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.

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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 orchectomy 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.
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 α-fetoprotein of 3.8 μg/L (reference: less than 5 μg/L). Ultrasound of the testes demonstrated a well-defined, oval, 0.8 cm × 1.2 cm × 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, α-smooth muscle actin (α-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 hydrocele (H).
keeping with a fibrous pseudotumor. No evidence of recurrence was found at 2 months follow-up.

3. Discussion

Besides fibrous pseudotumor, the differential diagnosis of a paratesticular mass in children includes other benign lesions (such as lipoma, adenomatoid tumor, leiomyoma, hemangioma, and lymphangioma) and malignant lesions (most commonly rhabdomyosarcoma, rarely leiomyosarcoma and fibrosarcoma). Fibrous pseudotumor is rare, but represents the third most common tumor of the para
testicular tissues, after lipoma and adenomatoid tumor [2]. The etiology is unclear but is thought to be related to prior trauma, although some have recently included this entity in the spectrum of IgG4-related sclerosing disease [3]. It occurs most frequently in adults in the 3rd decade of life. It is very rare in the pediatric population, with only few cases described in the literature [4–11] including one of bilateral fibrous pseudotumors in an adolescent African American with a history of medulloblastoma and meningioma [11]. The clinical and pathologic features of pediatric fibrous pseudotumor overlap with those found in adult cases. Patients present with scrotal swelling that is painless or of mild discomfort. History of preceding trauma or infection is common, but not invariable. A single or sometimes multiple small nodules may be found on palpation. Transillumination is negative. Serum β-human chorionic gonadotropin and α-fetoprotein are within normal limits, which helps to differentiate this tumor from germ cell tumors.

Preoperative characterization of pediatric scrotal masses is best accomplished with ultrasound because it provides exceptional anatomic detail and distinguishes intratesticular from extratesticular lesions. In the diffuse type of fibrous pseudotumor, a band of tissue involving the tunica vaginalis surrounds the testis. Indentation of the tunica albuginea and displacement of the testis in the scrotal sac may be found. In the nodular type, one or more nodular masses are seen adjacent to the testis, which may be indented or partially obscured by the nodules. The nodules may appear well-margined, lobulated, or poorly-defined. They may appear hyperechoic or hypoechoic compared with the adjacent testicle, depending on the proportion of collagen, cells, and presence of calcification. Posterior acoustic shadowing can occur in the absence of calcification owing to dense stromal collagen. The nodules usually exhibit a small to moderate amount of vascularity by color flow Doppler, but may be avascular. Detachment of the nodules produces floaters or scrotal pearls in the tunical space [12]. Hydrocele is an associated finding in about 50% of cases. Occasionally, diffuse low-level echoes suggestive of hematocoele or proteinaceous debris may be seen. The tunica albuginea may appear normal or focally

Figure 2  (A) Well-circumscribed tumor (H&E, 40×). (B) Abundant collagen (Trichrome, 40×). (C) Spindle cells, blood vessels, lymphocytes, and plasma cells in dense collagenous stroma (H&E, 200×). (D) H&E, 400×.

Figure 3  (A) Occasional IgG4-positive plasma cells (200×). (B) AE1/AE3-positive spindle cells (200×). (C) Strong, diffuse vimentin-positive spindle cells (200×). (D) Nuclear WT1-positive cells (200×). (E) Cytoplasmic CD99-positive spindle cells (200×). (F) CD31-positive spindle cells (200×).
thickened. The underlying testicular parenchyma usually appears normal or with mass effect related to adjacent tumor. When ultrasound evaluation of scrotal lesions is inconclusive, magnetic resonance (MR) imaging may be useful as an adjunctive modality for further tissue characterization. Fibrous pseudotumor is expected to demonstrate low signal intensity on both T1- and T2-weighted images because of the presence of fibrosis [13].

Intraoperative consultation is helpful in guiding the most appropriate management at the time of surgery. However, definitive diagnosis of paratesticular fibrous pseudotumor at the time of frozen section is challenging and often only a descriptive diagnosis is rendered [14]. Historically, the diagnosis of fibrous pseudotumor was usually established after radical orchiectomy. However, the nodular type of fibrous pseudotumor is frequently amenable to excision with testicle sparing. Radical orchiectomy may be unavoidable for the diffuse type. Microscopic examination and immunophenotyping usually confirm the diagnosis, but genetic studies may be helpful in situations where spindle cell lesions with diagnostically-useful cytogenetic findings, such as inflammatory myofibroblastic tumor, desmoplastic small round cell tumor, and synovial sarcoma, are being considered in the differential diagnosis. Electron microscopy may also be diagnostically useful in establishing the fibroblastic/myofibroblastic nature of fibrous pseudotumor.

4. Conclusion

The urologist should be aware of the occurrence of paratesticular fibrous pseudotumors in pediatric patients as its consideration in the preoperative diagnosis may help avoid unnecessary radical orchiectomies.

Conflicts of interest

The authors declare no conflict of interest.

References

[1] Parker PM, Pugliese JM, Allen Jr RC. Benign fibrous pseudotumor of tunica vaginalis testis. Urology 2006;68:427.e17–e19.
[2] Germaine P, Simerman LP. Fibrous pseudotumor of the scrotum. J Ultrasound Med 2007;26:133–8.
[3] Bösmüller H, von Weyhern CH, Adam P, Alibegovic V, Mikuz G, Fend F. Paratesticular fibrous pseudotumor—a IgG4-related disorder? Virchows Arch 2011;458:109–13.
[4] Corcione N, Mancini P, Cecchi M, Pingitore R. Fibrous pseudotumor of tunica vaginalis. Report of a case. Pathologica 1988;80:723–7.
[5] Yates TS, Remmler-Fisch C, Smilow PC, Fleisher MH. Benign fibrous testicular pseudotumors in children. J Urol 1993;150:1886–8.
[6] Atahan O, Atahan S, Kayigil O, Metin A. Fibrous pseudotumour of tunica vaginalis testis in childhood. J Pediatr Surg 2001;36:1057–8.
[7] Zannoud M, Ghadouane M, Alami M, Benissa L, Amil T, Abbar M. Intrascrotal inflammatory pseudotumor (a case report). Ann Urol (Paris) 2002;36:322–5.
[8] Kern SQ, McMann LP. Bilateral fibrous pseudotumors of the tunica albuginea in a pediatric patient. J Pediatr Urol 2012;8:e1–3.
[9] Yang DM, Kim HC, Lim SJ. Sonographic findings of fibrous pseudotumor of the tunica vaginalis. J Clin Ultrasound 2012;40:252–4.
[10] Cassiday FH, Ishioka KM, McMahon CJ, Chu P, Sakamoto K, Lee KS, et al. MR imaging of scrotal tumors and pseudotumors. Radiographics 2010;30:665–83.
[11] Gordetsky J, Findes-Hosey J, Erturk E, Messing EM, Yao JL, Miyamoto H. Role of frozen section analysis of testicular/paratesticular fibrous pseudotumours: a five-case experience. Can Urol Assoc J 2011;5:E47–51.