World Journal of Clinical and Medical Case Reports

Case Report
Published: 21 May, 2024

Clinicopathological Analysis of a Case of Perineal Lipoblastoma in an Infant

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Abstract

Background: Lipoblastoma is a benign tumor originating from embryonic white adipose tissue, which occurs mostly in the limbs and trunk, and perineal lipoblastoma is clinically rare.

Case Report: We report a case of perineal lipoblastoma in a child and review the relevant literature.

Conclusion: We have analyzed the clinical manifestations and pathological features of this disease and the conclusions of this report will be of high value to clinicians.

Keywords: Infant; Perineal lipoblastoma; Tumor; Clinical treatment; Pathological characteristics

Background

Lipoblastoma is a rare benign tumor that originates from embryonic white adipose tissue. It primarily occurs in children under the age of 3 and is most commonly found in the limbs and trunk. However, it can also be found in other areas of the body such as the head and neck, heart, intra-abdominal parenchyma, mesentery, and perineum. The initial symptom is typically a painless mass that gradually increases in size or causes clinical symptoms due to compression of surrounding organs and tissues. Complete resection of the tumor is the preferred treatment option due to its tendency to recur.

Case Presentation

The child is a male, age 19 months, parents unintentionally found a mass in the perineum behind the left testicle, about the size of yolk, without pain, the mass gradually increased in recent time. On examination, there was a localized bulge behind the left testicle, the height of the bulge was about 1.0 cm, the skin texture was normal, there was no redness, no swelling, no ulceration, no hair growth on the surface, the skin temperature was normal, and the soft mass could be palpated, the size of it was about 4.0 cm × 2.5 cm × 1.5 cm, the border was clear, the surface was smooth, there was no tenderness on palpation and it was still movable, there was no adherence to the surrounding tissues, and there was no abnormality of the opposite side of the testicle. Preoperative ultrasound (Figure 1) showed that the surface of both testes was smooth and echogenic, and the left testis was isoechoic in the perineum, with a size of about 4.3 cm × 2.7 cm × 1.9 cm, clear boundary, regular morphology, and deformation was visible on the probe, and there was no obvious blood flow signal. Sixty-four (64)-row spiral CT scan showed that the perineum could be seen in the form of a nodular soft-tissue density shadow. After completing the relevant examinations, surgery was performed, and a curved incision was made at the lower edge of the left scrotum. During the operation, it was seen that the mass was located between the spongy body of the urethra and the rectum, with no adhesion to the surrounding tissues, and the tumor was yellowish and fat-like, with the peritoneum intact. Intraoperatively, the tumor was completely excised (Figure 2), its morphology was irregular, and it was sent for pathological examination. It was diagnosed as lipoblastoma by the Department of Pathology of our hospital and the Department of Pathology of the First Affiliated Hospital of Nanjing Medical University, a remote consultation and quality control platform, and the pathology showed the following: Yellowish nodule seen by the naked eye, V: 4.1 cm × 3 cm × 1.5 cm, with peripheral membrane on the surface, slightly foliated, with yellowish solid texture and soft and greasy sensation on the cut surface, and the local foci of greyish-yellowish semi-transparent-like area, measuring 1.7 cm × 1.2 cm, and the remaining nodules with scattered semi-transparent-like areas, with diameters of: 1.0 cm to 2.0 cm (Figures 1-3).

OPEN ACCESS

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Received Date: *15 Apr 2024*
Accepted Date: *16 May 2024*
Published Date: *21 May 2024*

Citation:
Youbo X, Junzhao Z, Runpeng Y, Yinan Z, Lexiang S. Clinicopathological Analysis of a Case of Perineal Lipoblastoma in an Infant. World J Clin Med Case Rep. 2024; 2(1): 1010.

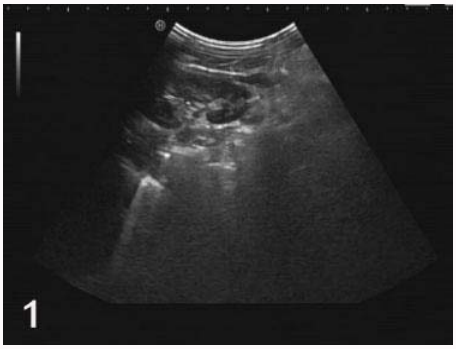

Figure 1: Preoperative ultrasound results.

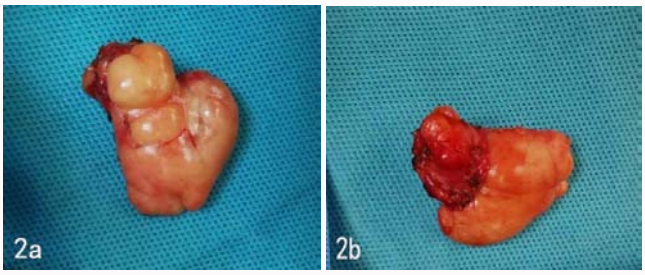

Figure 2a, 2b: Postoperative resection of the tumor.

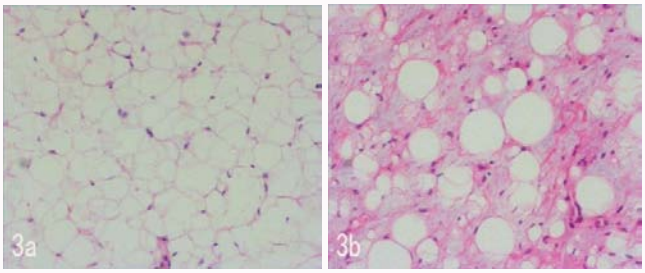

Figure 3a, 3b: Postoperative pathological results.

Discussion

Lipoblastoma is a tumor originating from embryonic white adipose tissue, the term lipoblastoma was first used by Jaffe who described it as a tumor of immature adipocytes. In 1958 VELLIOS used the term lipoblastomatosis to describe his discovery of a benign tumor resembling adipose tissue [1]. Benign diffuse tumor [1]. Chung and Enzinger later classified the tumor into two types, limited and diffuse; the limited tumors, called lipoblastomas, tend to be located in superficial areas of the body, are soft or tough, and have good mobility, whereas the diffuse tumors, called lipoblastomatosis, tend to be located in deeper areas of the body, are hard, may infiltrate and grow into the surrounding tissue, and have poor mobility [2]. Lipoblastoma is a rare benign tumor and there are no reports of tumor metastasis or malignancy. It occurs predominantly in children up to three years of age and is usually detected before the age of ten years, but a few cases have been found in older children and even in adults, with variable rates of growth, with both faster and slower growth reported [3,4]. The incidence is higher in males than in females, with a male-to-female ratio of 1.5-2.8:1 [3,5]. According to previous literature, the most common sites are the limbs and trunk, and a few cases have been reported in the mediastinum, head and neck, perineum, heart, parenchyma, mesentery, and other parts of the body. The trunk is the most common site, followed by the limbs, with relatively few cases in the remaining sites [6-9]. The length and diameter of the tumor is mostly between 3 cm and 5 cm, and masses located on the surface of the body are easier to be detected, while masses located in deeper parts of the body are mostly detected due to compression of the surrounding tissues or organs causing clinical symptoms or increasing in size to a certain extent that can be seen as a mass on the surface of the body. In 2019, Fanna reported the case of an abdominal mass in a 5-year-old boy, which was found to be a lipoblastoma located in the liver, with the size of 16.7 cm × 12 cm × 8.5 cm [10].

This case reports a case of lipoblastoma located in the perineum, which has been found to be relatively rare in China and abroad. Tumors in the perineum can exert pressure on the urethra, rectum, testes and epididymis, and accurate preoperative diagnosis as well as complete resection of the tumor to minimize possible pressure on the surrounding tissues and testicular development are the main focuses in the management of tumors in the perineum. Imaging tests such as ultrasound, CT or MRI are mostly applied for the diagnosis of tumors in children, and invasive tests are less frequently used in children. The ultrasonographic manifestations of superficial site lipoblastoma are mainly clear boundary, regular morphology, and internal non-uniform slightly high echogenicity, while cases in deeper site can be seen as weak echogenicity or heterogeneous echogenicity, and about 60% of cases can be seen as striated strong echogenicity segregation, which is the manifestation of intratumorally fibrous bands, while lipoblastomatosis is manifested as slightly high echogenicity with irregular boundary and morphology, and the rest of them are in line with lipoblastomatosis, and the case reported in our operation. The preoperative ultrasound reported in this case showed an isoechoic pattern, which is not consistent with the literature [11]. Ultrasonography has a major advantage in the diagnosis of tissues located superficially in the body. CT of intra-abdominal lipoblastoma showed an inhomogeneous density shadow with CT values of -121 Hu to -20 Hu, which was higher than the density of subcutaneous fat at the same level (-140 Hu to -90 Hu) [12]. MRI of lipoblastoma shows a lobulated fat mass with high signal in T1WI and low signal in T2WI, which can be accompanied by different degrees of non-enhancing cystic changes, with fibrous septa of varying thickness, and the presence of nodular and flocculent soft-tissue signal shadows. Meanwhile, MRI has the advantage of better soft-tissue resolution compared with CT and is radiation-free, so that it can be used for better observation of the characteristics of the tumor and its invasion of the surrounding normal tissues or organs, and is useful for preoperative surgery. It can better observe the characteristics of the tumor, the invasion of the surrounding normal tissues or organs, and has a good effect on the preoperative diagnosis, the formulation of surgical plan and the postoperative follow-up [12,13]. MRI is more advantageous for deeper sites and tumors in the abdominal and thoracic cavities. The only treatment for this tumor is surgical resection, and complete removal of the tumor is an effective way to avoid recurrence, which is almost always due to positive margins in recurrent cases. Because lipoblastoma has an envelope and limited growth, complete excision of the tumor is usually possible with a low recurrence rate, whereas lipoblastomatosis often infiltrates into adjacent tissues and complete excision of the tumor is not possible, resulting in a higher recurrence rate in lipoblastomatosis than in lipoblastoma. According to a cohort study conducted by Dao on

329 patients with follow-up information, the recurrence rate of a complete excision of the tumor was found to be 8.8%, and the recurrence rate of an incomplete resection had a recurrence rate of 60.7% [14]. It has been suggested that an appropriately wide resection can be done with protection of surrounding vital structures [15,16]. It is a benign tumor and so far, no metastasis or malignancy has been reported and there is a possibility of spontaneous regression and transformation into a mature lipoma [17]. Therefore, some researchers do not recommend extended resection to avoid damage to the surrounding tissues or organs, and staged resection can be adopted to treat lipoblastomatosis, and some tumors with deep infiltration into the surrounding tissues have a high recurrence rate even if the resection is extended, but not completely resected, and can also cause collateral damage to the surrounding tissues or organs [18]. Regardless of how the resection range is selected, protection of the surrounding normal organs and functions is the first priority, and the specific resection range can be considered according to the site of the tumor and the degree of infiltration. Due to its recurrence, it is currently recommended that the follow-up time should be at least five years, every six months to one year, using imaging examinations such as ultrasound or MRI, and tumors located superficially can be used to provide health education to their parents to determine whether there is any recurrence by touching the site of the disease in normal times [18,19]. Recurrent tumors can be treated by re-surgical resection.

Lipoblastoma appears nodular to the naked eye, microscopically it is composed of mature adipocytes and immature lipoblasts at various stages of differentiation and primitive round and spindle shaped mesenchymal cells in a mucinous background, separated into lobules by fibrous bands, the mucinous stroma and fibrous bands contain tufts of capillaries [20]. Lipoblastoma needs to be differentiated from mucinous liposarcoma, firstly by the difference in the age of the disease, mucinous liposarcoma tends to occur in older populations, and secondly microscopically, the mature fat cells in mucinous liposarcoma tend to occur peripherally, whereas in lipoblastoma they tend to be located in the center of the lobules [21]. In addition, mucinous liposarcoma shows poor lobulation, atypical nuclei and abnormal mitoses [22]. Cytogenetically, lipoblastoma was found to have a chromosome 8 abnormality, a rearrangement of q11 to q13 on chromosome 8 resulting in a transcriptional upregulation of the gene carrying PLAG1 (Pleomorphic Adenoma Gene 1) in the child, which could be detected by Fluorescence *in Situ* Hybridization (FISH). Liposarcoma, on the other hand, exhibits t(12;16)(q13;p11) leading to a (CHOP) gene fusion [19]. Additionally, lipoblastoma shows CD34 (+) as well as S-100 (+) by immunohistochemical examination [6].

Conclusion

Lipoblastoma is an extremely rare condition that is poorly understood by surgeons and carries a risk of tumor recurrence or overtreatment if misjudged during clinical practice. In younger children, lipoblastoma or lipoblastosis should be considered in the presence of a painless mass or a diffusely growing mass at a deeper site, and when ultrasound demonstrates internal heterogeneity of slightly elevated echoes with strongly echogenic striated septa or MRI suggests lipoid tissue with fibrous septa. Preoperative diagnosis is based on ultrasound or MRI, which provides the clinician with a preliminary assessment of the beingness or malignancy of the disease, and pathological and cytogenetic examinations can accurately draw conclusions, with the pathological diagnosis as the gold standard. For tumors with intact envelope, they can be sent for pathological diagnosis after complete resection. For tumors with incomplete or no envelope, intraoperative frozen pathological diagnosis is feasible to decide whether to adjust the scope of resection. Lipoblastoma is treated by complete excision of the mass, while lipoblastomatosis is treated according to the location of the tumor, infiltration of surrounding tissues or organs in order to decide on the surgical plan, and to try to treat the disease without causing collateral damage.

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