



Oncology

An Adult Case of Paratesticular Spindle Cell Rhabdomyosarcoma[☆]Joan C. Delto^a, George F. Wayne^c, Sofia Garces^b, Ana M. Medina^b, Alan M. Nieder^{a,*}^a Mount Sinai Medical Center Columbia University Division of Urology, Miami Beach, FL, USA^b Mount Sinai Medical Center, Department of Pathology, Miami Beach, FL, USA^c Florida International University, Herbert Wertheim College of Medicine, Miami, FL, USA

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ABSTRACT

Paratesticular rhabdomyosarcoma (RMS) occurs more frequently in children and is rare in adults. Embryonal RMS is the most common subtype of paratesticular RMS. Spindle cell is a rare variant of embryonal RMS and is associated with a favorable prognosis in children. Data in adults is lacking. We present a case of paratesticular RMS in a 24-year-old man.

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Case Presentation

A 24-year-old healthy man presented with a painless right scrotal mass for 3 weeks. He denied any difficulty with urination, dysuria, or hematuria. He denied any history of recent trauma, sexually transmitted diseases, or erectile dysfunction. His examination was significant for a nontender mobile epididymal mass measuring approximately 1 cm. He had no hernias. An ultrasonography examination revealed a small right epididymal tail mass with questionable vascularity. Bilateral testicles were otherwise normal. Given a low suspicion for malignancy, the patient was placed on surveillance. He returned for follow-up 3 months later, and the mass was found to be enlarged. A repeat ultrasonography demonstrated an enlarged right epididymal tail lesion with 2 new additional masses measuring up to 3 cm. These lesions were hypervascular and the testicles were without masses. Tumor markers were normal, and a computed tomography scan did not suggest any lymphadenopathy or metastatic lesions.

The patient elected to undergo right epididymal exploration via an inguinal approach and was consented for possible radical orchiectomy. On exploration, the masses were limited to the epididymal head. The diagnosis on frozen section was inconclusive, revealing a predominantly spindle cell neoplasm. The differential diagnosis included a sarcoma. The decision was made to preserve the testis and the remainder of the cord. The surgery was

uncomplicated. The final pathologic diagnosis was a paratesticular embryonal rhabdomyosarcoma, spindle cell type (Fig. 1A,B; Fig. 2). Immunohistochemical staining was positive for myogenin and desmin, indicating skeletal muscle differentiation (Fig. 1C,D). Staining was negative for Human Melanoma Black-45, smooth muscle actin, and S-100. Margins of resection were negative for malignancy.

Given his diagnosis of clinical T1N0M0 paratesticular rhabdomyosarcoma, spindle cell type, the patient was seen by a medical oncologist who recommended radical inguinal orchiectomy with ipsilateral nerve-sparing lymph node dissection followed by chemotherapy. The patient declined further surgery and was initiated on chemotherapy.

Discussion

Tumors in the paratesticular region are rare. They typically present as a rapidly growing and painless intrascrotal mass, eliciting only anatomically associated symptoms. Ultrasonography is the diagnostic imaging modality of choice.¹ The lesion is typically hypoechoic and may demonstrate increased vascularity on Doppler ultrasonography. The mass may be associated with a hydrocele, which may lead to a diagnosis of epididymitis rather than malignancy.¹ Most commonly found in the paratesticular region are benign solid masses (ie, adenomatoid tumors, leiomyomas) and cysts (ie, epididymal cysts, spermatocele). Adenomatoid tumors are round well-defined masses with variable echogenicity that do not demonstrate increased vascularity. Leiomyomas are solid and hypoechoic or heterogeneous masses that may have calcification.

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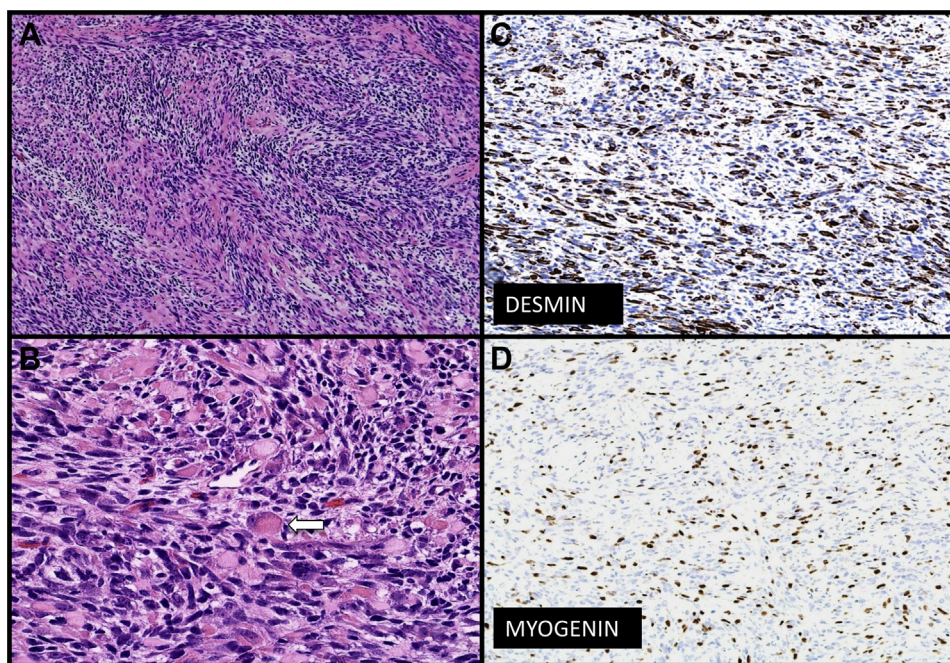


Figure 1. (A,B) Heterogeneous cellular proliferation including spindle and strap-like cells with rhabdomyoblastic differentiation (arrow). (C) Tumor cells strongly positive for desmin. (D) Tumor cells showing nuclear expression of myogenin.

An estimated 70% of paratesticular tumors are benign and 30% are malignant.² Sarcomas are the most common malignant neoplasms in the paratesticular area, and rhabdomyosarcomas (RMSs) represent 24% of adult sarcoma cases.² Of these RMS, 7%–10% involve the paratesticular region.³

Paratesticular RMS include several subtypes, namely embryonal, alveolar, and pleomorphic.⁴ Most cases are of embryonal histology. Spindle cell type is a rare variant of embryonal RMS, and was first documented in 1992 by Cavazzana.⁵ Adult spindle cell cases account for 3% of all RMSs.⁶ Notably, only a few case reports exist which describe a paratesticular lesion in an adult.

Gross dissection has often shown a white or tan whorled appearance occasionally accompanied by necrosis or cystic degeneration.⁴ Microscopically, spindle cell RMS often displays fascicular elongated cells with central nuclei and eosinophilic fibrillary cytoplasm with a small proportion of rhabdomyoblasts, featuring more eccentric nuclei, striations, and sharper eosinophilia. Interspersed collagen fibers have also been noted. The utility of frozen section to definitively diagnose these lesions is difficult as was noted in this case. Histologically, spindle cell rhabdomyosarcoma may be considered in stains positive for desmin and is virtually confirmed (specificity close to 100% and sensitivity ~95%) when

positive for myogenin and MyoD1–2 transcription factors involved in rhabdomyogenesis.⁴

Given a dearth of evidence and cases, treatment modalities have not been differentiated from those of other paratesticular RMSs.⁴ A diagnosis of paratesticular RMS of any variant typically warrants initial treatment with radical inguinal orchiectomy followed by systemic chemotherapy.⁷ Patients aged >10 years have been shown to have a higher risk of lymph node involvement.^{8,9} Dang⁹ reported that retroperitoneal lymph node dissection in these patients led to an improved 5-year survival from 64% to 86%. Radiation therapy for positive lymph nodes appears to improve the 5-year overall survival in older patients.^{8,9}

Generally, spindle cell–type paratesticular RMS is associated with a favorable prognosis, with 5-year survival rate of 95%.¹⁰ Additionally, lymph node metastasis is less common in the spindle cell variant compared with non–spindle cell variants (16% vs 36%).¹⁰

Conclusion

Paratesticular RMS is a very rare tumor with suspicious findings on ultrasonographic examination including solid masses with increasing size and vascularity. Urologists should have a high level

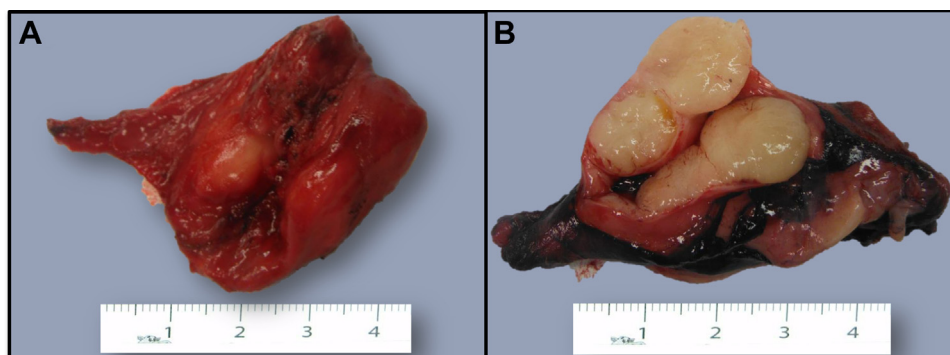


Figure 2. (A) Multinodular epididymal mass. (B) Three nodules with a rubbery, white-yellow cut surface.

of suspicion for these lesions and need to consider it in the differential diagnosis. Optimal treatment may be radical orchiectomy followed by adjuvant chemoradiation. Embryonal spindle cell–type RMS has an overall good prognosis. However, secondary to small number of cases, randomized prospective data is lacking.

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