Radiologic Pathologic Correlation of Primary Paratesticular Rhabdomyosarcoma

Vignesh Kumar Mohan[1] Dillibabu Ethiraj[1,] Venkatraman Indiran[1,] Immanuel Judah Pandiaraj[1] Adarsh Murugan[1] Prabakaran Maduraimuthu[2]

[1]Department of Radiodiagnosis, Sree Balaji Medical College and Hospital, Chennai, Tamil Nadu, India
[2]Department of Radiology, Sree Balaji Medical College and Hospital, Chennai, Tamil Nadu, India

Address for correspondence Venkatraman Indiran, MD, DNB, Department of Radiodiagnosis, Sree Balaji Medical College and Hospital, 7 Works Road, Chromepet, Chennai, Tamil Nadu 600044, India (e-mail: ivraman31@gmail.com).

J Gastrointestinal Abdominal Radiol ISGAR:2020;3(suppl S1):S80–S84

Abstract

Rhabdomyosarcoma is the most common soft tissue sarcoma in childhood and adolescence, arising from the mesenchymal tissue. It is an intrascrotal tumor localized in the paratesticular structures such as the epididymis or spermatic cord. The majority of this tumor occurs in the first two decades of life. Ultrasound is considered the imaging modality of choice for evaluating an intra-scrotal abnormality. We present a case of a 4-year-old boy with a 2-weeks history of rapidly increasing left scrotal swelling which was clinically diagnosed as hydrocoele. We present the ultrasound, elastography and MRI features of a case of paratesticular embryonal rhabdomyosarcoma with pathologic correlation and also emphasize need to consider sarcoma as a differential diagnosis for rapidly growing scrotal swelling.

Keywords
- rhabdomyosarcoma
- embryonal
- testis
- myogenin

Introduction

Soft tissue sarcomas account for up to 3% of the childhood cancers and up to 1% of the adult cancers.[1] The annual worldwide incidence of rhabdomyosarcoma is 4.5 cases per million.[2] Embryonal Paratesticular Rhabdomyosarcoma (EPR) arises from the mesenchymal elements of the testes, epididymis and the spermatic cord. Classically, it presents as a painless scrotal mass. It is regarded as a highly malignant tumor with frequent recurrence. Ultrasound (USG) examination is the first-line investigation, but can be inconclusive. In this setting, the magnetic resonance imaging (MRI) provides more useful information about location, extension, lymph node involvement; however, its role in defining the diagnosis of the rhabdomyosarcoma is not clear. High inguinal orchidectomy, followed by adjuvant chemotherapy, is the current recommended treatment.

Case Report

A 4-year-old boy presented with a 2-week history of a left painless scrotal swelling with rapid growth. There was no significant past medical history, scrotal trauma and infection. Physical examination revealed a scrotal mass occupying the entirety of the left hemiscrotum. The mass was tense and diffusely firm in consistency. There was no warmth or tenderness. Trans-illumination test was positive. Bilateral cord structures were palpable and it was possible to get above the swelling (►**Fig. 1**). There were no palpable inguinal lymph nodes. Ultrasound of scrotum (USG) demonstrated a large, mildly heterogeneous, well-defined solid lesion with moderate internal vascularity with low resistance noted in the left hemiscrotum just abutting the left testis (►**Fig. 2**). On Real time elastography (RTE) performed using Siemens Acuson S2000 scanner, mass appeared completely inelastic with a visual elasticity score (VES) score of 5, which was considered suspicious of malignancy (►**Fig. 3**). The right testis and epididymis were normal. The Magnetic resonance imaging of pelvis (MRI) showed a large well-defined ovoid, mildly heterogeneous lesion measuring ~ 4.2 × 4.1 × 3.9 cm and appearing predominantly hypointense on T1 weighted images and hyperintense on T2 weighted images. The lesion was seen inferior to the left testis and epididymis, abutting them closely. A vertical T2 hypointense septation was seen

DOI https://doi.org/10.1055/s-0040-1715776
ISSN 2581-9933.

Thieme Medical and Scientific Publishers Pvt. Ltd., A-12, 2nd Floor, Sector 2, Noida-201301 UP, India

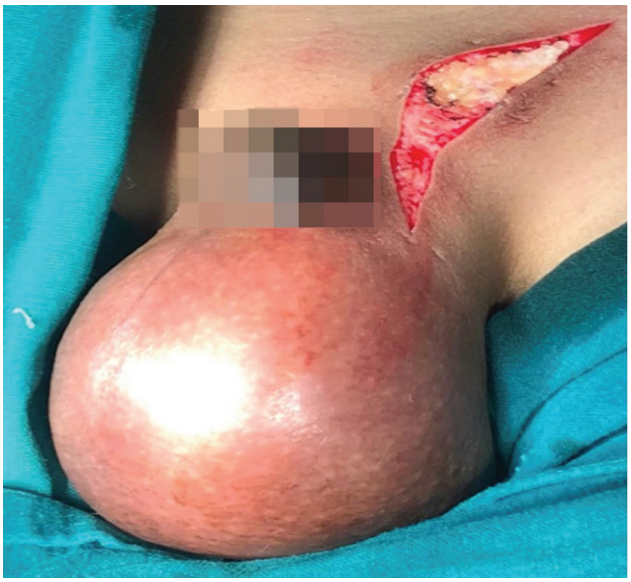

Fig. 1 Image shows a scrotal mass occupying the entirety of the left hemiscrotum.

within the lesion (►**Fig. 4 A**, **B**). MRI abdomen and chest radiograph showed no evidence of metastases. Inguinal lymph nodes appeared unremarkable. Laboratory test results including α fetal protein, human chorionic gonadotropin were all within the normal limits. Left radical orchiectomy was performed. There was a time gap of 2 weeks between MRI and surgery. The surgical specimens obtained include the spermatic cord measuring ~ 5 cm in length, the immature testis measuring ~ 1.1 cm and a globular soft tissue mass measuring ~ 7 × 7 x 5 cm. All the specimens were sent for histopathological analysis. The globular mass showed greyish white areas, with few gelatinous areas in the upper pole, which covered more than 95% of the testicular parenchyma. No necrosis was identified (►**Fig. 5**).

Histological study demonstrated cellular neoplasm composed of round to oval cells with irregular, enlarged, dark stained nuclei and drawn out eosinophilic cytoplasm, arranged in clusters and sheets in a cellular and hypocellular distribution. The stroma appeared edematous with myxoid areas. Mitotic figures (>20/10 HPF), thin-walled

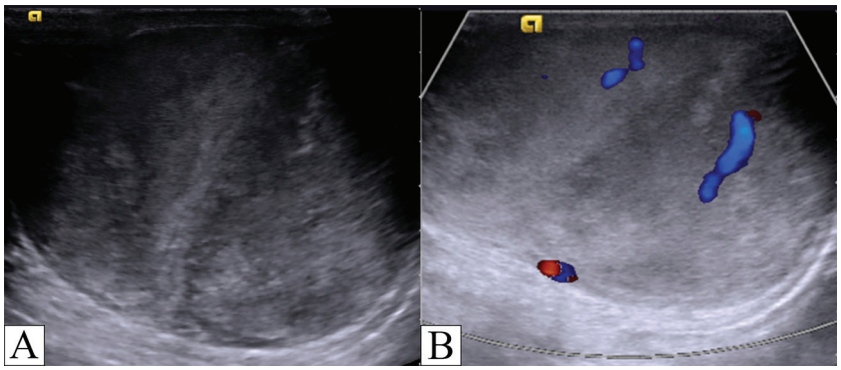

Fig. 2 (**A**) shows large, mildly heterogeneous, well defined, solid lesion noted in the left hemiscrotum just abutting the left testis. (**B**) Doppler shows moderate internal vascularity within solid lesion in the left hemiscrotum.

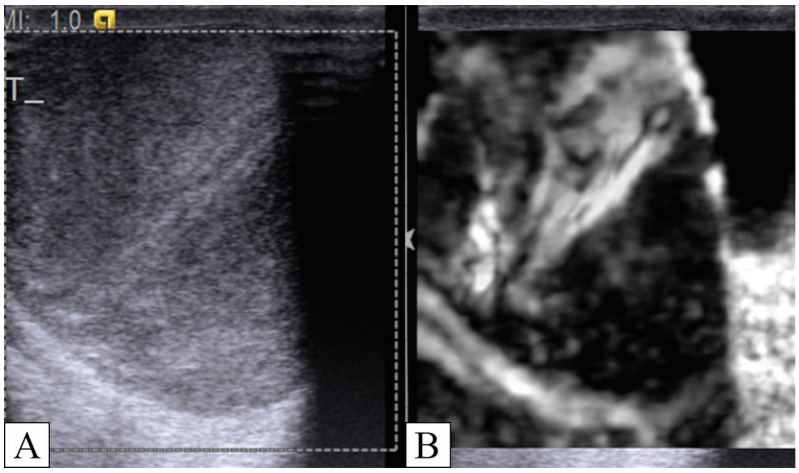

Fig. 3 (**A and B**) Real Time Elastography shows a completely inelastic mass with a visual elastic score of 5.

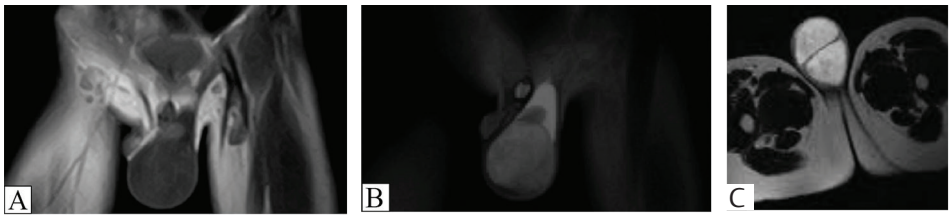

Fig. 4 MRI of pelvis. Coronal T1 weighted image (**A**) pelvis shows well defined ovoid mildly heterogeneous predominantly hypointense lesion. Coronal (**B**), axial (**C**) T2 weighted images show well defined ovoid hyperintense lesion obliquely T2 hypointense septations, inferior to left testis and epididymis, abutting them closely.

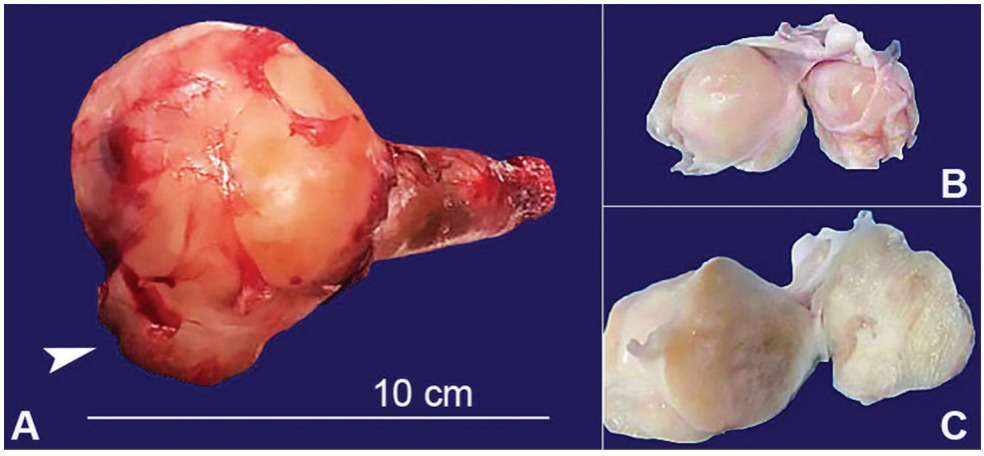

Fig. 5 (**A**) Surgical specimen with spermatic cord and immature testis with globular soft tissue mass (white arrow head) (**B, C**) Cut section of surgical specimen. The globular mass appeared greyish white with few gelatinous areas in the upper pole.

blood vessels in the stroma and rhabdomyoblast-like cells with cytoplasmic granularity were demonstrated (►**Fig. 6 A**, **B**). Immunohistochemistry (IHC) showed positivity for Desmin and Myogenin confirming the diagnosis of embryonal rhabdomyosarcoma (►**Fig. 7**). Post-operative period was uneventful. Chemotherapy with vincristine and actinomycin regimen was started 2 weeks after surgery and the patient was scheduled for follow-up.

Discussion

Rhabdomyosarcoma occurs predominantly in the head and neck followed by the genitourinary location.[2] Paratesticular rhabdomyosarcoma is a rare tumor comprising only 4% of all rhabdomyosarcoma cases.[3] Paratesticular rhabdomyosarcoma derives from the mesenchymal tissue of the epididymis, spermatic cord and testes.[3] There is a bimodal age distribution with peak incidences at 5 years and 16 years. Patients typically manifest with a short history of a hard, painless swelling in the scrotum.[4] USG is the first-line investigation imaging method for scrotal masses in distinguishing intratesticular from extratesticular lesions. USG commonly shows a large, well-defined, heterogeneous vascular mass. RTE allows further characterization of lesions as either non-neoplastic lesion or malignant neoplasms. Those lesions with VES > 3 may represent malignant lesions (sensitivity ~ 81.1–100%) with exceptions being testicular epidermoid and dermoid.[5,6] MRI can be used to differentiate benign and metastatic lesions with high accuracy.[4] EPR is regarded as an aggressive tumor with 26–71% of cases presenting with metastases to para-aortic and iliac nodes, hematogenous spread to lung, liver and bones.[4] Hence Computed Tomography of the chest, abdomen and pelvis is essential to staging the disease. Rhabdomyosarcoma is staged according to the TNM system. T1 tumors are confined to the organ and T2 tumors are locally infiltrating. T1 and T2 are further divided into (a) or (b) subset, depending on whether they are less than or greater than 5 cm. N0 is no nodal involvement, while N1 represents regional lymph node involvement. M0 represents no metastasis, with M1 defining distant metastasis. TNM staging of our patient was T1b, N0, and M0. The final diagnosis was established by histological study after the tumor excision.[7] Mimickers of rhabdomyosarcoma includes chronic inflammatory processes such a chronic epididymitis, leiomyoma and adenomatoid tumors.[8] Presence of variable echogenicity in a well-defined round shaped mass predominantly is seen in adenomatoid tumor which arises in the third and fourth decade of life. Sharp shadows caused by the transition zones between different tissue components of a solid hypoechoic or heterogeneous mass, is specific for leiomyoma.[9-11] Imaging cannot discriminate paratesticular rhabdomyosarcoma from other paratesticular sarcomas such as leiomyosarcoma, liposarcoma and fibrosarcoma. However, these entities have not been reported in the pediatric age group.

Histologically, according to the International Classification of Rhabdomyosarcoma for child age, rhabdomyosarcoma is divided into those with better prognosis (Botrytis

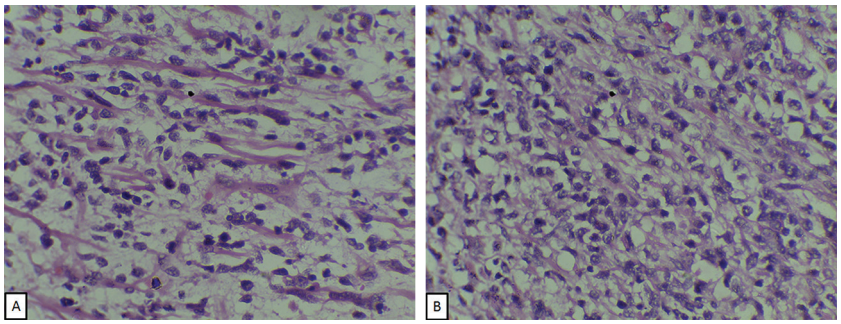

Fig. 6 (**A**) Histopathology shows round to oval cells with irregular, enlarged, dark stained nuclei and drawn out eosinophilic cytoplasm, arranged in clusters and sheets in a cellular and hypocellular distribution (H and E, ×40). (**B**) Shows edematous stroma, myxoid areas and mitotic figures (> 20 /10 HPF). Rhabdomyoblast like cells with cytoplasmic granularity are seen (H and E, ×40).

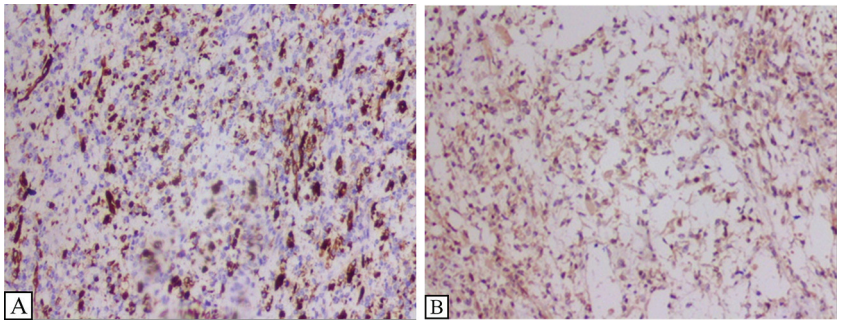

Fig. 7 (**A**) IHC shows positivity for Desmin with strong cytoplasmic positivity and (**B**) shows positivity for Myogenin with moderate nuclear positivity in 60% cells.

Rhabdomyosarcoma and Spindle Cells Rhabdomyosarcoma), intermediate grade (Conventional Embryonal Rhabdomyosarcoma) and high grade (Alveolar Rhabdomyosarcoma, Undifferentiated Rhabdomyosarcoma). The most common histologic type is the embryonic type.[12] The microscopic appearance of embryonal rhabdomyosarcoma is the presence of sheets or clusters of round to oval cells with thin cytoplasmic process that produce a 'tadpole', racquet' or 'strap cell appearance and collagenous stroma with myxoid foci as seen in our case. The presence of rhabdomyoblasts though characteristic is not a necessary criterion for diagnosis of rhabdomyosarcoma. Hence, confirmatory diagnosis is provided by immunohistochemistry.[13] IHC markers such as Myogenin, Desmin and MyoD1, are crucial for the confirmation of the diagnosis of rhabdomyosarcoma. Desmin is positive in more than 90% of the rhabdomyosarcoma although it can be positive in 50 to 70% of the leiomyosarcomas.[14] Immunostaining with MyoD1 has high sensitivity and specificity for alveolar rhabdomyosarcoma compared with embryonic ones. The Myogenin gene is highly specific for embryonic rhabdomyosarcoma. Rhabdomyosarcomas are staged based on TNM system.[15] Treatment strategies includes radical high inguinal orchidectomy, concurrent chemotherapy, radiotherapy and retroperitoneal lymph node dissection. Patients with disease localized to the scrotum have good prognosis.[4] Prognostic factors for children include tumor size, resectability, age and lymph node involvement.[14,16]

Conclusion

For patients with testicular or paratesticular mass, USG is essential to evaluate intrascrotal abnormalities. USG findings for benign diseases such as adenomatoid tumors, leiomyoma's and fibromas may mimic rhabdomyosarcoma. RTE and MRI can be used to differentiate benign and metastatic lesions with high accuracy[4] and this can help in early surgical intervention and chemotherapy, crucial to survival rate improvement. However final diagnosis is by HPE.

Ethical Approval

This article does not contain any studies with animals performed by any of the author(s). All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards. Informed consent was obtained from all individual participants included in the study.

Funding

None.

Conflict of Interest

None declared.

References

1 Kelly B, Lundon D, Rowaiye B, Daly P, Walsh K. Embryonal rhabdomyosarcoma of the testis. Can Urol Assoc J 2011;5(1):E7–E10
2 Rodríguez SS, Vinces A, Rey S, Cedeños JG, Solis O, Boccara H. Rare testicular tumor report of a case. Clin Oncol Case Rep 2018;1:1
3 Mak CW, Chou CK, Su CC, Huan SK, Chang JM. Ultrasound diagnosis of paratesticular rhabdomyosarcoma. Br J Radiol 2004;77(915):250–252
4 Shah T, Abu-Sanad O, Marsh H. Role of magnetic resonance imaging in the early diagnosis of paratesticular rhabdomyosarcoma. Ann R Coll Surg Engl 2016;98(5):e74–e76
5 Fang C, Huang DY, Sidhu PS. Elastography of focal testicular lesions: current concepts and utility. Ultrasonography 2019;38(4):302–310
6 Secil M, Bertolotto M, Rocher L, et al. Imaging features of paratesticular masses. J Ultrasound Med. 2017;36(7):1487–1509
7 Bouchikhi AA, Mellas S, Tazi MF, et al. Embryonic paratesticular rhabdomyosarcoma: a case report. J Med Case Reports 2013;7:93
8 Akbar SA, Sayyed TA, Jafri SZH, Hasteh F, Neill JS. Multimodality imaging of paratesticular neoplasms and their rare mimics. Radiographics 2003;23(6):1461–1476
9 Hricak H, Filly RA. Sonography of the scrotum. Invest Radiol 1983;18(2):112–121
10 Leonhardt WC, Gooding GA. Sonography of epididymal leiomyoma. Urology 1993;41(3):262–264
11 Hertzberg BS, Kliewer MA, Hertzberg MA, Distell BM. Epididymal leiomyoma: sonographic features. J Ultrasound Med 1996;15(11):797–799
12 Kumar R, Kapoor R, Khosla D, et al. Paratesticular rhabdomyosarcoma in young adults: A tertiary care institute experience. Indian J Urol 2013;29(2):110–113
13 Horn RC Jr, Enterline HT. Rhabdomyosarcoma:a clinicopathological study and classification of 39 cases. Cancer 1958;11(1):181–199 . (Ref. 13 “Horn, Enterline, 1958”)
14 Weiss SW, Goldblum JR, Tumors of soft tissues. Rabdomiosarcoma: Elsevier; 2010:595–633
15 Raney RB, Maurer HM, Anderson JR, et al. The Intergroup Rhabdomyosarcoma Study Group (IRSG): Major Lessons From the IRS-I Through IRS-IV Studies as Background for the Current IRS-V Treatment Protocols. Sarcoma 2001;5(1):9–15
16 Husain AN, Biopsy interpretation of pediatric lesions. Wolters Kluwer Health; 2014;323–413

