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Case Report

Paratesticular Ewing's sarcoma ☆,☆☆,★,★★

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ABSTRACT

This case report follows a 23-year-old man who presented with a painful right scrotal mass which was found to be a paratesticular vascular solid mass on ultrasound, and after uncomplicated orchiectomy, was revealed to be a high-grade extraskeletal Ewing's sarcoma. Diagnosis leading up to the orchiectomy was primarily clinical with only ultrasound used in identification and characterization of the paratesticular mass. Paratesticular masses are more commonly benign, and ultrasound is the first modality, with computed tomography and magnetic resonance imaging providing more definitive findings. We discuss imaging findings and histopathology of this rare tumor with an uncommon presentation.

Introduction

Paratesticular Ewing's sarcoma is a rare presentation of extraskeletal Ewing's sarcoma, an aggressive, highly recurrent malignancy. Among testicular masses, paratesticular tumors globally make up less than 5%-7% of tumors and can originate from the spermatic cord, epididymis, vestigial remnants, and tunica vaginalis [1]. Ultrasound is the imaging modality of choice in the initial radiological evaluation of a testicular mass and can help differentiate between intra- and extratesticular origin [2,3]. Further imaging with computed tomography (CT) and magnetic resonance (MR) imaging may help characterize the tumor by providing information on architecture, fat content, and clarifying origin as primarily to local tissues or extension from the abdomen [2]. With only a handful of similar cases discussed in the literature, we aim to provide a discussion on the presentation, imaging, and histological findings of this rare malignancy.

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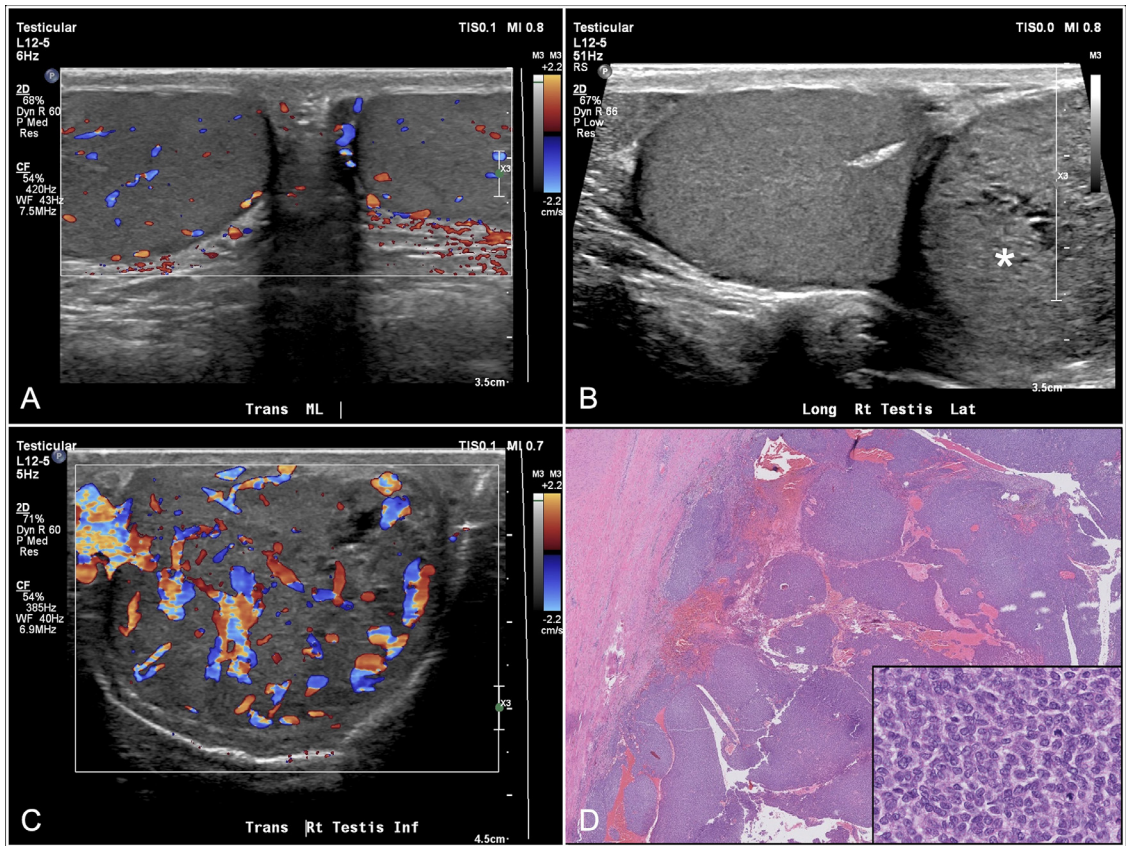

Fig. 1 – This figure shows multiple ultrasound and histopathology images. Gray-scale axial view (A) of both testicles (with color Doppler) shows uniform echogenicity and symmetric vascularity with no focal lesions. Panel B shows a long axis view of the right testicle and a mass inferior to it and separate from it (asterisk). The mass is oval, well defined, heterogenous, and hypervascular, as seen on image C. D shows representative section of the tumor confined to the tunica vaginalis and exhibiting small round blue cell cytomorphology (10×, inlay 200×; H&E).

Case report

A 23-year-old male with no significant past medical history initially presented to a urology clinic for a painful right-sided testicular mass. The patient first noticed the mass after removal of 2 penile implants 3 months prior. Physical examination showed well-healed incisions from implant removal, bilateral atrophic descended testes, and a large, approximately 2 inch soft, tender, mobile mass in the right testicle. Ultrasound at this visit revealed a 4 cm extratesticular solid vascular mass (Fig. 1).

Patient underwent uncomplicated right orchiectomy for right paratesticular mass, 13 months after his initial presentation. The pathology report showed a stage IIIa extraskeletal Ewing's sarcoma involving the parietal and visceral layers of the tunica vaginalis. Whole body bone scan showed no evidence of osteoblastic metastasis. Due to the high-grade nature of the tumor, the patient was started on adjuvant chemotherapy with vincristine/doxorubicin/cyclophosphamide and ifosfamide/etoposide. Follow-up CT of the abdomen and pelvis after surgery was negative for metastatic disease.

Discussion

Ewing's sarcoma is a rare malignant tumor that usually presents in the diaphysis of long bones, most commonly in the pediatric population, with an occurrence of approximately 300 new cases per year [4]. Although most cases of Ewing's sarcoma present in the bone, 15%-20% of cases have also been found to originate in soft tissue, with this presentation known as extraosseous Ewing's sarcoma [5–7]. This sarcoma is characterized by a chromosomal translocation, with around 85% of cases presenting with a t(11:22) (q24:q12) translocation between the EWS gene and FLI1 gene, leading to a fusion gene that affects transformation, differentiation, cell growth, and signal transduction of multiple genes.

Histologically, Ewing's sarcoma presents as anaplastic sheets and lobules of uniform cells, which can be round or oval, and blue in color. There are 2 cell types mainly seen, with most of the cells staining lightly and the rest staining darker, with this pattern being classically characteristic for Ewing's. Spindle cells and glycogen may also be present [4]. Contrary to the established histological features of Ewing's, the origin of

the cell type involved is still something debated. There have been many hypotheses regarding the cell type, with studies revealing markers such as neuron-specific enolase and S-100, which are associated with neuroectodermal lineage, being expressed in the tumor [5,8]. There have also been studies showing that the CD99 marker, which is relatively specific for Ewing's sarcoma, have been detected in mesenchymal stem cells and upregulated by EWS-FLI1 [5,9].

Ewing's sarcoma is a rare malignancy, as discussed earlier, with its most common manifestations in bone. There have been only 6 cases currently reported in the literature about Ewing's sarcoma in the testis. Patient ages have ranged from 17 to 46, with the main treatment option being orchiectomy followed by chemotherapy, with some patients presenting with no relapse of symptoms and 1 patient passing away 9 months after the initial diagnosis [10–14]. There has also been a case report about a 3-year-old boy that presented with the sarcoma in a scrotal mass [15]. Due to the rarity of the tumor, one must be well-informed to consider Ewing's sarcoma in the differential, as well as to effectively navigate through other paratesticular conditions when diagnosing a patient that presents with a mass.

The anatomy of the paratesticular region comprises the epididymis, vestigial remnants, tunica vaginalis, and spermatic cord along with its contents—the vas deferens, testicular and cremasteric arteries, pampiniform plexus, and lymphatic vessels. Pathological tumors of the paratesticular area are mostly benign lesions, histologically originating from epithelial, mesothelial, and mesenchymal elements of the contents of the paratesticular region. Lipomas of the spermatic cord are the most common benign tumors, closely followed by adenomatoid tumors and leiomyomas [2,3]. Primary solid malignancies are rare, with sarcomas accounting for most of these. Some rarely encountered pathological paratesticular mass differentials are desmoplastic small cell tumors, melanotic neuroectodermal tumors, and Ewing's sarcoma [16]. The paratesticular region is also the recipient of metastases from prostate, renal, and gastrointestinal primary neoplasms.

Patients presenting with paratesticular tumors are commonly asymptomatic, presenting with a slow-indolent, nontender mass. The diagnosis is often pinpointed by noting the morphological features and specific radiological findings and correlating them to patients' clinical history. The US as the first imaging modality, has nonspecific findings, but can differentiate a testicular from a paratesticular mass, and further characterize it as solid vs cystic and vascular vs nonvascular lesion. On ultrasound, the adenomatoid tumor is predominantly cystic and isoechoic, leiomyomas can be solid or cystic and may contain calcifications, and lipomas have a homogeneous hyperechoic appearance. For malignant tumors like rhabdomyosarcoma, liposarcoma, and leiomyosarcoma, US imaging findings are nonspecific and demonstrate varying echogenicity. At conventional US, rhabdomyosarcoma has a heterogeneous appearance due to hemorrhage and necrosis, liposarcoma appears as a hyperechoic area and leiomyosarcoma demonstrates a predominantly hypoechoic area [2,3].

The characterization of morphological features and delineation of staging is better established with CT and MR findings. Characteristic attenuation and signal intensity patterns of these tumors are often used to narrow the differential diagnosis. For example, low attenuation of a paratesticular mass at CT is suggestive of a fat-containing tumor. MR imaging is often employed to achieve a high-contrast spatial resolution and cover a wider anatomical space to precisely localize a tumor, define its anatomic relations and characterize the tissue to correlate with the histologic origin of the paratesticular mass to establish a diagnosis [16,17]. MR imaging differentiates solid from cystic lesions and testicular from paratesticular mass [2,3]. On MRI, lipomas demonstrate a high signal intensity on T1- and T2-weighted images, the adenomatoid tumor is seen as a relatively hypointense mass on T2-weighted images, whereas leiomyosarcoma show a marked enhancement. Metastasis presents as multifocal lower-intensity images on T2-weighted images [17]. CT and MR imaging also aid in the pelvic staging of malignant masses like liposarcoma and leiomyosarcoma to facilitate the planning of definitive surgical treatment.

Extraosseous Ewing's sarcoma can present on CT as a soft tissue mass with attenuation like that of skeletal muscle. On ultrasound, its presentation usually is a mass that is hypoechoic, heterogeneous, and has internal vascularity. On MRI, specifically T1-weighted imaging, the mass can demonstrate a signal intensity like that of skeletal muscle. On T2-weighted imaging, the mass can show a heterogeneous and hyperintense signal [18–20]. Treatment for Ewing's sarcoma slightly differs depending on patient presentation with regards to age and spread of the tumor but will typically include an alternating chemotherapy regimen of vincristine/doxorubicin/cyclophosphamide and ifosfamide/etoposide. Hematopoietic growth factor support may also be given. Surgical approaches as well as radiation therapy may also be considered. Due to the risk of relapse or recurrence, patients should follow-up closely over a long-term period with a multidisciplinary team [20]. In our case, the patient underwent surgical resection of the right testicle followed by chemotherapy and is currently receiving treatment.

Conclusions

Although most paratesticular tumors are determined to be benign, the aggressive and rare nature of paratesticular Ewing's sarcomas stress the importance of early and accurate recognition of this diagnosis. Imaging plays a vital role in differential diagnosis and subsequent patient management.

Patient consent

A written informed consent was obtained from the patient for the publication of this case report.

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