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Case Report

# A case of lipoblastoma in a pediatric patient ☆

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ABSTRACT

Lipoblastoma is a benign soft tissue tumor that originates from embryonic white fat. Lipoblastoma presents as a slow-growing mass that commonly occurs in the extremities of young children. Histological examination remains the gold standard in confirming lipoblastoma; however, radiology examination can help identify and evaluate the extent and characterization of the mass prior to the excision. Here, we report a 7-year-old male patient who presented with a painless mass in the right popliteal extending to the proximal cruris areas, and the imaging modalities suggested the presence of fat within the mass. The patient then underwent complete excision, and histopathology examination revealed lipoblastoma. This study highlights the possibility of lipoblastoma in older children and the role of imaging examinations in the diagnosis.



## Introduction

Lipoblastoma is a soft tissue neoplasm of embryonic adipose cells [1]. Lipoblastoma is also known as fetal lipoma, embryonic lipoma, and infantile lipoma [2]. Lipoblastoma occurs mostly in young children and is extremely rare in adults, with most cases are found in children less than 3 years of age [3,4]. Preoperative diagnosis of lipoblastoma can be difficult because it is heterogeneous and clinically similar to other lipomatous tumors [5,6]. Here we present a rare case of a 7-year-old patient with lipoblastoma at the right lower extremity and the role of imaging modalities in the diagnosis.

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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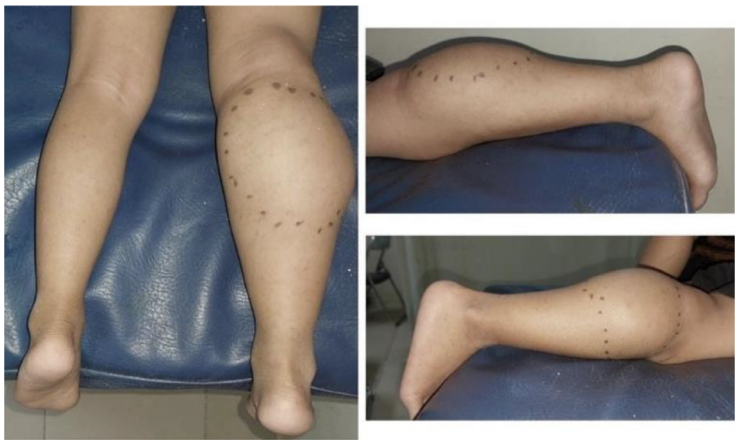

**Fig. 1 – The physical finding of a mass at patient's right popliteal and cruris area, which was noncompressible, painless, and immobile.**

## Case presentation

Our patient was a 7-year-old boy who came with a mass in his right leg, which had been present since he was 7 months old. The mass kept gradually increasing in size and made it difficult for him to walk. Physical examination revealed an $8 \times 6 \times 4$ cm mass at the right popliteal extending to the proximal cruris area Fig. 1. The mass was non-compressible, painless, and immobile. Ranges of movement (ROM) of the knee joint were limited due to the mass, but there was no neurological symptom. Laboratory examinations were within normal ranges.

Plain radiography showed a localized mass with no calcification at the right popliteal until the proximal cruris, with soft tissue and fat densities Fig. 2. Further examination using a contrast-enhanced computed tomography (CT) scan revealed a heterogeneous mass in the right popliteal region extending to the proximal cruris with fat component Fig. 3. The mass was lobulated with a clear margin and thin septa. There was also no evidence of bone destruction or erosion.

Magnetic resonance imaging (MRI) of the mass revealed a multilobulated, well-defined, heterogeneous mass with multiple thin septa. The mass was inhomogeneous hyperintense on the T1-weighted and T2-weighted images, but not on the fat-suppressed T2-weighted image, which confirmed the fat component of the mass Fig. 4. On the contrast-enhanced T1-weighted image, the mass showed minimal inhomogeneous enhancement, especially on the septa. The mass seemed to displace and compress the surrounding muscles and vascular structure.

The patient then underwent total excision of the mass; macroscopically, the mass was soft, lobulated, and had a yellowish white color (Fig. 5). Histopathological examination showed a lobular mass with mature fat cells and multivacuolated lipoblasts, separated by fibro-collagenous septa (Fig. 6). There were also myxoid and hyaline, as well as lymphocytes and dilated vessels. There were no malignant cells found. One year after the surgery, the patient remained symptomless and the follow-up plain radiography of the patient's right knee showed no sign of recurrence (Fig. 7).

## Discussion

Lipoblastoma is a rare benign tumor of embryonic adipose cells, at different maturation stages, which commonly occurs in infancy and early childhood [1]. The term *lipoblastoma* was first introduced by Jaffe in 1926 to describe a lipomatous tumor and distinguish it from other lipomatous tumors with no lipoblast [7], and another term *lipoblastomatosis* was coined by Vellios et al. to define an infiltrating lipomatous lesion [8]. According to Chung and Enzinger, there are 2 types of lipoblastomatous tumors, 1) lipoblastomas, which are more common, well circumscribed, and more superficially located; and 2) lipoblastomatosis, which is poorly circumscribed, diffuse, infiltrative, and deeply situated [9].

Lipoblastoma commonly occurs in regions with the largest amount of immature fat, including the trunk and the extremities. Other reported locations include the cervical, paravertebral, mediastinum, and retroperitoneum [3,9]. Lipoblastoma is a benign lesion that is usually asymptomatic. However, it may grow to a considerable size and the resulting mass effect can cause various symptoms [3,10]. The exact pathogenesis of lipoblastoma is still unknown; however, it has been postulated that lipoblastoma is caused by chromosomal abnormalities. Gisselson et al. found that some patients with lipoblastomas have alterations in the PLAG1 oncogene on chromosome 8q12, which are suspected to transform mesenchymal progenitor cells into lipoblastoma cells in various degrees of proliferation and differentiation [11].

Macroscopically, typical lipoblastoma appears as a light yellow or creamy white mass, commonly mottled with pale-pink homogenous gelatinous or myxoid areas [9]. The mass is also lobulated, encapsulated, and soft [9,12]. Microscopically, both circumscribed and diffuse forms of lipoblastoma have a lobular architecture comprising fat cells in varying degrees, separated by fibrous septa, which are often rich in capillaries and venules arranged in a plexiform pattern [2,9,12–15]. The fat cells range from primitive stellate or spindled mesenchymal cells, mono or multivacuolated lipoblasts and mature adipocytes, which are more located in the center of the lobules [3,12,14]. Lipoblastoma also contains an abundant

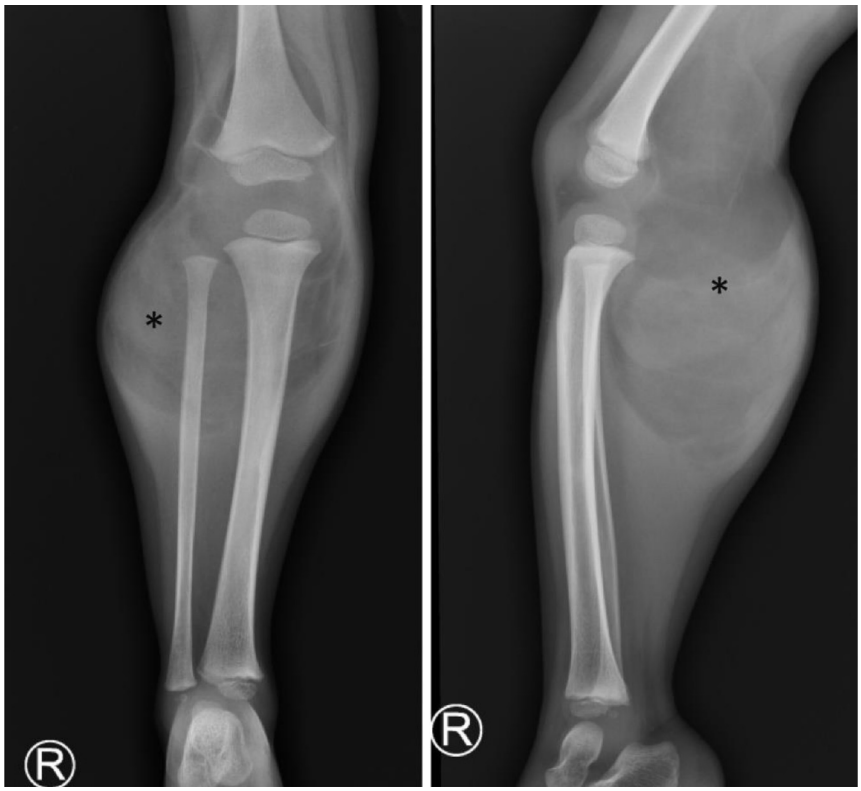

**Fig. 2 – Plain radiography of the right cruris. The anteroposterior (left) and lateral (right) images showed the presence of a heterogenous soft tissue mass (asterisk) without calcification in the right popliteal fossa extending to the proximal cruris.**

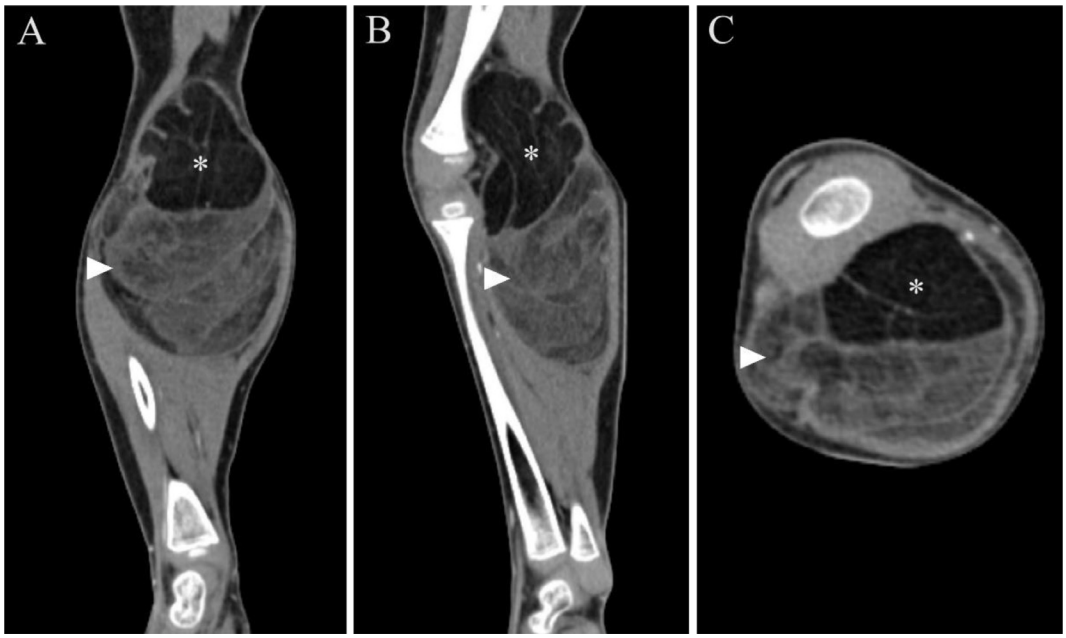


**Fig. 3 – (A) Coronal, (B) sagittal, and (C) axial sections of contrast-enhanced CT scan of the right leg revealed a huge mass from popliteal to proximal cruris areas. The mass was multilobulated with mixed fat (asterisk) and solid soft tissue (arrow head) components.**

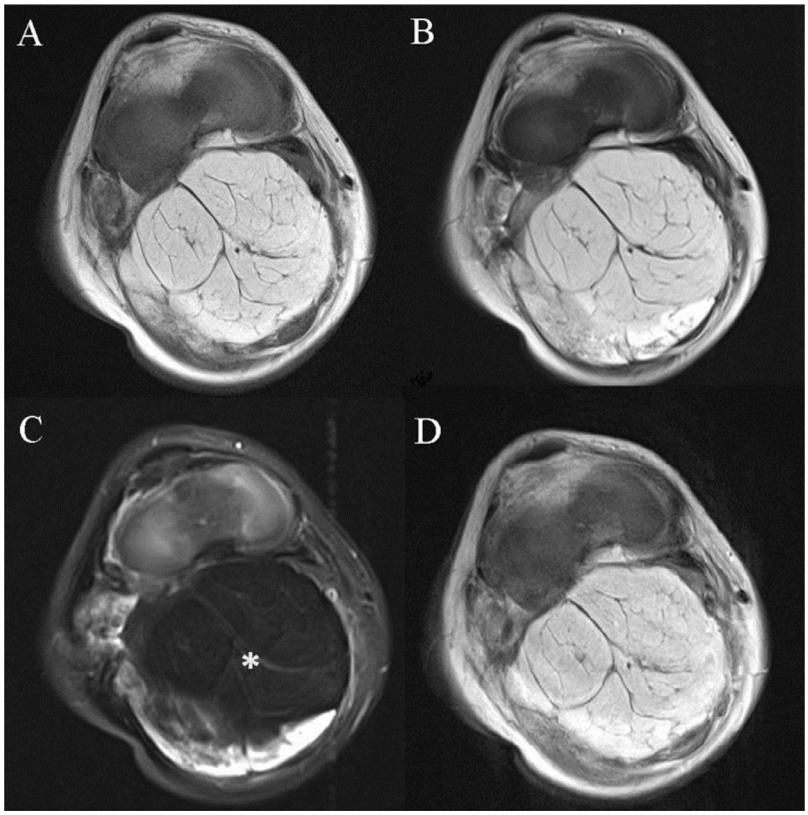


**Fig. 4 – The presence of fat component was shown by the hypointense signal on fat-suppressed T2-weighted image (asterisk). (A) T1-weighted, (B) T2-weighted, (C) Fat suppressed T2-weighted, and (D) Contrast-enhanced T1-weighted images of the MRI.**

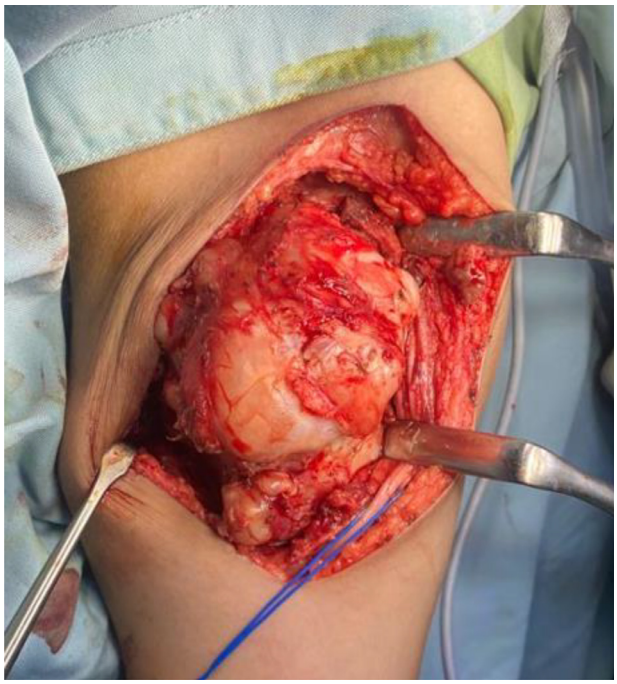

**Fig. 5 – The gross appearance of the mass, which was lobulated and yellowish white in color.**

myxoid matrix, which can be more prominent in very young patients, with little activity of mitosis [1,2,13,14]. The diffuse pattern of lipoblastomatosis has a less lobulated architecture and may contain skeletal muscle fibre corresponding to its infiltrative growth pattern [1].

Lipoblastoma may be clinically hard to differentiate from other lipomatous tumors, and the main differential diagnoses include lipoma, liposarcoma, and teratoma [2]. Lipoma is rare in young children and can be excluded in tumors that contain non-fatty components, while teratoma commonly has calcification [2]. Myxoid liposarcoma is extremely rare in children under the age of 10 years [13]; however, it remains the most important differential diagnosis since it has a similar clinical presentation [14,15]. Moreover, both lipoblastoma and myxoid liposarcoma have T2-hyperintense components on MRI which correspond to the abundant myxoid matrix, which makes it difficult to differentiate both entities based on imaging alone [2].

The true diagnosis relies on a histopathological examination. As compared to liposarcoma, lipoblastoma is more lobulated and has a more uniform growth pattern. Lipoblastoma also lacks nuclear atypia or mitotic activity, as well as the distinctive microcystic spaces, which are found frequently in liposarcomas [12,14]. In difficult cases, karyotyping may be beneficial in diagnosing lipoblastoma, which has an anomaly of chromosome 8, while myxoid liposarcoma usually has a clonal chromosomal anomaly of t(12;16)(q13:p11) [16]. On immuno-

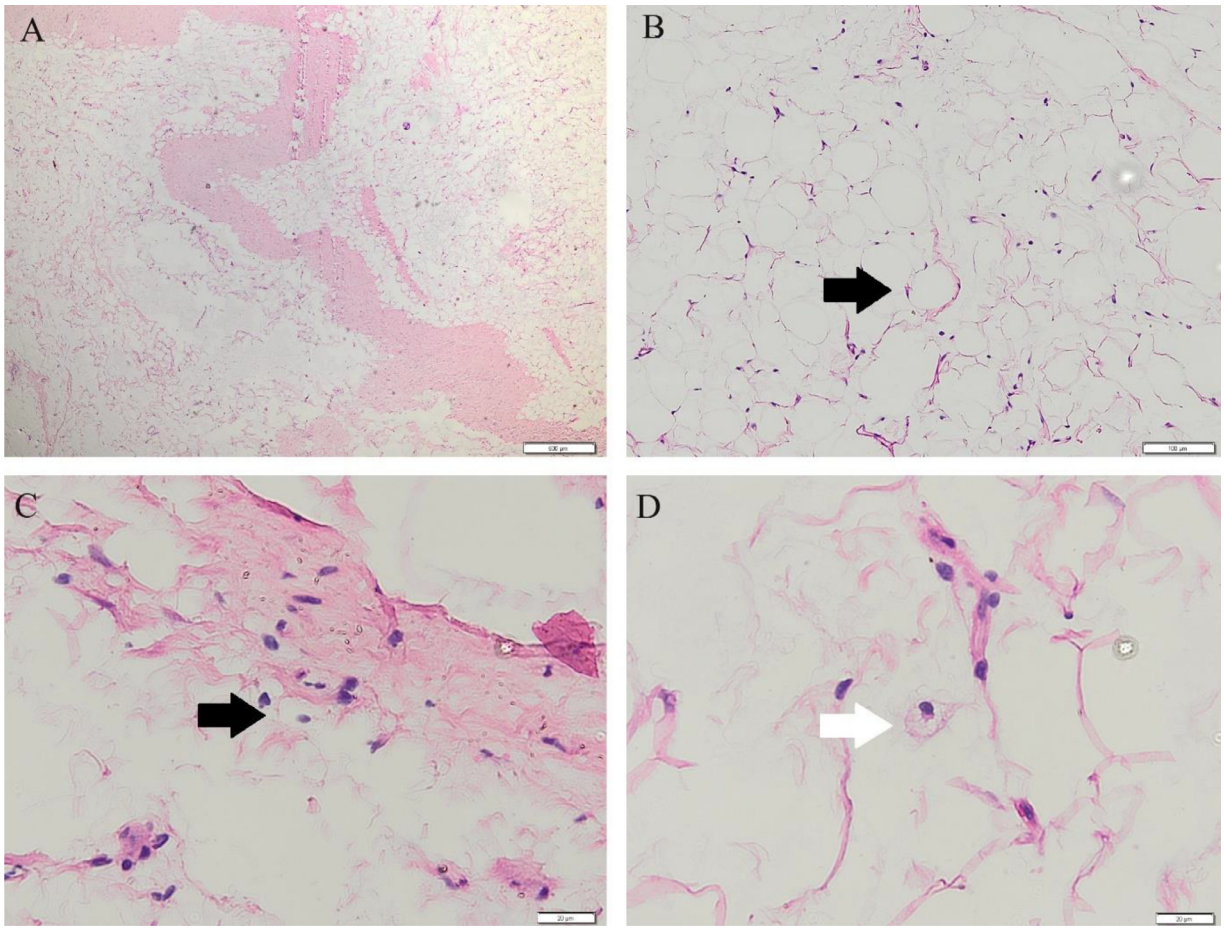


**Fig. 6 – Hematoxylin and Eosin (H&E) staining. (A) H&E 20x showed lobules of mature fat cells and lipoblasts separated by fibrous septa, (B) H&E 100x showed uni-vacuolated mature adipocytes with nuclei located at the periphery (black arrow), (C, D) H&E 400x showed mature adipocytes (black arrow) and multi-vacuolated lipoblast with hyperchromatic nucleus in the centre (white arrow).**

histochemistry examination, lipoblastoma has cells that are positive for vimentin and S-100 protein, while negative for cytokeratin, CD34, desmin, and NKIC32 [5].

Radiological imaging can be useful in assessing the extent of disease and as an aid in planning surgical excision; however, its use is limited to differentiate lipomatous tumor [17]. Ultrasonography (USG), computed tomography (CT) scans, and magnetic resonance imaging (MRI) all have a complementary role in the diagnosis of lipoblastoma. The imaging features of lipoblastoma reflect the characteristics of its predominantly fat cells, which are echogenic mass on USG, low attenuated areas on CT scan, and areas with hyperintense signal on T1 while being hypointense on fat-suppressed signal on MRI [1].

USG is often the first-line modality because it is the fastest and safest modality to evaluate masses in children, with a lack of need for sedation for examination [2,18,19]. Lipoblastoma on USG appears as a hyperechoic mass with internal septation; however, the mass will be predominantly hypoechoic if the myxoid area is abundant [1,2]. USG can also detect any fluid collection and assess the tumor vascularity using color Doppler [20]. However, USG is limited in defining the full extent of large masses within the soft tissue [20], and due to the variability of its appearance, USG i.s not sensitive or specific enough in diagnosing lipoblastoma [21].

CT scans provide more accurate anatomical detail and allows better identification of the fat components within the mass. On CT, lipoblastoma appears as a well-encapsulated or to have ill-defined margins (as in its diffuse form) with multiple thin-enhancing septations [1,6,19,22]. However, in some cases, the appearance can be varied depending on the amount of adipose and soft tissue elements, as well as the necrotic areas that may be found within the mass [22]. Calcifications are rarely found [22].

MRI is the preferred modality for localizing and characterizing lipoblastoma, without being invasive or radiating to the patients [5,19,23]. MRI is useful in determining the anatomical extent, tissue involvement, and mass characteristics; as well as providing information regarding other tissue characteristics such as cystic components, vascularity, and contrast enhancement [2,24]. MRI also allows analysis of the multiplanar space of the relations of the tumor with the adjacent organs and the vasculo-nervous structures [5,23]. On MRI, lipoblastoma appears as a solid and lobulated fatty mass with heterogenous hyperintense signal on T1-

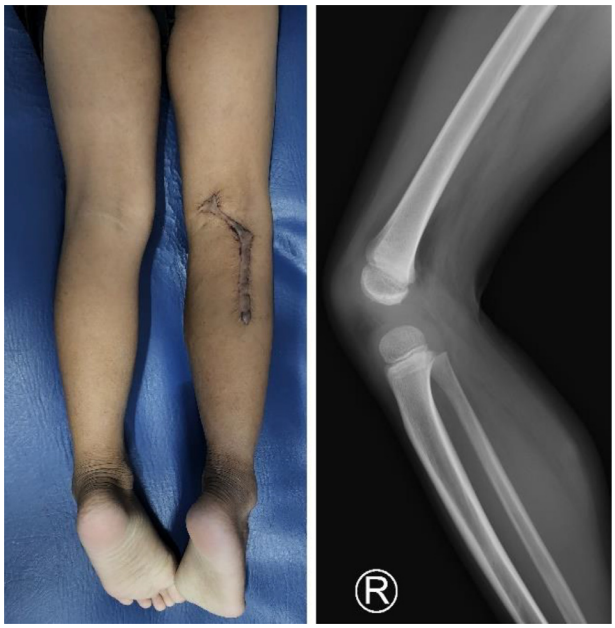

**Fig. 7 – The patient presented with no mass recurrence one year after the excision (left), which was then confirmed by the follow-up plain radiography (right).**

and T2-weighted images due to varied proportions of fat cells, myxoid, and fibrous tissue [5,19,21,25]. Lipoblastoma will appear hypointense on T1-fat saturation images and show enhancement on the septa and solid components after contrast administration [18]. The fat signal intensity of lipoblastoma correlates positively with the proportion of mature adipocytes, which have a comparatively higher signal on T1-weighted images than lipoblasts [2,26].

Lipoblastoma lacks malignant property; however, recurrence has been reported in 14% to 25% of cases, mainly due to incomplete removal [9,10]. If the patient presents with no symptoms, the clinician may only need to observe the patient; however, if the mass progresses rapidly and symptoms appear, the treatment of choice is total surgical excision with preservation of adjacent tissues [24,27,28]. However, total excision may not always be performed due to the risk of tissue and vessel injury, and a staged approach may be the preferred option to improve the preservation of the neurovascular bundle [4]. Post-operative follow up is needed to detect recurrent lesions and monitor the size of incompletely resected lesions, and a 5-year follow-up using MRI is recommended [10,28].

In this report, a 7-year-old boy presented with a soft tissue mass at his lower extremity, and the plain radiograph suggested a fat-density mass. Further examination using a CT scan also revealed a heterogeneous soft tissue mass and confirmed the predominantly fat composition. The presence of a lipomatous tumor was confirmed by MRI which revealed a lobulated and heterogeneous mass that was mostly hypointense on T1-fat suppressed image. The true diagnosis was confirmed by histopathological examination which showed lobules containing both immature and mature fat cells with thin septa. The heterogeneity of the mass can be related to the presence of myxoid and the varied maturity of the fat cells within lipoblastoma. This diagnosis is in accordance with the symptoms and radiologic findings, making it possible for an older child to have lipoblastoma, which is more commonly found in younger pediatric patients.

## Conclusion

Lipoblastoma is a benign lipomatous tumor which occurs predominantly in infancy and early childhood. Total excision of the tumor is recommended, and preoperative diagnosis is important in detecting and characterizing the mass as an aid in surgical planning. MRI is the preferred modality in confirming the fat component within the mass. The diagnosis of lipoblastoma in our patient was rather unusual as it occurred in an older age; however, the clinical symptom and radiological imaging supported the diagnosis.

## Patient consent

I, on behalf of all authors, confirm that complete written informed consent was obtained from the patient and his guardian (mother) for the publication of this study and accompanying images. Subject and his guardian voluntarily participated in the study. Subject and his guardian had been notified that his radiology examinations results would be published for scientific purpose. Subject had received a copy of this consent form signed by the authors.

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