Paratesticular rhabdomyosarcoma - A case report

Prakash Kayastha[1,2], MD; Sharma Poudel[1,2], MD; Rajan Mani Bhandari[2], MBBS; Prem Raj Gyawali[3,4], MS; Shova Banstola[5], MD

[1]Department of Radiology and Imaging, Grande International Hospital, Kathmandu, Nepal
[2]Department of Radiology and Imaging, Maharajgunj Medical Campus, Institute of Medicine, TUTH, Kathmandu, Nepal
[3]Department of Urology & Kidney Transplant Surgery, Grande International Hospital, Kathmandu, Nepal
[4]Department of Urology, Maharajgunj Medical Campus, Institute of Medicine, TUTH, Kathmandu, Nepal
[5]Department of Pathology, Grande International Hospital, Kathmandu, Nepal

Corresponding author
Prakash Kayastha, MD
Email: dr_prakash_kayastha@hotmail.com

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ABSTRACT

Rhabdomyosarcoma is a malignant tumor of muscular origin. It comprises the most common soft tissue tumor in children accounting for approximately 5-8% of childhood cancers. Here we present a case of paratesticular rhabdomyosarcoma in a 13-year male child who was referred for ultrasonic examination (USG) to the department of radiology for evaluation of right scrotal swelling with pain for few months. Paratesticular rhabdomyosarcoma is a rare non germ cell tumor of scrotal sac in children and young adult/teens which can invade testis at presentation. We review the epidemiology, histology, clinical presentation, staging and prognosis of paratesticular rhabdomyosarcoma and discuss the role of radiology in their management.

Key words: **USG, rhabdomayosarcoma, testis**

Introduction

Soft tissue sarcomas are a heterogeneous group of malignant tumors of the mesenchymal origin. They are classified on the basis of tissue seen on histology. Sarcomas are found in young patients of less than 45 years of age, with approximately 65% diagnosed in patients under the age of 10 years[1]. They have a slightly higher male predominance (1.67:1) with Caucasian children affected more often than children of other races[2]. Childhood rhabdomyosarcoma (RMS) is the most common soft tissue sarcoma. They are found essentially anywhere in the body – head and neck (50%), genitourinary (25%), extremities (15%) and other locations like trunk, thorax and gastrointestinal tract (10%)[3].

Paratesticular rhabdomyosarcoma accounts for 7% of all RMS and is the most common soft tissue sarcoma in childhood in the genitourinary region according to the Intergroup RMS Study (IRS) Group[4]. It can arise from the spermatic cord, epididymis and the testicular envelopes. It generally presents clinically as rapidly enlarging, usually painless, scrotal mass. Hydrocele may be an associated feature. Due to the superficial location of the mass, it is easily identified by the patient and the clinician and is amenable to complete surgical resection. Robust clinical examination and sonography are the keys to early detection. MRI provides better soft tissue resolution of the lesion and helps in delineating its extent and for planning the surgical procedure. CT and PET-CT are helpful for detecting any metastasis to locoregional or distant organ system or lymph nodes. Bone scan and MRI are helpful in detecting marrow deposits. Histopathological diagnosis can be obtained by percutaneous, incisional or excisional biopsy.

Case report

A male child aged 13-years of age presented with history of right scrotal swelling associated with slight pain which was progressive over a period of 4 to 5 months. No positive family history was present. Ultrasound examination of the scrotum revealed a large circumscribed hypoechoic mass in the right paratesticular region measuring approximately 8.2 x 5.4 cm in size with significant internal vascularity without evidence of invasion into the surrounding structure (Fig. 1). Minimal associated right hydrocele was seen. Bilateral testis and epididymis had normal appearance. No obvious inguinal or retroperitoneal lymphadenopathy was seen. Other intraabdominal organs were normal sonologically. The serum tumor markers (LDH-193U/L, AFP-3.66IU/ml and total β-hcg-<2.39mIU/ml) were within normal limits. He underwent right high extended orchidectomy with no need for inguinal or retroperitoneal lymph node dissection. The histopathological examination (HPE) showed right paratesticular mass with maximum dimension measuring 9 cm surrounded by tunica albuginea.

The microscopy showed tumor with cellular and hypocellular areas, round to spindled cells with hyperchromatic nuclei and focal areas showing prominent nucleoli (Fig. 2). No necrosis was seen. Mitosis was 30/10 HPF. Strap cells and tadpole like cells were also seen. Tumor was limited to tunica with no extracapsular, testicular, lymphovascular or epididymal invasion and with free resected margins. The findings were morphologically compatible with rhabdomyosarcoma, embryonal variant. Immunohistochemistry was not performed. Post-operative period was uneventful and the patient was discharged with advice for regular follow up.

Discussion

RMS usually occurs in the extremities. A paratesticular location is rare. Paratesticular tumor develops from mesenchymal tissues of the spermatic cord and epididymis. There are three subtypes of RMS - embryonal (75%), alveolar (20%) and pleomorphic (5%)[2]. The embryonal type consists of spindle cell, botryoid and anaplastic subtypes. Rhabdomyoblast is the characteristic cell but is not essential for diagnosis[5]. RMS can have syndromic associations with NF I, Beckwith-Weidemenn and Li-Fraumeni syndromes[1].The tumor manifests as a hard painless inguinoscrotal swelling, the size and duration of development are varied and it rarely invades the scrotal skin. Differential diagnoses include testicular torsion, epididymo-orchitis, scrotal abscesss and testicular tuberculosis.

Radiologically, the appearance of the mass is non-specific and indistinguishable from other sarcomas. The location and demographics of the patient are most useful in narrowing down the differential diagnosis. Scrotal ultrasound is the first imaging modality for a scrotal mass. It shows heterogeneous well defined irregular mass of low to medium echogenicity. Inguinoscrotal extension can be studied. Plain radiographs can help identify calcifications in the mass, bony involvement and metastases. A thoraco-abdomino-pelvic CT scan allows for any deep invasion, locoregional nodal and distant organ metastasis. MRI using

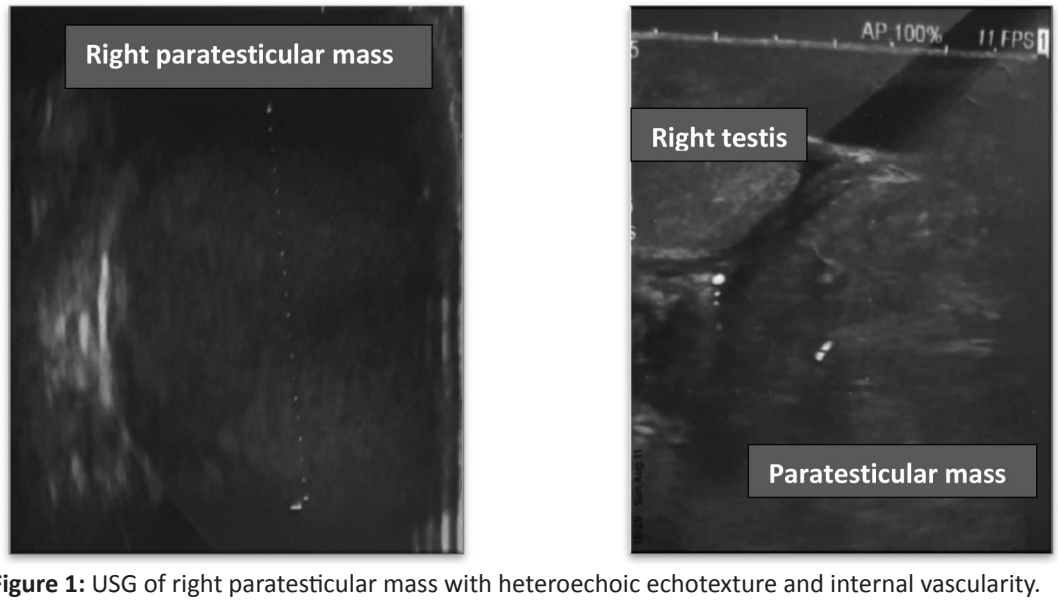

Figure 1: USG of right paratesticular mass with heteroechoic echotexture and internal vascularity.

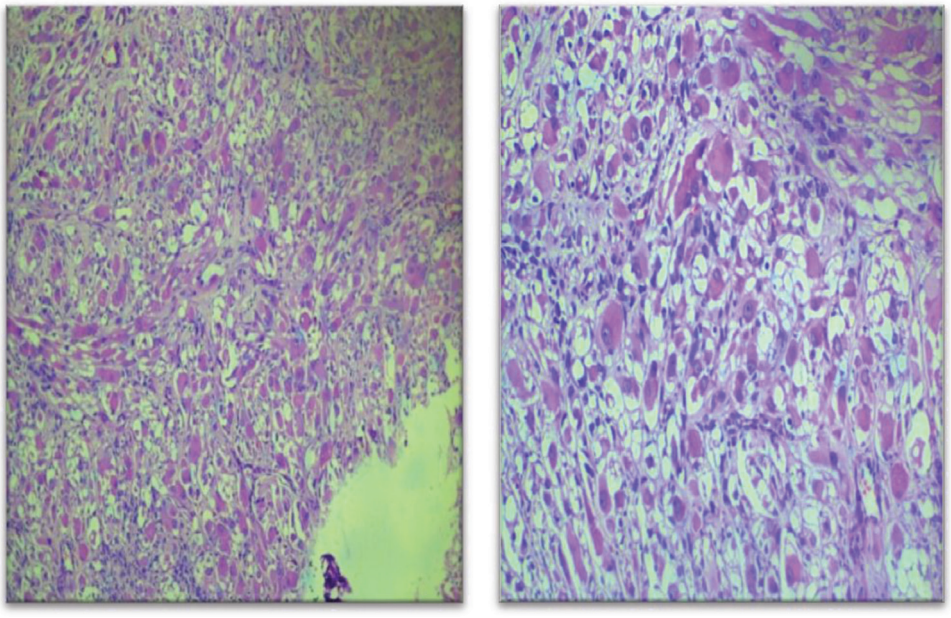

Figure 2: a- lower magnification b – higher magnification: Microscopy showing tumor with cellular and hypocellular areas, round to spindled cells with hyperchromatic nuclei, strap and tadpole like cells with focal areas showing prominent nucleoli.

surface coils is an efficient imaging modality. The tumor shows homogenously iso to low in T1 and heterogeneously high in T2 weighted images. The visualization of a low signal intensity of the tunica albuginea in T2 weighted images allows the clear demarcation of the mass from the testis.

The tumoral markers including AFP, B-hcg, LDH and CEA are usually normal. Same was the finding in our case. Whenever rhabdomyoblasts are not seen in the HPE, immunohistochemical investigations are conducted using a panel of antibodies including myosin and desmin[6].

The first step in treatment is radical orchidectomy by the inguinal route with first cord ligation regardless of the stage of the disease. Whenever local invasion or nodal involvement is seen, hemiscrotectomy is performed. Lymph nodal dissection should not be performed without CT imaging or lymphography. Some advocate routine chemotherapy with Actinomycin D, Vincristine and Cyclophosphamide since RMS is chemosensitive[6]. Radiotherapy is a complementary treatment of chemotherapy and surgery to eliminate residual foci and retroperitoneal lymph nodes.

Conclusion

Paratesticular rhabdomyosarcoma is a rare aggressive malignant tumor in children and young adults. However, localized forms have a good prognosis with early detection and treatment. Integrated treatment approach with surgery, chemotherapy and radiotherapy is required with locally advanced or late stage diseases. Patients are advised to follow up to detect recurrence or late distant metastasis.

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