Case Report

Advanced and recurrent testicular rhabdomyosarcoma in a young adult: A rare case report

L. Purnima Devi, Ritesh Kumar, Yookarin Khonglah[1], Akash Handique[2]

Departments of Radiotherapy and Oncology, [1]Pathology and [2]Radiodiagnosis, North East Indira Gandhi Regional Institute of Health and Medical Sciences, Shillong, Meghalaya, India

ABSTRACT

Rhabdomyosarcoma (RMS) is one of the most frequent soft tissue sarcomas. Pure testicular RMS is a very rare tumor and a few cases have been reported in the literature. We report a 16-year-old male patient with a painless right testicular swelling who underwent high inguinal orchiectomy and diagnosed as testicular embryonal RMS. The patient had a rare recurrence at the scrotal site with inguinal and retroperitoneal metastasis.

Key words: Chemotherapy, rhabdomyosarcoma, testicular

INTRODUCTION

Embryonal rhabdomyosarcoma (RMS) of the testis is a rare and aggressive tumor of children and young adult.[1] It develops from the mesenchymal tissue of the spermatic cord, epididymis, or tunica vaginalis. The testicular RMS accounts for 10% of testicular tumor in children. We report a case of embryonal RMS of the testis with recurrence at the scrotal site with inguinal and retroperitoneal metastasis.

CASE REPORT

A 16-year-old male had a swelling in the right scrotum for 2 months, for which he underwent right high inguinal orchectomy. Histopathological examination showed small ovoid to spindle tumor cells with vesicular to hyperchromatic nuclei arranged in sheets [Figure 1]. Mitotic figures were evident with occasional strap cells having cross striations. A diagnosis of testicular embryonal RMS was made.

Access this article online	
Quick Response Code:	Website: www.ccij-online.org
	DOI: 10.4103/2278-0513.151943

The patient reported to the Oncology Department after 2 months with recurrent swelling at the scrotum and pain in right lower abdomen. Clinical examination revealed a right irregular scrotal mass measuring 6 cm × 5 cm with ipsilateral inguinal lymphadenopathy extending up to the right iliac fossa. Ultrasonography of the whole abdomen and scrotum shows large heterogeneous lesion with necrotic areas noted in the right par aortic and bilateral iliac region, largest lesion measuring 9.7 cm × 7.5 cm. Serum alpha fetoprotein and beta human chorionic gonadotropin were within normal limits. Metastatic work up was done. Chest X-ray was within the normal level. Magnetic resonance image (MRI) of whole abdomen and pelvis revealed right testicular heterogenous mass measuring 4.2 cm × 3.5 cm × 2.8 cm with intraperitoneal extension and retroperitoneal lymph node displacing most of intra-abdominal space, compressing and displacing the bowel loops and other intra-abdominal contents causing right sided mild hydronephrosis [Figure 2]. No evidence of hepatic or pulmonary metastasis was seen. The patient was staged as T2 N1 M0 with clinical group III. The patient received 6 cycles of systemic chemotherapy 3 weekly comprising of ifosfamide and epirubicin. The patient tolerated chemotherapy well. Postchemotherapy MRI abdomen and pelvis showed significant reduction in the size of T2 hyperintense heterogenous mass lesion with multiple lobulation in the pelvis in the recto-vesical pouch and the bladder dome and infiltrating the prostate with multiple inguinal nodes. The patient was planned for radiation but

Address for correspondence: Dr. Ritesh Kumar, Department of Radiotherapy and Oncology, North East Indira Gandhi Regional Institute of Health and Medical Sciences, Shillong, Meghalaya, India. E-mail: riteshkr9@gmail.com

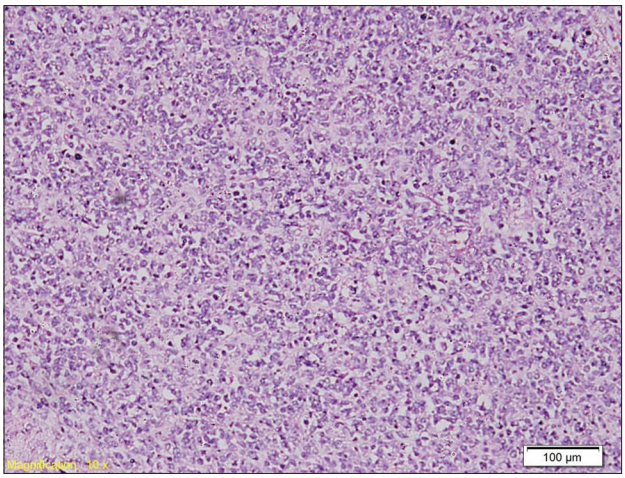

Figure 1: Photomicrograph showing a tumor composed of sheets and scattered round-to-oval cells with moderate pleomorphism, hyperchromatic nucleus and scanty eosinophilic cytoplasm in a background of collagenous stroma

lost to follow-up. Almost 5 months after patient reported to us again with progressive disease. The patient was planned for the second line palliative chemotherapy, followed by radiotherapy (RT) to the residual disease.

DISCUSSION

Rhabdomyosarcoma is the most common type of sarcoma of children. A testicular localization is rare and represents only 7% of RMS. Testicular RMS is a rare aggressive tumor manifesting in children and very young adults.[1] The peak incidence is between 1 and 5 years of age and is rare in adolescents and young adults.[2] The index case presented at an age of 16 years. The tumor derives from mesenchymal elements of the testis envelope, epididymis, and spermatic cord. The tumor manifests as a hard painless inguinoscrotal swelling, the size and duration of development are varied, and it rarely invades the scrotal skin. The mass might evolve within the external inguinal ring away from the scrotal contents.[3,4] Our patient presented with 2 months history of testicular swelling and a painless mass in the left scrotum.

Embryonal RMS is the predominant histological subtype in 90% of testicular RMS and has a good prognosis.[5] Myogenin and MyoD1 are important immunohistochemical (IHC) markers which aid in the confirmation of RMS histology. These IHC markers could not be done in the index case due to unavailability in the institute. RMS is regarded as a highly malignant tumor with frequent recurrence. Spread of the tumor is mostly by lymphatics to the iliac and para-aortic nodes in approximately one-third of cases with testicular lesions.[6] The index case had both iliac and para-aortic lymph node metastasis at presentation. Hematogenous metastases to lungs, liver, bone and bone marrow are present in 20% of patients at initial presentation.[7]

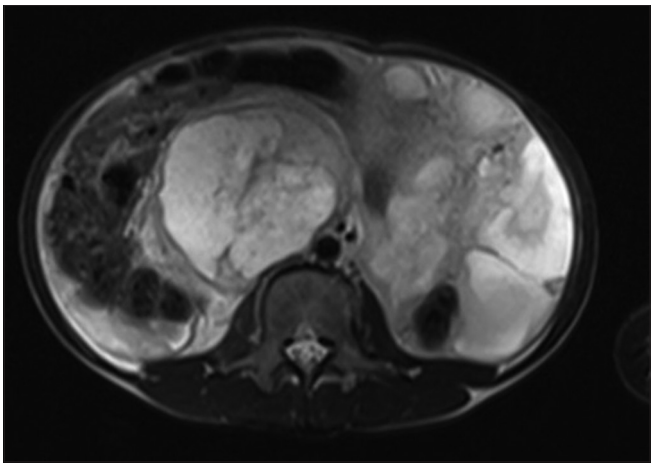

Figure 2: T1-weighted magnetic resonance image of abdomen showing large retroperitoneal lymph node

The optimal management of paratesticular RMS remains unclear because of the rarity of the disease in adults. Treatment strategies reviewed in the literature include radical high inguinal orchidectomy, concurrent chemotherapy (CCT), RT and retroperitoneal lymph node dissection (RPLND).

There is a definitive role for adjuvant CCT in RMS. Ferrari *et al.* reported that CCT was effective to treat childhood RMS, in adjuvant setting.[5] Vincristine, dactinomycin, cyclophosphamide, adriamycin, epirubicin, ifosfamide, carboplatin and etoposide were used in different combinations, and with varying dose schedules, in the cited study. Advanced stage and lymph nodal involvement are indications of adjuvant CCT. About a third of patients with paratesticular sarcomas die from metastatic disease, thus highlighting the importance of CCT. The index case received systemic CCT in view of advanced stage and lymph nodal involvement. The combined modalities of surgery, chemotherapy, and radiation therapy have greatly improved the survival rate in testicular RMS in young adults without significant long-term complications.[8]

The role of RPLND still remains controversial. Ferrari *et al.* reported on 44 patients with testicular RMS who did not undergo RPLND.[9] The authors considered that RPLND was unnecessary for localized disease because of the sensitivity afforded by computed tomography, the low rate of the retroperitoneal recurrence, and the presumed efficacy of CCT in controlling of microscopic disease.

In a study evaluating RMS at all sites in 2600 patients, adults with RMS experienced significantly worse prognosis than the children.[10] In another study, in which adult RMS patients also fared more poorly than children, the 5-year adult progression-free survival and overall survival rates were 28% and 40%, respectively.[11] Important prognostic

factors include tumor histology, diameter, stage and location, patient age, response to CCT and metastases status.[12]

Testicular RMS is rare and aggressive neoplasm in young adults. Radical high inguinal orchidectomy is the primary treatment. These patients have frequent recurrences and metastases. Palliative and supportive approaches in the recurrent and metastatic setting improves the quality of life of these patients.

REFERENCES

1. Stewart LH, Lioe TF, Johnston SR. Thirty-year review of intrascrotal rhabdomyosarcoma. Br J Urol 1991;68:418-20.
2. Kumar R, Bharti S, Khosla D, Kapoor R. Long term survival in paratesticular rhabdomyosarcoma. Clin Cancer Invest J 2012;1:31-2.
3. Mason BJ, Kier R. Sonographic and MR imaging appearances of paratesticular rhabdomyosarcoma. AJR Am J Roentgenol 1998;171:523-4.
4. Trippitelli A, Rosi P, Selli C, Carini M, Turini D. Rhabdomyosarcoma of spermatic cord in adult. Urology 1982;19:533-5.
5. Ferrari A, Bisogno G, Casanova M, Meazza C, Piva L, Cecchetto G, *et al.* Paratesticular rhabdomyosarcoma: Report from the Italian and German Cooperative Group. J Clin Oncol 2002;20:449-55.
6. Shapiro E, Strother D. Pediatric genitourinary rhabdomyosarcoma. J Urol 1992;148:1761-8.
7. Demir A, Onol FF, Türkeri L. Paratesticular pleomorphic rhabdomyosarcoma in an adult. Int Urol Nephrol 2004;36:577-8.
8. 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.
9. 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.
10. Sultan I, Qaddoumi I, Yaser S, Rodriguez-Galindo C, Ferrari A. Comparing adult and pediatric rhabdomyosarcoma in the surveillance, epidemiology and end results program, 1973 to 2005: An analysis of 2,600 patients. J Clin Oncol 2009;27:3391-7.
11. Ferrari A, Dileo P, Casanova M, Bertulli R, Meazza C, Gandola L, *et al.* Rhabdomyosarcoma in adults. A retrospective analysis of 171 patients treated at a single institution. Cancer 2003;98:571-80.
12. Esnaola NF, Rubin BP, Baldini EH, Vasudevan N, Demetri GD, Fletcher CD, *et al.* Response to chemotherapy and predictors of survival in adult rhabdomyosarcoma. Ann Surg 2001;234:215-23.

Cite this article as: Devi LP, Kumar R, Khonglah Y, Handique A. Advanced and recurrent testicular rhabdomyosarcoma in a young adult: A rare case report. Clin Cancer Investig J 2015;4:428-30.

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