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Case Report

Liposarcoma of mesorectum presenting as chronic intestinal obstruction: a rare case

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ABSTRACT

The term 'liposarcoma' refers to a spectrum of neoplastic lesions that can be benign to malignant and likely to recur or metastasize. Liposarcoma is a common soft tissue sarcoma found in adults and occurs mostly in extremities especially thigh followed by retroperitoneum. Here we present a case of spindle cell variant of well differentiated liposarcoma of mesorectum and underwent Anterior resection for the same. The treatment of choice for liposarcomas of retroperitoneum remains debatable and there are no clear-cut guidelines available regarding the same. However, considering the higher rates of local recurrence we advise a complete surgical excision following anatomical principles.

Keywords: Liposarcoma, Mesorectum, Retroperitoneum, Recurrence

INTRODUCTION

Liposarcoma is a malignancy of connective tissue that resembles fat cells when examined under microscope. They are common in adults and account up to 20% of all sarcomas. They have higher incidence in 40 to 60 years age group without any difference in frequency among sexes. Nearly 75% of liposarcomas occur in limbs (mostly in thigh), 20% in retroperitoneum and very smaller percentage in uncommon locations like inguinal region, mesentry etc.[1] Etiology of liposarcoma remains unknown and they are mostly idiopathic. WHO classified liposarcoma into: well-differentiated (most common), pleomorphic, round-cell, myxoid and dedifferentiated type.[2] Most of the times they are asymptomatic, unless they grow very large and cause pressure symptoms. So, they are very difficult to diagnose in early stages They have higher rates of recurrence and it increases with the grade of the tumor. High grade tumors have poor prognosis and high metastatic potential. Surgical resection is the only accepted modality of treatment, chemotherapy or radiotherapy have no proven role. They have higher recurrence rates. Hence it is important to do proper surgical resection to prevent any chances of recurrence. The types of resection and radicality of the surgery depends upon the location of tumor and its relation to the surrounding structures. The peculiarity of this tumor lies in the infrequency of anatomical localization of tumor.

CASE REPORT

A 28-year-old female P3, L3 and tubectomies presented to our out-patient department with complaints of heaviness in abdomen for 2 months. No history of vomiting, Constipation or any other associated symptoms. No other co-morbidities were present. On examination, an ill-defined, mobile, firm mass felt in

hypogastrium extending up to 2 cm below umbilicus. On per vaginal examination, cervix and uterus were pushed anteriorly and mass was palpable posteriorly along with fullness in pouch of Douglas. On Per rectal examination, mucosa was found to be free of tumor and mass felt in posterior border suggesting an extrinsic mass. Contrast Enhanced Computerized tomography (CECT) of abdomen suggested a 13 × 12 cm cystic mass located in pelvis extending superiorly into abdomen (Figure 1).

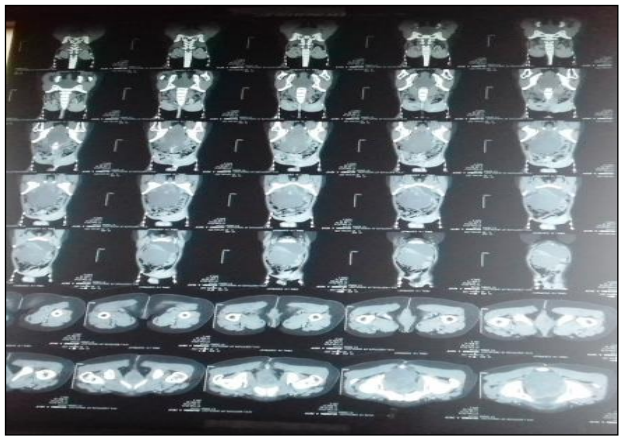

Figure 1: CT image showing mass in retroperitoneum.

MRI of pelvis was done which revealed that the mass pushed uterus anteriorly, abutting the sacrum posteriorly, rectum and sigmoid were displaced anterolaterally to the left side suggesting a retroperitoneal mass. Colonoscopy was normal and no evidence of any intraluminal extension. HCG, CA125 and CEA were within normal limits. All the routine blood investigations were performed and are normal. USG Guided FNAC was attempted but it was inconclusive.

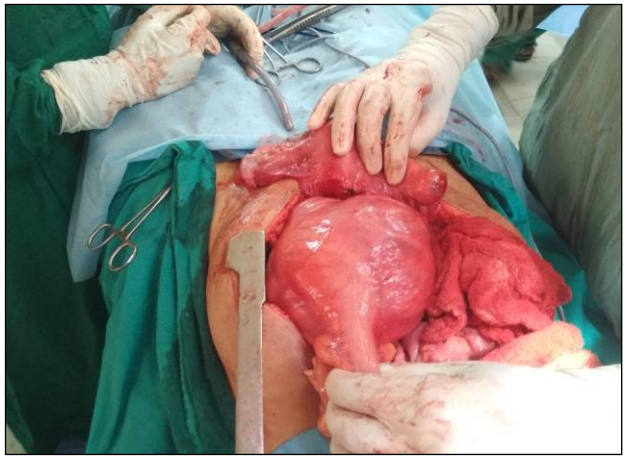

Figure 2: Intra-operative picture showing growth arising from mesorectum.

All the features suggestive of a retroperitoneal mass arising from mesorectum. Surgery was planned after routine examination for metastasis was found negative. Intra-operatively a firm solid mass was found to be arising from the mesorectum and mesocolon extending up to the pelvic floor. Wall of the mass was formed by the rectum and hence no plane for dissection could be attempted. Hence En-Bloc anterior resection of tumor along with rectum was done up to the pelvic floor, distal anal canal was closed, and a Sigmoid end colostomy was fashioned. B/L ovaries and uterus were spared. No macroscopically enlarged lymph nodes were found. Wound closure was done, and post-op was uneventful.

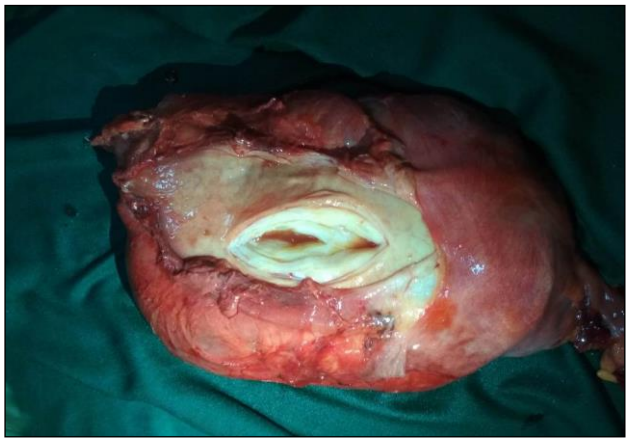

Figure 3: Resected specimen.

On Gross examination, 27 × 26 × 8 cm globular greyish white mass was found arising from intestine and the adjacent intestinal segment measuring 9 cm (Figure 2 and 3). The mass has solid component along with multiple cystic spaces measuring 1 to 5 cm filled with yellow gelatinous material. On Histopathological examination, all the margins were found to be clear of tumor and microscopy revealed the lesion consist mostly fibro collagenous tissue along with few congested blood vessels, focal areas show few spindle cells with elongated nucleus with moderate amount of cytoplasm. There are areas of fibro collagenous and fibro adipose tissue along with mild pleomorphism. All the features suggestive of a low-grade spindle cell tumor favoring liposarcoma. Immunohistochemistry was done, and the tumor was found positive for Bcl-2, vimentin, desman, CD34, CD56, CD10, and CD99 thus confirming diagnosis spindle cell liposarcoma which is a subtype of well differentiated liposarcoma. Patient was not advised any radiotherapy or chemotherapy as the margin clearance was satisfactory and patient was kept on regular follow-up.

DISCUSSION

Liposarcoma is a tumor derived from primitive cells that undergo adipose differentiation. The term 'liposarcoma' refers to a spectrum of neoplastic lesions that can be benign to malignant and likely to recur or metastasize. Evans was the first to characterize a liposarcoma in 1979. He described a combination of WDLPS and a non-lipogenic dedifferentiated sarcoma-like component.[3] They occur commonly in extremities followed by

retroperitoneum. Of all the retroperitoneal tumors, liposarcoma of retroperitoneum is the commonest sub-type with an incidence of approximately 2.5 per million.[4] It usually occurs in between 40 to 60 years of age with male to female ratio of 1:1.[5]

The exact etiology of liposarcoma remains unknown and more mostly idiopathic in nature. However, risk factors for development of liposarcoma include radiation, familial syndromes like Neurofibromatosis and gardener's syndrome, damaged lymphatic systems and chemicals like PVC and dioxin[1]. A number of cytogenetic correlations also have been made with liposarcoma, well differentiated liposarcomas have been found to associated with abnormalities of q13-15 region of chromosome 12.

According to WHO, Liposarcoma can be histologically subdivided into 5 subtypes: Well-differentiated, myxoid, round cell, pleomorphic and dedifferentiated. Round cell, pleomorphic and dedifferentiated subtypes are regarded as high-grade; whilst well-differentiated and myxoid liposarcoma are low-grade.[6,7] High grade tumors have a negative prognostic value, where as well differentiated liposarcomas have very low metastatic potential but are known to have local recurrence. Pleomorphic liposarcomas have high local recurrence.

Liposarcomas are usually asymptomatic unless they cause compression or invade into the surrounding organs. The large volume of intra-abdominal compartment allows the liposarcomas to grow very large without causing any symptoms. Therefore, its very unusual to identify liposarcomas in early stages. It is difficult to diagnose the liposarcomas especially in retroperitoneum based onclinical and local examination. Hence investigations like Ultrasonography, CT and MRI are very useful in arriving at a preliminary diagnosis. These investigations are helpful in assessing the extent, size, consistency of tumor and thus aid in planning of the surgical treatment. Ultrasonography helps in identifying the rough size and consistency of the tumor. On a CT scan, retroperitoneal liposarcoma usually appears as a large encapsulated mass containing variable amounts of fatty and soft tissue components.[8] MRI is important for the diagnosis of liposarcoma invasion of the abdominal aorta or inferior vena cava. Biopsy is not generally recommended due to the probability of tumorseeding.[9]

Complete surgical resection remains the mainstay of treatment especially considering the higher recurrent rates associated with the liposarcomas. Therefore, the aim of surgical treatment is to obtain a R0 resection. However complete resection is always a challenge especially In well differentiated subtype as margins are not always distinguishable and usually the tumor along with invaded organ is resected.[10] There is no evidence supporting the role of chemotherapy or radiotherapy in improving the overall surivival rate.[11] The prognosis for liposarcoma remains very poor, the 5 year survivial for well differentiated liposarcoma is 83% and for dedifferentiated sarcoma is 20%.[7] However, the survival rates in patients who received complete surgical treatment is much higher when compared to the patients who did not receive it and for this reason Complete surgical resection remains as the gold standard treatment of choice.[12] In order to detect any recurrence a regular follow up with necessary investigations should be advised to the patient after the surgical treatment.

CONCLUSION

Even though liposarcomas are common tumors in extremities of adults, they are relatively rare in mesorectum. Often patients present with vague symptoms and by the time diagnosis is made, it requires a major surgical resection. Surgical resection with clear margins remains the mainstay treatment. Role of chemotherapy or radiotherapy is not yet evident. Overall survival rate in patients is improved after surgical resection.

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