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Paratesticular spindle cell rhabdomyosarcoma: A case report of a rare tumor

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ABSTRACT

Spindle cell rhabdomyosarcoma (RMS) is a rare variant of embryonal rhabdomyosarcoma. This tumor commonly involves paratesticular region of young males. Herein we report a case report of right paratesticular spindle cell rhabdomyosarcoma in a 16-year-old boy, who was referred to us for painless right scrotal mass. Staging computerized tomography (CT) showed no nodal or distant metastasis. The right radical inguinal orchidectomy was performed. Diagnosis of paratesticular spindle cell rhabdomyosarcoma was confirmed by histopathological and immunohistochemical examination. Paratesticular spindle cell RMS is a rare tumor and morphologically it may mimic other spindle cell neoplasms. Immunohistochemistry plays a pivotal role in its diagnosis. Making a correct diagnosis is crucial owing to the different treatment modalities available for this tumor

Key Words: Paratesticular, spindle cell, embryonal rhabdomyosarcoma, myogenin, actin, desmin.

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Introduction

Being a highly malignant soft-tissue sarcoma, rhabdomyosarcoma (RMS) has an incidence of approximately 4.5 cases per 1 million children and young adults. Paratesticular RMS among children and young adults are relatively rare, accounting 7% of all RMS. Spindle cell RMS are rare variant of embryonal RMS; only 3% of all RMS cases reported in Intergroup Rhabdomyosarcoma Study Group (IRSG) [1-5]. Treatment modality consists of surgical

resection of tumor, radiotherapy, and/or adjuvant chemotherapy. Prognosis of spindle cell RMS is relatively poor [5]. Herein, we report a case report of right paratesticular spindle cell rhabdomyosarcoma in a 16-year-old boy, who was referred to us for painless right scrotal mass, due to its rarity and features in its diagnosis.

Case report

A 16 years old Chinese Malaysian boy was referred from district hospital for one-month history of painless right scrotal swelling. He had no tuberculosis contact before. He is a Form 5 student with uneventful perinatal period. He was adhered to standard immunization schedule set by Ministry of

Health Malaysia. Other than well-controlled childhood bronchial asthma, he has no other co-morbid, past surgical history or family history of cancer.

On clinical examination, there was a right scrotal mass measuring about 8x6cm. It was hard in consistency, has a regular surface and well-defined margin, but it was not tethered to the scrotal skin. The left hemiscrotum was normal. Abdomen was unremarkable, and no inguinal lymphadenopathy was noted. There was no syndromic facies or obvious musculoskeletal deformities as well. Clinical diagnosis of primary right testicular tumor was made. Laboratory investigations revealed normal range of serum α -fetoprotein (AFP), lactate dehydrogenase (LDH) and β -human chorionic gonadotropin (bhCG). Ultrasound examination of scrotum and testes revealed a well-defined homogenous hypoechoic right scrotal mass measuring 3.9x3.7x4.5cm with positive color Doppler signal which suggestive of right testicular tumor. The thoraco-abdominopelvic computerized tomography scan did not reveal any nodal or distant metastases. Right radical orchidectomy was performed via inguinal approach.

On gross examination, the tumor was globular and well-encapsulated measuring 7.9x6.2x5cm. On cut section, it has homogenous tan brownish surface with minimal area of hemorrhage. The tumor was found adhering to the right testis which is unremarkable on cut section. Microscopic examination demonstrated a tumor composed of spindle, neoplastic cells with ovoid to elongated nuclei and pale, eosinophilic cytoplasm (Fig. 1a). Scattered rhabdomyoblasts are also seen. In some areas, neoplastic cells were arranged in microalveolar pattern with dense stromal sclerosis, imparting a pseudovascular appearance (Fig. 1b).

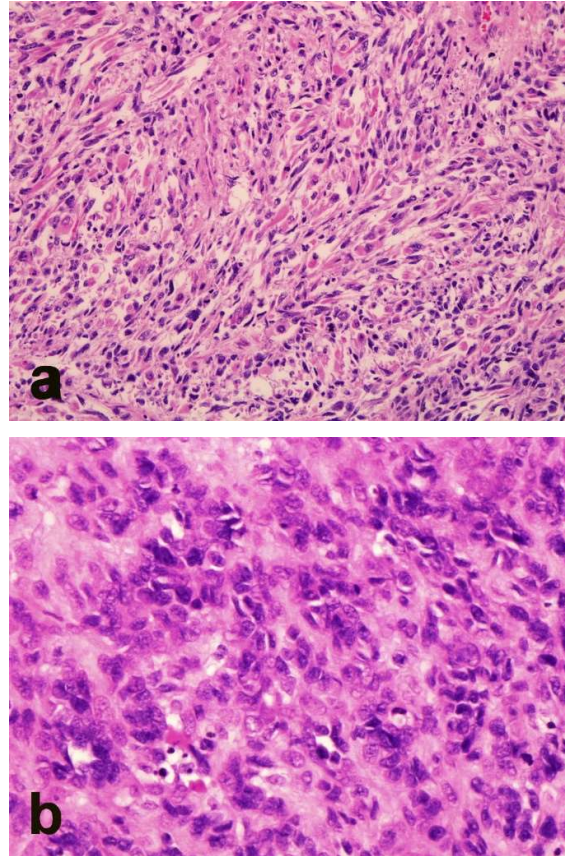


Fig. 1. Light microscopy shows (a) spindle cells in fascicles with occasional rhabdomyoblasts seen (H&E, 100x) and (b) sclerosing area of tumors imparting a pseudovascular appearance (H&E, 400x).

Mitoses were 10/10 hpf. The adjacent testicular parenchyma, rete testis and spermatic cord are unremarkable. On immunohistochemistry, the tumor cells are positive for Myogenin (Fig. 2a), Actin (Fig. 2b) and Desmin (Fig. 2c) and negative for Pancytokeratin, CD117, PLAP, SALL4, LCA and CD 20. Based on the fore mentioned histopathological features, diagnosis of right paratesticular RMS with spindle cell variant was made. The tumor was staged as stage 1 (T2 N0 M0) according to TNM staging system for RMS. His parents refused to subject him for adjuvant chemotherapy and he is still alive 6 months post-operatively.

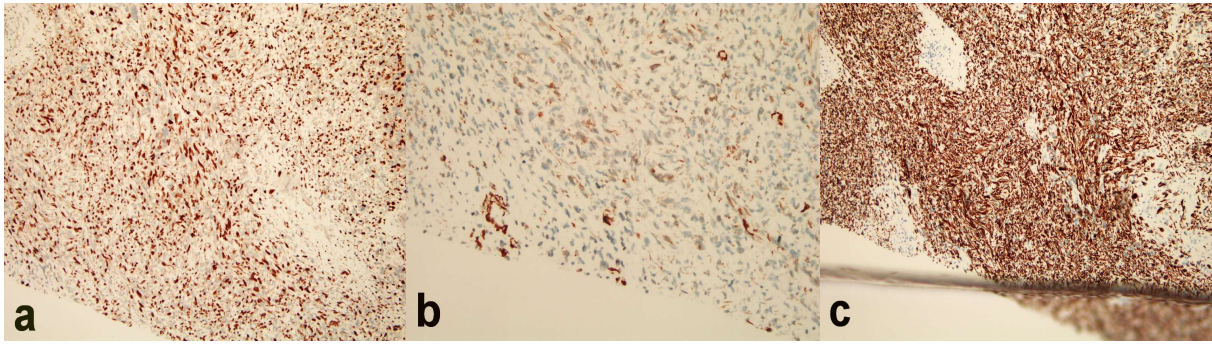


Fig. 2. On histochemistry, the tumour cells are positive for (a) Myogenin, (b) Actin and (c) Desmin.

Discussion

Being a highly malignant soft-tissue sarcoma, rhabdomyosarcoma (RMS) has an incidence of approximately 4.5 cases per 1 million children and young adults. Paratesticular RMS among children and young adults are relatively rare, accounting 7% of all RMS. Spindle cell RMS are rare variant of embryonal RMS; only 3% of all RMS cases reported in Intergroup Rhabdomyosarcoma Study Group (IRSG) [1]. Paratesticular spindle cell RMS has male preponderance [2,3] and is more common in children and adolescents with mean age of 7 years [4]. It is a rapid growing tumor but usually spares the scrotal skin [5]. Most cases of RMS are sporadic, nevertheless, there is approximately 7 to 8 percent of them may be associated with inherited syndrome such as neurofibromatosis, Li-Fraumeni syndrome, Beckwith-Wiedemann syndrome, and Costello syndrome [6-9].

On gross examination, it often displayed a white or tan whorled appearance occasionally accompanied by necrosis or cystic degeneration. On microscopic examination spindle cell RMS often shown fascicular elongated cells with central nuclei and eosinophilic fibrillary cytoplasm with a small proportion of rhabdomyoblasts [10]. Differential diagnoses of spindle cell RMS are leiomyosarcoma, myofibroblastic sarcoma,

fibrosarcoma, malignant peripheral nerve sheath tumor with rhabdomyoblastic differentiation and desmoplastic melanoma; owing to morphological similarities between these neoplasms. Of them, leiomyosarcoma is top in the list. Therefore, immunohistochemistry plays an important in making the accurate diagnosis and thence correct treatment. Spindle cell RMS consistently reacts with myogenic markers such as Desmin, Titin, Troponin D, Myoglobin, MyoD1, Myogenin and Actin [11]. Generally, RMS is staged by using Tumor, Node, and Metastasis (TNM) system. This system assigns one of four disease stages (from 1 through 4) based upon the site and size of the primary lesion, regional nodal involvement, and presence or absence of metastatic disease (Table 1). Favorable disease sites include the orbit and eyelid, other non-parameningeal head and neck locations, and non-bladder/prostate genitourinary tumors (e.g. paratesticular tumors). Extremities, bladder and prostate, cranial parameningeal sites, trunk and peritoneum, on the other hand, suggest unfavorable disease. In our case, it was staged as Stage 1 which points toward favorable disease.

Paratesticular RMS is currently being treated with protocols similar to other RMS because

Table 1. TNM Staging System for rhabdomyosarcoma.

Stage	Sites	T-invasiveness	T-size	N	M
1	Orbit, head and neck*, genitourinary [#]	T1 or T2	a or b	Any N	M0
2	Bladder/prostate, extremity, cranial parameningeal, other ^{\$}	T1 or T2	a	N0 or Nx	M0
3	Bladder/prostate, extremity, cranial parameningeal, other ^{\$}	T1 or T2	a b	N1 Any N	M0
4	All	T1 or T2	a or b	Any N	M1

T (tumour): T1, confined to anatomic site of origin; T2, extension; a, ≤ 5 cm; b > 5 cm in diameter. N (regional nodes): N0, not clinically involved; N1, clinically involved; Nx, clinical status unknown. M (metastases): M0, no distant metastases; M1, distant metastasis present. * Excluding parameningeal.

[#]Non-bladder/prostate. ^{\$}Includes trunk, retroperitoneum, etc.

they are rare and limited research has been done. Radical orchidectomy by the inguinal approach with spermatic cord ligation remains the vital act for histological diagnosis and constitutes the first step of treatment regardless of the stage of the disease [12]. Adjuvant chemotherapy should be administered since the tumor is chemosensitive. This therapeutic approach consists of giving actinomycin D, vincristine, and cyclophosphamide (IRS-IV protocol).

Conclusion

Paratesticular spindle cell RMS is a rare tumour occurring in children and adolescents. Morphologically it may mimic other spindle cell neoplasms. Thus immunohistochemistry plays a pivotal role in its diagnosis. Making a correct diagnosis is crucial owing to the different treatment modalities available for this tumour.

Compliance with ethical statements

Conflicts of Interest: None.

Financial disclosure: None.

Consent: Informed and written consent were taken from patient and her parents to publish this case report.

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