Liposarcoma of the spermatic cord in the child: A case report

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Abstract

Intoduction : Liposarcomas account for 7% of spermatic cord tumors. These are rare tumors that are most commonly found in adults. Tumors located in the spermatic cord are extremely rare in children. They account for only 0.7% of liposarcomas.

Case presentation: A 4-year-old boy who presented with a left scrotal mass that had been present for one month. A biopsy and histopathological examination revealed a low-grade myxoid liposarcoma. Six months after the biopsy, a local examination of the left scrotum revealed a large scrotal mass with ulceration at its apex and in the left inguinal region. Ultrasound of the mass revealed a large tissue mass measuring 150 mm × 12 mm with necrotic-hemorrhagic changes occupying nearly the entire left scrotum, with the left testicle not visualized. Staging tests revealed calcification of the abdominal aorta. Given the delayed diagnosis and the advanced stage of the disease, the patient was placed in palliative care, and the outcome was unfavorable, resulting in the patient's death.

Conclusion: The liposarcoma of spermatic cord is a rare tumor in children. Diagnosis is difficult due to its similarity to a persistent peritoneovaginal duct. When diagnosis is delayed, the prognosis is poor.

Keywords

- Liposarcoma
- spermatic cord

Introduction

Liposarcoma is a rare and unusual malignant mesenchymal tumor of the spermatic cord, accounting for 7% of such tumors. [1] Only 200 cases have been reported worldwide across all age groups, with a peak incidence between the ages of 40 and 60 [2,3]. The occurrence of liposarcoma in children is rare and accounts for 0.7% of all liposarcomas [4]. The prognosis is

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most often poor, characterized by a high risk of recurrence. Due to its rarity, few large series have been reported in the literature regarding management and prognostic factors. To our knowledge, this is the second reported case of liposarcoma of spermatic cord in the child after the one described by Romero Tenorio M et al [5].

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Case presentation

A 4-year-old boy was referred to us by a hospital in the suburbs of Abidjan for a left scrotal mass. The condition reportedly began one month prior with the appearance of a painless left scrotal mass that did not pulsate when the child cried, measuring 3 cm in diameter. The child was examined by a general surgeon. A diagnosis of a left spermatic cord cyst was made. The decision was made to remove the cyst. Intraoperatively, the mass was found to be poorly vascularized, arising from the left spermatic cord with areas of necrosis. The mass was excised. The surgical specimen was not submitted for pathological examination. Three months later, the left scrotal mass recurred, leading to the patient's admission to our pediatric surgery department. On physical examination, the patient was in good general condition, weighing 18 kg, with a temperature of 37°C, a heart rate of 109 beats per minute, oxygen saturation at room air of 99%, and a respiratory rate of 22 breaths per minute. Locally, a large, firm, irregular, painless, non-mobile left inguinal-scrotal mass measuring 15 cm × 10 cm was noted, with an irregular surface and collateral venous circulation (Figure 1a). The left testicle was not palpable. Scrotal transillumination was negative. There were no inguinal or axillary lymphadenopathies. Examination of the other systems revealed no abnormalities. Scrotal ultrasound revealed a large tissue mass measuring 150 mm × 12 mm with necrotic-hemorrhagic changes occupying almost the entire left scrotum, with the left testicle not visualized. (Figure 2). The right testicle was normal. Alpha-fetoprotein (AFP) and human chorionic gonadotropin (beta-hCG) levels were normal. One month later, while awaiting the results of the histopathological examination and due to financial difficulties in performing the staging workup, the course of the disease was marked by rupture of the scrotal mass at its base (Figure 1b). The results of the histology, immunohistochemistry, and staging evaluation were available 3 weeks after the rupture. The pathological examination revealed a low-grade myxoid liposarcoma (Figure 3a, b, c). The staging workup (thoracoabdominal-pelvic CT scan)

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revealed calcification of the abdominal aorta (Figure 4). The patient was placed on palliative care due to tumor spread to the abdomen and pelvis and tumor rupture. The outcome was unfavorable, and the patient died.

Discussion

Spermatic cord liposarcoma (SCL) is a rare malignant tumor in children [1]. The tumor may result from the malignant transformation of a lipoma or develop directly from the adipose tissue of the spermatic cord [1]. Macroscopically, the lesion is often large, multilobed, yellowish, firm or elastic, with areas of hemorrhagic necrosis and cystic degeneration [1]. These lesions were identified during surgery by the general surgeon in our case.

The diagnosis is histological and confirmed by immunohistochemistry [3]. The most commonly used histological classification is that of the WHO, which describes four categories based on the predominance of a cell type: well-differentiated, myxoid, pleomorphic, and round-cell. This classification has prognostic value, as well-differentiated and myxoid forms tend to remain localized and have a favorable prognosis, whereas round-cell and pleomorphic forms appear more aggressive, with a higher risk of local recurrence and a greater potential for metastasis to the lungs and bones [1; 6, 2]. In our case, a myxoid SCL was identified. In our patient, it presented as a firm, irregular, painless mass encompassing the testicle. According to Lachkar et al., the mass may be located away from the testicle, either within the scrotum or in the groin, or it may encompass both [1].

It often develops insidiously and is therefore frequently mistaken for spermatic cord cysts or inguinal hernias [1,7,8, 9,10].

This was the case in our observation, where the swelling was mistaken for a spermatic cord cyst. It may also be an incidental finding during surgery for hernia repair. It is in this latter scenario that the risk of tumor dissemination is highest. [10, 8] In our case, this surgical procedure led to a flare-up of the spermatic cord tumor. The first imaging test to perform is a scrotal ultrasound. It will reveal a heterogeneous, hyper vascular mass with hyperechoic areas. Although ultrasound can provide information on the size and consistency of the mass, there

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are no pathognomonic ultrasound features that allow for the differentiation of benign from malignant lesions, so a second-level imaging study is often necessary. Some authors recommend MRI as the first-line imaging modality, as it allows for the determination of the nature of the mass, its size, its topography, its anatomical relationships, and the exclusion of differential diagnoses [10]. In our case, computed tomography (CT) was performed as a last resort to determine the extent of the tumor. CT can provide the same information as MRI but involves radiation exposure [2].

As a general rule, surgical resection of the tumor is the primary treatment method, but it is not always possible, and sometimes, even when performed, it proves insufficient due to local recurrence. The indication for potential adjuvant therapy depends on the histological grade, degree of infiltration, and the presence of metastases.[6, 10]

The prognosis for SCL depends on the histological type as well as certain factors described by Lachkar et al.[1] Although the myxoid type has a favorable prognosis, when the tumor is 5 cm in size, accompanied by infiltration and an embolus in the abdominal aorta, the prognosis becomes poor. This was the case with our patient. Apart from these factors, the 5-year survival rate for well-differentiated SCL is approximately 85%, 77% for the myxoid subtype, and 20% for the other subtypes [3].

Conclusion

SCL is a rare and exceptional tumor in children. The diagnosis is often made retrospectively following treatment for an inguinal hernia. Any clinical suspicion should therefore be evaluated with imaging and percutaneous biopsy to obtain an accurate preoperative diagnosis.

Ethical Consideration

written informed consent was obtained from the parents for publication of this case report and any accompanying images.

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Not applicable

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Conflict of interests

There is no conflict of interest

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