

Giant Omental Lipoblastoma in a 21-Month-Old Infant

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Abstract

Case Report

Lipoblastoma is a rare benign neoplasm of fetal fat tissue that occurs almost exclusively in infants and children, to our knowledge less than 16 cases of omental lipoblastoma have been reported in the literature. We present a new case of omental lipoblastoma in a 21 months-old girl with a giant omental lipoblastoma measuring (17*8*16 cm). Complete excision of the tumor was performed. Histology confirmed the diagnosis of omental lipoblastoma. prognosis was good, with no recurrence during follow-up.

Keywords: Lipoblastoma; Omental tumor; Abdominal; CT scan; child; Histology, prognosis.

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INTRODUCTION

Lipoblastomas are rare, benign neoplasms of adipose tissue exhibiting a wide range of differentiation, including lipoblasts and mature adipocytes. They typically occur in infants and young children and are exceptionally rare in adults. The majority of lipoblastomas occur in the extremities, with smaller numbers occurring in the chest/abdomen, head and neck [1]. The omentum is a very unusual site, only 12 cases of omental lipoblastoma have been reported in the English literature [1-6]. We report a case of giant omental lipoblastoma in a 21-month-old girl, which was treated successfully by total excision.

CASE REPORT

It's about a 21-month-old infant, with no notable pathological history, admitted for progressive bloating in a context of apyrexia. Clinical examination revealed an apyretic child in good general condition. Abdominal examination: abdomino-pelvic mass involving the right flank and extending beyond the umbilicus, measuring 15cm, fixed in the deep plane, mobile in the superficial plane, with no inflammatory signs opposite, with collateral venous circulation and negative lumbar contact (Figure 1).

Abdominal ultrasonography showed a heterogeneous vasclerotic mass 17 cm long, with normal liver size and no dilatation of the intra- or extra-hepatic

bile ducts. Both kidneys were of normal size and well differentiated, and the spleen was of normal size.

AlphaFP and BHCG assays: negative.

Abdominal CT showed a voluminous intraperitoneal fatty mass (17*8*16 cm) with suspicious morphological features, initially suggestive of peritoneal lipoblastoma (Figure 2).

The patient underwent surgery, through an upper transverse abdominal incision, the exploration revealed a giant encapsulated mass that was carefully dissected from the greater omentum. Excision of the lmass was complete, and the specimen was sent for anatomopathological study (Figure 3).

Histologically, the study reveals adipose proliferation of diffuse and sometimes lobular architecture. In the center of the lobules, adipocytes are mature, with eccentric nuclei and large, optically empty cytoplasm. At the periphery of the lobules, adipocytes are immature, with less abundant cytoplasm and sometimes spindle-shaped or oval cells. Occasional foci of extramedullary hematopoiesis are noted (Figure 4).

The diagnosis of lipoblastoma was confirmed on the basis of these data. Postoperative recovery was uneventful, and discharge was possible one week after surgery. After 3 years of follow-up, no signs of clinical

or radiological recurrence were observed on routine abdominal ultrasound.



Figure 1: excessive abdominal distension in a child



Figure 2: Abdominal scan revealing a large intraperitoneal fatty mass

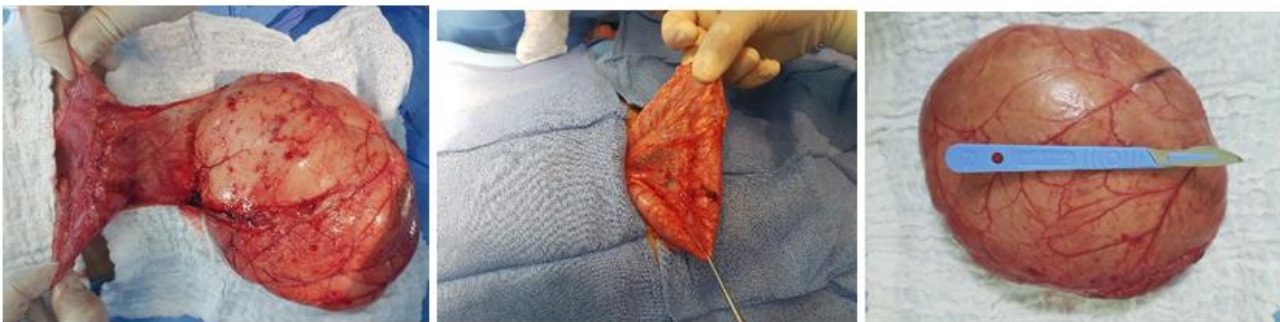


Figure 3: an encapsulated mass carefully dissected from the greater

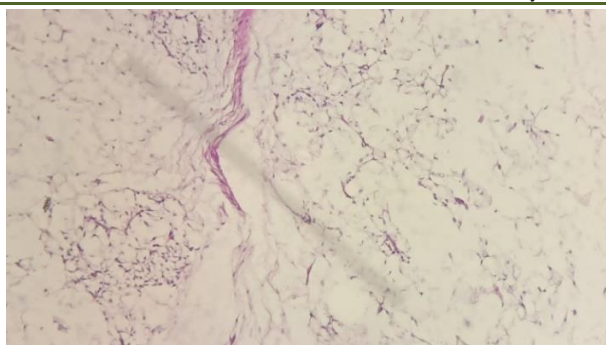


Figure 4: Lipoblastoma: tumor proliferation of mature adipocytes and lipoblasts (HESx100)

DISCUSSION

Lipoblastoma is a developmental disorder of embryonal fat that might manifest at birth or appear later in childhood. Of all the soft tissue neoplasms reported in children, lipoblastoma represents only 5 to 30%; intraperitoneal lipoblastoma is extremely rare and comprises 7% of these tumors [4]. Of the reported cases of intraperitoneal lipoblastoma, 85% are infants younger than 3 years and 40% younger than 12 months. Male-to-female ratio is 3:1 [3, 7, 8].

Lipoblastoma is well encapsulated [9]. In forms that infiltrate adjacent tissues, we speak of lipoblastomatosis [7, 8], the etiology of which is not yet well established. It probably involves an abnormal proliferation of fat cells and immature lobules - lipoblasts, not lipocytes - that lack the capacity to form fat.

Only 12 cases of omental lipoblastoma have been reported in the English literature [1-6, 10], however other cases have been reported in many countries : 2 cases japan [4, 6], spain [3], tunisie [8].

Lipoblastoma usually forms a slow-growing abdominal asymptomatic mass. It occasionally may cause chronic abdominal pain, discomfort, nausea and vomiting, distension, or intestinal volvulus. Solid omental masses can also produce acute clinical symptoms because of bleeding or intestinal infarction, requiring emergent surgery [7, 9, 11, 12].

These tumors are easily detected by ultrasound scan. Most patients undergo abdominal ultrasound as a first line screening imaging study. Sonography can differentiate cystic from solid tumors. However, it usually cannot identify the primary origin of the mass [13]. Abdominal CT scan is the diagnostic study of choice in omental tumors. Computed tomography usually shows a uniform, well-circumscribed fat-density mass (-30 to -70 Hounsfield units) and can identify the primary tumor site. Computed tomographic scans may also indicate compression on adjacent organs [8, 14]. Omental lipoblastomas are located in the anterior/superior compartment of the abdomen, usually anterior to the small bowel loops and transverse colon.

Magnetic resonance imaging is a valid method for establishing the differential diagnosis of soft tissue tumors. Magnetic resonance imaging usually shows a lobular pattern and high intensity on both T1 and T2-weighted images, similar to subcutaneous adipose tissue, and is excellent for demonstrating the anatomical detail of these masses. Unfortunately, it is not specific enough for differentiating between different adipose solid tumors [15].

MRI confirms the fatty nature of the tumour and its heterogeneous appearance, with enhancement of the fibrovascular septa [9, 15]. It also enables us to better study the tumor's relationship with adjacent organs, especially in the infiltrative form, in order to plan the most complete excision possible [9]. Lipoblastoma is treated surgically. Excision must be complete. Spontaneous regression has been reported [16]. Recurrence has been reported in cases of lipoblastomatosis [12].

The differential diagnosis may include lipoma, lipoblastoma, liposarcoma and teratoma. The latter is ruled out by the absence of calcifications. The presence of septa is not specific [7, 9, 13, 16, 17]. They may also be present in lipomas, which is why it is important to take the child's age into account. On imaging and even histology, it is difficult to differentiate between a lipoblastoma and a liposarcoma.

However, liposarcoma is very rare before the age of 5. Today, thanks to cytogenetics, the discovery of a structural rearrangement with deletion of the long arm of chromosome 8 is highly favorable to the diagnosis of lipoblastoma [12].

We preoperatively planned to perform laparotomy for extirpation of the tumor in the current case because the lesion was very large and solid. In contrast, Taizo F *et al.*, decided to perform laparoscopic abdominal exploration, Therefore, they successfully extirpated the large and heavy solid tumor laparoscopically by rotating and lifting it up. And by gradually rotating the tumor using forceps, they were able to proceed with the dissection of the tumor from the surrounding tissue. This maneuver was very useful for dissecting this movable solid tumor [6].

In principle, the primary treatment for lipoblastoma is complete resection. The local recurrence rate is reported to be between 14% and 25% [5]. In contrast, Mahour *et al.*, suggested that if a child with lipoblastoma is followed up long enough, the tumor will eventually differentiate into a mature lipoma [18]. A 12-month period without recurrence is generally regarded as curative [18, 19]. Therefore, lipoblastoma has an excellent prognosis. After surgery, the current patient displayed an increased appetite and weight gain. This is because the large lipoblastoma had compressed her intestines for a long time. We suggest that symptomatic patients undergo resection for lipoblastoma, even if the tumor is not malignant [6].

Our case remains well 3 years postoperatively, with no signs of recurrence.

CONCLUSION

Omental lipoblastoma can present with a wide clinical spectrum, but must be suspected in children under 3 years of age in good general condition. Abdominal ultrasound combined with CT scan guides the diagnosis, with confirmation remaining purely histological. Complete surgical excision offers the best chance of recovery without recurrence.

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