

Giant gluteal-pelvic lipoblastoma in a four-year-old girl. Report of a rare case

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Abstract— Lipoblastoma is a rare benign tumor, accounting for less than 1% of childhood neoplasms. It begins from embryonic fat cells, with approximately 80% of cases occurring before the age of 3 and 40% occurring before the age of 1. Herein, we present an extremely rare case of gluteal-pelvic lipoblastoma in a 3.6-year-old girl successfully treated by excision of the tumour, with no evidence of clinical recurrence 3 months postoperatively.

Keywords— giant lipoblastoma, diagnosis, surgical treatment, child.

1. Introduction

Adipose tumors are rare in childhood and are usually benign, such as lipomas, lipoblastomas, lipoblastomatoses, angiolipomas and myolipomas. Usually, they are localized in the subcutaneous tissue of the thorax (10-60%) and limbs (up to 40%), with the exception of giant ones with intraperitoneal or pelvic localization^{1, 5, 9, 11}. Because they are asymptomatic, they can reach significant sizes. Adipose tumors originate from embryonic fat cells and usually develop in children under the age of three. Locally aggressive adipose tumors are atypical lipomatous tumors and well-differentiated liposarcoma, while malignant tumors are represented by undifferentiated liposarcoma, myxoid liposarcoma, pleomorphic liposarcoma, and mixed types of liposarcoma⁶. Guanà et al.⁶ describe a successfully excised case of a lipoblastic tumor in the gluteal-pelvic region. Cascini et al.⁴ described three cases of this tumor with abdominal-pelvic localization.

2. Case report

A three-year-six-month-old girl born in a normal delivery was admitted to our clinic because soon after birth, the parents noticed asymmetry of the two gluteal regions with a slightly pronounced soft tissue formation on the right side (Figure 1). In the next three years, the tumour mass grew to the size of a large apple. Clinically, the formation had

a soft-elastic consistency, was painless, had a noninflammatory character and had non-enlarged regional lymph nodes. There was no pain or limited movement in the adjacent joints or functional disorders in the gastrointestinal and excretory systems. Imaging, including X-ray, ultrasound, and MRI (Figure 2), showed evidence of a giant lobulated lipoma, a large portion of which was located in the gluteal region with partial extension to the pelvis, and evidence of moderate compression of the bladder and rectum.

Operative technique. Surgical position as in obstetrics and gynaecology was used. With the approach along the course of the gluteal-femoral sulcus, the formation was reached layer by layer. It was found to be encapsulated and lobulated (Figure 3). A total excision of the tumour mass was performed. A drain and a compression dressing were applied. Histologically, the diagnosis was determined to be lipoblastoma. The operative wound was closed in a usual matter. Three months postoperatively, there was no evidence of local recurrence (Figure 4).



Figure 1. Preoperative photography

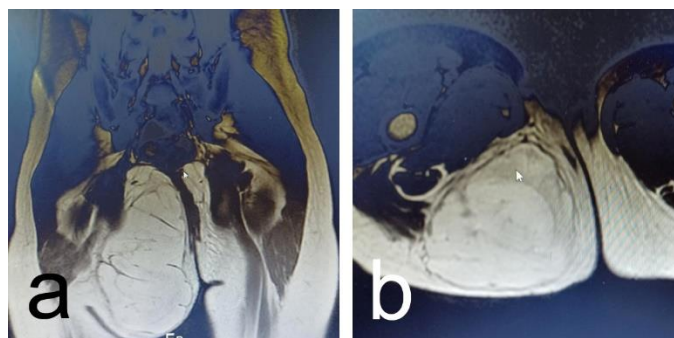


Figure 2. MRI preoperatively



Figure 3. Macroscopic view of the formation



Figure 4. Postoperative photography

3. Discussion

Lipoblastomas are benign soft-tissue tumors originating from embryonic fat cells that develop at an early age, predominantly affect males and are characterized by rapid growth. They are most often observed a month after birth and are found until 14 years of age (on average approximately 2 years after birth)⁷.

In 1926, Jaffe et al.⁸ described lipoblastoma for the first time as an atypical lipoma consisting of embryonic fat cells (lipoblasts). Two variants of lipoblastoma (well-demarcated encapsulated tumor) and lipoblastomatous (nonencapsulated, lobulated and infiltrative tumor) are described. Both variants are rare benign mesenchymal tumors. Lipoblastomas are more common in the head and neck region⁶.

MRI is the most valuable diagnostic method for determining the size of the tumor and distinguishing it from normal subcutaneous fat tissue. The tumor mass has high signal intensities and most often ranges from 20 to 25 cm^{3,11}.

The method of choice in the treatment is total excision with preservation of vital organs^{2-4, 6, 7, 9}.

Histologically, immature lipoblasts separated by thin fibrous septa and a peripheral pseudocapsule are demonstrated. In some cases of primary or recurrent lipoblastic tumors, puncture biopsy is appropriate³.

The long-term prognosis of lipoblastoma is good, and metastases are usually not observed. Potential recurrences after incomplete primary excision are possible in 9% to 22% of cases¹⁰.

In lipoblastomatosis, wide resection is recommended due to the higher recurrence rate².

Possible reported intraoperative complications associated with tumor resection are chronic bilateral venostasis and injury to the internal iliac artery and vein with subsequent reconstruction³.

A follow-up of at least five years after excision is recommended. Most authors report no recurrences in this period^{2-4, 6, 7, 9}.

4. Conclusion

Lipoblastoma is a rare benign soft tissue tumor with an extremely rare location in the pelvic region. The prognosis is good, with recurrences being rare with no risk of metastatic dissemination. However, continued ultrasound follow-up is recommended. Radical surgical excision is the gold standard in treatment and must be carefully performed to avoid potential recurrence.

5. References

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