Paratesticular Rhabdomyosarcoma – Spindle Cell Variant: Case Report of a Rare Tumor

Abstract

Testicular and paratesticular rhabdomyosarcomas (RMSs) in children are uncommon tumors. Histologically though embryonal RMS is common, the spindle cell variant is considered rare. Paratesticular RMS presents in children and adolescents with a unilateral, painless scrotal swelling or mass above the testis. We report the case of a 15-year-old boy, with a left paratesticular mass who underwent high inguinal orchidectomy. Histopathological examination of the specimen demonstrated spindle cell RMS (SC-RMS). Because of its morphological resemblance to spindle cell neoplasms such as leiomyosarcomas and fibrosarcomas, SC-RMS may pose diagnostic difficulties for the pathologist. This problem can be overcome by a careful search for rhabdomyoblasts in sections and immunohistochemistry for myogenin. We are reporting this case as paratesticular RMS itself is uncommon, and the spindle cell variant of embryonal RMS is all the more rare. There are lacunae in our knowledge about their presentation, diagnosis, response to treatment, and cure.

Keywords: *Chemotherapy, embryonal rhabdomyosarcoma, immunohistochemistry, paratesticular, rare, rhabdomyosarcoma, spindle cell variant*

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Introduction

Rhabdomyosarcoma (RMS) is a highly malignant soft-tissue sarcoma that arises from mesoderm with an incidence of approximately 4.5 cases/1 million children/adolescents.[1] Paratesticular RMS in children is uncommon representing approximately 7% of all RMS.[2] The spindle cell variant of embryonal RMS is considered rare (only 3% of all RMS cases in the Intergroup Rhabdomyosarcoma Study [IRS]).[3]

The index case is a 15-year-old male who presented with a large mass in the left paratesticular region that had developed over 3 months and on evaluation was found to be paratesticular spindle cell RMS (SC-RMS). We report this case as paratesticular RMS itself is uncommon, and the spindle cell variant of embryonal RMS is all the more rare.

Case Report

A 15-year-old boy consulted surgical outpatient department in May 2016 for a painless left scrotal mass that had evolved over 3 months. His Eastern Cooperative Oncology Group score was 1. On local examination, a firm testicular mass was found. The systemic examination was normal. Ultrasonography showed heterogeneous mass lesion measuring 3.6 cm × 4.9 cm × 5.3 cm in the left scrotal sac inferior to the left testis. On magnetic resonance imaging (MRI) scrotum, he was found to have a paratesticular mass with altered signal intensity measuring 5.2 cm × 4.4 cm × 6.9 cm and subcentimetric pelvic and inguinal lymph nodes. There was no evidence of metastasis in the chest or abdomen. MRI image is shown in Figures 1 and 2.

The patient was taken up for high inguinal orchidectomy in June 2016. Histopathological report of the specimen revealed RMS, shown in Figures 3 and 4. On immunohistochemistry (IHC), tumor is desmin, vimentin, and smooth muscle actin positive, confirming the diagnosis of RMS, spindle cell variant.

Postoperative positron emission tomography computed tomography (PET–CT) in August 2016 demonstrated the presence of metabolically active left para-aortic lymph node measuring 3.6 cm × 3.6 cm

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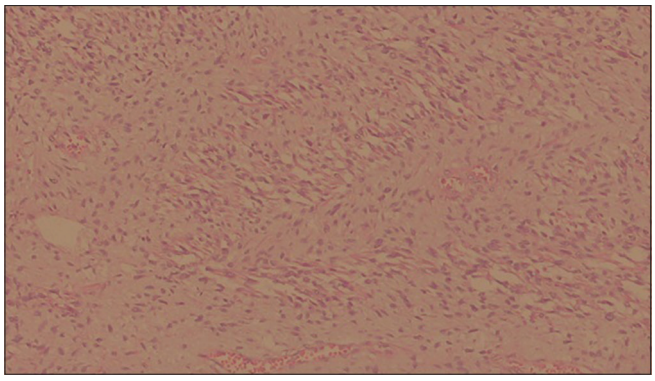

Figure 1: Microscopic view of tumor composed of spindle cells arranged in the form of sheets and fascicles showing mitosis and cellular atypia

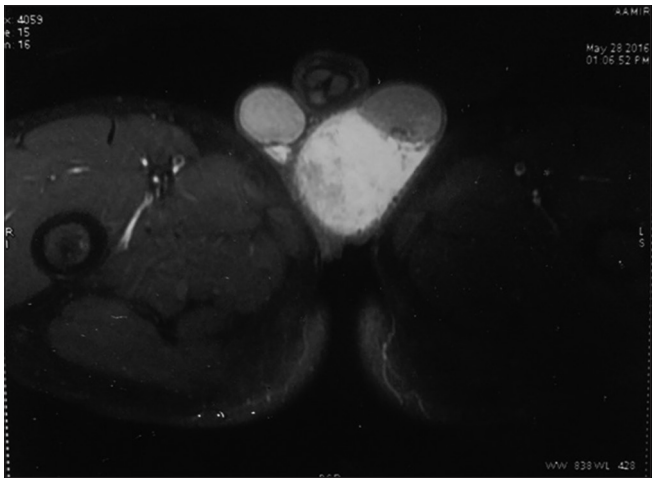

Figure 3: Preoperative magnetic resonance imaging image showing heterogeneously enhancing mass in the paratesticular region in axial view

at L2 vertebral level. The patient received six cycles of chemotherapy with ifosfamide 2.25 g and Adriamycin 90 mg, every 21 days at medical oncology department of our hospital. PET–CT for response evaluation at the end of six cycles of chemotherapy in February 2017 showed metabolic resolution of the left para-aortic lymph node with little change in size. The patient was then lost to follow-up and presented with pain abdomen 3½ months after completion of chemotherapy. PET–CT showed an fluorodeoxyglucose-avid large-conglomerated left para-aortic lymph nodal mass measuring 8.3 cm × 10.7 cm × 12.6 cm at the level of D12 to L3 vertebra suggestive of progressive disease. There was no other abnormal hypermetabolic focus elsewhere in the body. Difference between rhabdomyosarcoma at paratesticular and other locations is depicted in Tables 1 and 2.

The issue of disease progression was discussed in the multidisciplinary clinic, and a consensus was reached to start him on vincristine, actinomycin D, and cyclophosphamide as per the IRS-IV protocol at the department of radiation oncology. He received six cycles of chemotherapy, completing the same in December

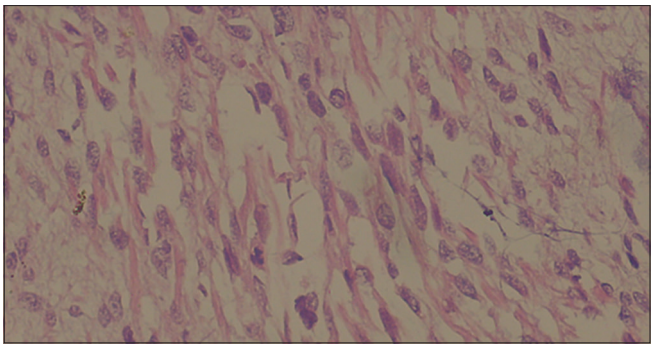

Figure 2: Microscopic view of tumor cells – high power

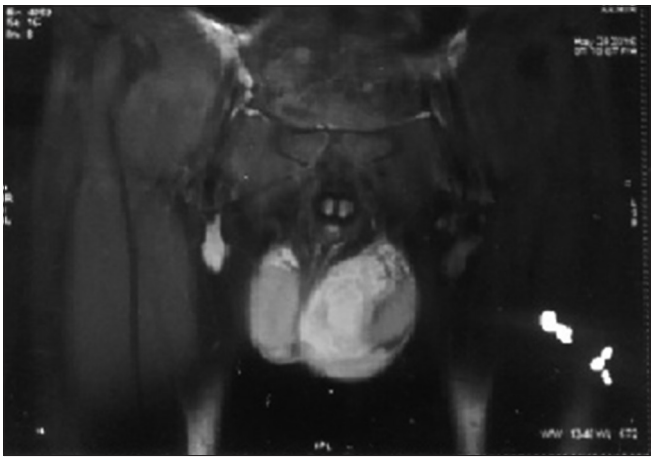

Figure 4: Preoperative magnetic resonance imaging image showing heterogeneously enhancing mass in the paratesticular region in coronal view

2017. However, further disease progression was detected in contrast-enhanced CT done in January 2018.

Discussion

Paratesticular RMS may arise anywhere along the spermatic cord, from the intrascrotal area through the inguinal canal. Most patients are <10 years of age at diagnosis, presenting with early-stage disease as a unilateral, painless scrotal swelling or mass above the testis. A local and systemic examination should be done to detect all possible disease sites. Differential diagnoses include testicular torsion, orchiepididymitis, scrotal abscess, and testicular tuberculosis. The tumor can be completely resected usually because of its superficial location and localized nature of the disease and hence has a good prognosis. With paratesticular lesions, about one-third of cases have lymph node metastasis.[4]

RMS has three histologic subtypes: embryonal, alveolar, and pleomorphic. The embryonal subtype, which is the most common, has three variants such as spindle cell, botryoid, and anaplastic.[5] The spindle cell variant was first recognized, in 1992 by German-Italian Cooperative Soft Tissue Sarcoma Study as a rare entity having a

Table 1: Depicting differences between para testicular rhabdomyosarcoma and rhabdomyosarcoma at other sites

	Paratesticular RMS*	Other RMS*
Incidence	7%	Orbit - 9% Other head and neck - 7% Para meningeal - 25% Genitourinary - 31% Extremity - 13% Trunk - 5% Retroperitoneum - 7% Other sites - 4%[10]
Site	Along spermatic cord, from intrascrotal area through the inguinal canal	As above
Prognosis by site	Unfavorable	Favorable Orbit Head and neck excluding Parameningeal Genitourinary excluding bladder and prostate Biliary tract Or unfavorable Bladder and prostate Extremity Head and neck parameningeal Others
Presentation	Mostly early stage	Early or late
Lymph node Involvement	33%	Head and neck - 15% Extremity - 24%[10]
Hematogenous metastasis	15%[10]	Common in orbit
Local evasion	Seen	Para meningeal - Base of skull erosion, cranial nerve palsy, and direct extension to CNS§

*RMS: Rhabdomyosarcoma, CNS§: Central nervous system

Table 2: Depicting treatment differences between paratesticular rhabdomyosarcoma and rhabdomyosarcoma at other sites

	Paratesticular RMS	Other RMS
Treatment	Inguinal orchidectomy With RPLND (International RMS Study Group) Without RPLND (European Investigators) Scrotal irradiation or hemiscrotectomy if scrotal irradiation	**Orbit** - Vincristine actinomycin D cyclophosphamide or vincristine cyclophosphamide with RT beginning between 3rd and 12th week **Head and neck** - Chemotherapy + CSI (50.4 Gy/28# for known CNS dissemination) No surgery indicated **Bladder and prostate** - Cisplatin + Adriamycin followed by RT after 6 weeks **Surgery** - Historically anterior pelvic exenteration **Gynecologic** - Surgery + Chemotherapy followed by RT if R1 or R2, RT and chemotherapy if lymphatics involved
RT dose	Total dose 50.4 Gy at 1.8 Gy per fraction, for R1 dose=41.4 Gy For R1 without lymph node involvement=36 Gy	**Orbit** - Total dose=45 Gy **Head and neck parameningeal sites** - CSI 50.4 Gy/28# **Other sites** - Total dose 50.4 Gy at 1.8 Gy per fraction, for R1 dose=41.4 Gy For R1 without lymph node involvement=36 Gy

RPLND: Retroperitoneal lymph node dissection, RMS: Rhabdomyosarcoma, CNS: Central nervous system, RT: Radiotherapy, CSI: Craniospinal irradiation

male predilection, propensity for occurrence in the paratesticular, and head, and neck regions, and a low malignant potential.[6]

Morphologically, SC-RMS has varied differential diagnosis including leiomyosarcoma, fibrosarcoma, malignant peripheral nerve sheath tumor, and malignant fibrous histiocytoma.[7] Considering the large number of entities included in the differential diagnosis of SC-RMS, IHC has an important role in its diagnosis. SC-RMS reacts consistently with myogenic markers such as desmin, myoglobin, MyoD1, and myogenin.[7]

Paratesticular SC-RMS is currently being treated with protocols similar to other RMS. This is because they are rare, and not enough research has been done on an alternative therapeutic approach. Radical orchidectomy by the inguinal route with spermatic cord ligation remains the essential act for histological diagnosis and constitutes the first step of treatment regardless of the stage of the disease. When there is nodal involvement, regional lymph node irradiation covering periaortic and ipsilateral iliac nodes is indicated.[8] For surgical violation of the scrotum or tumor extension to the structure, hemiscrotectomy, or less commonly scrotal irradiation is recommended. Chemotherapy should be routinely administered since the tumor is chemosensitive. This therapeutic approach consists of administrating actinomycin D, vincristine, and cyclophosphamide according to the IRS-IV protocol.[9]

Conclusion

Paratesticular SC-RMS is a rare tumor occurring in children and adolescents. Clinical setting and morphology should trigger appropriate immunohistochemical workup, to distinguish this entity from multiple other spindle cell neoplasms. It is possible to obtain successful results with a well-defined treatment protocol. More research should be carried out to get data about the presentation, diagnosis, tumor response, and treatment of these neoplasms.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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