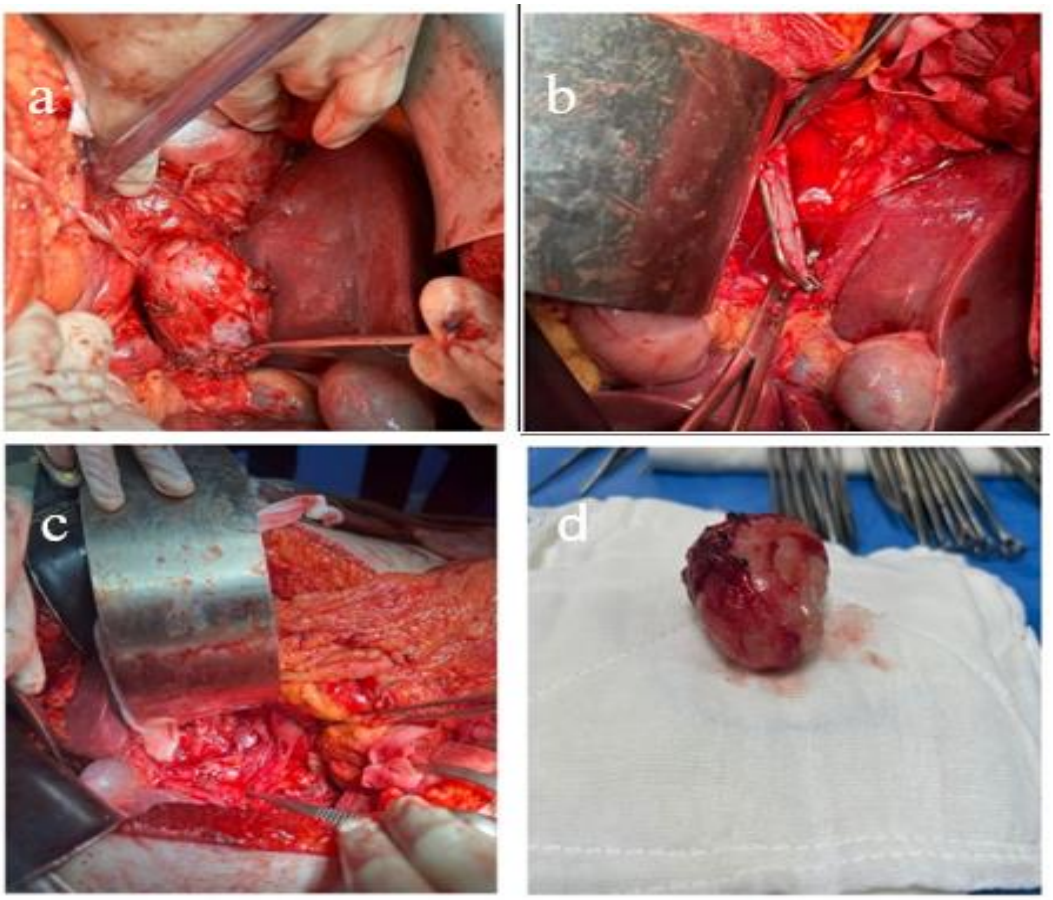

Figure-2:

DISCUSSION

Leiomyosarcoma of the IVC, first described by Perl et al in 1871, is the most common primary tumour of the IVC, with fewer than 600 cases reported worldwide. These tumours of mesenchymal origin typically arise from smooth muscle cells in the tunica media. They account for 0.5% of all adult tissue sarcomas and affect <1/100,000 of all adult malignancies.[5] Surgical resection with clear margins is the definitive treatment, although the prognosis is generally poor. Symptoms are often nonspecific, with abdominal pain being the most common. This case highlights a successfully treated locally advanced LM of the IVC, emphasising the collaborative efforts of gastroenterologists, surgeons, and radiologists in diagnosis and treatment planning.

Leiomyosarcoma predominately arises in females with its incidence peaking in the 5th and 6th decade of life[6]. Notably, our patient also mirrors this demographic profile, being a female in a similar age group. Our patient presented with nonspecific abdominal pain, which is the most common symptom reported in the literature.[7]

Improved survival rates up to a 5-year survival rate of 49.4% are reported with radical tumour dissection compared to those who are managed with medical therapy only.[8] Histopathological analysis of the surgical specimen unveiled negative margins. Hines et al. have documented this outcome as a good prognostic indicator, noting a 5-year survival rate of 68%, in contrast to the zero percent survival observed among patients with positive margins.[9]

Surgical resection is currently the only curable treatment for LM of the IVC, due to the rarity of the disease and lack of data regarding alternative options like chemotherapy, radiotherapy, and chemoradiotherapy. Improved median survival has been reported with adjuvant radiation and chemotherapy as compared to surgical resection alone.[9] While metastasis at the initial presentation is uncommon, instances have been documented where patients present with metastases. The lungs are the most prevalent sites, with subsequent occurrences in the thigh and shoulder muscles, liver, and bones. After achieving complete resection, the likelihood of recurrence at distant sites surpasses that of local recurrence.[10]

Leiomyosarcoma of the IVC remains a significant challenge for timely diagnosis and optimal treatment due to factors like its vague presentation, low incidence, and the absence of established management guidelines. Given these hurdles, we propose that further studies should be conducted to improve patient outcomes and develop evidence-based protocols for the effective management of this condition.

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