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CASE REPORT

Lipoblastomatous Tumours, a Rare Entity: Report of Two Cases in Children

DD Rasalkar, WCW Chu

Department of Imaging and Interventional Radiology, The Chinese University of Hong Kong, Prince of Wales Hospital, Shatin, Hong Kong

ABSTRACT

Lipoblastomatous tumours are rare neoplasms derived from embryonic white adipose tissues. We report on two children with different types of lipoblastomatous tumours at rare sites. The first had a localised lipoblastoma within the peritoneal cavity, successfully resected without recurrence. The second patient had diffuse lipoblastomatosis infiltrating the paraspinal muscles and spinal canal, which recurred two years after surgery. Radiologically, both lesions had characteristic fat density signals. The former lesion was benign-looking with a well-defined border, while the latter was infiltrative and had raised the suspicion of malignant liposarcoma. The definitive diagnosis was derived from characteristic histopathological features.

Key Words: *Lipoma; Lipomatosis; Liposarcoma; Magnetic resonance imaging; Retroperitoneal neoplasms; Tomography, X-ray computed*

中文摘要

罕見的脂肪母細胞腫瘤：兩個小兒病例

DD Rasalkar、朱昭穎

脂肪母細胞瘤是一種少見的腫瘤，源自於胎兒的白脂肪組織。本文報告兩名兒童的脂肪母細胞瘤發生在罕見位置。第一名兒童在腹膜腔處發現局限性脂肪母細胞瘤，經切除後未有復發。另一名兒童的瀰漫性脂肪母細胞瘤已滲入了脊旁肌及椎管，術後兩年復發。兩名兒童的病灶均出現脂肪密度的放射影像特徵。第一名兒童的病灶邊緣規則清晰，似乎屬良性。第二名兒童出現滲入性病灶，懷疑是惡性脂肪肉瘤。兩個病例均憑組織病理學特徵確診。

INTRODUCTION

Lipoblastomatous tumours are rare benign adipose tumours arising from immature adipocytes with varying degrees of differentiation.[1] These lesions are considered tumours of infancy and early childhood; about 90% occur in the first five years of life and occasionally at birth[2,3] with 2:1 male predominance. Sporadic cases have been reported in older children and adults. The most common are the upper and lower extremities, where they present as a painless nodule or mass. The head and neck, mediastinum, lung, trunk, mesentery, peritoneal cavity, retroperitoneum, intramuscular, and inguinal canal are less common sites.[2,3] The diffuse infiltrative entity (lipoblastomatosis) is more often noted than the circumscribed form of localised lipoblastoma. The former, which accounts for about 66% of cases,[4]

Correspondence: *Dr Winnie CW Chu, Department of Imaging and Interventional Radiology, The Chinese University of Hong Kong, Prince of Wales Hospital, Shatin, Hong Kong.*
Tel: (852) 2632 2299; Email: winnie@med.cuhk.edu.hk

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has a tendency to recur post-surgery.[1] We describe the imaging findings of the two different types of lipoblastomatous tumours in children, which occurred at rare sites.

CASE REPORTS

Case 1

A 14-year-old girl presented with swelling and discomfort in the lower abdomen. Bedside ultrasound showed a huge well-marginated hypoechoic mass occupying the central peritoneal cavity. Subsequent computed tomography (CT) abdomen revealed a capsulated, ellipsoid mass with fat attenuation in the central abdomen. It had no calcification, solid or cystic intralesional component. The mass was located anterior to the aorta and the inferior vena cava, displacing small bowel loops peripherally, hence suggesting its mesenteric origin (Figure 1a, b). The patient underwent radical resection of the lesion. Histologically, the mass was composed of a mixture of mature lipocytes against a background of myxoid tissue and a few mesenchymal cells (Figure 1c). There was no evidence of cellular atypia or mitotic activity and hence it was diagnosed to be a lipoblastoma. Follow-up imaging has shown no recurrence to date.

Case 2

A 1.5-year-old girl presented with an enlarging non-tender right buttock swelling from birth. There was no lower limb or sphincter dysfunction. The initial ultrasound showed a large 7-cm avascular hypoechoic mass at the right gluteal region. Subsequent CT confirmed a subcutaneous fatty tumour infiltrating the right paravertebral muscles and extending into the spinal canal from T12 to L3 levels, resulting in slight leftward displacement of the thecal sac. Magnetic resonance imaging (MRI) substantiated the CT findings and better delineated the extent of the lesion. The fatty lesion involved the right gluteus maximus, erector spinae and part of the latissimus dorsi muscles, which all appeared to be atrophic and distorted. Anteriorly, the lesion extended to the posterior pararenal space and displaced the right kidney forwards. Medially, the lesion infiltrated the right psoas muscle and tracked along its paraspinal insertion into the spinal canal via the T12/L1, L1/L2 and L2/3 neural foraminae. This intraspinal fatty component was contiguous with the normal epidural fat (Figure 2a, b). Radiologically, in view of its infiltrative appearance the lesion was indicative of lipoblastomatosis or liposarcoma. The patient underwent radical excision of the mass. On histology, the section was composed of variable mixtures of mature lipocytes against a background of myxoid tissue and a few mesenchymal cells (Figure 2c). In some areas, skeletal muscle fibres infiltrated with lipomatous cells were noted. No cellular atypia or mitotic activity was evident; the features being consistent with lipoblastomatosis. The patient developed another swelling two years after the surgery and MRI findings were consistent with a local recurrence. Conservative treatment was adopted after discussion with the parents. The lipoblastomatosis has remained the same size over the next five years.

DISCUSSION

Lipoblastomatous lesions usually present as soft, non-tender, lobular masses of variable size that grow slowly, although rapid growth has also been reported.[1] On plain radiographs, lipoblastomas at extremities usually present

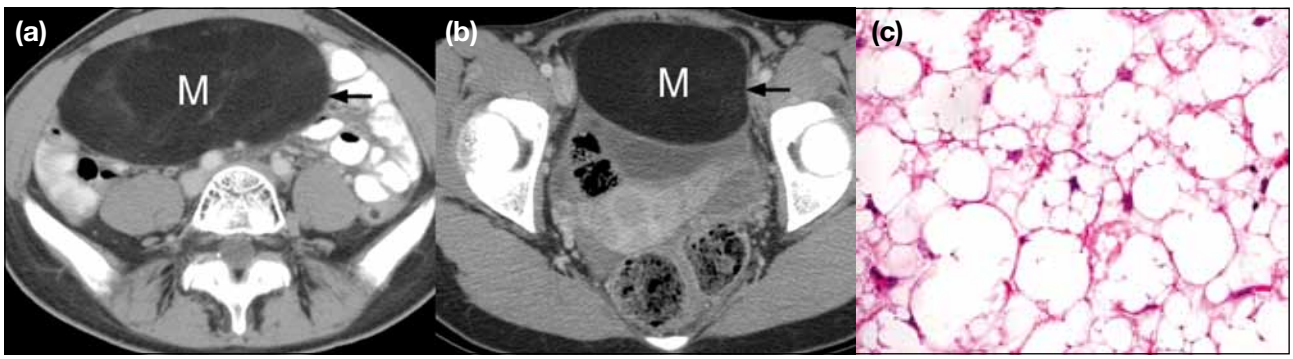

Figure 1. A 14-year-old girl presented with swelling and discomfort in the lower abdomen. (a, b) Axial computed tomographic images of the abdomen showing a well-defined peritoneal mass with fat attenuation (M) occupying the central abdomen and extending into the pelvis. The bowel loops are displaced peripherally. The lesion has a well-defined capsule (black arrows). It has been histologically confirmed to be lipoblastoma after complete resection. (c) A high-magnification histology slide shows multivacuolated lipoblast among lipocytes (H&E, x 600).

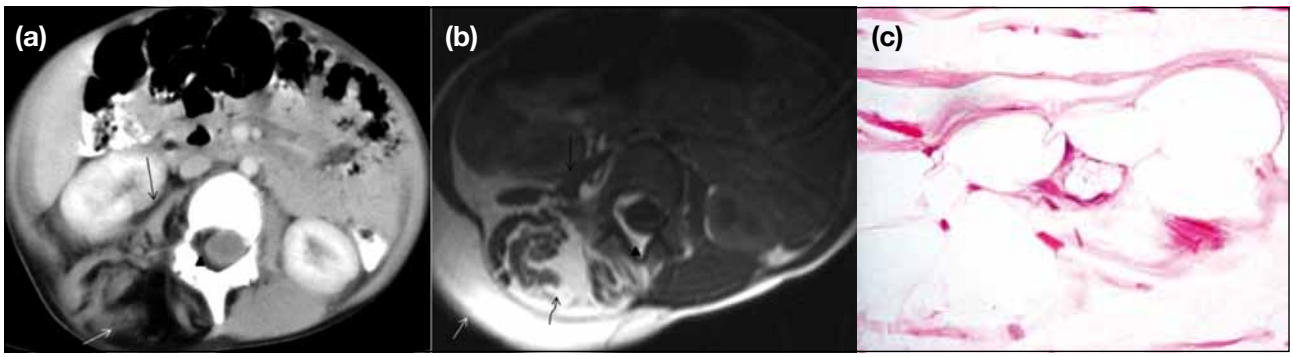

Figure 2. A 1.5-year-old girl presented with non-tender enlarging right buttock swelling since birth. (a) An axial computed tomographic abdomen image shows a fat attenuation lesion infiltrating the right paraspinal (white arrow) and psoas muscles (black arrow). There is forward displacement of right kidney and extension into the right spinal canal (black arrowhead). (b) An axial T1-weighted magnetic resonance image shows a T1 hyperintense lesion involving part of the latissimus dorsi muscles (curved black arrow) which appears atrophic and distorted. The signal of the lesion is the same as the subcutaneous fat (white arrow). Anteriorly, the lesion extends to the posterior pararenal space and displaces the right kidney forward. Medially, the lesion infiltrates the right psoas muscle (black arrow) and extends into the spinal canal (black arrowhead). The lesion has been histologically confirmed to be lipoblastomatosis, which has recurred two years after resection. (c) A high-magnification histological slide shows many multivacuolated lipoblasts with nuclei indented by the vacuoles (H&E, x 600).

as a non-specific soft-tissue mass without calcification or erosion of adjacent bones. On ultrasound, the lesion is lobulated and of mixed echogenicity. With large lesions, adjacent structures are usually displaced but not significantly compressed.

The lipomatous features of the lesion, which strongly depend on the pervasiveness of mature lipocytes,[1] are usually evident on CT and MRI that are useful imaging modalities for delineating the lesion. On CT, lipoblastomatous lesions are of low attenuation and isodense to subcutaneous fat. On MRI, the tumours are usually isointense to subcutaneous fat, and appear hyperintense on both T1 and T2 fast spin echo sequences, while the fatty signals are completely attenuated on fat-suppressed sequences. Occasionally, increased cellularity (myxoid and mesenchymal components) of lipoblastomas might lead to lower T1-weighted signal and make the lesions appear more heterogeneous than normal subcutaneous fat.[5]

Morphologically, the localised type of the lesions has well-delineated margins while the diffuse type has infiltrating margins. In the latter type, differentiation from lipoma or liposarcoma might be difficult if solely based on imaging. The main differential diagnosis of lipoblastoma / lipoblastomatosis is myxoid liposarcoma, which has a peak incidence between the third and sixth decades of life. Myxoid liposarcoma is, however, uncommon in adolescents and exceedingly rare in children aged younger than 10 years. Therefore, when imaging identifies a fatty infiltrative lesion in a paediatric patient, lipoblastoma / lipoblastomatosis should be regarded as more likely than its aggressive malignant counterpart.

Histologically, both lipoblastoma and myxoid liposarcoma can appear lobulated. They both contain lipoblasts and spindle cells, deposited in a myxoid matrix with a prominent plexiform vasculature. However, lipoblastomas have a more pronounced lobulated pattern than myxoid liposarcoma, and cytology suggestive of malignant potential (nuclear atypia, mitotic figures, hyperchromasia, and hypercellularity) is relatively uncommon.[6]

The clinical course of lipoblastoma is benign, and complete excision usually leads to an excellent prognosis. Patients with a focal lipoblastoma (as in case 1) are unlikely to require further surgery after initial resection. Recurrence is restricted to diffuse lipoblastomatosis (as in case 2) and attributed to incomplete tumour removal. Approximately 9 to 22% of patients with lipoblastomatosis have recurrent disease, usually within two years of presentation. Therefore, patients with the diffuse type of lesion warrant close clinical monitoring.[6] Due to rarity of this disease, there is no standardised treatment protocol with regard to chemotherapy and radiotherapy of the condition. O'Brien et al,[7] however, reported a case of cervical spine lipoblastomatosis, which responded remarkably to chemotherapy (six cycles of vincristine, ifosfamide

and actinomycin according to the International Society of Paediatric Oncology Malignant Mesenchymal Tumor 89 protocol).[8] They speculated that the tumour cells within the cervical spine underwent rapid re-growth following the initial surgery, but were obliterated by chemotherapy. However, chemotherapy failed to prevent subsequent proliferation of lipoblastoma cells within the extraspinal component of the same lesion. In this particular case report, why intraspinal and extraspinal components of the tumour responded differently remains unclear. Adjuvant local therapy such as radiation can be an alternative intervention, but is generally limited by the young age of the patients, and similar considerations apply to cytotoxic chemotherapy and ionising radiation for this rare tumour of children and teenagers.

In conclusion, this paper illustrates the radiological appearance of two different presentations of lipomatous lesions in children. In both localised and diffuse types, the most characteristic imaging feature is the fatty density / signal within the lesion. In the diffuse type of lipoblastomatosis, there might be varying features of infiltration mimicking malignant liposarcoma. Although the definitive diagnosis relies on histology, imaging is important for preoperative planning, as the mainstay of treatment is complete surgical resection.

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