CASE REPORT

Recurrent lipoblastoma of upper extremity in a 9-year-old boy

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SUMMARY

Lipoblastoma is a benign soft-tissue tumour of infancy and early childhood. The location of the tumour varies from that of the extremities, trunk, retroperitoneum, mediastinal, inguinal and scrotal regions. The most common symptoms are painless mass with or without increasing size. Complete surgical excision is the gold standard for managing this entity. Though rare, the recurrence occurs usually because of incomplete excision. We report a case of recurrent lipoblastoma of the upper extremity in a 9-year-old boy.

BACKGROUND

Lipoblastoma is a rare benign soft-tissue tumour found in infancy and early childhood.^{1 2} The common presenting symptom is a painless, slowly growing mass usually involving the extremities or trunk but can be found in an unusual region such as the scrotum, retroperitoneum or mediastinum. Though they are benign, they can also invade locally and grow rapidly, warranting a complete wide surgical excision and close follow-up period of 2 years. We report the case of a 9-year-old boy who presented with recurrent lipoblastoma of the upper extremity along with a review of the literature.

CASE PRESENTATION

A 9-year-old boy presented in our paediatric surgical outpatient department with symptoms of a slow growing mass over the left shoulder for the last 2 years. A history of attempted surgical excision 6 months previously in another centre was given by the parents. The child also received corticosteroids for 6 weeks prior to surgical resection. Post surgery

swelling recurred within 6 months and was progressively increasing in size. Physical examination revealed a non-tender, irregular, firm and mobile mass measuring 5×4 cm at the left shoulder region just above the acromioclavicular joint. The rest of the physical examination, including systemic examination, was unremarkable.

INVESTIGATIONS

Serum chemistry, complete blood count and α -fetoprotein were within normal limits. Radiological investigations included chest X-ray, ultrasonography and MRI. Histological slides available from previous surgery were reviewed to confirm the diagnosis of lipoblastoma. The MRI T1-weighted image of the left shoulder showed a high signal bright fat containing lesion (figure 1A) in the subcutaneous plane without any extension into the shoulder joint or thoracic cavity. MR angiography (MRA) showed the lesion to be supplied by a feeding vessel from the external carotid artery (figure 1B).

DIFFERENTIAL DIAGNOSIS

- ▶ Lipofibromatosis/infantile fibromatosis
- ▶ Myxoid liposarcoma
- ▶ Well-differentiated liposarcoma
- ▶ Fibrosarcoma

TREATMENT

Elective surgical excision was carried out under general anaesthesia. A complete wide excision of the mass was achieved successfully without any damage to the capsule of the shoulder joint.

Figure 1 (A) MRI T1-weighted image of the left shoulder showed a high signal bright fat containing lesion (arrow) in the subcutaneous plane without any extension into the shoulder joint or thoracic cavity. (B) MR angiography showed the lesion (asterisk) to be supplied by a feeding vessel (arrow) from the external carotid artery.

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Figure 2 (A) Photomicrograph of the histopathology of the excised mass showing hypocellular lobules of adipocytes in various stages of differentiation. (B) Lobular architecture with prominent fibrous septa.

OUTCOME AND FOLLOW-UP

The result of the histopathology of the resected specimen revealed features suggestive of lipoblastoma (figure 2A,B). Margins of the resected specimen were free from tumour. The postoperative period was uneventful. The patient had undergone postoperative MRI after 3 months of surgery, which showed no recurrence or residual disease. Presently, the child is symptom free with no evidence of recurrence after the follow-up period of 1 year.

DISCUSSION

Lipoblastoma in children is usually a rare benign neoplasm that arises within the adipose tissue and contains a spectrum of fat cells ranging from immature lipoblasts to mature adipocytes. The term lipoblastoma was coined by Jaffe in 1926.^{3 4} The ability of lipoblastoma to differentiate into a common lipoma was recognised by Van Meurs⁵ in 1947, supporting the concept that lipoblastoma was a result of continuous proliferation of immature fat cells and lobules in the postnatal period. Lipoblastoma is encountered mostly during the first year of life and it is more frequent in boys than girls.^{6 7} The tumour hardly ever occurs after 5 years of age. On the other hand, the malignant tumour, fibrosarcoma, rarely occurs below the age of 5. This fact is important in preparing excisional biopsy for surgeons.

Clinically, lipoblastoma appears as a painless, slowly growing mass while histologically lipoblastomas tend to appear as hypocellular lobules of adipocytes in various stages of differentiation having lobular architecture with prominent fibrous septa.⁸ When tumour is unifocal, it is called lipoblastoma and multicentric ones are called lipoblastomatosis.⁹ Although they may arise almost anywhere within the soft tissue, the extremities and the trunk are the most common sites.^{10–12}

Most of the lipoblastomas had rearrangement of the 8q11~q13 region. This rearrangement targets the PLAG1 gene and has been reported in only 3% of conventional lipomas and never in myxoid liposarcoma.¹³ Cytogenetic analysis or fluorescent in situ hybridisation for the PLAG1 gene can provide useful differential diagnostic information.

In our case, the location was the left shoulder just above the acromioclavicular joint. Recurrence occurs within 6 months of surgery, confirming the fact that a high recurrence rate of 14–25% is reported despite surgical excision.^{9 14} This case also highlights the point that medical treatment in the form of corticosteroids has no role to play in managing such cases and that a regular and close follow-up is necessary for years together because of the high recurrence rate. Some lipoblastomas develop rapidly and become very large, a reminder of the fact that not all rapidly growing large tumours of the extremity are malignant.

Learning points

- ▶ Extremities and the trunk are the most common sites for lipoblastoma.
- ▶ A painless, slow growing mass is the most common presentation.
- ▶ Though benign, they can invade locally and grow rapidly.
- ▶ Complete surgical excision is the key treatment for lipoblastoma.
- ▶ Despite surgical excision, a recurrence rate of nearly 25% is reported within 6 months of surgery.
- ▶ A regular and close follow-up is necessary for years after surgery.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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