

A Giant Mesenteric Lipoblastoma in a Two-Year Girl: A Case Report

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Abstract:

Back ground: Abdominal lipoblastomas which are benign but uncommon soft tissue tumors in children rarely arise from the mesentery. However, benign, these may attain in large size and cause compression symptoms. Complete resection and close post-operative monitoring are necessary for avoidance of recurrence. Histopathology with immunohistochemistry in conjunction with the morphology is the gold standard for diagnosis. Herein such a rare case of mesenteric lipoblastoma with huge abdominal distention is reported.

Case presentation: A 2-year-old girl presented with an enormous abdominal swelling occupying almost all quadrants of the abdomen with repeated dull abdominal pain. The swelling was a non-tender and tensely cystic. USG and CT Scan could not distinguish the proper origin or location of the mass but demonstrated as a large encapsulated and lobulated fatty mass measuring about (16.4x10.0x18.6cm) involving right hypochondriac, epigastric, bilateral ilio-lumber, umbilical & hypogastric regions, pushing the bowel loops upward & laterally with mild mesenteric thickening of perilesional mesentery and separated from the liver & both kidneys and other intraabdominal organs. After exploratory laparotomy there revealed a well capsuled large (about 15x11x14cm) soft yellow mass displacing the entire intra-abdominal contents, connected by a single vascular pedicle with the mesentery and loosely enclosing the mesentery as a whole. Histology and immunohistochemistry of completely resected mass confirmed the presence of mature adipocytes and immuno-morphology was compatible with Lipoblastoma.

Conclusion: This case represents one of the largest mesenteric lipoblastomas in accordance to the patient's age that histologically, mostly composed of mature adipocytes with few lipoblasts.

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Introduction:

Lipoblastomas are rare benign mesenchymal neoplasms of embryonal fat cells, comprising of adipocytes and lipoblasts usually encapsulated and occurs in infants and

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young children with having a tendency of local recurrence^{1,2,3,4,5,6}. Majority (80-90%) of the cases are detected under the age of 3 years with a male predominance (3:1)⁷. The sites of a lipoblastoma varies in percentage from study to study. Some studies revealed extremities are the most common site (61%) and this is followed by trunk (15%), abdomen (14%) and in head and neck region (11%) whereas, other studies demonstrated a preponderance of the tumor site is in the trunk (44%-48%) and these may also arise in the spine, mediastinum, lungs, trunk, mesentery, peritoneal cavity, retro peritoneum and inguinal canal^{1,8,3,9,10,11,12,13,14}. Among them abdominal lipoblastomas which are uncommon comprising around 7% of all reported lipoblastomas and most commonly found in the retroperitoneum. Only less than 15 reported cases abdominal lipoblastoma were noticed to arise in the mesentery^{15,16}. We report such a rare childhood soft tissue tumor that was noticed in the abdomen of a 2 years female

child presented with huge abdominal distension along with intermittent abdominal pain. Exploratory laparotomy revealed a giant intraabdominal mesenteric mass and both histopathological and immunohistochemistry jointly reported this as a case of lipoblastoma. The most important clinical aspect to report this case is to avoid the diagnostic dilemma regarding diagnosis of other similar intraabdominal childhood neoplastic conditions and accurate diagnosis of such a rare variant of childhood soft tissue mass that is mesenteric lipoblastoma.

Case Report:

A 2 years old female child, presented with a gradually developing huge abdominal swelling occupying almost whole abdomen associated with dull aching intermittent pain, occasional nausea and non-bilious, non-projectile vomiting for last 8 months. The child had history of anorexia along with occasional constipation and gradual weight loss during the last 5 months of her illness. She did not have any urinary complaints related to any other systemic involvement. On examination the child was ill-looking, afebrile, mildly anaemic, mildly dehydrated, normotensive and non-icteric and abdominal examination revealed, there was a large softly cystic nontender mass measuring about (18x16cm) in diameter occupying about all quadrant of abdomen, mostly prominent on right side. The mass was with smooth surface but ill-defined in margin and slightly movable from side to side. The whole abdomen was distended with fullness of right flank, dull on percussion, but shifting dullness & fluid thrill were absent. Liver was just palpable below the right costal margin; spleen was not palpable & kidneys were not

ballotable. External genitalia showed normal findings with intact hernial orifices. There was no regional lymphadenopathy and the other systemic examination along with per rectal examination revealed no abnormality.

All laboratory reports (hematological and biochemical) including Urine R/E, Urinary V.M.A (4.23mg/24hrs), Alpha-fetoprotein (1.77ng/ml), β HCG (>1mIU/ml) level were within normal limit except a small raise of LDL level (650U/L). Abdominal radiographs showed nothing abnormality except dis-tended bowel loops in the upper abdomen with air filled rectosigmoid region. As 1st the USG was inclusive because of huge abdominal distention with distended loops of intestine, repeat one was done and that revealed a soft tissue echo large lobulated mass occupying the right lower lumbar region & the umbilical area, pushing the bowel loops upward & laterally, mass was separated from the liver & both kidneys, size of mass could not be well evaluated due to huge size & in single scan could not be measured, bowel loops are dilated, loaded with gas & fecal matter. The CT Scan of abdomen described as- a well delineated encapsulated soft tissue mass with fatty component measuring about (16.4x10.0x18.6cm) occupying right hypochondriac, epigastric, bilateral ilio-lumber, umbilical & hypogastric regions. There was also noted moderate inhomogeneous attenuation of the capsule of the lesion and mild mesenteric thickening of perilesional mesentery. The mass was noted to compresses the intestinal loops towards the left side though the demarcating fat plane was seen in between the mass and regional intestinal loops.

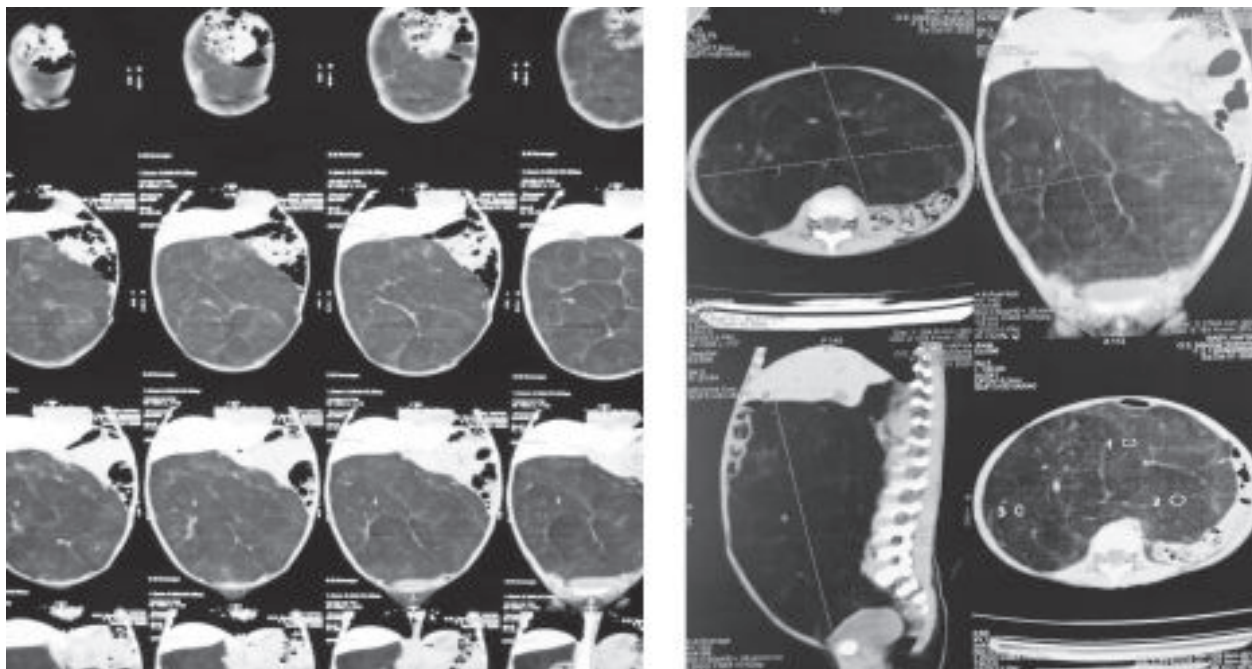
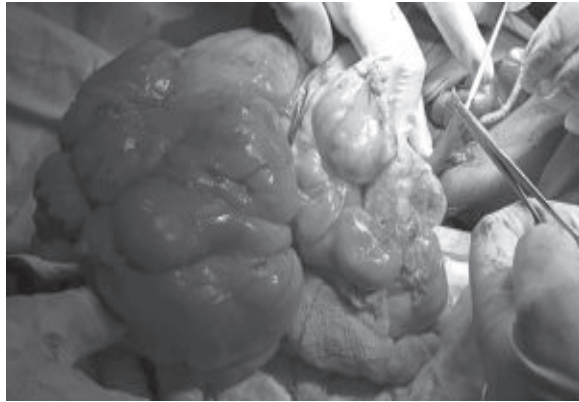


Fig: CT scan of abdomen

After exploration there was delineated that an encapsulated, lobulated, irregular large mass approximately ((15x11x14cm)), with smooth yellowish fatty outlook, involving almost all quadrant of abdomen & loosely adherent with surrounding structure particularly mostly with mesentery and was easily separable from surrounding structures. Complete excision of the mass was done and the whole mass with its capsule was sent for histopathology and immunohistochemistry separately.



Per-operative images:



Per-operative images (after anesthesia):

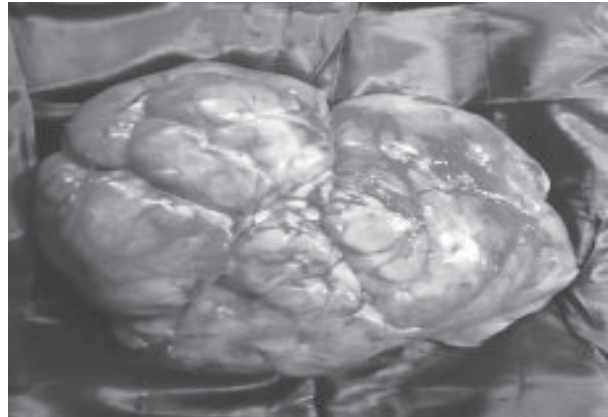


Image after complete excision:

Histopathology report revealed one tan yellowish nodular soft tissue, measuring about (14x11.5x5cm) with smooth surface, slimy & capsulated. The neoplasm composed of adipocytes & of various sizes, some of cells are vacuolated & some others have eosinophilic cytoplasm and this is suggestive of a case of Lipoblastoma. Immunohistochemistry (IH-1229/20) was performed on formalin fixed, two para-embedded block of a mass using standard techniques with antibodies to S-100 protein, CD34, and Mib-1. Histologic sections revealed circumscribed neoplasm composed of adipose tissue having various stage of maturation with few lipoblasts. The tumor cells on immunohistochemistry show- Immunophenotype S100L: positive, CD34: positive, Desmin: Negative. Immuno-morphologically consistent with Lipoblastoma.

Discussion:

Lipoblastoma which is a rare benign soft-tissue tumor, vast majority (80%) are detected in children under 3 years of age and less majority (40%) before 1 year of age. Among the abdominal lipoblastomas, mesenteric one is very rare and slow-growing, usually mobile, soft and do not infiltrate the surrounding organs^{15, 17} Studies^{2, 3} however, reported a male predominance of lipoblastoma the present case is

described with the complain of a female child. Literature revealed, majority of tumors are of more than 5 cm in size and the mean size is 10.8 cm. which may be related to late presentation owing to poor socio-economic conditions and health facilities². Clinically most of the lipoblastomas are painless but sometimes intraabdominal or mesenteric lipoblastomas may present with the features of pain, vomiting and gastrointestinal obstruction^{9, 18}. In present reported case, a 2 years old girl presented with a in initial slow growth of abdominal mass that rapidly grew for the last five months along with recurrent abdominal pain. As the abdomen was hugely distended because of the large size of the tumor during her presentation at our institution the mobility of the mass was restricted only in side to side with difficulty in measuring the size and the consistency was almost tensely cystic.

The accurate pre-operative diagnosis of lipoblastoma is very difficult¹⁸. However, USG may help to outline the mass as a highly-echogenic or sometimes with hypo-echogenic patches, and CT scans can delineated a non-enhanced, fat-density and sharply marginated mass with few internal septations but cannot give any conclusive diagnosis^{18, 19}. Besides, image techniques may be indefinite or equivocal and do not always reliably

differentiate benign from malignant tumors²⁰. In our case there was also diagnostic difficulty because of inconclusive evidences of diagnostic reports.

Lipoblastoma, though recognize as a benign neoplasm, however, has a tendency of recurrence rate 14% to 25 %²¹ because of its locally invasive nature. Complete Surgical excision without sacrificing the surrounding vital structures or ablation of any tissue that could lead to major deformity is the basic principle of treatment except in those infiltrating tumors requiring mutilating excision²². Hence, in this reported case, considering the basic principle of treatment option and the inconclusive preoperative diagnostic evidences exploratory laparotomy was performed. At laparotomy an encapsulated yellowish adipose tissue mass was noted loosely adherent with mesentery and this was completely excised carefully without extirpation of any intraabdominal structure.

According to literature, histopathology with immunohistochemistry in conjunction with the morphology is the gold standard in diagnosis of lipoblastoma and its differentiation from other tumors specially myxoid liposarcoma as myxoid morphology in lipoblastoma is very uncommon.^{22,23} In accordance to this, in present reported case, the histopathological and immunohistochemistry reports of excised tumor separately suggested as lipoblastoma without any evidence of malignant transformation or any other tumor morphology.

Conclusion:

Intraabdominal lipoblastomas arising from mesentery however rare should be considered as a differential diagnosis of a child presenting with an abdominal mass. Complete surgical excision with a regular and periodic postoperative follow-up is suggested for a sound (post-operative) consequences.

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