Case Report

Paratesticular embyronal rhabdomyosarcoma in an adolescent: A rare case report

B. R. Vani, K. Geethamala, V. Srinivasa Murthy, M. U. Thejaswini, K. P. Padmaja

Department of Pathology, ESIC Medical College and PGIMSR, Rajajinagar, Bengaluru, Karnataka, India

ABSTRACT

Embyronal rhabdomyosarcoma (RMS) accounts for approximately 49% of all RMS. After head and neck, this tumor is most commonly found in genitourinary region, which includes paratesticular RMS. Paratesticular RMS is rare constituting 4–7% of all RMS in children and young adults. It has been regarded as highly malignant tumor with frequent recurrence. The management protocol is of multimodal approach of surgery, chemo, and radiotherapy. We herein report a case of left paratesticular RMS in an 18-year-old male, which posed a diagnostic dilemma clinically and by imaging. Histopathology with added immunohistochemistry brought out the confirmatory diagnosis. The patient was successfully treated and on follow-up is disease free until date.

Key words: Histopathology, paratesticular, rhabdomyosarcoma

INTRODUCTION

Rhabdomyosarcomas (RMS) are the heterogeneous soft tissue sarcoma.[1] They arise from primary mesenchymal cells committed toward skeletal muscle differentiation and occur in a variety of organs lacking skeletal muscle.[2] After head and neck, RMS is most commonly found in genitourinary region. Genitourinary RMS include tumors originating in the urinary bladder, prostate, testis, paratesticular sites, penis, perineum, vagina, and uterus occurring in children in the first two decades of life with a median age of 14 years. Paratesticular RMS is a collective term for primary tumor arising from spermatic cord, testis, penis, and epididymis.[3] They have an aggressive course if not treated with combined modalities of surgery, chemo and radiotherapy.[1,4,5] Herein, authors report a case of left paratesticular RMS in an 18-year-old male, which posed a diagnostic dilemma clinically and by imaging. Histopathology with added immunohistochemistry (IHC) brought out the confirmatory diagnosis.

Access this article online	
Quick Response Code:	Website: www.ccij-online.org
	DOI: 10.4103/2278-0513.142688

CASE REPORT

An 18-year-old college student presented to the surgical outpatient departments with a history of painless progressive swelling in the left scrotum since 2 months. On examination, a palpable firm nontender, nontransilluminating mass measuring 11 cm × 8 cm felt in the left scrotal sac was noted. Other scrotal side was normal. Perabdomainal examination found no mass or oragnomegaly. There were no other local or system complaints. Clinically a diagnosis of left testicular tumor was offered. Routine urine and blood investigation were normal. Ultrasonogram (USG) scrotum revealed enlarged left scrotal mass with decreased echogenicity; however, no discernible testis or epididymal structures identified and also showed inguinal lymph node mass possibly metastatic deposit; hence, diagnosis was retained as testicular carcinoma with inguinal lymph node metastasis. Special investigation of beta human chorionic gonadotropin (β-HCG) (<100 mIU/L) and alpha-fetoprotein (<0.5 IU/ml) considering germ cell tumors were within normal limits. With prior preoperative investigation, left radical orchidectomy with retroperitoneal lymph node dissection was carried out. The resected specimen was a circumscribed lobulated mass measuring 11 cm × 8 cm × 6 cm along with attached spermatic cord and a single lymph node mass was identified. Cut surface revealed grey-white firm lesion with intervening microcysts, hemorrhage, necrosis and myxoid foci. On extensive sampling periphery of the mass showed compressed

Address for correspondence: Dr. B. R. Vani, Department of Pathology, ESIC Medical College and PGIMSR, Rajajinagar, Bengaluru, Karnataka, India. E-mail: vanibr@yahoo.in

normal testis measuring 2 cm × 2 cm with retained positive string test. However, morphology of epididymis could not be made out. The excised inguinal lymph node mass also showed similar features [Figure 1]. Histopathological examination revealed neoplasm with pleomorphic cells arranged in sheets, nodules, cords separated by fibrous septae. These pleomorphic cells were round to oval with vesicular nucleus, prominent nucleoli and scant cytoplasm. Foci of clear cells having dense eosinophilic cytoplasm, few spindle cells with cytoplasmic tails were noted. The tumor exhibited varied cellularity with hypocellular myxoid areas, microcysts, and hemorrhage [Figures 2-4]. In addition large areas of necrosis, mitotic figures of 10–12/high power field and infiltration into epididymal tissue seen. Adjacent testis, spermatic cord, surgical margins were free of tumor. Single lymph node mass identified showed metastatic deposit of similar morphology. IHC performed was positive for vimentin, desmin, myogenin [Figure 4 inset] and negative for smooth muscle actin. Considering IHC and histomorphology we arrived at a final diagnosis of embryonal paratesticular RMS with inguinal lymph node metastatic deposits. Further metastatic workup was done with chest X-ray, computed tomography abdomen pelvis, whole body magnetic resonance imaging and found no other local or distant metastasis and hence, the patient was staged as Stage 1 (favorable site of paratesticular region T1a, N1, M0). Patient was given six cycles of chemotherapy with VAC regimen comprising of vincristine, Adriamycin, and cyclophosphamide. Follow-up of the patient with periodic radiological examination showed no recurrence until date and boy is doing well.

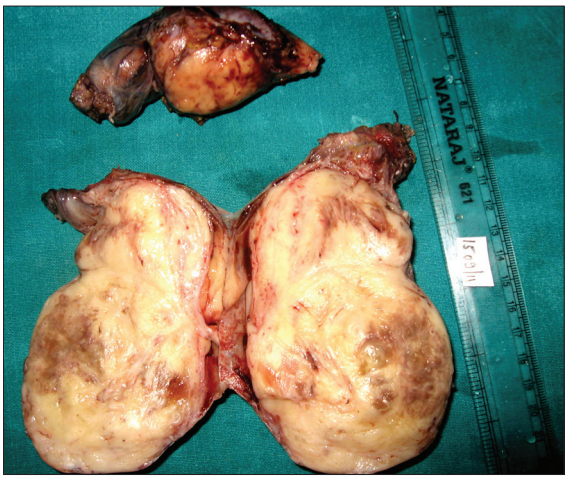

Figure 1: Gross photograph of the excised mass with a grey-white lobulated surface and areas of myxoid change. Further sectioning showed compressed normal testis. The excised inguinal lymph node mass also showed similar features

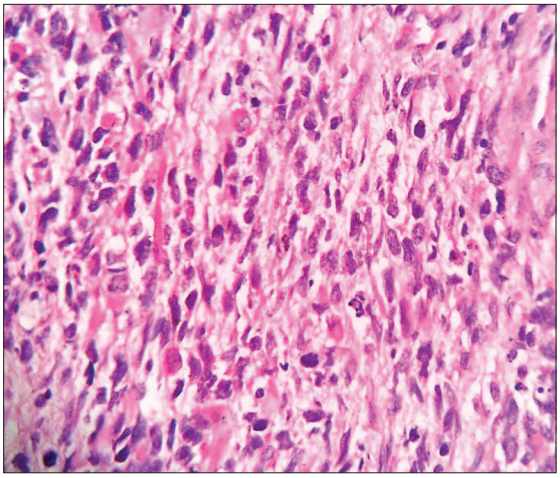

Figure 3: Photograph showing mitotic figures and tadpole cells (H and E, ×400)

DISCUSSION

The first description of RMS was by Weber in 1854. However, the "definitive" publication was considered by Arthur Purdy Stout in 1946, 92 years later.[6] RMS represents 5-10% of malignant solid tumor and 15-30% of genitourinary RMS in childhood.[3,7] Paratesticular RMS is rare constituting 4-7% of all RMS in children and young adults.[3,5,8] In scrotum, they are the most common nongerminal malignant lesions with

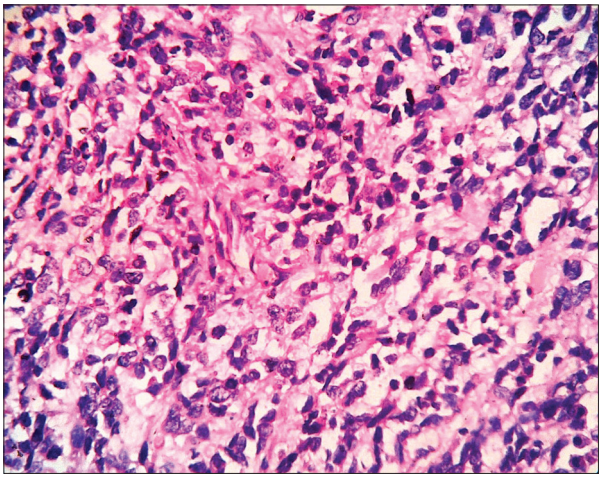

Figure 2: Photograph showing small cells with hyperchromatic nucleus, scant cytoplasm and spindle cells (H and E, ×400)

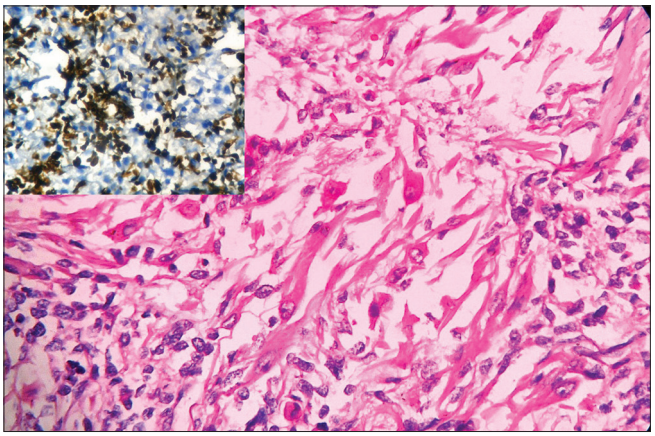

Figure 4: Photograph showing strap cells or ribbon cells (H and E, ×400), inset showing immunohistochemistry myogenin positivity (×400)

the peak incidence between 1 and 5 years.[1,8] Nevertheless, bimodal age distributions with the second peak during adolescence do occur with a median age of 14 years.[3]

The most common clinical manifestation being short duration of painless scrotal mass often ignored.[4,8] Rarely can also present as tender scrotal mass, wherein clinically can be misdiagnosed as inguinal hernia, hydrocele or epididymitis.[4] The tumor has no racial or right/left scrotal side predilection.[8] Kumar *et al*.,[5] in their study of 10 paratesticular RMS, found median age being 16.5 years, while mean duration of symptoms were of 5 months. In the present case, patient was 18-year-old boy with a history of left scrotal painless swelling of 2 months duration.

Ultrasonogram can be a screening effective modality of investigation.[3,9] However, many authors have deferred in opinions saying due to varied echogenicity of the lesion USG poses diagnostic dilemma.[3,4,9] Color and duplex Doppler sonographic evaluation of paratesticular RMS exhibit characteristic hyperemia and high diastolic flow in the mass.[3,4] In the present case the patient was subjected to only USG and with varied echogenicity diagnosis of testicular carcinoma was offered. Moreover considering the age, yolk sac tumor was thought of and serum markers like β-HCG, alpha-fetoprotein were done and found to be normal. Hence considering the clinical and radiological diagnosis of testicular carcinoma left radical orchidectomy with retroperitoneal lymph node dissection was carried out.

The gross morphology of RMS is variable. Paratesticular RMS is often well circumscribed by tunica and cut surface reveals firm, fleshy, lobulated, myxoid, necrotic and hemorrhagic areas.[3,10,11] However, thorough sampling is required for the identification of normal testicular structure as noted in our case.

The classic histology features is of varied representation of maturational stages of rhabdomyoblasts (fetal muscle cells). Hence, poorly differentiated tumor is composed of small round or spindle shaped cells with hyperchromatic nuclei and indistinct cytoplasm and differentiated rhabdomyoblasts are either absent or localized to small foci. Well-differentiated tumors composed of eosinophilic cells ranging from slender spindle shaped cells to large eosinophilic cells with strap, ribbon, tadpole or racquet shape and one to two centrally placed nuclei with prominent nucleoli with or without cross-striations in the cytoplasm.[3,9,10] Added to this IHC plays an important role wherein cells are positive for desmin, myogenin and negative for smooth muscle actin.[3,4,6-8]

Rhabdomyosarcoma is a highly aggressive malignant tumors with frequent recurrence.[1,4,5] First and most common pathway of spread is via lymphatics to paraaortic, paracaval and inguinal lymph nodes.[8] Hematogenous spread to the bone marrow, lung and liver seen in 20% of patients at the time of initial presentation.[8,12] In the present case, patient had inguinal lymph node metastasis and no distant spread. Cytogenetics of embryonal RMS exhibit consistent loss of heterozygosity at chromosome 11p15.5. Spindle cell variant (composed of > 80% elongated spindle cells) of embryonal RMS, is more common in the paratesticular region and has a better prognosis.[6] Due to financial constraints cytogenetics was not done in our case.

Rhabdomyosarcoma being a rare entity, the prognosis is determined by clinical group, stage, histology and age at presentation. Three pediatric cancer groups joined in 1972 and designed the Intergroup Rabdomyosarcoma Study Group, which formulates an important treatment protocol and can predict prognosis.[2] However, later by Newton *et al*. framed another classification called International Classification of Rhabdomyosarcoma which is easily reproducible and predictive of outcome among patients with differing histologies.

Radical orchidectomy procedure with negative surgical margins is the gold standard in the treatment protocol of these paratesticular RMS.[8] Addition of chemotherapy and radiotherapy has elevated survival rates from 30% to 90%, respectively.[1,8] Hence emphasizes on the multimodal approach to the usual tradition treatment regimen. In the present case, the patient was carried with the surgery followed by chemotherapy and doing well until date.

CONCLUSION

Paratesticular RMS is a rare aggressive neoplasm, which needs multimodal treatment regimen to increase disease free survival. In this background, the present case attempts to highlight the paramount of histopathology along with IHC even when preoperative varied investigations pose diagnostic dilemmas.

REFERENCES

1. Kumar R, Bharti S, Khosla D, Kapoor R. Long term survival in paratesticular rhabdomyosacoma. Clin Cancer Investig J 2012;1:31-2.
2. McCarville MB, Spunt SL, Pappo AS. Rhabdomyosarcoma in pediatric patients: The good, the bad, and the unusual. AJR Am J Roentgenol 2001;176:1563-9.
3. Agrons GA, Wagner BJ, Lonergan GJ, Dickey GE, Kaufman MS. From the archives of the AFIP. Genitourinary rhabdomyosarcoma in children: Radiologic-pathologic correlation. Radiographics 1997;17:919-37.
4. Mak CW, Chou CK, Su CC, Huan SK, Chang JM. Ultrasound diagnosis of paratesticular rhabdomyosarcoma. Br J Radiol 2004;77:250-2.
5. Kumar R, Kapoor R, Khosla D, Kumar N, Ghoshal S, Mandal AK,

et al. Paratesticular rhabdomyosarcoma in young adults: A tertiary care institute experience. Indian J Urol 2013;29:110-3.
6. Weiss SW, Goldblum JR. Rhabdomyosarcoma. In: Enzinger and Weiss's Soft Tissue Tumors. 5th ed. Saunders: Mosby Elsevier; 2008. p. 595-629.
7. Raney RB, Tefft M, Hays DM, Triche TJ. Rhabdomyosarcoma and the undifferentiated sarcomas. In: Pizzo PA, Poplack DG, editors. Principles and Practice of Pediatric Oncology. Philadelphia: Lippincott; 1993. p. 769-94.
8. Resim S, Okur N, Bakarıs S, Kilic AO, Altunoluk B. Paratesticular embryonal rhabdomyosarcoma; report of a case. Iran J Pediatr 2009;19:430-4.
9. Lang P, Johnston JO, Arenal-Romero F, Gooding CA. Advances in MR imaging of pediatric musculoskeletal neoplasms. Magn Reson Imaging Clin N Am 1998;6:579-604.
10. Horn RC Jr, Enterline HT. Rhabdomyosarcoma: A clinicopathological study and classification of 39 cases. Cancer 1958;11:181-99.
11. Ferrari A, Casanova M, Massimino M, Luksch R, Piva L, Fossati-Bellani F. The management of paratesticular rhabdomyosarcoma: A single institutional experience with 44 consecutive children. J Urol 1998;159:1031-4.
12. Newton WA Jr, Gehan EA, Webber BL, Marsden HB, van Unnik AJ, Hamoudi AB, *et al*. Classification of rhabdomyosarcomas and related sarcomas. Pathologic aspects and proposal for a new classification: An Intergroup Rhabdomyosarcoma Study. Cancer 1995;76:1073-85.

Cite this article as: Vani BR, Geethamala K, Murthy VS, Thejaswini MU, Padmaja KP. Paratesticular embyronal rhabdomyosarcoma in an adolescent: A rare case report. Clin Cancer Investig J 2014;3:554-7

Source of Support: Nil, **Conflict of Interest:** None declared.