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CASE REPORT

# Pediatric fibrous pseudotumor of the tunica vaginalis testis

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**Abstract** We describe a 16-year-old male with ultrasound evidence of a 1.3 cm right paratesticular nodule, which was managed by intraoperative frozen section and excisional biopsy. The pathologic findings were consistent with benign fibrous pseudotumor of the tunica vaginalis testis, which is a very rare lesion in the pediatric population. Consideration of fibrous pseudotumor in the differential diagnosis of pediatric paratesticular masses may help prevent unnecessarily aggressive therapy.


## 1. Introduction

Paratesticular fibrous pseudotumor is a rare, non-neoplastic, fibroproliferative lesion that arises most commonly from the tunica vaginalis, occasionally from the epididymis, and rarely from the spermatic cord and tunica albuginea [1]. It has been variably described in the literature as fibroma, nodular fibropseudotumor, inflammatory pseudotumor, fibrous mesothelioma, non-specific peritesticular fibrosis, nodular fibrous periorchitis, chronic proliferative periorchitis, reactive periorchitis, pseudofibrous periorchitis, and peritesticular fibromatosis. The wide variety of terms reflects its presentation, which is either that of a gray–white nodule (i.e., pseudotumor) or a thick, fibrotic band that encases the testis (i.e., periorchitis). Although fibrous pseudotumor is benign, it is clinically important because it may mimic malignant tumors, such as rhabdomyosarcoma, leiomyosarcoma, and desmoplastic small round cell tumor for which radical orchiectomy is indicated. Occurrence of this lesion in the pediatric population is exceedingly rare and it may not be considered in the clinical differential diagnosis, leading to unnecessary treatment. We describe a teenage patient with a fibrous pseudotumor of the tunica vaginalis testis.

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## 2. Case report

A 16-year-old male without significant medical history presented to hospital with a right testicular lump of 1-month duration. He played football in school, but did not report a history of testicular trauma. On physical examination, both testes were equal in size, but with an easily visualized and superficially palpable mass on the right side, concerning for a tumor. Bloodwork showed total human chorionic gonadotropin of less than 1 IU/L (reference: less than 2.5 IU/L) and $\alpha$-fetoprotein of 3.8 $\mu$g/L (reference: less than 5 $\mu$g/L). Ultrasound of the testes demonstrated a well-defined, oval, 0.8 cm $\times$ 1.2 cm $\times$ 1.3 cm soft tissue nodule (Fig. 1A, B) over the inferior surface of the right testicle. It was exophytic in relation to the adjacent right testicle and epididymal tail. The nodule appeared iso- to slightly hypoechoic compared to the adjacent testicle with internal vascularity (Fig. 1C) and areas of posterior sound attenuation. Acute angles were formed between the mass and testicle and vessels were identified traversing in between. A small amount of adjacent hydrocele containing low-level echoes was identified (Fig. 1D). No enlarged right inguinal nodes were found. The sonographic findings were suggestive of a tumor of tunical/epididymal origin, including adenomatoid tumor among others.

The presence of a paratesticular nodule was confirmed intraoperatively. Frozen section was interpreted as suggestive of a benign connective tissue lesion. The nodule was well-circumscribed from the adjacent testicular and paratesticular tissue and was excised with a 0.5 cm rim of tunica albuginea. On microscopic examination, the nodule was comprised of spindle cells, lymphoplasmacytic inflammation, and scattered thin-walled blood vessels in dense collagenous matrix with occasional less dense myxoid areas (Fig. 2). The spindle cells were focally clustered with oval nuclei, single nucleoli, and open chromatin. Mitotic activity and necrosis were inconspicuous. The lymphoplasmacytic infiltrate was most prominent around vessels. Occasional multinucleated plasma cells were identified and IgG4-positive plasma cells were present but rare (16 per 10 high-power fields) (Fig. 3A). The spindle cells were immunoreactive for cytokeratin AE1/AE3 (Fig. 3B), vimentin (Fig. 3C), Wilms tumor-1 (WT-1) (Fig. 3D), CD99 (cytoplasmic) (Fig. 3E) and CD31 (Fig. 3F). There was no immunoreactivity for anaplastic lymphoma kinase-1 (ALK-1), CD34, $\alpha$-smooth muscle actin ($\alpha$-SMA), desmin, and epithelial membrane antigen (EMA). The findings were in

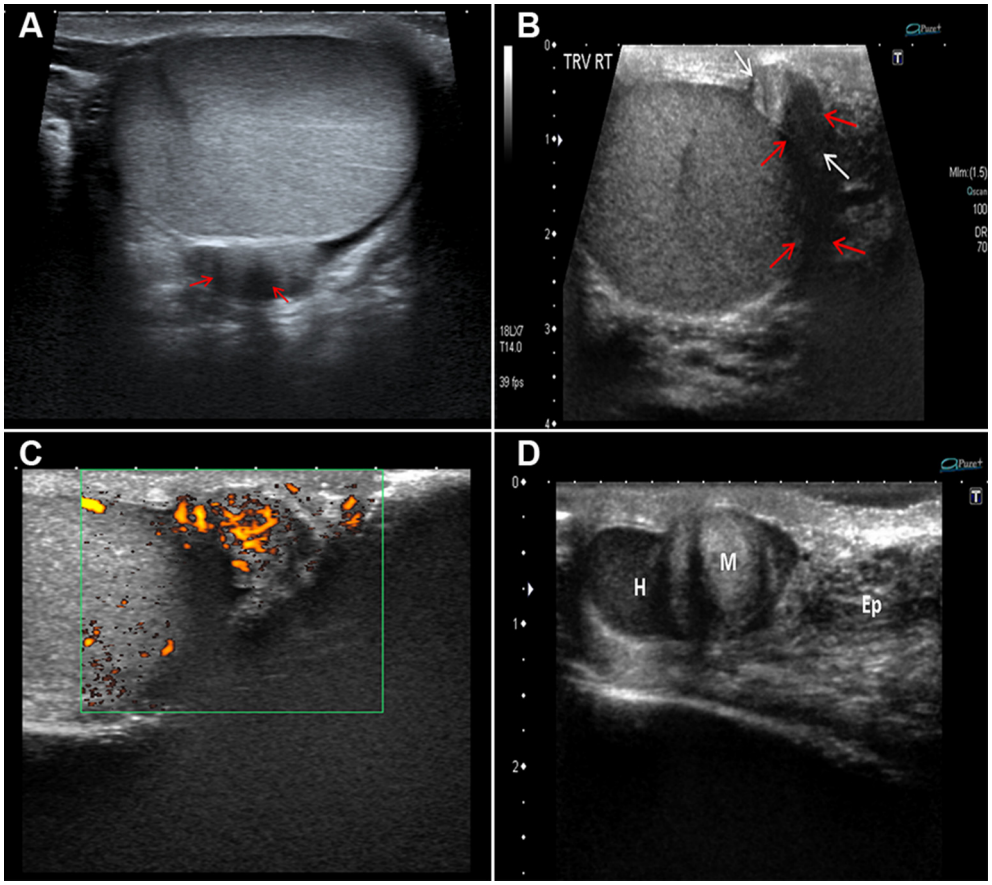


**Figure 1** (A) Transverse sonogram shows an oval, iso-hypoechoic soft tissue nodule posterior to the right testicle. The nodule has poorly-demarcated areas of distal shadowing in keeping with dense fibrous stromal component (red arrows). (B) The mass is well-demarcated from the testicle (white arrows). Shadowing fibrous component obscures part of the mass and testicle (red arrows). (C) Doppler demonstrates vascularity within the mass. (D) Paratesticular mass with whorled pattern (M), epididymal tail (Ep) and hematocele (H).