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Case report

A rare case of inflammatory myofibroblastic tumor of the vulva in a newborn ☆,☆☆

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ABSTRACT

Inflammatory myofibroblastic tumor (IMT) is an uncommon neoplasm that rarely arises in the genitourinary system. IMTs in the vulva in infants are extremely rare in the literature. The tumor consists of myofibroblastic spindle cells accompanied by inflammatory cell infiltration. In this article, we aimed to describe the case of IMT in the vulva. A newborn girl presented with a mass in the vulva detected in the prenatal period. The patient was treated with surgery and chemotherapy. Follow-up 8 months after surgery showed no signs of recurrence. In conclusion, IMT has a variable clinical presentation, surgery is the optimal approach, but in cases without complete resection, chemotherapy is essential.

Introduction

Inflammatory myofibroblastic tumor (IMT) is a rare mesenchymal tumor with unknown etiology [1]. The lungs, liver, and gastrointestinal tract are the most common sites for IMT; however, in rare cases, IMT can affect the genitourinary tract [1]. In the genitourinary tract, the kidney, urethra, prostate, ureter, and testis may be affected, but the bladder is the most frequently affected site [2]. IMT tends to present in children and young adults [1]. IMT can become malignant, and metastasis presents with an incidence of less than 5% [3]. The final diagnosis is typically based on histopathological findings. Here, we intended to report a rare case of IMT of the vulva to share the experience of diagnosis and treatment.

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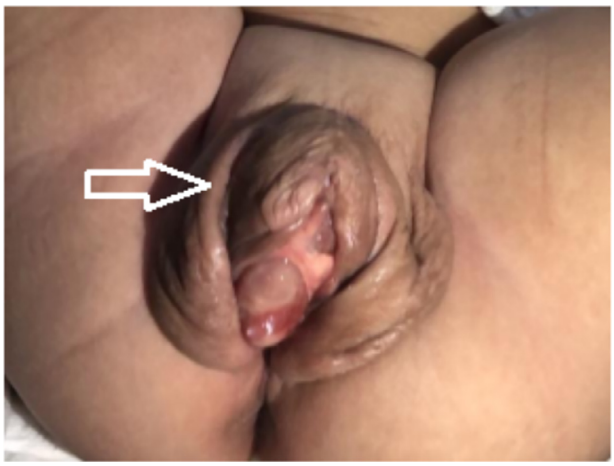

Fig. 1 – There was a mass in the vulva (arrow).

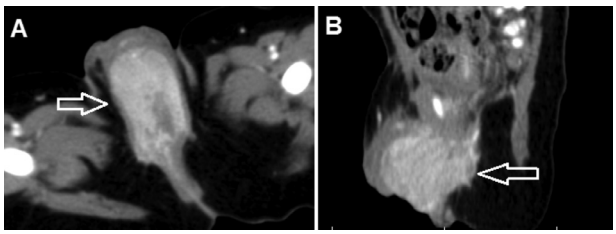

Fig. 2 – Axial (A) and sagital (B) CT scanner of the pelvis. This mass was well-circumscribed and hyperenhancing (arrows).

Case report

A newborn girl was hospitalized with a mass located in the vulva, which was diagnosed in the prenatal period (Fig. 1). Physical examination showed the soft mass with difficult movement in the vulva. Defecation and urination were normal. Her growth and development were normal. This patient had been suspected of congenital genitourinary abnormalities since the third trimester and perinatal period. No abnormality was detected in her family history. Ultrasound revealed a mass located in the vulva, which was hypoechoic and homogeneous, with well-defined borders, the uterus and ovaries were normal. Pelvic computed tomography (CT) scans showed a mass with well-defined margins, measuring 2.5 × 4 cm, with hyperenhancement, located in the vulva (Fig. 2). This patient underwent surgery at the age of 4-month; however, the surgeons were unable to remove the tumor completely. Postoperative histopathological results showed that the tumor tissue consisted of spindle cell proliferation, composed of fibroblasts with angiogenesis, and acute inflammatory cells, eosinophils, and lymphocytes in a collagenous stroma (Fig. 3). Immunohistochemistry results revealed that the tumor cells were negative for CD31, actin, myogenin, and desmin, and the Ki-67 index was 10%. No rearrangement of the anaplastic lymphoma kinase (*ALK*) gene was detected by fluorescent *in situ* hybridization (Fig. 3). Due to incomplete tumor removal, the patient was treated with 6 cycles of methotrexate and vincristine, with complete response. After 8 months of treatment, the patient remains stable with no recurrence.

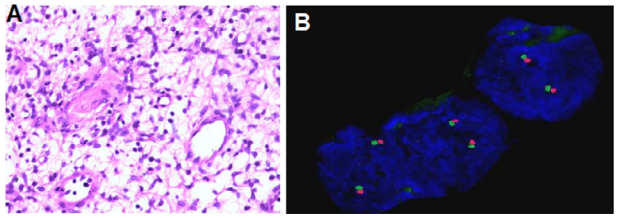

Fig. 3 – (A, ×200) Hematoxylin and eosin staining sample showed spindle cell proliferation, composed of fibroblasts with angiogenesis and inflammatory cells in a collagenous stroma. (B) No rearrangement of the *ALK* gene was detected by fluorescence *in situ* hybridization (FISH)

Discussion

IMTs are extremely rare neoplasms with malignant potential due to a low rate of metastasis [3]. The etiology and pathogenesis of IMT remain unknown; however, Epstein-Barr virus and human herpes virus-8 have been associated with IMT occurrence [4]. IMTs in the genitourinary tract are uncommon, and the case of IMT in the newborn vulva reported here represents the first such reported case in the literature.

On imaging, IMTs located in the urinary system are similar to those found in other locations [5]. On CT scans, the tumors present as variable and nonspecific. They can be low-density, isodensity, or high-density, with homogeneous or heterogeneous enhancement [5]. On magnetic resonance imaging, the tumors appear homogeneous, isointense, or slightly hypointense on T1-weighted imaging, hyperintense on T2-weighted imaging, and homogeneous or heterogeneous enhancement can be observed after contrast injection [6].

Microscopically, IMTs are characterized by a mixture of acute and chronic inflammatory cells and myofibroblastic spindle cells in a collagenous stroma [7]. The immunohistochemical staining of IMTs can be variable, and the tumor cells can be positive for smooth muscle actin and vimentin and negative for CD34, S100 protein, CD15, and CD3 [1].

In approximately 50% of cases, IMTs are associated with the rearrangement of the *ALK* gene, located on chromosome 2p23 [8]. ALK expression correlates with *ALK* rearrangement, and distant metastases of IMTs occur more frequently in tumors associated with *ALK* rearrangement than in ALK-negative tumors; however, local recurrence is not significantly correlated with ALK status [9]. Patients with *ALK* gene rearrangement can benefit from targeted therapy [10]. The differential diagnoses for IMT on histopathology include pseudo lymphomas, malignant lymphomas, fibrous tumor, fibrosarcoma, and leiomyosarcoma, rhabdomyosarcoma [3,11]. However, some tumors had several individual features, such as rhabdomyosarcoma has a multiseptated cystic appearance. Furthermore, imaging signs such as CT or magnetic resonance imaging can provide features of malignant tumors such as

invasive, distant, or lymph nodes metastasis which were extremely rare in IMT.

The recommended treatment of IMT depends on the location, size, and extent of the disease [11]. IMTs can be treated using various treatment modalities. Although surgical excision is the recommended first-line therapy, other treatment options include radiotherapy, chemotherapy, ALK-targeted therapeutic drugs [1,3]. Generally, IMT has a good prognosis, but most of the tumors cannot be removed completely, which can result in recurrence or metastasis [6]. Due to the rarity of IMT, no standard chemotherapy treatment regimens have been established for this disease [12]. Patients may be treated with vinorelbine, methotrexate, vincristine, cyclophosphamide, doxorubicin, 5-fluorouracil, cisplatin, carboplatin, paclitaxel, ifosfamide, or etoposide [12].

For the case reported here, the initial diagnosis after birth was congenital genitourinary abnormalities; however, the CT scans suggested the presence of a tumor. Due to incomplete tumor resection, this patient received adjuvant chemotherapy, including methotrexate and vincristine. After 8 months, the patient responded to treatment and was discharged from the hospital.

Conclusion

This case describes a rare IMT located in the vulva of the genitourinary tract. Due to the rarity of this presentation, genitourinary IMTs can be misdiagnosed as congenital genitourinary abnormalities. A definitive diagnosis should be based on histopathological results. In the case of incomplete excision, chemotherapy should be considered as an adjuvant treatment.

Patient consent

Informed consent for patient information to be published in this article was obtained.

Ethical statement

Appropriate written informed consent was obtained for publication of this case report and accompanying images.

Author contributions

Bui-Van L and Nguyen MD contributed equally to this article as co-first authors. All authors have read the manuscript and agree to the contents.

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