



Atypical location of lipoblastoma in a 5-year-old girl

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ABSTRACT

We reported a perineal lipoblastoma in a 5-year-old girl located in the right labia mayor. The lesion gradually increased within 6 months. Ultrasound and magnetic resonance imaging (MRI) showed a limited solid heterogenous tumor with fatty component. After it had been surgically removed, the anatomopathological examination confirmed that it was a lipoblastoma.

Lipoblastoma is a rare benign mesenchymal tumor of infancy and early childhood. Symptoms vary depending on localization; signs of compression of adjacent organs may be seen. This type of unusual soft tissue tumors occurred most often in under 3 years old. The localization of lipoblastomas is predominantly in the extremities but may be also found in other sites including the head and neck, trunk, mediastinum, kidney, mesentery, retroperitoneum and perineum. The suspicion should be considered according to ultrasound and MRI findings.

Keywords: *perineum; lipoblastoma; magnetic resonance imaging; child.*

doi: <http://dx.doi.org/10.5546/aap.2022-02853>

To cite: Bi Z, Dong P, Sun C, Shi Y, et al. Atypical location of lipoblastoma in a 5-year-old girl. *Arch Argent Pediatr* 2023;121(6):e202202853.

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Funding: This work was funded by "Taishan Scholar" Project (NO.tsqn202103197) and a Shandong Province Natural Science Foundation grant [ZR2022MH274].

Zhaojun Bi and Peng Dong contributed equally to this work.

Conflict of Interest: None.

Received: 9-13-2022

Accepted: 2-7-2023



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INTRODUCTION

Lipoblastomas are rare benign soft tissue tumors that occur in infants and children.¹ The vast majority of this tumors are found under the age of 3. Although lipoblastomas are benign, they rapidly enlarge enough to be discovered or cause symptoms. The most common symptom is a painless mass.² Magnetic resonance imaging (MRI) is an important tool for determining the anatomical extent, tissue involvement, and composition of tumors and designing a treatment strategy. The best surgical treatment is a complete resection including the edge of the tumor and long-term follow-up is recommended in case of recurrence.³

Lipoblastomas are frequently discovered as a soft tissue lesion of extremities, which account for 70% of total cases, and lipoblastomas of the lower limbs are more common than those of the upper limbs.³ They can also appear in deep soft tissues, such as the peritoneum, chest wall, oral cavity, mesentery, trunk, and neck.⁴ Perineum is a rare location of lipoblastomas with only 7 cases

been reported in the literature.

Case report

A 5-year-old girl presented with a peanut-sized “mass” in her right labia majora, there was no history of trauma, pain or discharge. The painless “mass” had gradually increased to approximately 5 cm in diameter in 6 months. No treatment was performed. Physical examination revealed a 5 cm × 2 cm × 3 cm cystic mass with high tension and acceptable mobility. Magnetic resonance imaging (MRI) of pelvis revealed a solid mass (5.3 cm × 2.6 cm × 3.3 cm) with fatty component and heterogeneous hyperintensity signal on axial T2 (*Figure 1A*) and T1 (*Figure 1B*). and reduced signal at fat suppression sequences demonstrating the fatty component of the lesion (*Figure 2*). The Doppler sonography showed little blood supply in the lesion (*Figure 3*). The tumor was completely removed by surgery. The pathologist confirmed that the mass was a lipoblastoma (*Figure 4*).

FIGURE 1 A-B. Large mass with HETEROGENEUS signal intensity on T2 AND T1 weighted imaging in the right labia majora

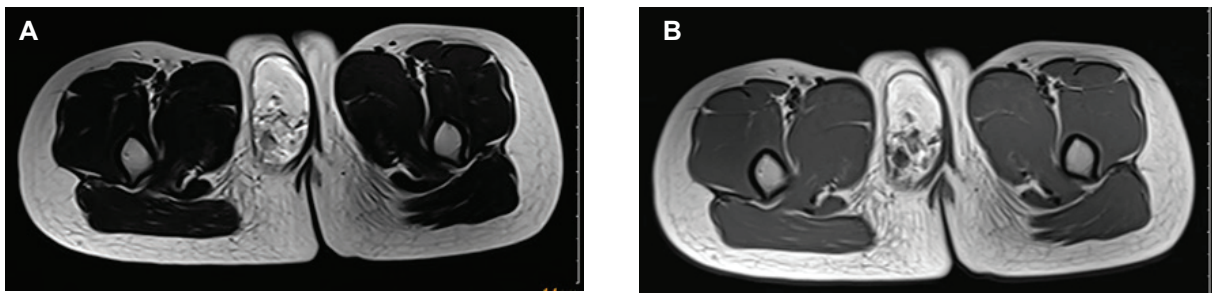
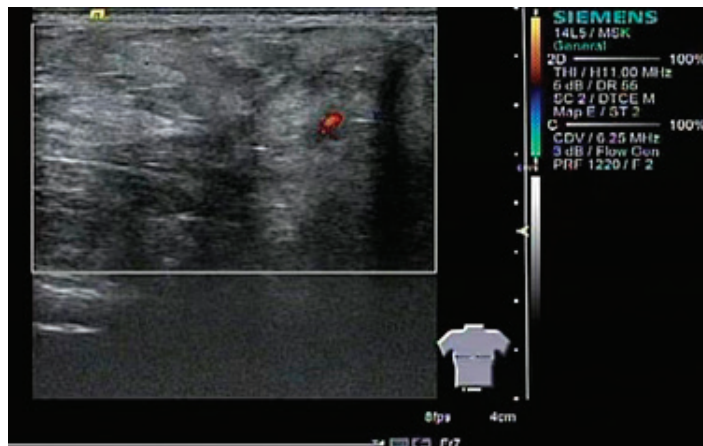
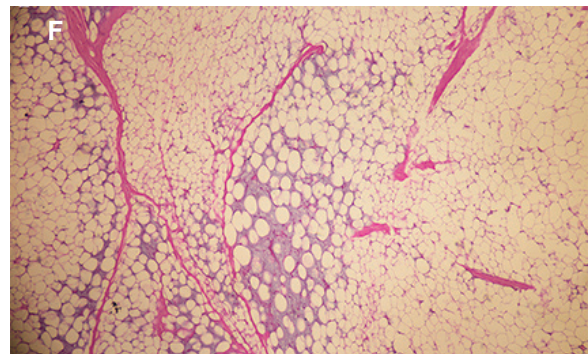
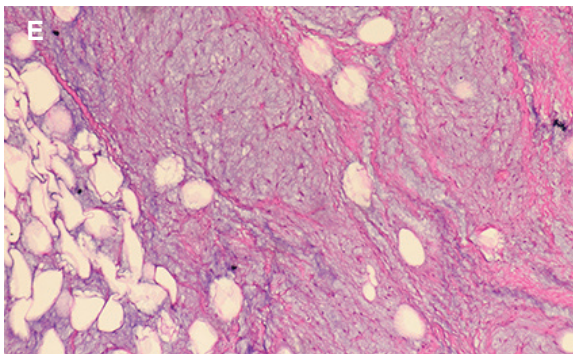


FIGURE 2. Fat-suppression imaging demonstrating reduced-signal intensity



FIGURE 3. Doppler sonography showing little blood supply in the lesion**FIGURE 4. Histological analysis shows adipoblastas (HE×100 and HE×10)**

DISCUSSION

In the present study we report a lipoblastoma on the right labia majora. The ultrasonic image shows that the echo is located in the fat layer and that the boundary is unclear. The interior is slightly hyperechoic and shows fiber strip separation. Doppler sonography shows little blood supply in the lesion. The information provided by ultrasound image allows us to understand the internal structure of lipoblastoma. Lipoblastoma can be divided into two pathologic types:⁵ local and diffuse. Localized and diffuse lipoblastoma differ in color Doppler ultrasound findings. The blood flow classification of localized lipoblastomas⁶ is mainly grade 0 or I, but the blood flow shunt of diffuse lipoblastoma is usually grade II or III. In short, the latter has more abundant blood flow, which explains why it is more prone to malignant transformation than the former.

Radiologically speaking, MRI multiple sequence imaging plays an indispensable role

in the location and characterization of tumor. MRI can definitely identify fat within a tumor. Especially fat-suppression techniques⁵ are valuable in the evaluation of lipoblastomas because even standard T1 and T2 cannot show the presence of adipose tissue in the tumor.

To understand lipoblastomas better, we need to be familiar with myxoid liposarcomas, which are similar to them. On the one hand, lipoblastomas commonly occur in infants and young children, whereas myxoid liposarcomas are common among adults. On the other hand, compared with those of lipoblastomas, the lobular structure of myxoid liposarcoma is not obvious, their fibrous septa are incomplete, and their mature adipocytes are concentrated in the lobular margin.⁷

Lipoblastomas are benign tumors unlikely to become malignant and distant metastases have never been reported. Depending on the location, lipoblastomas may oppress surrounding nerve and organs.⁸ Thus, total resection is the

treatment of choice with preservation of adjacent structures. Lipoblastomas have an excellent prognosis. However, some cases may recur, with a recurrence rate of approximately 14%-25%;⁹ therefore, ultrasound and MRI are the currently recommended for follow-up, especially in cases of incomplete excision or previous recurrence.

CONCLUSION

To date, lipoblastomas have rarely been described in the labia majora. In the present case, ultrasound and MRI revealed a regular mass with fat content in the right labia majora. Total resection is the optimum choice for diagnosis confirmation and treatment. In cases of incomplete excision or local recrudescence long-term observation must be performed to detect recurrence. ■

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