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CASE REPORT

Lipoblastoma: Different Features on Magnetic Resonance Imaging

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ABSTRACT

Lipoblastoma is a rare benign adipocytic tumour that usually occurs in children younger than 3 years, with male predominance. Lipoblastoma usually presents as a slowly enlarging soft tissue mass in the extremities although other sites of involvement have been reported. We report three cases of lipoblastoma in a lower limb with different magnetic resonance imaging features, some of which are shared by malignant soft tissue tumours.

Key Words: *Lipoblastoma; Magnetic resonance imaging; Thigh*

中文摘要

脂肪母細胞瘤磁共振成像的多種表現

石家偉、鄭珊珊、謝健燊、黎國忠、陳文光

脂肪母細胞瘤是一種良性脂肪細胞腫瘤，好發於三歲以下的兒童，男性患者居多。脂肪母細胞瘤大部分病變出現緩慢增大的軟組織包塊，四肢為最常累及的部位，也有發生於其他部位的報導。本文報告出現在下肢的脂肪母細胞瘤的三個病例，它們均有不同的磁共振成像表現，其中一些表現與惡性軟組織腫瘤相似。

INTRODUCTION

Lipoblastoma is a rare benign adipocytic tumour that usually occurs in children younger than 3 years, with male predominance. Lipoblastoma usually presents as a slowly enlarging soft tissue mass in the extremities, although other sites of involvement have been reported. We report three cases of lipoblastoma in a lower limb with different magnetic resonance imaging (MRI) features.

CASE REPORTS

Case 1

In December 2008, a neonatal boy presented on day 7 of birth with rapidly progressing left knee swelling without any sign of inflammation. Plain radiographs showed non-calcified soft tissue swelling over the anterior aspect of the knee. MRI showed a well-circumscribed mass with predominantly T2-weighted (T2W)-hyperintense and T1W-hypointense non-fatty components over the prepatellar subcutaneous tissue, without musculotendinous or joint invasion. The mass contained several small T1W-hyperintense locules with suppression on short T1 inversion recovery (STIR) sequence, suggestive of small lipomatous components. No significant contrast enhancement was seen (Figures 1 and 2).

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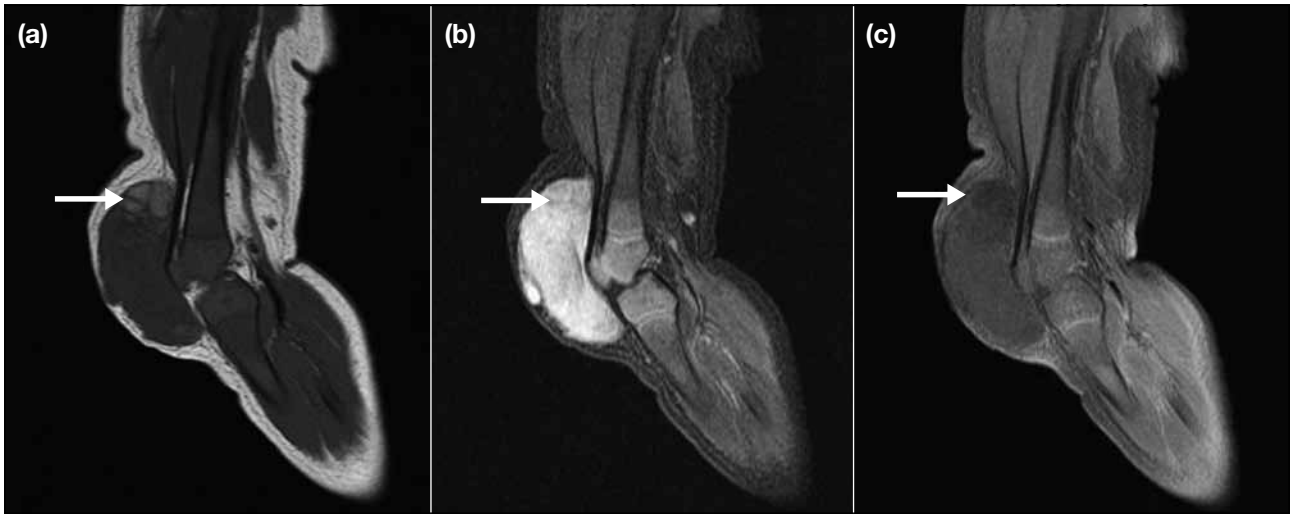

Figure 1. Sagittal magnetic resonance images of the left knee of a neonatal boy (patient 1) show lipomatous components (arrows) at the superior aspect (T1-weighted hyperintense and suppressed on fat-saturated sequence) and no contrast-enhancing soft tissue component. (a) T1-weighted, (b) T2-weighted and short T1 inversion recovery, and (c) post-gadolinium T1-weighted fat-saturated images.

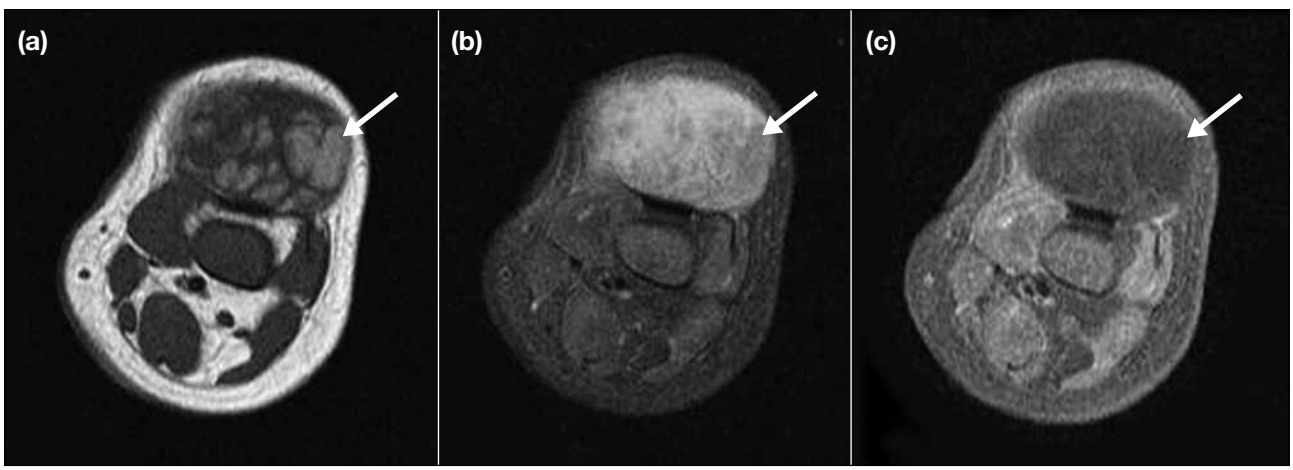

Figure 2. Axial magnetic resonance images of the left knee of a neonatal boy (patient 1). The lesion shows lipomatous components (T1-weighted hyperintense and suppressed on fat-saturated sequence) and no contrast-enhancing soft tissue component (arrows). (a) T1-weighted, (b) T2-weighted and fat saturation, and (c) post-gadolinium T1-weighted fat-saturated images.

Ultrasonography (USG) examination of the mass showed two components, one hyperechoic component at the superomedial aspect corresponding to the T1W-hyperintense lipomatous areas on MRI, and another large hypoechoic component. USG-guided core-needle biopsy of the mass showed lipoblastoma. The patient subsequently underwent excision of the mass. Gross specimen inspection showed homogeneous and vaguely nodular, light yellowish, firm fibrofatty tissue. The histology confirmed the diagnosis of lipoblastoma, demonstrating diffuse and lobular growth of myxoid mesenchymal tissue with myxoid pools and aggregates of adipocytes.The mass was vascular, but without significant cytological atypia. Focal fibrous bands transversing the fatty islands were also present.

Complete resection was achieved. This patient had no tumour recurrence at clinical follow-up after 5 years.

Case 2

In March 2010, a 33-month-old girl was referred for progressive swelling over the medial aspect of her left thigh. Physical examination found a firm mass attached to muscle. Plain radiographs showed a non-calcified

radiolucent lesion over the medial aspect of the mid-thigh. MRI showed a well-demarcated intramuscular mass inside the adductor magnus muscle (Figure 3). The mass demonstrated predominantly T1W-hyperintense signal with suppression on STIR sequence, suggestive of lipomatous signal. The mass contained numerous septa of varying thickness, and non-septal T2W-hyperintense signals. There was no significant internal enhancement, except for septal and patchy non-septal enhancement.

Subsequently, USG-guided core-needle biopsy of the mass was performed. Biopsy revealed lipoblastoma. This patient underwent complete excision of the mass. Histological examination identified lobules of predominantly adipose tissue separated by thin fibrous septa. The adipose tissue largely comprised adipocytes, which was interspersed by foci of lipoblasts within myxoid stroma. Clinically, the patient remained tumour-free at follow-up after 3 years.

Case 3

In May 2012, a 26-month-old girl presented with insidious onset of a painless left popliteal mass. The mass was firm in consistency, and not fixed to skin. USG examination revealed a hyperechoic mass medial to the medial head of the gastrocnemius muscle, and the popliteal vessels were displaced laterally. The USG finding excluded a diagnosis of popliteal cyst. MRI of the mass showed predominantly T1W-hyperintense signal and hypointense signal on fat-suppression STIR sequence, compatible with fat signal (Figure 4). The mass contained multiple hypointense septa of varying thickness, and multiple patchy and nodular T1W-hypointense and T2W-hyperintense non-fatty areas with contrast enhancement scattered within the mass.

USG-guided biopsy of the mass was performed and the results suggested lipoblastoma. Subsequently, the patient underwent complete excision of the mass. Pathological examination revealed a partially encapsulated nodular fatty mass containing occasional areas of haemorrhage. Lobules of adipose tissue with admixed lipoblasts and matured fat cells, with intervening fibrous septa and myxoid stroma, were observed on microscopy. The diagnosis of lipoblastoma was made. There was no clinical sign of tumour recurrence at follow-up after 2 years.

DISCUSSION

Lipoblastoma is a lobulated lipomatous tumour resembling fetal adipose tissue.[1] Lipoblastoma is also known as fetal lipoma, embryonic lipoma, and infantile lipoma. The condition most commonly occurs in children younger than 3 years, with male predominance. The trunk, followed by the extremities, and the head and neck region are the most commonly involved sites.[2] Other reported sites include the mediastinum and retroperitoneum.[3-5] Clinically, lipoblastoma presents as a slow-growing mass over the extremities. The presenting symptoms are related to the location and size of the tumour. Histologically, lipoblastoma contains an admixture of mature and immature adipocytes, and lipoblasts at various stages of development.[6] The proportion of lipoblasts varies with age and can sometimes be scanty.[6] Variable prominent lobulations

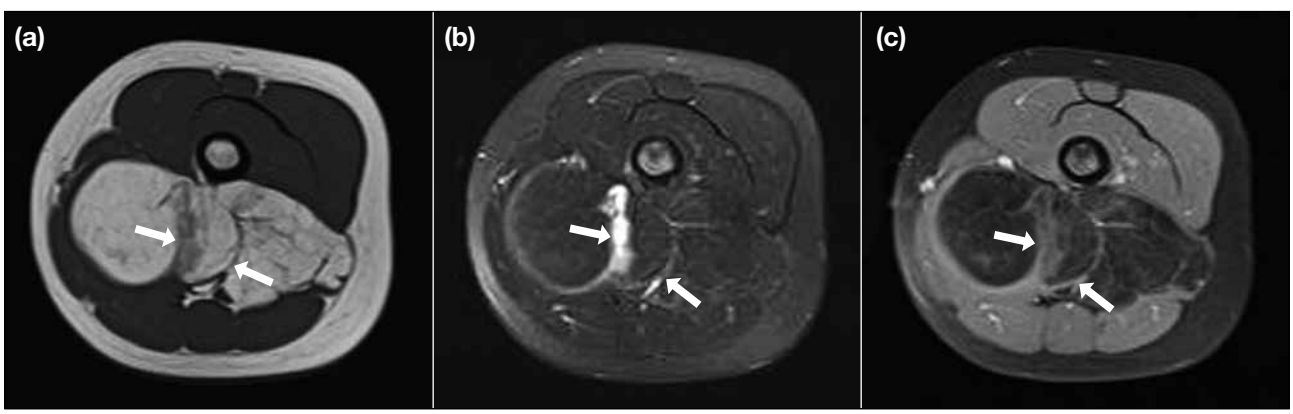

Figure 3. Axial images of the left thigh of a 33-month-old girl (patient 2). The lesion is predominantly lipomatous with multiple septations of varying thickness (arrows). The lesion also comprises non-septal areas of T2-weighted hyperintense signals. There was no significant internal enhancement, except septal and patchy non-septal enhancement. (a) Axial T1-weighted, (b) T2-weighted plus fat saturation, and (c) post-gadolinium T1-weighted fat-saturated images.

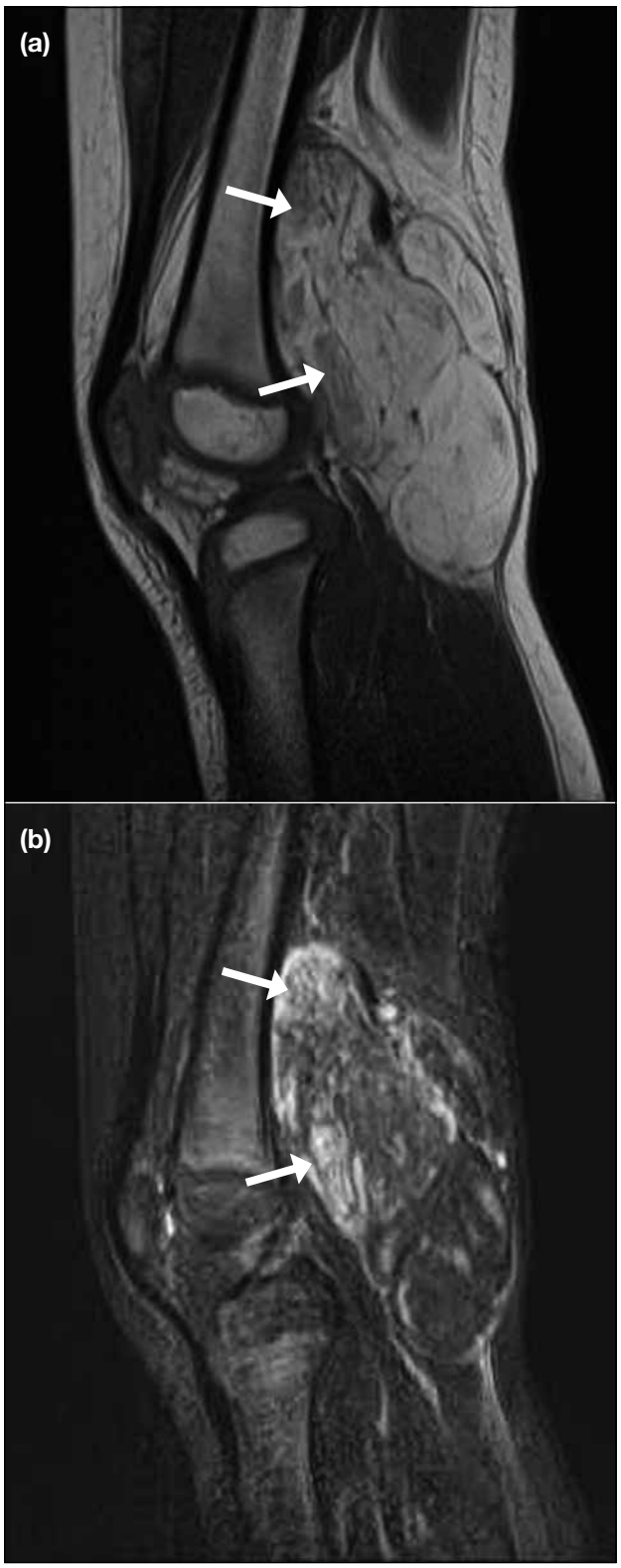

Figure 4. Sagittal images of the left knee of a 26-month-old girl (patient 3). The lesion contains multiple septa of different thicknesses, and multiple scattered patchy or nodular T1-weighted hypointense and T2-weighted hyperintense non-fatty areas, which demonstrate contrast enhancement (arrows). (a) T1-weighted, and (b) T2-weighted and short T1 inversion recovery.

and myxoid changes, demarcated by fibrous bands, are typical in lipoblastoma. The matrix usually consists of abundant myxoid matrix and primitive mesenchymal cells, and has a plexiform vascular pattern.[2,6]

USG is usually the first-line investigation for patients with suspected lipoma because of its ready availability and lack of need for sedation for examination. Lipoblastoma contains variable proportions of lipomatous components depending on the composition of lipocytes of different maturity and myxoid stroma.[2] Fatty components are usually shown as hyperechoic tissue on USG, which can provide a clue to the possible lipomatous lesion, prompting further evaluation with other imaging modalities. However, characterisation of tissue nature based on echogenicity is non-specific, particularly in solid lesions. As illustrated by patient 1, the lesion had both hyperechoic and hypoechoic components on USG (Figure 5), in which the hyperechoic components were subsequently shown to be lipomatous on MRI. Nonetheless, USG plays a key role in guiding the biopsy site for operative management.

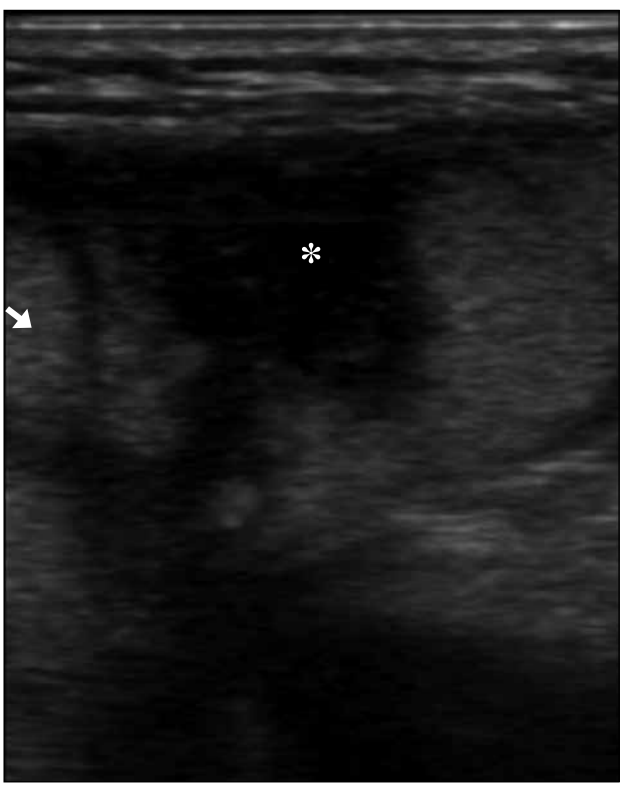

Figure 5. An ultrasonography image of patient 1 shows a hyperechoic component (arrow) at the superior aspect and the hypoechohic component (asterisk) of the mass.

MRI has the advantage of excellent radiation-free tissue characterisation and tissue contrast resolution. Fatty components of lipoblastoma can be readily distinguished on MRI. Other tissue characteristics such as cystic components, vascularity, and contrast enhancement can also be seen. These features correlate well with pathological findings.[7]

The main differential diagnoses of lipomatous soft tissue tumour include lipoblastoma, lipoma, liposarcoma, and teratoma. True lipoma is rare in young children and the presence of non-fatty components excludes lipoma. Absence of calcification or ossification is also uncommon in teratoma. Differentiation of lipoblastoma from liposarcoma based on imaging alone is usually difficult, in particular differentiating lipoblastoma from myxoid liposarcoma in the presence of apparently 'cystic' components that correspond to the abundant myxoid matrix, resulting in T2-hyperintense components. Diagnostic confusion may arise between lipoblastoma and myxoid liposarcoma, as in patients 1 and 3. However, liposarcoma is exceedingly rare in children younger than 10 years.[6] A lipomatous lesion with non-fatty components in a young child suggests a lipoblastoma rather than a malignant lesion.[8] Pathological analysis of myxoid liposarcoma typically shows evidence of nuclear atypia and absence of a prominent lobulated pattern as in lipoblastoma.[6]

Sometimes, a myxoid matrix engulfing the cells, with few lipoblasts, can be encountered, causing diagnostic confusion with myxoid liposarcoma. Nevertheless, mitosis is almost non-existent to rare in lipoblastoma.[2] The absence of nuclear atypia together with a lobulated appearance help differentiation of lipoblastoma from myxoid liposarcoma.[3,9] On the other hand, lipoblastoma can sometimes mimic atypical lipoma / well-differentiated liposarcoma, as illustrated in patient 2. The amount of fat will vary with the maturity of the tumour cells and the fat signal intensity correlates positively with the proportion of mature adipocytes. Finally, different from the infiltrative growth pattern of lipoblastomatosis and lipomatosis, both of which also occur in early childhood, in which lipoblastoma usually presents with a focal lobulated soft tissue mass or nodule.[6]

CONCLUSION

Lipoblastoma is a rare benign soft tissue tumour predominantly occurring in children younger than 3 years. These patients illustrate that lipoblastoma may present with a variable amount of fatty component on MRI, sometimes with cystic-like components or internal septations. Even in the case of rapid enlargement, lipoblastoma is the most likely diagnosis for a lipomatous tumour in a young child. Preoperative USG-guided biopsy can help to ascertain the diagnosis and surgical planning.

REFERENCES

1. Coffin CM. Lipoblastoma: an embryonal tumor of soft tissue related to organogenesis. Semin Diagn Pathol. 1994;11:98-103.
2. Coffin CM, Lowichik A, Putnam A. Lipoblastoma (LPB): a clinicopathologic and immunohistochemical analysis of 59 cases. Am J Surg Pathol. 2009;33:1705-12. cross ref
3. Collins MH, Chatten J. Lipoblastoma/lipoblastomatosis: a clinicopathologic study of 25 tumors. Am J Surg Pathol. 1997;21:1131-7. cross ref
4. Kerkeni Y, Sahnoun L, Ksia A, Hidouri S, Chahed J, Krichen I, et al. Lipoblastoma in childhood: about 10 cases. Afr J Paediatr Surg. 2014;11:32-4. cross ref
5. Rasalkar DD, Chu WC. Lipoblastomatous tumours, a rare entity: report of two cases in children. J Hong Kong Col Radiol. 2010;13:209-12.
6. Fletcher CD, Unni KK, Mertens F, editors. World Health Organization classification of tumors. Pathology and genetics of tumors of soft tissue and bone. Lyon: IARC Press; 2002.
7. Moholkar S, Sebire NJ, Roebuck DJ. Radiological-pathological correlation in lipoblastoma and lipoblastomatosis. Pediatr Radiol. 2006;36:851-6. cross ref
8. Chen CW, Chang WC, Lee HS, Ko KH, Chang CC, Huang GS. MRI features of lipoblastoma: differentiating from other palpable lipomatous tumor in pediatric patients. Clin Imaging. 2010;34:453-7. cross ref
9. Bolen JW, Thorning D. Benign lipoblastoma and myxoid liposarcoma: a comparative light- and electron-microscopic study. Am J Surg Pathol. 1980;4:163-74. cross ref