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Diagnosis and Management of Retroperitoneal Myxoid Liposarcoma: A Case Report

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Abstract

Retroperitoneal tumours are one of the rarest malignant tumours encountered in clinical practice. It is usually diagnosed by a combination of clinical, radiological and histopathological analysis. There are multiple histological types of liposarcoma and each type varies in terms of response to radiotherapy and incidence of recurrence. Surgical excision is the mainstay of treatment and providing negative margins is the key, which may even involve resection of adjacent organs. Here we describe a case of a huge retroperitoneal myxoid liposarcoma and how we managed the patient comprehensively.

Keywords: Liposarcoma, Myxoid liposarcoma, retroperitoneum

Introduction

Mesenchymal tumors of the retroperitoneum usually appear with an advanced illness and have a dismal prognosis. Due to their anatomical placement and rarity, these malignant tumors might provide multiple therapeutic hurdles as well as a diagnostic conundrum. They typically require long-term and frequently indefinite follow-up because they are linked to a high rate of recurrence even after anatomically complete resection.

These are uncommon tumors, making up only 1%–2% of all malignancies that are solid. Most sarcomas don't occur inside the retroperitoneum. Retroperitoneal sarcomas account for only 10%–20% of sarcomas, with a frequency of 0.3%–0.4% per 100,000 people. Though they can happen at any age, the fifth decade of life is when they are most common.[1]

These patients usually present later in life because these tumors grow quite large without causing symptoms since they originate in the enormous potential spaces of the retroperitoneum. Furthermore, when symptoms do arise,

they are often vague (e.g., fullness and pain in the abdomen) and can be written off as the result of unrelated, less serious conditions. As a result, retroperitoneal sarcomas typically have a very large size at the initial diagnosis.[2]

Case Report

51 year old male, a known smoker and alcoholic for the past 20 years, presented to hospital with complaints of mass per abdomen for 8 months, diffuse abdominal pain for past 4 months, insidious in onset, gradually progressed to current state with sharp shooting pain, aggravated at night, associated with nausea and altered bowel movements. He had an episode of fever 15 days ago and He had unintentional weight loss of 12 kg in the past 6 months.

On examination, abdomen appeared distended, umbilicus in midline. No scars, sinuses, dilated veins or visible pulsations. A firm 15x20 cm mass was palpable, extending medially from the midline to left mid axillary line laterally, inferiorly up to umbilical line, superior border not palpable, smooth surface with areas of nodularity. Tenderness present in the epigastric, left hypochondrium and left lumbar region. No guarding, rigidity or rebound tenderness. Bowel sounds were heard.

Figure 1: Image showing large mass abdomen

Blood investigations were within normal limits. CT whole abdomen showed a large ill-defined heterogeneously (predominantly hypodense) enhancing soft tissue density mass with probable few septations, probably arising from the left side of the peritoneum with fat contents, causing displacement of fundus and body of stomach and oesophago-gastric junction to the right as shown in the Figures 2,3.

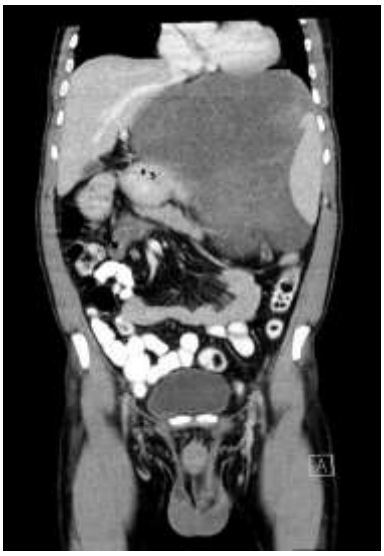

Figure 2: Coronal section showing the huge retroperitoneal tumour

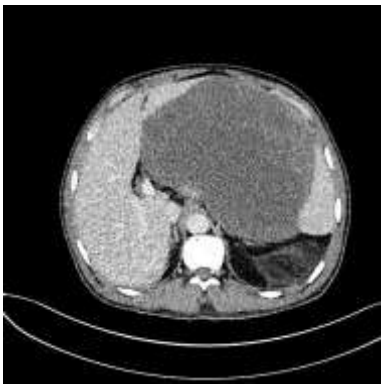

Figure 3: Axial section showing the huge retroperitoneal mass

Ultrasound guided trucut biopsy of the mass showed low grade epitheloid to spindle cell neoplasm. From the history, clinical examination and the diagnostic tests, a

provisional diagnosis of retroperitoneal sarcoma/liposarcoma was made.

The patient then underwent retroperitoneal lesion excision biopsy under general and epidural anesthesia. Roof top incision was made to access the peritoneal cavity (Figure 4). Retroperitoneum was opened by Mattox maneuver and retroperitoneal tumor was identified. Blunt and sharp dissections carried out all around the mass and tumor pseudo capsule was identified and dissected out. Adhesions were released and tumor in the intra peritoneum excised separately along with the spleen, by dividing the splenorenal ligament and splenic pedicle. A large intraperitoneal mass approximately weighing 3kg, whitish, lobulated, firm to hard with smooth surface, with dense adhesions between the mass and the anterior/ lateral abdominal wall, spleen, diaphragm and left lobe of liver (Figure 5). Adhesions were excised along with a smaller, whitish, lobulated, firm to hard swelling with a smooth surface with dense adhesions to the retroperitoneum. Gerota's fascia was free from tumor. Lesion appeared to be dual in nature- one in retroperitoneum and one in lesser sac, both with similar gross features; lesser sac seeding of retroperitoneal tumor with separate retroperitoneal lesion (Figure 6).

Figure 4: Roof top incision

Figure 5: Lesion being dissected

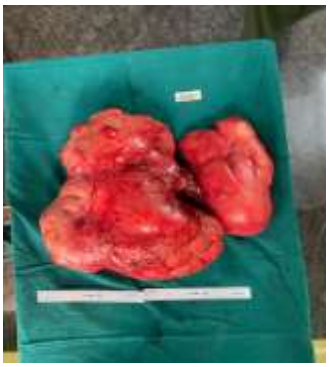

Figure 6: Two separate tumours after excision

Histo-pathology report showed that the larger specimen measured 30.5x23x10.5 cm and smaller specimen 19x14x7cm.Sections studied showed neoplasm composed of mature adipocytes, spindle and stellate shaped cells, many signet ring and multivacuolated lipoblasts (Figure 7) embedded in pool of loose myxoid stroma (Figure 8). Sections from splenic hilar vessels and parenchyma show normal histology and are free of tumor.

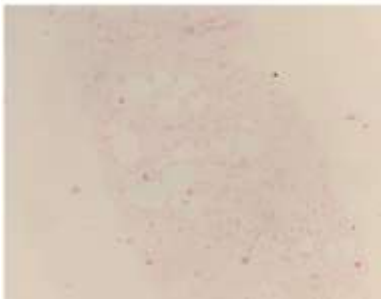

Figure 7: Microscopic image showing multivacuolated lipoblasts

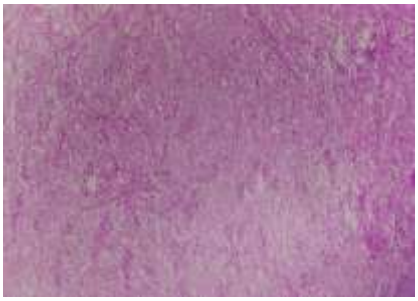

Figure 8: Microscopy showing myxoid stroma

The neoplastic cells were negative for immunohistochemical markers SMA, Myogenin, Desmin, CD 34, S 100. Ki 67 less than 1 percent.

These findings were suggestive of myxoid liposarcoma, with no identifiable necrosis and lymphovascular invasion. All margins were negative for tumor.

Postoperative course was uneventful and patient was advised on the medications to be continued, wound care, vaccination against OPSI. Radiotherapy was not advised due to large area to be irradiated. He is thus on regular follow-up.

Discussion

Retroperitoneal liposarcoma (RPLS) is an uncommon tumour that can be difficult to operate on, and with the right care, a medium-term survival is possible. The recommended course of treatment is surgery because there is no definite proof that chemotherapy or radiation therapy are curative.[3]

The four forms of liposarcoma that are recognized now are determined by morphologic characteristics and chromosomal abnormalities. The four categories are as follows: (1) myxoid/round cell; (2) de-differentiated; (3) pleomorphic. For patients with liposarcoma (LS) following resection, the degree of differentiation, as shown by histological grade, continues to be the most significant predictor of the clinical course and final outcome. The anatomical distribution seems to be closely associated with its histologic type: well-differentiated/dedifferentiated LS primarily occurs in the retroperitoneum, while myxoid/round cell and pleomorphic LS preferentially affect the extremities.

The round cell component's extent determines the histological grade of myxoid/round cell liposarcoma, which is a morphologic continuum. Patients with low-grade myxoid liposarcoma, which is defined as <5% round cell areas, have a 90% 5-year survival rate, according to recent research from Memorial Sloan-Kettering Cancer Center (MSKCC). Patients with high-grade myxoid/round cell liposarcoma, which is defined as >5% round cells, have a 60% 5-year survival probability.[4]

Because of the enormous intraabdominal space, liposarcoma can grow without squeezing important organs, leading to delayed diagnosis. When the retroperitoneal liposarcoma does show clinical symptoms, it has typically spread to engulf adjacent organs and become very big.[5]

Crucial information on tumor grade can be obtained from preoperative CT. Particularly interesting are the encasement of major veins, involvement of nearby organs, and detection of metastases to the liver and lungs. While CT is a valuable tool for characterizing liver lesions, ultrasound and magnetic resonance imaging (MRI) provide more in-depth details regarding the involvement of the muscles and neurovascular system.[6]

Even after the macroscopic illness has been completely eliminated, these sarcomas have a significant probability of recurrence. Reluctance to get macroscopic clearance is typically associated with the tumor's size and the requirement for visceral excision. The majority of patients eventually experience recurrent disease and almost certainly leads to death, even though it may take

a long time for the tumor to return. Technically more challenging, subsequent surgery is only recommended as a palliative measure.

There are differences between left- and right-sided LPS in terms of the order in which the surgical procedures are performed and where the tumor is in relation to the abdominal viscera.

Right side: To determine whether the IVC is involved, a right medial visceral rotation en bloc with the tumor may be necessary for right LPS. The duodenum can be retracted and the infrahepatic cava vein can be fully extended using a wide Kocher technique. The duodenum and the pancreatic head can both be preserved with this method. The goal of the surgery at this point is to pass through the tumor pseudocapsule (i.e., marginal dissection) and release it from these organs because duodenum-pancreas resection does not improve disease-free survival and is instead linked to the highest complication rates. However, in cases where the dissection of the pancreatoduodenal junction from the tumor results in duodenal perforation because the wall has been thinned by tumor invasion or compression, partial resection ought to be taken into consideration. Very seldom, a duodenopancreatectomy is necessary.

Left side: It's crucial to release the Treitz angle widely in the left LPS without harming the duodenum. The third duodenal segment should be removed and rebuilt via a duodeno-jejunostomy when there is obvious infiltration. A distal spleno-pancreatectomy or possibly a diaphragm resection is necessary for malignancies located in the left upper quadrant.[7,8]

Radiosensitivity varies among the different types of retroperitoneal sarcomas. To assess the viability and results of pre-, intra-, and postoperative radiation therapy in the treatment of retroperitoneal sarcomas, a number of observational and retrospective studies have been reported. Preoperative radiation therapy has several benefits, such as the tumor's clear demarcation for radiotherapy planning, its displacement of some radiosensitive nearby organs, and the possibility of a reduced therapeutic dosage of radiation in the preoperative context. With postoperative radiation, patients with the highest recurrence risk can be identified based on their grade and margin status. On the other hand, the risk of radiation-related toxicities will increase in the postoperative context as the surrounding organs will migrate toward and attach to the tumor bed.[9]

Adjuvant chemotherapy is not very helpful for low-grade, well-differentiated liposarcoma since these lesions have a very low mitotic rate. When treating higher grade subtypes like dedifferentiated, myxoid/round cell, or pleomorphic liposarcoma with adriamycin and ifosfamide chemotherapy, up to 50% of patients experience partial responses but their overall survival is not significantly improved. Furthermore, fewer than 10% of patients have shown complete response rates to treatment. Thus, the most crucial aspect of therapy remains total surgical excision.[10]

Conclusion

Thus, myxoid retroperitoneal liposarcoma, is an extremely rare tumor with prognosis depending on the location, histological type and size of the tumor. Surgical resection of the tumor is the mainstay of treatment. However, obtaining negative margins does not reduce the incidence of recurrence of the tumour. Radiotherapy is recommended based on the histological type of tumour. However, in cases of extremely large tumours, owing to the large area to be irradiated, its most often not recommended.

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