Case Report

Giant mesenteric myxoid liposarcoma: Challenges of diagnosis and treatment✩✩✩

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Myxoid liposarcoma is the most frequent form of liposarcoma, frequently localized in the extremities. Abdominal liposarcomas more commonly arise from the retroperitoneum. However, primary mesenteric localization is extremely rare. To the best of our knowledge, 22 cases have been reported in English literature. The diagnosis is often delayed by the insidious evolution of this tumor. On radiological examination, primary mesenteric liposarcoma presents as a large mass that entrapped the small bowel loops. The treatment requires complete tumor resection. We report an observation of mesenteric myxoid liposarcoma in a 64-year-old female patient, revealed by an abdominal mass associated with abdominal pain. The treatment consisted of complete tumor removal. Histologically, the tumor proved to be a myxoid liposarcoma. No adjuvant treatment was considered necessary. The patient recovered well after the operation. The aim of reporting this case is to present an uncommon clinical entity because of its huge dimensions, the rarity of the site, and histological pattern.

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Introduction

Liposarcoma is a malignant tumor of mesenchymal origin [1]. It is the most common histological subtype of soft tissue sarcomas [2]. The Myxoid subtype is the most frequent liposarcoma (56.2%) [3]. They usually arise in the soft tissues of the extremities [4]. Abdominal liposarcoma is a rare lesion and accounts for approximately 20% of all mesenchymal malignancies in adults. They commonly occur in the retroperitoneum [5]. A primary mesenteric origin is an unusual occurrence and has been reported to occur from the small bowel mesentery and transverse mesocolon [4]. It has been reported in 22 cases in the literature [6]. Here, we describe a rare case of mesenteric myxoid liposarcoma revealed by an abdominal mass. A mesenteric origin was discovered during surgery.

Abbreviation: LPS, liposarcoma; MDM2, Mouse Double Minute 2; CD117, Cluster of differentiation; DDIT3, DNA Damage Inducible Transcript 3.

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Case report

A 69-year-old female presented with a 4-month history of sensation of heaviness associated with an abdominal mass. Past medical history included diabetes, hypertension, and atrial fibrillation managed with Biguanides, Angiotensin-converting enzyme inhibitor, and direct oral anticoagulant. Physical examination found abdominal distension. The waist circumference was 118 cm. On palpation, the patient had a painless abdominal mass extending from the epigastrium to the hypogastrium. An abdominal ultrasound scan showed a large, heterogeneous, hypoechoic intra-abdominal mass measuring 22 cm in size. It also shows left renal cortical atrophy with several stones and hydronephrosis. An abdominal computed tomography scan showed a giant solido-cystic mass containing a mesenchymal component and a liquid component with septa, primarily suggestive of a gastrointestinal stromal tumor (Fig. 1A). It was pushing back the small intestine (Fig. 1C), the

Fig. 1 – Axial section of abdominal CT scan: showing a 21.3 x 15.8 x 22.5 cm heterogeneous and hypodense mass in the peritoneal cavity (A, B, C). Bowel loops are located between the tumor and the anterior abdominal wall (B, C, E). The lesion displaced and entrapped the small bowel loops and aorta (C, D).
men revealed reduced renal parenchyma with hydronephrosis over multiple kidney stones. A distant staging work-up, including thoracic, abdominal, and pelvic CT scans, didn’t show metastatic lesions. No adjuvant therapy was considered necessary. The patient was symptom-free with good wound healing. She remained free of recurrence 6 months after her treatment.

**Discussion**

Liposarcoma (LPS) is a malignant soft tissue tumor of mesenchymal origin, developed from immature lipocytes at various stages of differentiation. It represents 14%-18% of all malignant soft tissue tumors [7]. There are 5 subtypes of liposarcoma according to the WHO classification: myxoid LPS (56.2%) followed by well-differentiated LPS (21.9%), pleomorphic LPS (17.8%) and dedifferentiated LPS (6.8%) [3].

It occurs mainly in the deep soft tissues of the limbs (75%), in particular in the thigh, popliteal fossa, and also in the retroperitoneum [8]. Intra-abdominal localization is uncommon. Cases have been reported in the greater omentum, the small bowel, colon, and mesorectum. Primary tumors of the mesentery are uncommon, and fewer than 25 cases have been reported so far. The mean age is 42 years old (18-67) [9]. There is also a masculine predominance [7]. In our case, the patient is a female, 64 years old.

The discovery is often delayed due to an adaptation of the abdominal cavity to the tumor volume. Patients may present with complaints of gradual abdominal distention, abdominal pain, or complications such as urinary obstruction or occlusive syndrome [4]. Our patient developed abdominal pain without occlusive syndrome despite the giant volume.

Ultrasound remains the first-line examination in cases of abdominal localization. It usually shows a multinodular mass of variable size with an echogenic and heterogeneous appearance [10]. In our case, a large, heterogeneous, hypoechoic intra-abdominal mass was found on abdominal ultrasound.
The diagnosis is sometimes difficult on computed tomography (CT) scans due to similarities with gastro-intestinal stromal tumors. The tumor is frequently hypodense with heterogeneous enhancement [6, 11]. The mesenteric origin is suggested by the presence of intestinal loops between the tumor and the anterior abdominal wall [10]. In the present case, CT scans revealed a giant solido-cystic mass containing a mesenchymal component enhanced by contrast and a liquid component with septa, developed in intestinal loop space, pushing back the small intestine and vessels.

Magnetic resonance imaging is important to assess the extent of the tumor and to characterize its nature. It typically shows a nodular, inhomogeneous mass with pseudo cleavage and well-limited [6].

The anatomopathological examination is important to confirm the diagnosis and to specify the prognostic factors: histological grade, tumor size, tumor necrosis, invasion of intra-tumoral vessels, tumor depth, and the quality of the resection margins, which is a major prognostic factor for local tumor control in most series in the literature [3].

Macroscopically, liposarcoma is often characterized by a soft, fleshy, or firm appearance. The surface is shiny, gelatious, and homogeneous. Tumors that are 5 cm or larger and deep-seated, firm and fixed to underlying structures are usually considered suspicious [10].

Myxoid liposarcoma is histologically defined as a tumor composed of lipoblasts and myxoid stroma with a characteristic vascular pattern of fine, branched, plexiform, and arborescent vessels [11]. In the presence of a typical morphology, there is no indication for immunohistochemical examination. Immunohistchemistry usually shows the absence of HMG2, MDM2, and S100 protein expression [7]. Myxoid liposarcoma is defined genetically by the presence of a translocation involving the DDIT3 gene. The t (12; 16) (q13; p11) translocation is the most frequent (95%), leading to the FUS-DDIT3 fusion gene [5]. The alternative translocation t (12; 22), leading to the EWSR1-DDIT3 fusion gene, has also been found in some patients [5].

The prognosis of a liposarcoma depends on the location and the histological type. In abdominal localization, the prognosis is worse than in other localizations. The median 5-year disease-free survival rates of liposarcomas in the abdominal cavity versus extremities were 41.9% versus 66.7%, and the 5-year overall survival rates were 64.5% versus 84.5%, respectively [1].

The treatment of choice for these tumors is complete surgical excision with wide surgical margins, followed by radiation with or without adjuvant chemotherapy in high-risk patients [11]. Radiotherapy (RT) is typically added to surgery as part of the standard treatment of high-grade (G2-3) lesions. Preoperative RT can offset the negative prognostic impact of R1 margins much more than post-operative RT. Chemotherapy is indicated for patients with unresectable tumors or metastatic disease. In our case, the patient’s treatment consisted of complete surgical resection of the tumor, and no adjuvant treatment was indicated [12].

Preoperative chemotherapy has been reported by one group to be successful in shrinking a large ileocolonic mesenteric liposarcoma. They indicated that the key drugs to be used were doxorubicin, dacarbazine, and ifosfamide [13].

Conclusion

Primary mesenteric myxoid liposarcoma is extremely rare. The diagnosis is often late because of the insidious evolution of the tumor. The overall 5-year survival of this tumor remains low, and surgery appears to be the only option for effective treatment. Clinicians should be aware of the likelihood of primary mesenteric liposarcoma in the case of an abdominal mass to avoid misdiagnosis and suboptimal treatment.

Declaration of figures authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

Author contributions

RM was involved in the analysis of the data and the literature search and wrote the manuscript. FB and GR helped with the patient management and revision the manuscript. MM contributed to the preparation of this manuscript, and interpretation of the case. All the authors have read and approve the final version of the manuscript.

Patient consent

Written informed consent was obtained from the patient’s next of kin for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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