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Case Report

Mesenteric Dedifferentiated liposarcoma presenting as a massive abdominal lump: A Rare Case

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Abstract

This case study examines a sixty five-year-old male presenting with a distended abdomen, lower back discomfort, and dyspnea on lying down, progressing over three months. Physical examination revealed multiple hard palpable lumps in the abdomen. Initial radiological evaluation indicated a connective tissue tumor, leading to a trucut biopsy revealing a spindle cell neoplasm of uncertain malignant potential. The tumor was found to be resistant to immunotherapy and neoadjuvant chemotherapy. Surgical resection unveiled multiple mesenteric tumors along the small bowel and also involving the descending colon, diagnosed as a dedifferentiated liposarcoma. Liposarcoma encompasses five histological subtypes, with dedifferentiated being the least common and comprised of poorly differentiated cells. Although liposarcomas typically originate from the retroperitoneum, the occurrence of dedifferentiated liposarcoma in the small bowel mesentery is exceedingly rare. This case report aims to underscore this uncommon occurrence, discuss diagnostic and treatment strategies, and advocate for further research into targeted therapies addressing specific immunohistochemistry markers to enhance outcomes for such patients, who generally face a grim prognosis

Keywords – *dedifferentiated liposarcoma , mesenteric tumors, spindle cell malignancy, mesenchymal tumors of GIT.*

Introduction

We present the case of a sixty-five-year-old male with a prominently distended abdomen and palpable masses. Initially suspected to originate from the retroperitoneum based on a trucut biopsy showing a spindle cell neoplasm, surgical exploration later identified the tumor as mesenteric. Histopathological examination confirmed the diagnosis of dedifferentiated liposarcoma. Liposarcoma is a rare type of cancer that originates from adipose tissue, and dedifferentiated liposarcoma is a specific subtype

characterized by the presence of both well-differentiated liposarcoma and high-grade undifferentiated sarcoma components[1]. Dedifferentiated liposarcomas in the mesentery are particularly noteworthy due to their aggressive nature[2] and the challenges they pose in management, with as few as seven cases reported in the literature. Surgical resection remains the mainstay of treatment for these tumors, but the infiltrative nature and anatomical location of mesenteric dedifferentiated liposarcomas can make complete resection challenging[3]. Here, we highlight the need for the development of other novel therapies for the treatment of this aggressive variant.

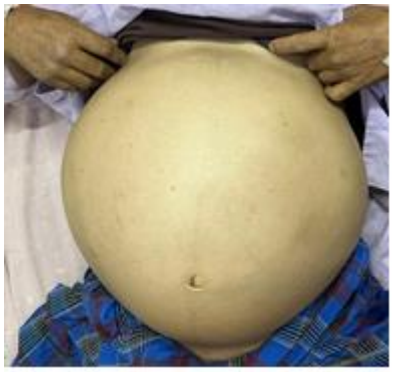

Figure 1: This pictures represents the patient at the time of presentation, showing gross uniform abdominal distenstion.

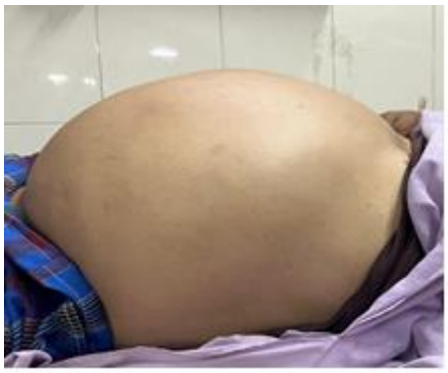

Figure 2: Figure showing gross abdominal distention in all regions and the absence of a smiling umbilicus as seen in ascites.

Case

A 65-year-old male presented with progressive abdominal distention and discomfort over the past three months (Figures 1, 2). The distension had an insidious onset, gradually worsening, without associated pain, skin changes, visible veins on the abdomen, fever, weight loss, or anorexia. He also experienced difficulty breathing while lying down and back pain for the past two weeks, coinciding with the progression of distension. He had no known comorbidities, prior surgeries, or history of alcohol consumption, but did have a history of smoking (ten pack-years). There was no significant family history.

On abdominal examination, multiple well-defined globular lumps were palpable in the umbilical, right lumbar, and left iliac fossa regions, which were firm in consistency. The umbilical lump did not move with respiration, while the others were mobile. The overlying skin was normal, there was no tenderness on palpation, and no local rise in temperature.

The patient underwent routine hematological workup, along with a chest X-ray, ECG, and pulmonary function tests to rule out chronic obstructive pulmonary disease and other potential causes for dyspnea while lying down, as well as for pre-anesthetic evaluation. Initially, an ultrasonography was performed, but since it did not provide sufficient information, an abdominal contrast-enhanced CT scan was conducted (Figures 3, 4, 5). The scan revealed a large, ill-defined lobulated soft tissue density mass in the intra-abdominal and pelvic cavity, with loss of fat planes between the ileal loops and displaced mesentery, without involvement of major vessels. An ultrasound-guided trucut biopsy of the mass was performed, which reported a spindle cell lesion of uncertain malignant potential. The tumor markers Vimentin and MSA were positive, while other markers such as CD117, S100, Desmin, Beta Catenin, and Ki67 were negative except for 1% expression.

No metastatic lesions were observed on the CT scan, and the patient did not exhibit symptoms or signs suggestive of any possible paraneoplastic syndromes. Based on this immunohistochemistry

profile, neo-adjuvant chemotherapy or radiotherapy was not indicated, thus the decision was made to proceed with surgical excision of the tumor. Prior to surgery, it was confirmed that major vessels were not involved, and the goal was to achieve maximal resection possible. Our differential diagnosis included a Gastrointestinal Stromal Tumor (GIST) or a bulky retroperitoneal tumor with intra-peritoneal extension. Considering the loss of fat planes between the ileal loops on imaging, the possibility of gut resection with anastomosis or stoma formation was also anticipated.

During surgery (Figures 6, 7, 8), large, multiple well-defined tumor masses were found in the mesentery of the small bowel, involving the descending colon. Debulking of the tumor and resection with anastomosis of the descending colon were performed. The mass at the root of the mesentery was not excised to avoid devascularization of the small bowel. Postoperatively, the patient experienced an 8 kg weight loss, relief from back pain and respiratory distress, and was discharged after seven uneventful days. He tolerated oral solids and remained afebrile.

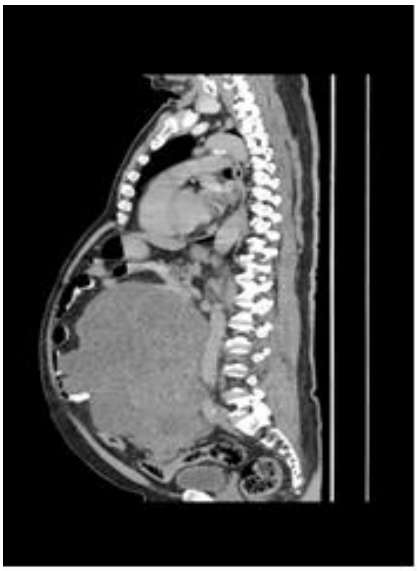

Figure 3: CT - Computed tomography- Figure shows a large heterogenous irregular soft tissue mass with possible retroperitoneal origin

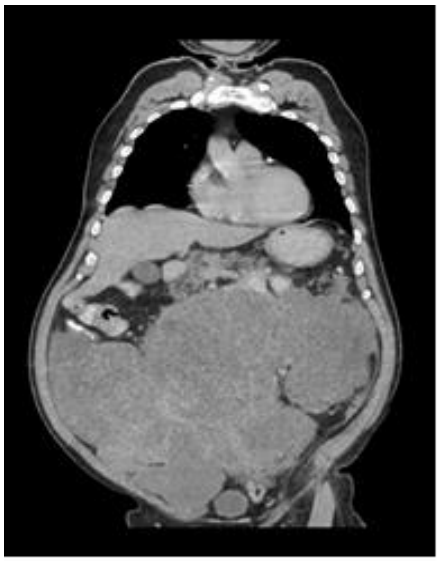

Figure 4: Multilobulated mass showing fat and solid components

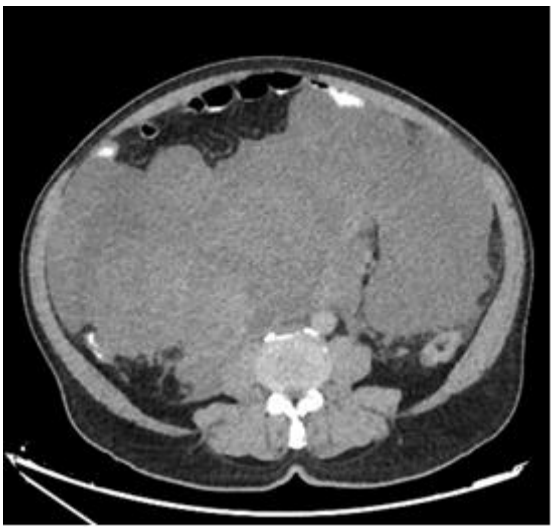

Figure 5: Displaced small bowel loops seen

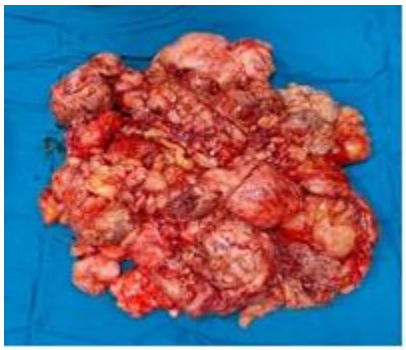

Figure 6: Figure shows the mass involving small bowel mesentery and the colon

Figure 7: Figure shows the multilobular mass excised from the mesentry and segment of descending colon which was also involved

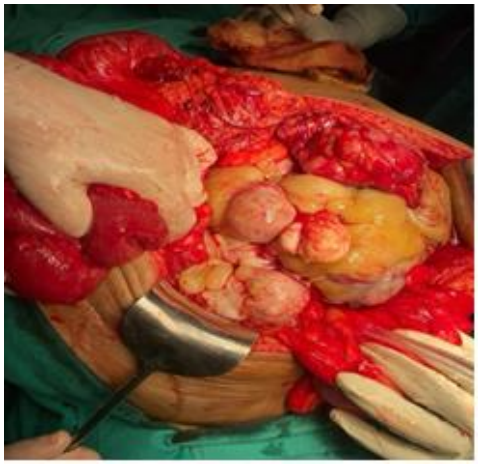

Figure 8 Figure shows the mass comprising of areas of fibrous connective tissue as well as fat on a gross appearance.

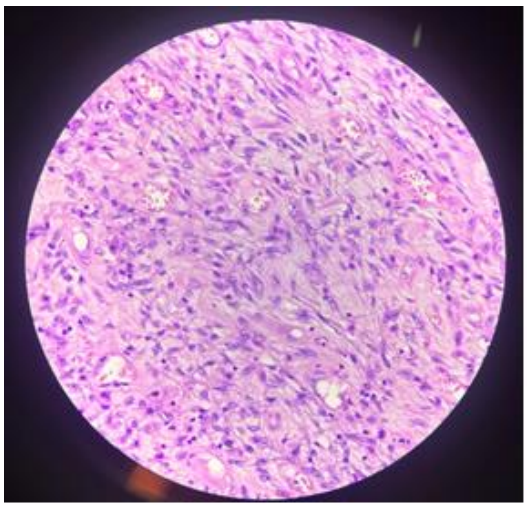

Figure 9: Figure shows Histopathological image of spindle cells on a background of lipomatous areas

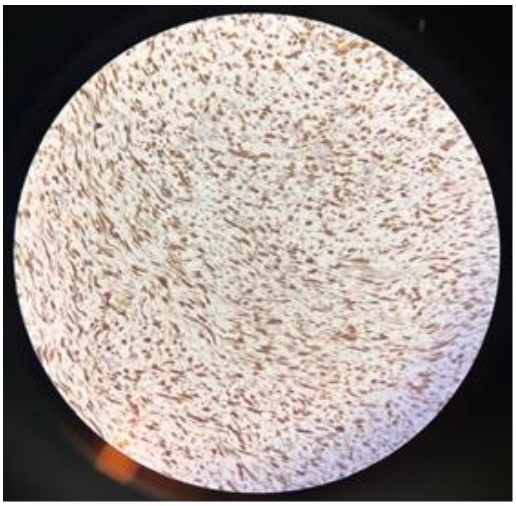

Figure 10: Figure shows Spindle cells staining positive for S100 and MDM

Histopathological examination of the excised lump reported a dedifferentiated liposarcoma of the mesentery with immunohistochemistry markers positive for MDM 2 and S100(Fig 9,10), confirming our intra operative findings. The patient was further referred to Medical Oncology for further treatment options.

Discussion

Liposarcoma is the second most common sarcoma, primarily occurring in the retroperitoneum and extremities. Primary mesenteric liposarcoma constitutes a rare entity. Liposarcoma is broadly classified into five categories: well-differentiated, myxoid, round cell, pleomorphic, and dedifferentiated types[4], with the myxoid variant being the most common. 'Dedifferentiated liposarcoma' was first termed by Evans in 1979 and exhibits histological features of both well-differentiated and poorly differentiated liposarcoma, with non-lipomatous areas—spindle-cell or pleomorphic—observed in our patient showing the spindle cell pattern[5]. It is hypothesized to arise when low-grade tumor cells undergo transformation into higher-grade cells.

Immunohistochemically, tumor cells typically stain strongly positive for MDM2 and CDK4, with MDM2 being a more consistent marker, as seen in our case. Spindle cell intra-abdominal

lesions can also include gastrointestinal stromal tumors (GIST), fibromatosis, retroperitoneal sarcomas, intra-abdominal desmoid tumors, or peripheral nerve sheath tumors[6]. Mesenteric tumors, commonly lymphomas, can arise from various mesenteric structures—blood vessels, fat, lymph nodes, or connective tissue[7]. Distinguishing between these entities based on clinical or radiological examination alone is challenging, underscoring the importance of molecular and histopathological testing to determine tumor origin.

Most GISTs are positive for KIT (CD117) and CD34, with the negative variants being epithelioid rather than spindle cell types[8]; both markers were negative in our patient. These tumors typically remain asymptomatic until they grow large, causing abdominal pain, discomfort, obstruction, or pressure symptoms. Our patient's tumor was exceptionally large (25.5 × 19 × 12.5 cm) and symptomatic. Following thorough imaging with abdominal CT to assess size, location, local invasion, and vessel involvement, a preoperative biopsy confirmed the tumor type.

On CT, these tumors often exhibit areas of fat attenuation, absent in high-grade liposarcoma. On MRI, dedifferentiated liposarcoma presents with necrotic areas and heterogeneous contrast enhancement, which can resemble other high-grade tumors[9-11]. Histologically, dedifferentiated liposarcoma stains positive for MDM2 and CDK4 [12], proto-oncogenes on chromosome 12, with high sensitivity and specificity—97% and 92% for MDM2, and 83% and 95% for CDK4, respectively. Overexpression of the cell cycle regulator p16 has also been noted.

Complete surgical resection remains the cornerstone of treatment, with extensive radical dissection warranted for tumors invading adjacent organs, such as our case where a segment of the colon was excised. Failure to achieve R0 resection is associated with high rates of local recurrence. The five-year survival rate for dedifferentiated liposarcoma is 44%, contrasting with 93% for well-differentiated liposarcomas[13]. Despite a low rate of visceral metastasis (5-10%), overall survival remains poor with a mean of 15 months, and effective systemic therapies are yet to be established. Anthracycline-based regimens have shown modest response rates of 12%, while CDK4 inhibitors and MDM2 antagonists are currently in development[14]."

Conclusion

This case highlights the complexity of evaluating diffuse abdominal lumps and the diverse range of differential diagnoses that must be considered. A comprehensive preoperative workup is essential, involving thorough imaging to anticipate surgical challenges. Particularly challenging are poorly differentiated, high-grade liposarcomas originating from anatomically complex regions like the abdomen and retroperitoneum, where achieving complete (R0) resection is often difficult. There is a pressing need for extensive research into the management of these aggressive tumors. Current treatment options are limited, and there is a significant gap in effective systemic and immunotherapies that could improve outcomes. Detecting such tumors at an earlier, less invasive stage would lead to better prognoses, as complete surgical resection becomes feasible.

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References

1. Harhar M, Ramdani A, Bouhout T, Serji B, El Harroudi T: Myxoid Liposarcoma: A Case Report of a Rare Location in the Abdominal Wall. 10.7759/cureus.8715
2. Lee ATJ, Thway K, Huang PH, Jones RL: Clinical and Molecular Spectrum of Liposarcoma. 10.1200/JCO.2017.74.9598.
3. Priyanshu S, Mohsen R: 10.3389/fsurg.2023.1110580
4. Fletcher CDM: WHO: in classification of tumours of soft tissue and bone. In: Bridge JA, Hogendoorn PCW, Mertens F, eds. World Health Orgn. 2013, 468.
5. Evans, Harry L. M.D: Liposarcoma A study of 55 cases with a reassessment of its classification. The . American Journal of Surgical Pathology 3(6): p 507-524, December. 1979
6. Al-Nafussi A, Wong NA: Intra-abdominal spindle cell lesions: a review and practical aids to diagnosis. Histopathology. 2001, 38:387-402.
7. Dufay C, Abdelli A, Le Pennec V, Chiche L: Mesenteric tumors: diagnosis and treatment. J Visc Surg. 2012, 149:239-51.
8. Sbaraglia M, Businello G, Bellan E: Mesenchymal tumours of the gastrointestinal tract. Pathologica. 2021, 113:230-251.
9. Kim T, Murakami T, Oi H, Tsuda K : CT and MR imaging of abdominal liposarcoma . AJR Am J Roentgenol. 1996, 166:829-33.
10. Hunter JC, Johnston WH, Genant HK: Computed tomography evaluation of fatty tumors of the somatic soft tissues: clinical utility and radiologic-pathologic correlation. Skeletal Radiol. 1979, 6:79-91.
11. van Vliet, M., Kliffen, M. : Soft tissue sarcomas at a glance: clinical, histological, and MR imaging features of malignant extremity soft tissue tumors. EurRadiol19. 1499-1511.
12. Binh, Matthieu Bui Nguyen; Sastre-Garau: Xavier; Guillou: MDM2 and CDK4 Immunostainings Are Useful Adjuncts in Diagnosing Well-Differentiated and Dedifferentiated Liposarcoma Subtypes: A Comparative Analysis of 559 Soft Tissue Neoplasms With Genetic Data. The. American Journal of Surgical Pathology 29(10): p 1340-1347, October. 2005, 10.1097/01.pas.0000170343.09562.39
13. Dalal K.M. Kattan M.W. Antonescu C.R: et al.Subtype specific prognostic nomogram for patients with primary liposarcoma of the retroperitoneum, extremity, or trunk. Ann Surg. 2006, 244:381-391. 11 of 12
14. Advanced well-differentiated/ dedifferentiated liposarcomas: role of chemotherapy and survival . Italiano, A. et al. Annals of Oncology, Volume. 6:1601 - 1607.