

Thoracic wall lipoblastoma: a rare case with rare presentation

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Lipoblastoma is a rare benign tumor occurring in infancy and early childhood. It is usually located on the extremities but can be found anywhere on the body. Normally, it is well capsulated, but when it is multicentric in origin and diffused in nature it is known as lipoblastomatosis. We report a case of a 1-year-old boy who presented with a huge swelling on the back that, on postoperative histological examination, was diagnosed as lipoblastoma. *Ann Pediatr Surg* 8:56–58 © 2012 Annals of Pediatric Surgery.

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Introduction

Lipoblastoma is a rare lipomatous neoplasm encountered almost exclusively in infants and young children [1]. Adipose tumors comprise 10% of all pediatric soft tissue tumors, of which approximately 5–30% are of the lipoblastomatous type [1,2]. Lipoblastoma arises from embryonic white fat cells, which are usually well-circumscribed benign tumors with potential to produce mass effect on adjacent structures. Complete surgical resection is possible, but recurrence has been reported in up to 25% of cases [3]. We report a case of lipoblastoma in an infant and review literature pertaining to diagnosis and clinical management of these tumors.

Case report

A 1-year-old boy was presented at our institute with a huge mass on the back since 4 months. It had been rapidly increasing in size since its initiation as a small swelling. As the patient belonged to a remote tribal area of the country, previous documentation about the mass was unavailable. It extended from the upper part of the back to the neck (Fig. 1). There was no history of trauma, unusual rashes or infections, and no physical and developmental delay. There was no other similar swelling elsewhere. Family history was inconclusive. On palpation, the swelling was nontender, ill circumscribed, and without overlying skin changes. Complete blood count and metabolic profile were within normal limits. Chest radiograph was normal. Ultrasonography of the mass had lipomatous echogenicity and echo texture. MRI revealed a subcutaneous soft tissue mass in the upper back, which extended over the neck (Fig. 2) without intraspinal extension. Complete excision was performed without any complication. The gross specimen measured 18 × 14 cm and weighed 380 g (Fig. 3). It was not well encapsulated and was interspaced with fibrous septa. Its extension into the adjacent muscle was not clearly evident by MRI, but during surgery it was found to have minimal invasion in the adjacent muscle mass, which was removed by raising the flaps of muscles. Histology of the tumor (Fig. 4) was suggestive of lipoblastoma. The patient's 6-monthly

evaluation for 2 subsequent years (postoperative) was normal. His wound healed without any complication. No obvious signs of recurrence were noted, and the patient's next follow-up visit was rescheduled after 1 year.

Discussion

The term lipoblastoma was coined by Jaffe in 1926 [4] to describe atypical lipomatous lesions, which consisted of cells resembling embryonic white fat. This term was intended to differentiate these lesions from common lipoma, which contains no lipoblasts [4]. Lipoblastomas are usually well circumscribed. They can also be diffuse, called lipoblastomatosis, and are more difficult to excise fully. Otherwise, lipoblastoma and lipoblastomatosis display identical histological features.

Lipoblastomas exhibit a wide spectrum of cellular differentiation and maturation comprising primitive mesenchymal cells, spindle cells, lipoblasts, and mature adipocytes. The maturation pattern of lipoblastoma tends to be characterized by a greater proportion of mature adipocytes at the center of the lobule and with lipoblasts on the periphery. Fibrous septa are often interspersed among tumor lobules. Abnormal mitosis, which is characteristic of liposarcoma, is not found in lipoblastoma [1]. The tumor is often encapsulated but can invade the adjacent structures [5]. Lipoblastoma can cause mass effect on surrounding structures, causing abdominal pain, respiratory insufficiency, lethargy, and hernia [2,6].

A review of 84 cases from the literature found that 61% of lipoblastomas occur in the extremities, followed by 15% on the trunk, 14% in the abdomen, and 11% on the head and neck [7]. Lipoblastomas are generally found in pediatric patients, with an estimated 55% of cases diagnosed in patients under 1 year of age and 90% of cases becoming obvious by the age of 3 years [1,8]. Lipoblastomas are always benign, and it is imperative to differentiate these lesions from potentially malignant lipomatous tumors. Liposarcoma is relatively common in adults but uncommon in infancy and rarely observed in

Fig. 1



Lipoblastoma of the thoracic wall.

Fig. 3



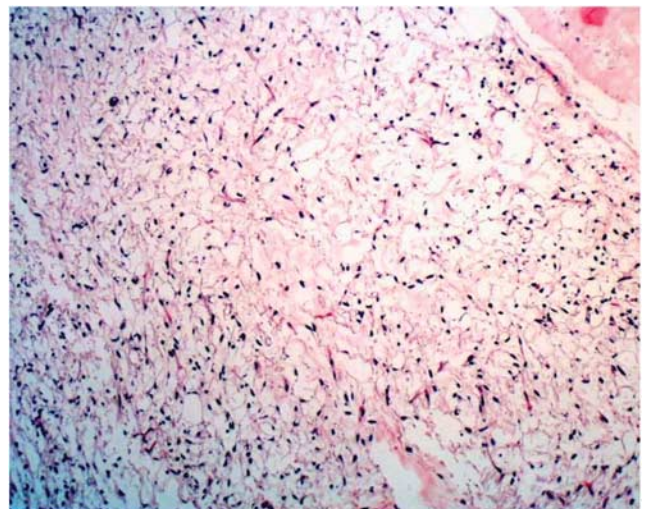
Resected specimen.

Fig. 2



MRI thorax showing a huge lipoblastoma on the back.

Fig. 4



Histology suggestive of lipoblastoma.

the first decade of life. Nuclear atypia and abnormal mitosis are characteristic of liposarcoma, but these findings are unusual in lipoblastoma. Complete surgical resection of tumor with microscopically healthy tissue margins yields excellent prognosis. Recurrence has been reported in 14–25% of cases, [3] usually due to incomplete resection

or diffuse disease. Lipoblastomatosis exhibits higher recurrence rate [1,3]. In our case, at 2-year follow-up, there was no sign of recurrence of tumor. As recurrence has been reported to occur for as long as 84 months postoperatively [2], a follow-up period of at least 3–5 years is recommended. Recurrent growth should be excised.

Conclusion

Lipoblastoma is a rare benign tumor most often seen in infants and young children and exhibits a wide range of cellular differentiation and maturation from lipoblasts to mature adipocytes. Complete resection to achieve optimal results is advised, but recurrence is possible.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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