Case Report

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A giant mesenteric lipoblastoma in an 18-month old infant: A case report and review of the literature

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ABSTRACT

Infantile lipoma (or lipoblastoma) of the mesentery is an extremely rare benign tumor of embryonal fat, with 15 cases reported in the English literature until today and only three of them arise from the ileum mesentery. We report an 18-month old boy presenting with a palpable intraabdominal mass arising from the ileum mesentery. Histopathologic and cytogenetic studies confirmed the diagnosis of mesenteric lipoblastoma (or infantile lipoma). Complete excision of the mass was performed. A follow-up examination consisting of physical examination and an abdominal ultrasound at 30 months postoperatively revealed no recurrence. We also present a review of the English literature regarding the presentation and management of mesenteric lipoblastomas in children.

Key words: Children, lipoblastoma, lipoblastomatosis, mesentery

INTRODUCTION

Lipoblastoma of the mesentery is an extremely rare benign embryonal fat tumor with only 15 cases reported in the English literature until today. There have been no reports of metastases, making the designation "blastoma" confusing, since this term is usually reserved for malignant tumors. The tumor may more accurately be described as an "infantile lipoma".[1] However, in order not to confuse the readers, the term "lipoblastoma" is used throughout the manuscript since this term has been widely used in previous reports.[2-15] It presents in two forms, a well-encapsulated one, called lipoblastoma and a diffuse one, known as lipoblastomatosis. It usually presents in children below 7 years of age, while 6 of them were <2 years old.[1-15] While it is primarily located in the soft tissues of the extremities and trunk, it is more rarely seen in the neck, face, buttock, perirectal area, and abdomen, and it is extremely rare in the abdominal mesentery and even more rare in the ileum mesentery. We report an 18-month old boy presenting with a palpable intraabdominal mass arising from the ileum mesentery. We also review the English literature regarding the presentation and management of mesenteric lipoblastomas in children.

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CASE REPORT

An otherwise healthy 18-month old boy was admitted to our hospital due to abdominal distention and an incidentally palpable intraabdominal mass. All blood tests (including complete blood count, liver function tests, erythrocyte sedimentation rate, protein-C, α-fetoprotein, carcinoembryonic antigen, and β-human chorionic gonadotropin) were normal, except for a platelet count of 450,000/ml. An abdominal ultrasound examination showed a homogenous hyperechoic mass ranging 14 × 11 × 8 cm^3 in size. The Doppler ultrasound revealed a right hepatic vein thrombosis. Following these, an abdominal CT scan revealed a large 11 × 12 × 14 cm^3 homogenous fatty dense tumor separated by septa extending from the hilum of the liver to the lower pelvis [Figure 1]. The chest X-ray was normal.

The boy was admitted for surgery. During the operation, a fatty mobile and encapsulated mass was found arising from the mesentery of the ileum with part of it adherent to the mass [Figure 2]. The mass was excised *en bloc* together with the affected segment of the ileum and the bowel continuity was restored with a primary end-to-end anastomosis. The gross specimen measured 16 11 × 8 cm^3 in size and its weight was 1500 g.

The histopathologic examination revealed a single

fragment of adipose tissue with a maximum diameter of 16 cm with a prominent myxoid appearance and mild- to moderate cellularity. The neoplasm consisted mainly of spindle and stellate cells with a limited eosinophilic cytoplasm and lipoblasts of a unilocular and multilocular type with a mild-focal moderate atypia and rare mitoses (0-2 mitoses/10 hpf). Rare multinuclear stellate cells and focal obvious apoptotic bodies were seen. Characteristic features of the histopathologic specimen were: (a) the presence of delicate sinusoidal network in the myxoid stroma, (b) the extensive presence of mucin pools surrounded by spindle cells and lipoblasts, and (c) the absence of lobulation and areas of maturing adipose tissue. The focal presence of imperfect fibrous septa at the periphery of the tumor and the absence of the *t* [16;22] FUS/CHOP and *t* [12;22] EWSR1/CHOP translocations favored the diagnosis of lipoblastoma [Figure 3].

A regular follow-up consisting of physical examination and ultrasound 30 months postoperatively revealed no recurrence.

DISCUSSION

Lipoblastoma was first described as a benign, soft tissue, solid tumor composed of immature embryonal fat by Jaffe in 1926, while lipoblastomatosis was first reported by Vellios *et al.*, in 1958. In 1973, Chung and Enzinger described the lipoblastoma as a well-circumscribed lesion and benign lipoblastomatosis as the diffuse type of this.[4] It is primarily located in the soft tissues of the trunk and extremities, and it is rarely reported in the face, neck, buttock, perirectal area, and abdomen.[14] Approximately 200 cases have been reported in the literature with occurrence at the mesentery in only 15 cases presented in the English literature and only three of them at the ileum mesentery (1.5% of all lipoblastomas) [Table 1].[16] The age distribution of all these children with mesenteric



Figure 1: Abdominal CT of the infant showing a mass with fatty tissue density

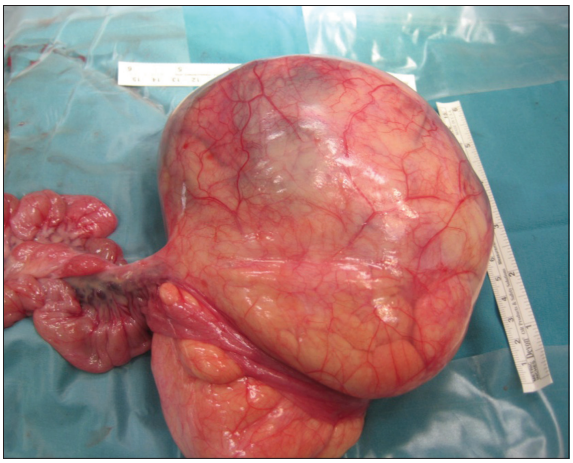


Figure 2: The gross specimen composing of the mesenteric mass and part of the small bowel

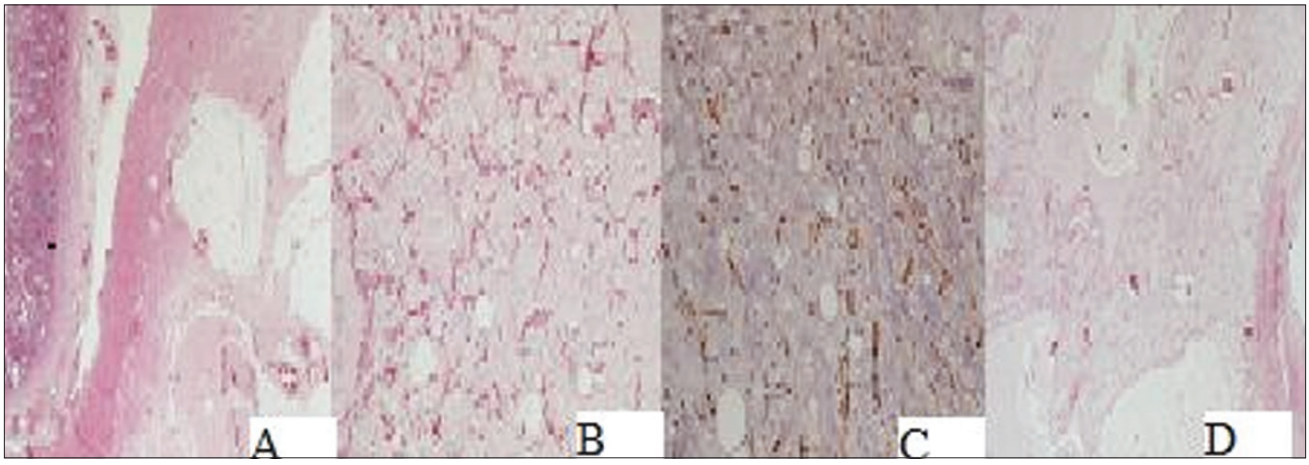


Figure 3: (A) The wall of the small intestine with the adjacent tumor (×200). (B) Lipoblastoma in higher magnification. Lipoblasts and the sinusoidal network are obvious (×200). (C) lipoblastoma CD 34 outlines the sinusoidal network in the tumor (×200), and (D) lipoblastoma pools of mucin (×200).

Table 1: Reports of mesenteric lipoblastoma in children.

REF	Age	Sex	Size (cm)	Site	Symptoms	Surgery	Follow-up	RECUR
1	5 mo	F	14.5 × 11 × 10.5	Transverse colon	Abdominal distension, diarrhoea	Excision of the mass	6 mo	NR
2	13 mo	M	17 (diameter) 12 × 10 8	Ileum	Abdominal distension, palpable mass	Excision of the mass and 26 cm small bowel	1 yr	NR
3	11 mo	M	12 × 10 × 0.8	Jejunum	Abdominal distension, palpable mass	Excision of the mass and part of jejunum	3 yrs	NR
4	2 yr	F	NI	NI	NI	Excision of the mass and 23 cm small bowel	NI	NR
5	4 yr	F	18 (diameter)	NI	Pain, vomiting	Excision of the mass and part of intestine	2 yr	NR
6	2 yr	M	23x19x9	NI	Abdominal distension, palpable mass	Excision of the mass	NI	NI
7	2 yr	M	NI	NI	Abdominal distension, palpable mass	Excision of the mass	NI	NI
8	10 mo	M	13 10 × 6	Ileum	Abdominal distension, palpable mass	Excision of the mass	2 yr	NR
9	16 mo	F	13 10 × 9	Ileum	Abdominal distension, palpable mass	Excision of the mass and 26 cm small bowel	NI	NI
10	2 yr	M	8 × 6 × 5	Jejunum	Obstruction, midgut volvulus	Excision of the mass	NI	NI
11	17 mo	M	9.7 × 7 × 5.5	NI	NI	NI	23 mo	NR
12	7 yr	F	10 × 8 × 6	Jejunum	Obstruction, midgut volvulus	Excision of the mass and part of the small bowel	1 mo	NR
13	3 yr	M	15 × 4.5 × 10	Jejunum	Abdominal distention, fullness, intermittent pain	Excision of the mass and part of the small bowel	2 yr	NR
14	4 yr	M	18 × 15 × 10	Ileocecal junction	Intermittent abdominal pain	Excision of the mass	4 mo	NR
15	4 yr	M	NI	NI	NI	NI	NI	NR

REF: References; RECUR: Recurrence; NR: Not Reported; NI: Not Indicated; mo: months; yr: years

lipoblastomas (including our case) is from 5 months to 7 years old with a male predilection.[1-15]

The exact aetiology is not known.[10] Although most cases are asymptomatic at presentation, lipoblastomas can present, as in our case, with a palpable abdominal mass and progressive abdominal distention, when originated at the ileum mesentery; however, on the other side with bilious vomiting, loss of appetite, mild abdominal pain or diarrhea for jejunal and ileocecal valve mesenteric lipoblastomas.

An accurate preoperative diagnosis is rarely made. Ultrasonography, computed tomography (CT), and magnetic resonance imaging (MRI) complement each other, but seldom can they determine whether the tumor is benign or malignant.[17]

Our patient was admitted to the operating theater based on his clinical picture, physical examination, and CT findings, but the histopathologic and cytogenetic studies established the diagnosis. Lipoblastoma has prominent lobulation, no nuclear atypia, or pleomorphism.[14]

The differential diagnosis between other lipogenic tumors can only be made histologically. In addition, gene rearrangement and subsequent upregulation of the development of zinc-finger gene PLAG1 is common in lipoblastoma, but not in other lipogenic tumors.[12]

The most difficult differential diagnosis is with liposarcoma, mainly its myxoid type. Liposarcoma is extremely rare under 10 years old, and it is histologically diagnosed by lack of lobulation, variable growth pattern, increased nuclear atypia, and abnormal mitoses.[12]

In histologically borderline cases, the tumor karyotype establishes an accurate diagnosis. Chromosomal translocations within chromosome 8 are a specific marker for lipoblastoma. On the other hand, myxoid liposarcoma demonstrates a specific clonal chromosomal anomaly t(12;16)(q13:p11) or t(12;22)(q13;p11).[7,10,13,14]

In our patient, the primary results of Reverse-Transcriptase-Polymerase Chain Reaction have not revealed the above translocations. Fine-needle biopsy before surgery is not indicated because there is increased risk of abdominal cavity contamination with tumor cells. On the other hand, radiation and chemotherapy have no role in the treatment of lipoblastoma.[13]

Therefore, we suggest that the treatment-of-choice should

comprise of complete excision of the lipoblastoma. If the excision is complete, there should be no recurrence or metastases.[7,12]

In conclusion, lipoblastoma should be included in the differential diagnosis when a young child under 7 years old presents with abdominal distention and palpable, easily mobile abdominal mass of the mesentery. US, CT and MRI could prove helpful preoperatively. No metastasis has been reported until today.[17] Complete surgical excision is curative with no recurrence. In histologically borderline cases, the tumor karyotype is of diagnostic value in order to differentiate with liposarcoma. When symptoms restricted in abdominal distention and palpable mass most possible arise from the ileum mesentery. In the cases where complete surgical excision is achieved and the histological report certifies lipoblastoma as the diagnosis, a follow-up period of more than 3 years is recommended for patients with this condition.[11]

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