A Giant primary mesenteric Liposarcoma in an adolescent male: A rare case with presentation in unusual age and location

Vandana Rana¹, Ranjan Praveer¹, Singh Giriraj², Rathi Khushi Ram¹

ABSTRACT
Primary mesenteric Liposarcoma is a rare neoplasm and only few documented cases are there in literature that too in adults. We report a case of giant primary mesenteric Liposarcoma in an adolescent male. Our patient is a 17 yrs old male who presented with gradual abdominal distension and was found to have huge multilobed and heterogeneous abdominal tumor arising from transverse mesocolon on imaging. The excised tumor weighed 19 kg and was reported as well differentiated Liposarcoma (WDLPS) on histomorphology. Atypical Lipomatous Tumor and WDLPS are synonyms describing lesions that are identical morphologically, karyotypically and in terms of biological potential. In deep visceral sites where wide excision margin are difficult to attain, the term WDLPS is preferred. Myxoid Liposarcoma is the most common variant of LPS seen in children and young adults with lower extremities being commonest site of occurrence. Our case is unique in rarity of lesion, its dimensions, location and uncommon histomorphology for age.

Key Words: Well differentiated Liposarcoma, Atypical lipomatous Tumor, Mesentery, Giant, Adolescente.

Key Messages: Liposarcoma is usually a malignancy of adults and constitutes <3% of sarcomas in children. Lower extremities and retroperitonium are its usual sites of occurrence. Retroperitoneal Liposarcoma are known to grow to large sizes. Primary mesenteric Liposarcoma is a rare entity and can also reach great dimensions and should be kept as one of the differential of gradual abdominal distension.

INTRODUCTION
Liposarcoma (LPS) is the most common type of soft tissue sarcoma in adults.¹ According to WHO classification it is divided in to four subtypes—Atypical lipomatous tumor (ALT)/ Well differentiated LPS (WDLPS), Myxoid/ Round cell, Pleomorphic and Dedifferentiated LPS.² Among these WDLPS are the commonest in adults and are seen more in the deep soft tissue of the limbs (75%) followed by retroperitoneum.³ The common age group involved is between 50 to 70 years.¹ These sarcomas are very rare in childhood and constitute about 2% of all childhood soft tissue sarcoma with peak incidence in second decade of life. Myxoid histology is most common in children with lower extremity as common primary site.⁴ Primary Intraperitoneal or mesenteric LPS are very rare and only few cases that too in adults have been reported in literature.⁵ Retroperitoneal and abdominal LPS are known to grow to large sizes however, after exhaustive search of written English literature; we report most probably the first case of a huge intra- abdominal LPS in an adolescent male.

CASE HISTORY
The patient is a 17 years old male presented in our institute with complaints of abdominal distension and pain since last three years. Distension was insidious in onset, gradually progressive and associated with pain in abdomen which was dull aching and associated with two to three episodes of vomiting initially. He was thin built and was not gaining weight. There was no history of altered bowel habits, malena or urinary complaints. General examination showed thin built and presence of pallor. On per abdominal examination, he was having grossly distended abdomen with dilated veins and flat, stretched umbilicus (Figure 1). On palpation there was a huge, firm and painless lump filling almost the whole of abdomen. The lump was smooth, with irregular margins and moved with respiration. No systemic examination was normal. Serum Tumor markers were not raised. On USG it appeared ill defined heterogeneous mass lesion extending from epigastric region to the pelvis. No significant vascularity noted on Doppler. Differential of intraperitoneal benign teratoma was offered. Plain and contrast CT abdomen showed...
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intraperitoneal bizarre soft tissue mass measuring 20.5×28×38 cm (AP X TR X CC) extending from the epigastric, filling both the hypochondrias and reaching to pelvic region (Figure 2). It was having density of soft tissue, fat, fluid and multiple areas of calcification. Multiple septations were also noted. The mass was seen displacing the abdominal organs superiorly and posterior-laterally. It is seen extending through the transverse mesocolon. Severe compression and displacement of mesenteric vessels also present. No organomegaly, signs of obstructive uropathy, significant lymphadenopathy or free fluid was present. Exploratory laparotomy and excision of the tumour was done. Per operative findings confirmed the origin from transverse mesocolon. No infiltration into the surrounding structures was seen. The excised specimen was received in our lab as multiple nodular masses with largest mass measuring 45×37×14 cm and the whole specimen weighed 19 kg (Figure 3) The specimen was multilobated, ivory to yellow coloured, soft to firm with semitransparent membranous capsule. Most of the lobes on cut sections showed yellowish gelatinous appearance with many of them showing the secondary changes in the form of haemorrhage, calcifications, focal cystic degeneration and few myxoid looking areas. Microscopy revealed encapsulated lesion comprising of mature adipocytic areas with transgressing capillaries and fibrous septas along with many sclerosing areas having scattered bizarre stromal cells and some multinucleated floret cells (Figure 4). Occasional multivacuolated lipoblast seen. Areas of hemorrhage, cystic degeneration, focal calcification and myxoid change present. Few foci show perivascular presence of scanty lymphocytic cell infiltrate. No areas of necrosis or mitotic activity seen. The lesion was reported as Atypical lipomatous tumor/ Well differentiated LPS (WDLPS) and patient is kept on follow up.

DISCUSSION

Liposarcoma is one of the most common soft tissue sarcomas of adult life with relative incidence of 9.8% to 16% among other sarcomas but they rarely occur in children.[1] The common age group involved is between 50 to 70 years.[1] Our patient was just 17 yrs of age at presentation with a history of progressive distension of abdomen for last three years. There was a history of growth compromise and inability to gain weight. Although, approximately less than hundred cases of LPS in children and adolescents has been reported in written literature till date still they account for <3% of all paediatric sarcomas.[4] One of the largest case series of LPS in young patients found lower limb to be the commonest involved location with only three cases to be of abdominal LPS out of 82 cases. All the three cases were histologically proven myxoid LPS.[7] Intra abdominal or primary mesenteric LPS are rare occurrence and approximately less than 20 cases mostly in adults have been reported in English literature till date.[5,4] Our patient was detected to have primary mesenteric WDLPS which is highly unusual for this age and anatomic location.

The most recent WHO classification of soft tumors recognizes four categories of LPS: ALT/WDLPS (Intermediate grade), Dedifferentiated, Myxoid/round cell and pleomorphic LPS.[2] WDLPS is the most common form of LPS accounting for 40–45% of all LPS and are encountered in late adult life and 75% of the cases are known to develop in the deep muscles of the extremities, 20% in the retroperitonium and the remaining in the groin, spermatic cord and miscellaneous sites.[1,3] ALT and WDLPS are synonyms describing lesions that are identical morphologically, karyotypically and in terms of biological potential. In deep visceral sites like retroperitonium and mediastinum where wide excision margin are difficult to attain, the term WDLPS is recommended and justifiable.[2] In our case the anatomic site being intraabdominal, the lesion was reported as WDLPS. ALT/WDLPS show three main morphological patterns: adipocytic, sclerosing and inflammatory and they all can be seen in same lesion as seen in our case also.
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Histologic grade is one of the most important predictors of outcome, with low grade myxoid tumors having significant improved survival rates than round cell, pleomorphic and dedifferentiated types. ALT/ WDLPS are histologic grade 1 tumors with usually low Ki 67 proliferative index. They are characterized by supernumerary ring and giant marker chromosomes which can be detected by cytogenetic and FISH studies. Murine double minute 2(MDM2) gene is consistently amplified and over expressed in cases of ALT/WDLPS. MDM2 and or CDK4 nuclear immunopositivity is present in most cases and also found to be correlated with higher Ki 67 proliferation index.[2,4] In our case the Ki 67 index was low and no areas of necrosis or dedifferentiation was present. Dedifferentiation occurs in up to 10% of WDLPS with risk being higher in deep sited LPS.[5] There is substantial amplification of MDM2 in Dedifferentiated LPS.

LPS are known to grow in large dimensions especially in retro peritoneum due to presence of potential space and delayed symptoms. Reports of many giant retroperitoneal LPS are there in literature with largest reported to be weighing 42 kg.[8] However, largest abdominal or mesenteric LPS reported in literature is not more than 15 Kg.[9,10] Sato et al found that the tumor size greater than 20 cm predicted a significantly poorer prognosis.[10] The tumor in present case was huge and weighed together as 19 kgs. After thorough search of literature, ours is most probably the first case of giant intra abdominal WDLPS in an adolescent male.

Surgical resection with clear margins is the treatment of choice for primary mesenteric LPS. Role of Radiotherapy and systemic chemotherapy remain to be established. Our patient is presently asymptomatic and is on follow up after surgical removal of tumor.

CONCLUSION

Primary mesenteric Liposarcoma though a rare entity and more so in children, should be kept as one of the differential of gradual abdominal distension. Awareness about lesion and correlation with clinical presentation, Imaging and histopathology helps to clinch the diagnosis.

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CONFLICT OF INTEREST

Nil.

REFERENCES


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