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CASE REPORT

Paediatric Mesenteric Lipoblastoma: A Remarkable Rarity

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ABSTRAK

Lipoblastoma menyumbang kepada 5-30% daripada tumor tisu lembut kanak-kanak. Ia adalah tumor yang jarang berlaku, berasal dari sel lemak embrio, dan kebanyakannya memberi kesan kepada bayi serta kanak-kanak. Walaupun biasanya ditemui pada anggota badan, ia boleh muncul di mana-mana bahagian badan. Kami melaporkan kes seorang kanak-kanak lelaki berusia 2 tahun dengan lipoblastoma mesenterik yang mengalami distensi dan ketidakselesaan abdomen selama 3 bulan. Imbasan tomografi berkomputer (CT) toraks, abdomen dan pelvis menunjukkan massa intraabdominal yang besar dengan komponen lemak, yang didiagnosis sebagai lipoblastoma mesenterik. Pesakit menjalani pembedahan laparotomi, pembuangan tumor, reseksi usus dan anastomosis primer dengan jayanya. Histopatologi mengesahkan diagnosis lipoblastoma. Pemulihan selepas pembedahan berjalan dengan lancar.

Kata kunci: *Lipoblastoma mesenterik; massa intraabdominal; tomografi berkomputer toraks, abdomen dan pelvis (CTAP)*

ABSTRACT

Lipoblastoma accounts for 5-30% of paediatric soft-tissue tumours. It is a rare tumour originating from embryonic fat cells, mainly affecting infants and young children. While it is commonly found in the extremities, it can occur anywhere in the body. We reported a case of a 2-year-old boy with mesenteric lipoblastoma presenting with a 3-month history of abdominal distension and discomfort. Computed tomography (CT) scans of the thorax, abdomen, and pelvis revealed a large lobulated intraabdominal mass with fat components, diagnosed as mesenteric lipoblastoma. The patient

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underwent successful laparotomy, tumour excision, bowel resection and primary anastomosis. Histopathology confirmed the diagnosis of lipoblastoma. Post-operative recovery was uneventful.
Keywords: Computed tomography thorax, abdomen and pelvis (CTAP); intraabdominal mass; mesenteric lipoblastoma

INTRODUCTION

Lipoblastoma is a rare benign tumour that arises from immature fat cells, mostly affecting infants and young children, with over 90% of cases diagnosed before age three. Despite being non-cancerous, these tumours can grow rapidly, compressing nearby structures, which can cause symptoms like abdominal distension and constipation. Early recognition is important, especially in cases like this 2-year-old boy, where the tumour's large size and location in the mesentery caused significant compression of the bowel and pancreas (McRae et al. 2021). Mesenteric lipoblastomas are particularly rare, making this case notable.

CASE REPORT

A 2-year-old Malay boy with no known medical illness initially presented in the clinic with abdominal distension 3 months ago. The symptom was associated with unspecified minimal abdominal discomfort and constipation for the past two weeks. Otherwise, before the current presentation, he had a normal bowel output. Physical examination revealed a huge abdominal mass palpable at the right lumbar region of 8 x 10 cm. An abdominal radiograph at the clinic revealed a huge mass on the right side of the abdomen with no dilated bowel. He was referred to a nearby government hospital for further management. An urgent ultrasound of the abdomen showed a large abdominal-pelvic hyperechoic mass with no significant vascularity on Doppler signal, sonographic diagnosis of lipomatous mass was made (Figure 1). Thus, computed tomography thorax, abdomen and pelvis

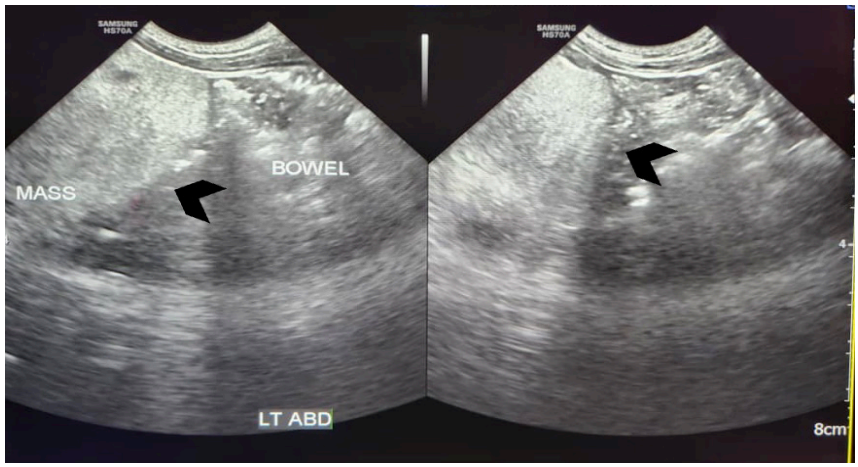

FIGURE 1: Abdominal ultrasound showed a large lobulated hyperechoic mass occupying abdomen and pelvis (black arrowhead)

(CTAP) was performed, and a huge lobulated and multiseptated hypodense mass occupied the intraperitoneal spaces measuring 6.1 cm x 15.2 cm x 14.5 cm (AP x W x CC) (Figure 2). The mass predominantly consisted of fat components. It pushed the liver upwards and inferiorly it extended until the L5 vertebra level. The mass displaced the bowel posteriorly and peripherally compressing the pancreas (Figure 3). Radiologically, the possibility of it being a mesenteric lipoblastoma was made associated with observable mass-related effects.

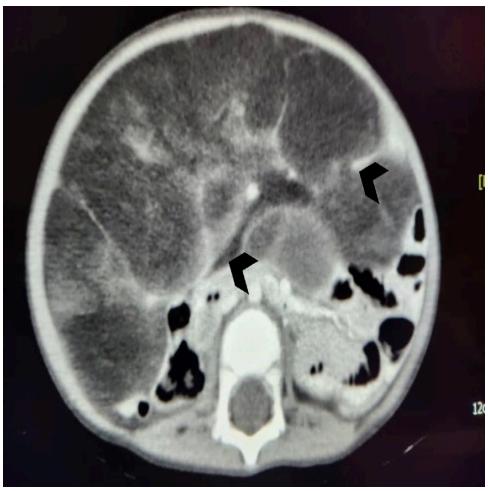

FIGURE 2: CT TAP axial view showed a huge lobulated and multiseptated hypodense mass occupying the intraperitoneal space (black arrowhead)

The patient underwent laparotomy, intraoperatively a lobulated mass with fatty component about 20 x 20 cm arising from the root of small bowel mesentery. En bloc resection of the tumour involving resection of 35 cm of small bowel with primary bowel anastomosis was done. Histopathology examination was consistent with lipoblastoma with no evidence of malignant changes. Post-operation patients recovered well under paediatric surgical follow-up.

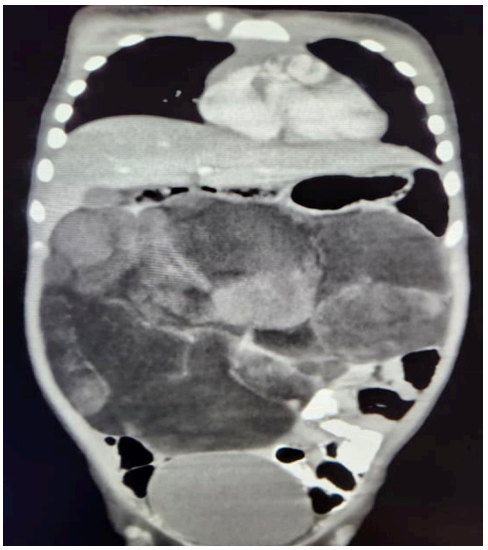

FIGURE 3: CT TAP coronal view showed the mass occupying the entire abdominopelvic region that pushes the liver upward and displaces bowels peripherally

DISCUSSION

Lipoblastoma is a soft tissue tumour composed of immature adipocytes, believed to originate from abnormal embryogenesis of white fat (McRae et al. 2021). It is a rare benign mesenchymal tumour of embryonal fat that exclusively affects children and infants (Kerkeni et al. 2014). Over 80% of cases are discovered in children under the age of 3, and 40% are seen in infants younger than 1 year old (Hashizume et al. 2020). In the largest published series, 88% of individuals were diagnosed while they were under 3 years old, with a median age of 1 year (Chung & Enzingek 1973). Males are predominantly affected than females (Moholkar et al. 2006). Due to the fact that there have only been 15 cases described in the English literature of this extremely rare benign embryonal fat tumour and only three of them arise from the ileum mesentery (Gentimi et al. 2011). Despite being benign, risk for complications

can occur because of the tumours' bulk effect and they are sometimes found in surgically difficult anatomical site that increase the risk of complications (McRae et al. 2021).

Depending on its size and location, lipoblastoma may induce a variety of vague and nonspecific symptoms, sometimes it discovered in asymptomatic patient (Hashizume et al. 2020). The majority of lipoblastomas appear asymptomatic at early presentation, they can present as a growing painless palpable mass and progressive symptoms of various organ compression depending on localisation (Cempaka et al. 2023). In this case, the patient presented with abdominal distension for 3 months duration and non-specific abdominal discomfort. Due to the mass effect on the bowel that affects peristalsis movement, the patient also presented with constipation with stool Bristol 4.

When a child presents with abdominal distension and clinically there is a palpable intrabdominal mass, ultrasonography is frequently used as the initial imaging technique as it is able to detect abdominal lipomatous tumours (McRae et al. 2021). Sonographically, lipoblastoma can appear as homogenous echogenic masses; however, they might display a perplexing image, ranging from a homogenous to a heterogeneous echo pattern (Moholkar et al. 2006). Due to this appearance, ultrasound cannot be used to accurately diagnosed lipoblastoma because it lacks sufficient specificity and sensitivity (Gentimi et al. 2011).

Computed tomography (CT) scan offers definitive identification of the fatty component within lipoblastomas, along with the potential presence of intra-tumoral stranding and other soft tissue-density elements (Moholkar et al. 2006). From CT findings, differential diagnoses may include liposarcoma, lymphangiomas, and cavitating mesenteric lymph node syndrome, particularly when visualised fat attenuation processes associated with the mesentery are observed (Moholkar et al. 2006).

While CT scans are frequently employed in lipoblastoma assessment, magnetic resonance imaging (MRI) emerges as the preferred imaging modality due to its superior ability to delineate relationships with adjacent vascular and muscular structures. It facilitates precise identification of lipomatous components and aids in preoperative strategy. Utilising fat suppression techniques can further confirm the presence of fatty elements. Nonetheless, the definitive diagnosis hinges upon histopathological examination (Moholkar et al. 2006). Macroscopically, lipoblastomas manifest as lobulated masses with a light yellow or creamy hue, often exhibit mottled pink area (Chung & Enzingek 1973). Moreover, lipoblastomas typically display a more uniform growth pattern, marked by prominent lobulation, and their lipoblast exhibit no nuclear atypia and pleomorphism (Moholkar et al. 2006).

For the management of mesenteric lipoblastoma, regardless of location the definitive treatment is complete surgical resection and when there is a high risk of impingement on nearby structures, treatment should not be postponed (Cempaka et al. 2023). These lesions demonstrate a propensity for recurrence even following presumed complete excision, hence necessitating a recommended minimum follow-up period of 5 years (McVay et al. 2006).

Lipoblastoma typically has excellent long-term prognosis (Gentimi et al. 2011). Recurrence rates have been found to be between 9 to 22%, which is due to insufficient initial tumour resection (Gentimi et al. 2011). A definite diagnosis eventually requires thorough clinical, radiological and pathological examinations.

CONCLUSION

This case underscores the need to recognise rare peadiatric tumors like lipoblastoma and the crucial role of CT and MRI in their diagnosis and surgical planning. Learning points include, that despite being benign, their size can cause significant symptoms, requiring timely and precise surgical intervention especially when vital structures are at risk. Moreover, careful follow-up is needed to monitor for recurrence.

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