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Lipofibromatosis Arising in a Pediatric Forearm -A Case Report-

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ABSTRACT

Lipofibromatosis is a rare pediatric tumour described by Fetsch et al. in 2000. There have been few reports about this tumour and few descriptions of its imaging features. We report a case of this tumour forming a slowly growing, painless mass arising in the forearm of a ten-month-old boy. This is the first report of lipofibromatosis to present the radiological findings before surgery. Plain radiography and computed tomography demonstrated a tumour located just beneath the fascia. Ultrasonic study showed a hyperechoic tumour of 33 x 30 x 7 mm in size. Magnetic resonance imaging demonstrated a multilobular mass with high signal intensities on both T1 / T2 weighted images. Macroscopically, the tumour was poorly marginated. Microscopically, the tumour was composed of abundant adipose tissues transversed by fibroblastic bands. The adipose tissue occupied over 70% of the tumour. Adipocytes and fibroblastic element infiltrated into the normal skeletal muscle tissues. Recognition of this clinical entity is needed, especially in cases of lipomatous tumours arising in pediatric hands and feet.

INTRODUCTION

Lipofibromatosis is a rare pediatric tumour described by Fetsch et al. in 2000 [1]. Since then, there have been few reports about this tumour and few descriptions of its image findings. We report a case of this tumour occurring in the forearm of an infant together with the radiological and histological findings.

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Key words: lipofibromatosis, lipomatous tumour, infant, radiological finding

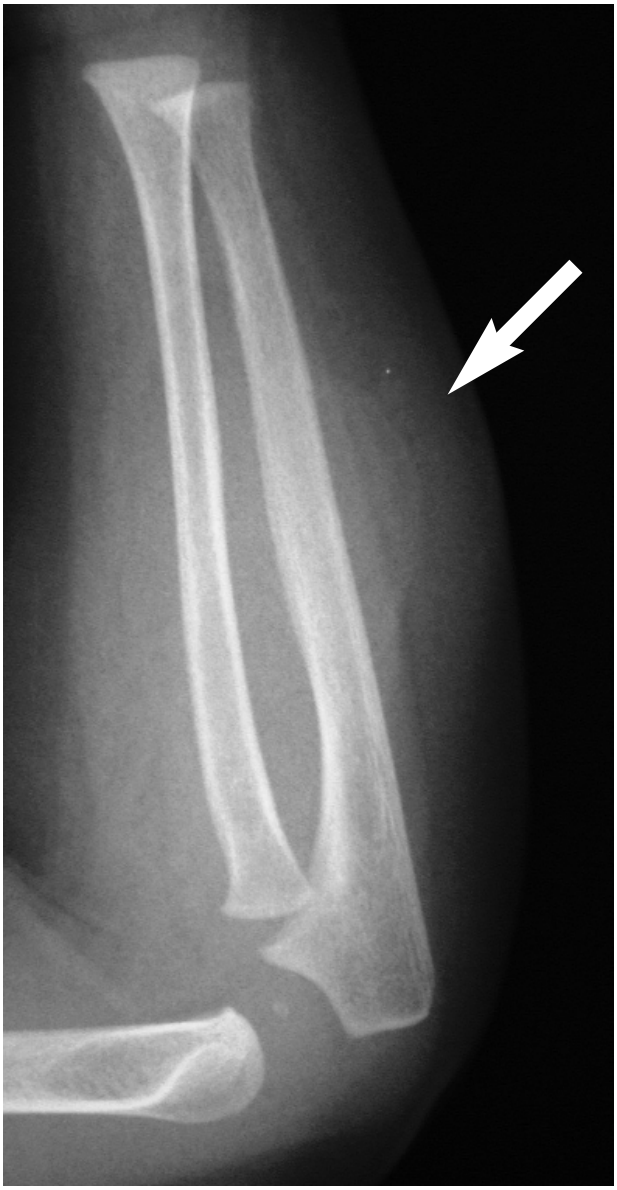

Fig 1. Plain radiogram showing soft tissue swelling (arrow).

CASE REPORT

A ten-month-old boy was referred to our clinic with a painless and slow-growing tumour in the right forearm. The tumour, 25 x 20 mm in size, had a smooth surface, clear margin, and was soft. Initial plain radiogram showed a mass beneath the fascia (Fig 1). Ultrasonic study showed a hyperechoic tumour of 25 x 22 x 6 mm in size. During the follow-up for five months, the mass increased in size to 33 x 30 x 7 mm. Computed tomography (CT) showed a tumour located just beneath the fascia, the density of which was similar to the subcutaneous fat (Fig 2). Magnetic resonance imaging (MRI) revealed a multilobular mass of homogenous high signal intensities on both T1 / T2 weighted images.

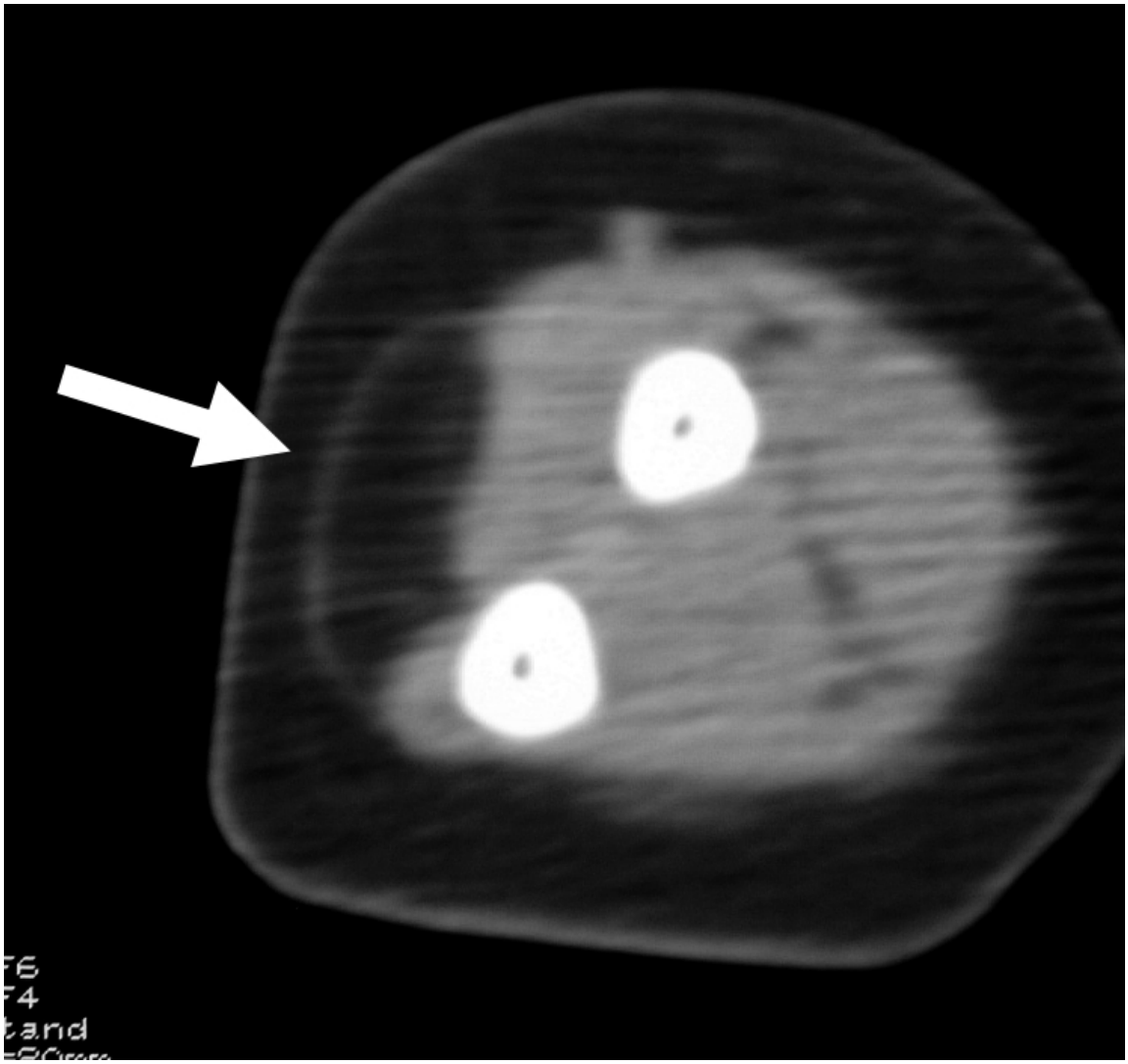

Fig 2. CT scan demonstrated a tumour just beneath the muscular fascia (arrow).

Radiological differential diagnosis was intramuscular lipoma, lipoblastoma, and other kind of lipomatous tumours. An attempt was made at marginal excision, but the tumour was found to be not encapsulated and to have continuity with the adjacent tissues. Removal of the tumour together with the surrounding normal muscles was performed.

Grossly, the tumour, 30 x 25 x 10 mm in size, was a mass yellowish in color, soft in consistency, and poorly demarcated, involving the muscle (Fig 4). Microscopic examination revealed abundant adipose tissue transversed by fibroblastic bands (Fig 5a). The fibroblastic component had relatively high cellularity with an immature appearance, no cellular atypia, and no nuclear pleomorphism (Fig 5b). The adipose tissue occupied over 70% of the tumour. The adipocytes did not have lipoblastic appearances. Univacuolated cells were present between the fibroblastic fascicles and the mature adipocytes (Fig 5c). Around the margin, adipocytes and fibroblastic

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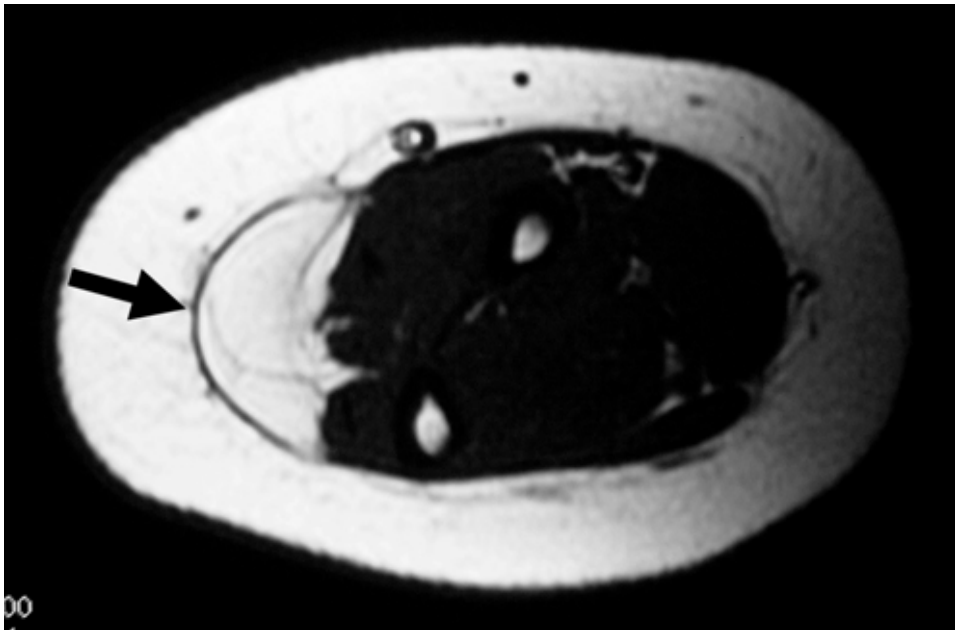

Fig 3. MR images. a) T1 weighted image. b) T2 weighted image. The tumour had homogenously high signal intensities on both T1 / T2 weighted images (arrow).

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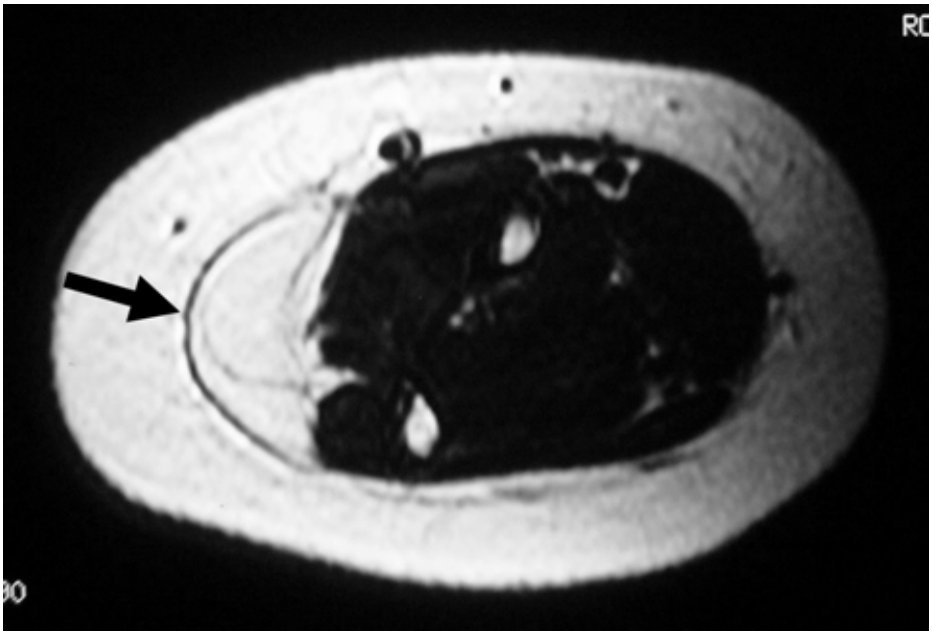

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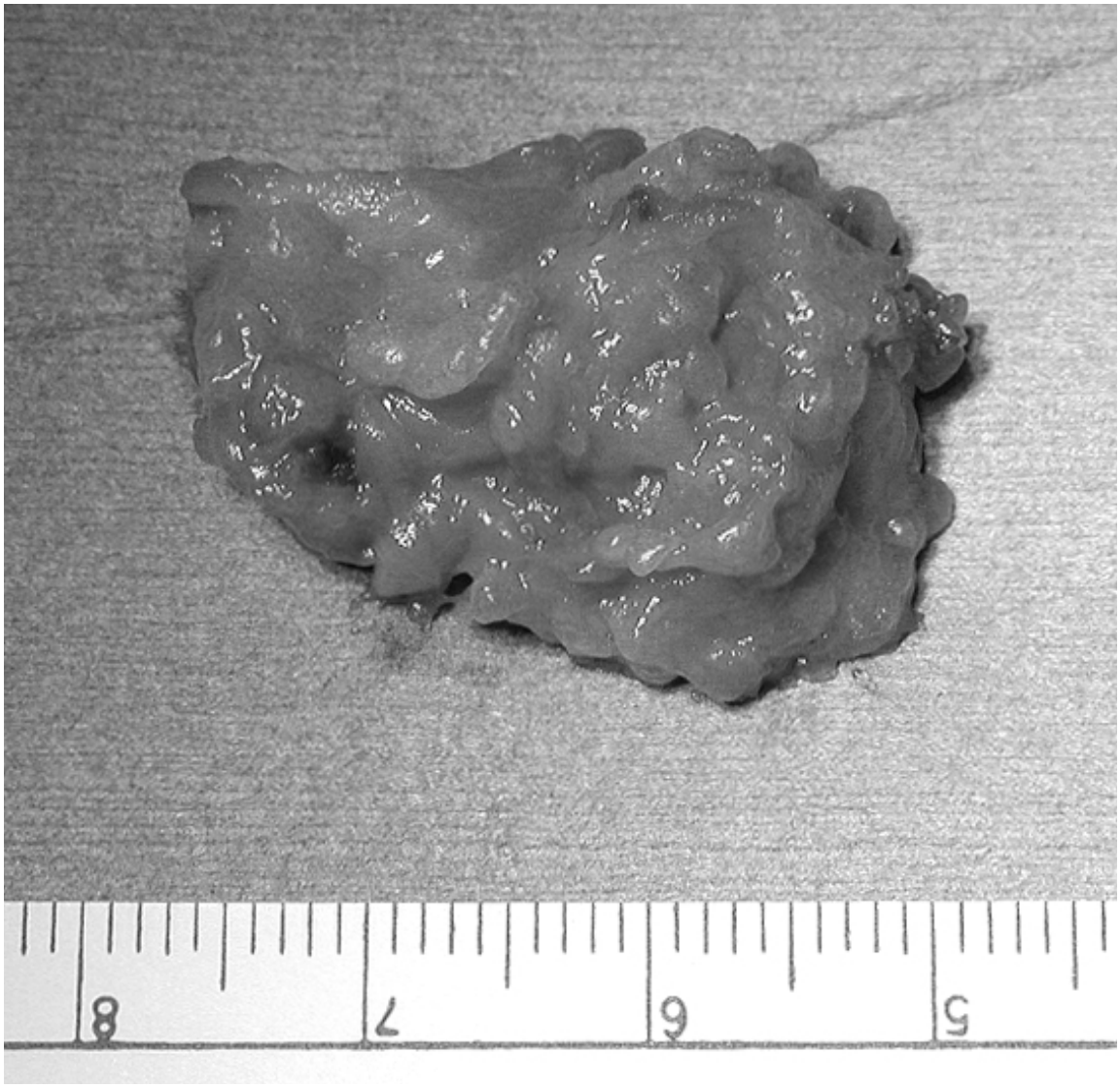

Fig 4. Gross appearance of the specimen.

cells infiltrated into the normal skeletal muscle tissue (Fig 5b). Immunohistochemical study showed that the fibroblastic cell exhibited immunoreactivity for S-100 (focal), CD99, CD34, alpha SMA (focal), HHF-35 (focal, weak), bcl-2 (focal, weak). No reactivity was detected for EMA, melan A, HMB-45. Adipocytes exhibited a strong immunoreactivity for S-100, but negative reactivities for the other antibodies. The rate of positive immunoreactivity for Ki67 was about 7-8% in the areas of fibroblastic components.

The tumour was diagnosed as lipofibromatosis. The patient has had no local recurrence for one year after the operation.

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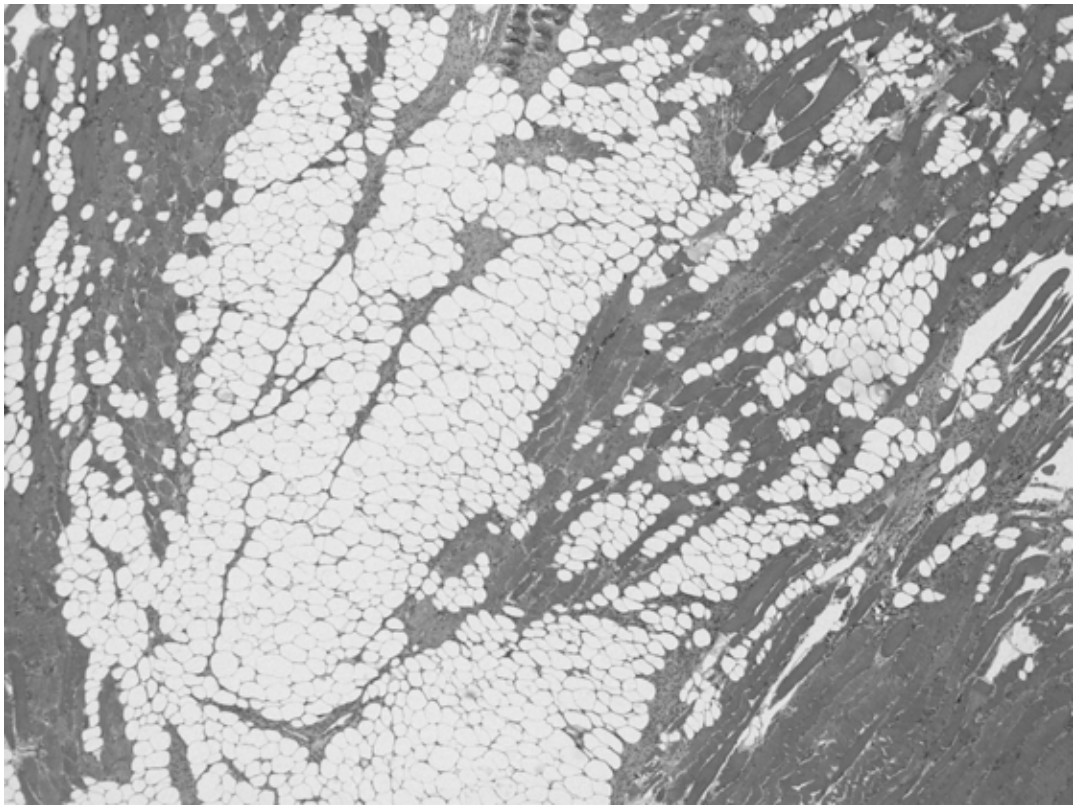

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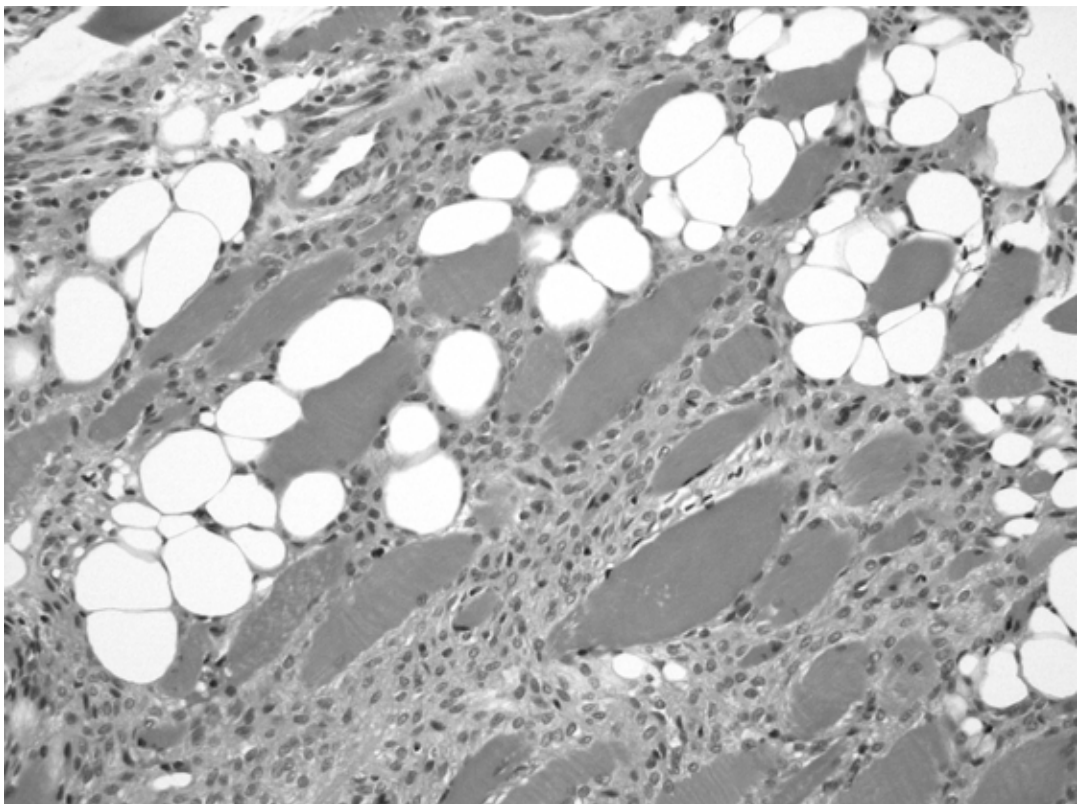

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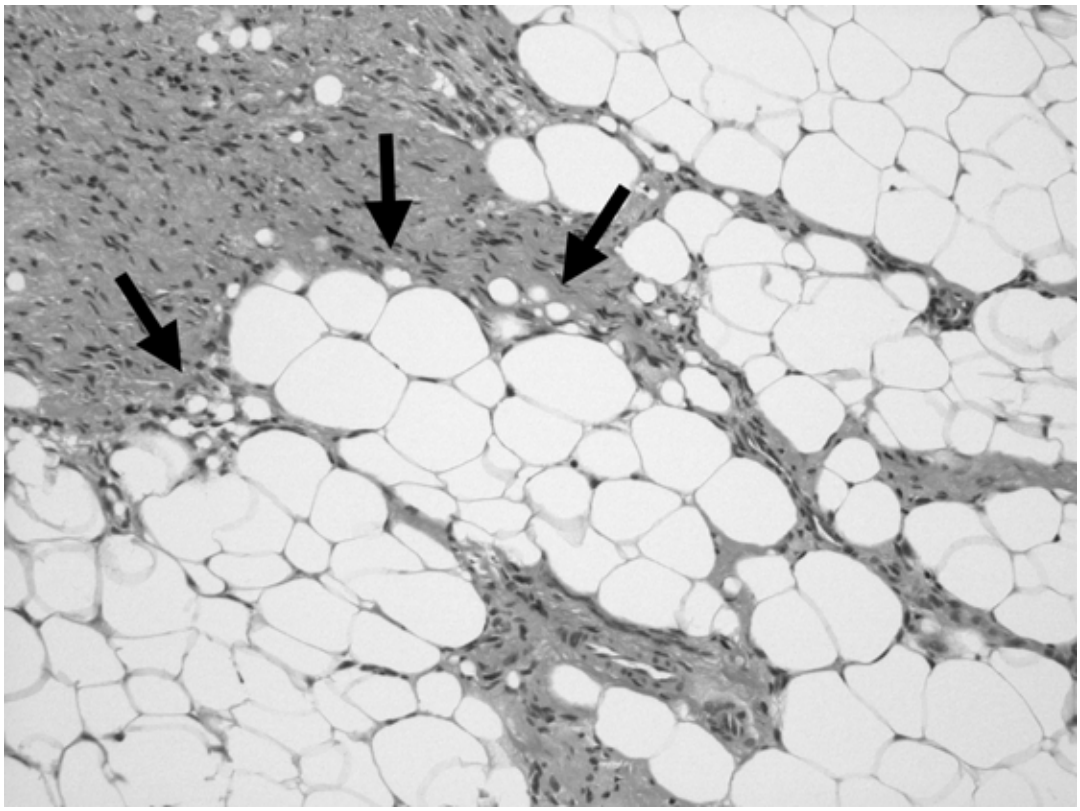

Fig 5. Histology of the specimen. a) Low-power view. Abundant fat involving dense fibrous tissue was noted. b) High-power view of mature adipocytes and spindled fibroblastic element. The skeletal muscle was remarkably infiltrated. c) Univacuolated cells were noted between them (arrow).

DISCUSSION

Lipofibromatosis was proposed by Fetsch et al. in 2000. They classified it as a tumour consisting of abundant adipose tissue with a spindled fibroblastic element involving the septa of fat tissue. It forms a slowly growing painless mass, especially arising in the hands and feet of children, and in some cases congenitally. The ages ranged from 11 days to 12 years (median age, 1 year) at the time of initial biopsy or resection. There was a more than 2:1 male predominance [1,3].

Histopathologically, adipose tissue is an integral component of this tumour, typically comprising more than 50% of the tumour. In the present case, the adipose tissue occupied more than 70% of the tumour. On the other hand, true fibromatoses exhibit a more solid, sheet-like growth of the fibroblastic element and do not contain fat as an integral component [1,3]. Fetsch et al. described that lipofibromatosis had been interpreted as a type of infantile or juvenile fibromatosis, a variant of fibrous hamartoma of infancy, a calcifying aponeurotic fibroma, and a fibrosing lipoblastoma. The image findings of this tumour were not described in their report. An extensive survey of the literature revealed that there was only one case report of recurrence presenting an intramuscular, poorly circumscribed, huge mass with calci-

fication, showing heterogeneous MR signal intensities [2]. Our case is the first report to present the image findings of lipofibromatosis before operation. The imaging features of the present case were not different from those seen in lipomas, although microscopically the tumour had a highly cellular fibroblastic component. Therefore, it seems difficult to diagnose lipofibromatosis by its imaging features alone. Fetsch et al. reported that 72% of such patients had regrowth of their tumour or had persistent disease, and most of whom had been managed by incomplete removal. Recognition of this clinical entity is needed, especially in cases of lipomatous tumours arising in pediatric hands and feet.

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