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Lipoblastoma in childhood: About 10 cases

Yosra Kerkeni, Lassaad Sahnoun, Amine Ksia, Saida Hidouri, Jamila Chahed, Imed Krichen, Mongi Mekki, Mohsen Belghith, Abdellatif Nouri

ABSTRACT

Background: Lipoblastoma is a rare benign mesenchymal tumour of embryonal fat that occurs almost exclusively in infants and children. This determined the epidemiological, clinical and therapeutic aspect of this disease. **Materials and Methods:** A total of 10 cases of pathologically proven lipoblastoma from 2003 to 2012 were reviewed. **Results:** There were six boys and four girls ranging in age from 7 months to 9 years. A soft-tissue mass was the main complain in nine patients. The various locations of the mass were mediastinal, thigh, buttock, inguino-scrotal, the greater omentum and the Latissimus dorsi. Lesions measured 5-15 cm. complete excision was done. The median time of follow-up was 42 months (ranges between 18 and 84 months). There were no recurrences. **Conclusion:** It is important to consider lipoblastoma in the diagnosis of a rapidly enlarging fatty mass in children. Complete resection is the only definitive treatment and should not be delayed when impingement on surrounding structures is imminent. There is a tendency for these lesions to recur despite presumed complete excision. Therefore, follow-up for a minimum of 5 years is recommended.

Key words: Adipose tumour, child, lipoblastoma, lipoblastomatosis

INTRODUCTION

Lipoblastoma is a rare benign neoplasm of the adipose tissue occurring most commonly in infants and young children, usually those younger than 3 years.[1] There is a slight male predominance.[2] This tumour presents as a localised well-circumscribed lesion (lipoblastoma) or as a multicentric lesion (lipoblastomatosis).[2] Most lipoblastomas occur in the superficial tissues of the arms and legs but may also arise in the head and neck, parotid, eyelid, tonsillar fossa, trunk, mediastinum and retroperitoneum regions. <200 cases have been reported in the literature since its first description in 1926.[3]

Department of Pediatric Surgery, Monastir Hospital, Monastir, Tunisia

Address for correspondence:
Dr. Yosra Kerkeni,
Department of Pediatric Surgery, Monastir Hospital, 5000 Monastir, Tunisia.
E-mail: yosrakerkeni@yahoo.fr

MATERIALS AND METHODS

A total of 10 cases of pathologically proven lipoblastoma treated at Monastir University Hospital from 2003 to 2012 were reviewed. There were six boys and four girls, ranging in age from 7 months to 9 years. The clinical presentation, laboratory findings, radiologic investigations, surgical treatments, pathological findings and follow-up were reviewed.

RESULTS

The clinical data are summarised in Table 1. The age of 10 patients at the time of operation ranged from 7 months to 9 years. Lipoblastoma occurs almost exclusively in infants and children. Our data show that eight of our patients were younger than 3 years. There were six boys and four girls. Of the 10 tumours studied, seven tumours presented as painless superficial soft-tissue masses. One tumour was mediastinal [Figure 1]. Another tumour was mesenteric and was discovered due to abdominal pain [Figure 2]. Tumours occurred on the thigh in two patients, on the buttock in two patients, on the back, in the axilla and inguino-scrotal-area in one patient each. Lesions measured 5-15 cm diameter in greatest dimension. The largest tumour was in the axilla and measured 15 cm. The computed tomography exam showed regions of fat density, separated by septae of soft-tissue density and does not enhance following contrast administration [Figure 3]. In seven cases, the lesions were well-circumscribed. Complete excision was done in all patients. Follow-up ranged from 18 months to 84 months. Histological features showed lobules of mature and immature fat cells, primitive mesenchymal cells and typical lipoblasts

Table 1: Patient characteristics

Case no.	Sex	Age	Site	Size	Histological features	Follow-up
1	F	3 years	Omental	10 cm	Lipoblastoma	84 months
2	F	7 months	Latissimus dorsi muscle	6 cm	Lipoblastoma	84 months
3	M	3 years	Left inguino scrotal area	5 cm	Lipoblastoma	48 months
4	M	3 months	Left thigh	9 cm	Lipoblastoma	36 months
5	M	2 years	Right buttock	6 cm	Lipoblastoma	36 months
6	M	1 years	Mediastinal	5 cm	Lipoblastomatosis	30 months
7	F	4 years	Right thigh	10 cm	Lipoblastoma	30 months
8	M	9 years	Right buttock	12 cm	Lipoblastomatosis	30 months
9	M	2 years	Right axilla	15 cm	Lipoblastoma	24 months
10	F	1.5 years	Right inguinal	10 cm	Lipoblastoma	18 months

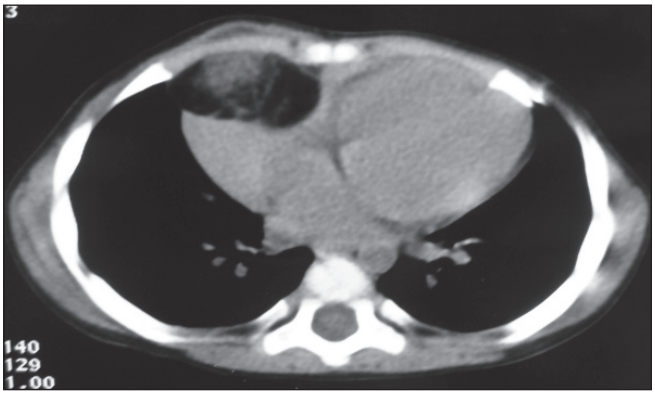


Figure 1: Mediastinal lipoblastoma

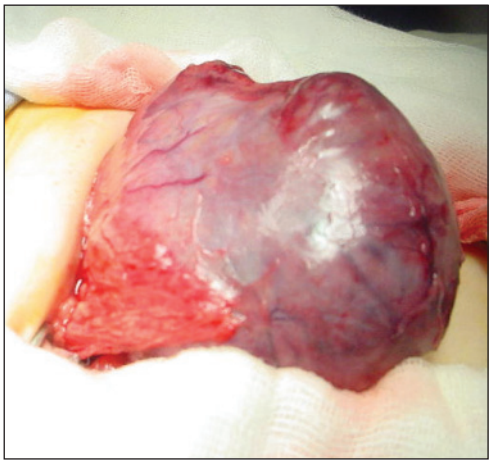


Figure 2: Mesentery lipoblastoma

of the monovacuolated and multivacuolated types showing varying degrees of differentiation. The lobules were separated by fibrous trabeculae that contained numerous small blood vessels and variable proportions of hyaline collagen and fibroblasts corresponding to lipoblastoma. In two patients (case 6 and 8), the lesions were poorly circumscribed, diffuse and infiltrative corresponding to another variety of lipoblastoma named: Lipoblastomatosis.

DISCUSSION

Of all the soft-tissue neoplasms reported in children, lipoblastoma represents only 5-30% of them. It may be single or multiple. Lipoblastoma occurs almost exclusively in infancy and early childhood, most commonly in boys and in the upper and lower extremities.[4,5] Occasionally, other sites (head, neck, trunk, retroperitoneum and mesentery) can develop lipoblastomas. In our series two interesting tumour locations were noted: Mediastinal and omental tumour. To the best of our knowledge only eight cases of omental lipoblastoma have been reported.[6]

Although some investigators have acknowledged the potential utility in using ultrasonography, computed tomography, or magnetic resonance imaging, surgical removal and pathologic evaluation is mandatory to exclude malignant processes.[5] Lipoblastomas have no known malignant potential; therefore, additional studies aimed at staging are currently not indicated in the absence of any histologic changes consistent with malignancy. However, recurrences have been reported;[4] therefore, periodic re-evaluation is indicated. Lipoblastomas represent a variant of lipoma that, when circumscribed and benign, can be designated as embryonal or foetal lipoma. When deep-seated and ill-defined, these lesions are designated as lipoblastomatosis.[7] It can be mistaken for other histologic types such as lipoma, hibernoma, rhabdomyosarcoma, or well-differentiated liposarcoma during frozen section analysis. This might result in inappropriate treatment. A lipoblastoma can be confused with a myxoid liposarcoma due to the presence of lipoblasts and the plexiform vascular pattern in a myxoid background. In these histologically borderline or difficult cases, cytogenetics can be of diagnostic value.[8] Rearrangements of chromosomal region 8q11-13 are diagnostic of lipoblastoma, whereas a distinctive translocation *t*(12;16) is observed with myxoid liposarcoma.[2-6] However, the young age

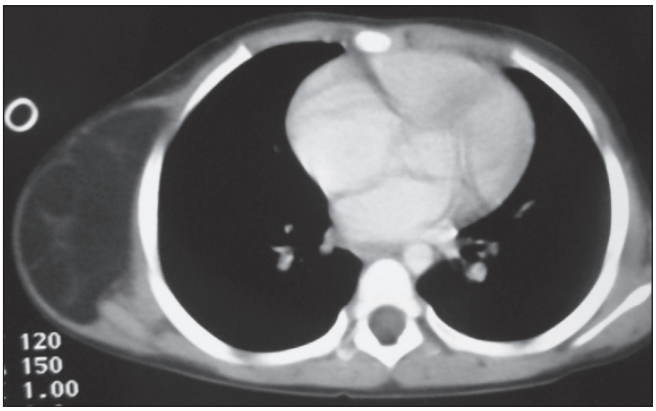


Figure 3: Computed tomography exam showing regions of fat density, separated by septae of soft tissue density

of the patient, the lobulation and the absence of pleomorphic cells tend to rule out the malignant diagnosis. These rearrangements of the consequences of 8q11-q13 target the PLAG1 gene, which becomes upregulated through promoter swapping.[9]

It is now well-established that lipoblastoma is a benign tumour best treated by complete surgical excision and that a radical operation is not necessary and may be mutilating, especially in children.

Patients with lipoblastoma have an excellent prognosis despite the tumour's potential to invade locally and grow rapidly to a considerable size. Metastases have never been reported.[5] The recurrence rate reported is between 14% and 25% respectively. [4,5] There were no recurrences for our 10 patients. However, this rate is an underestimate of the true recurrence rate because of the short period of follow-up. Moreover in our series, there were two cases of lipoblastomatosis, which is more likely to recur. Perhaps, if more patients had been followed-up longer, a tumour recurrence may have been diagnosed. Appropriate length of follow-up is controversial. Various minimum lengths of follow-up have been suggested, including 1, 2, 3 and 5 years.[5-10] Time-to tumour recurrence should determine the length of follow-up.

CONCLUSION

Lipoblastomas are rare benign tumours most often seen in infants and young children. They exhibit a wide range of cellular differentiation and maturation, from lipoblasts to mature adipocytes. Complete resection achieves optimal results, but recurrence is possible and close follow-up is mandatory.

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