

Paratesticular fibrous pseudotumor

Andrew A. Pridjian, MD, Adam Sharbaugh, BS, Amar Raval, MD,
David McGinnis, MD

Department of Urology, Sidney Kimmel Medical College - Thomas Jefferson University, Philadelphia, Pennsylvania, USA

PRIDJIAN AA, SHARBAUGH A, RAVAL A,
MCGINNIS D. Paratesticular fibrous pseudotumor.
Can J Urol 2017;24(1):8676-8678.

We present a rare case of fibrous pseudotumor of the tunica vaginalis. Discussion includes identification,

histopathologic findings and management. Proper understanding and preoperative identification of this benign disease allows for an organ-sparing surgical approach.

Key Words: fibrous pseudotumor, IgG4, testicular mass, benign

Introduction

Paratesticular fibrous pseudotumors are extremely rare fibroinflammatory growths with no metastatic potential. They usually present in the 3rd to 6th decade of life as unilateral painless scrotal nodule(s), often in the setting of trauma or infection. They are commonly found within testicular tunics and less often associated

with the epididymis and spermatic cord.^{1,2} Recent case reports have associated a subset of these tumors with IgG4 related systemic sclerosing disease.³

Case report

A 73-year-old male presented to our institution with progressive unilateral testicular discomfort and swelling. Examination revealed numerous non-tender nodules over the right testicle and epididymis. There was no inguinal lymphadenopathy and serum tumor markers were negative.

Scrotal ultrasound revealed multiple extratesticular hypoechoic mass-like lesions with peripheral

Accepted for publication December 2016

Address correspondence to Dr. Andrew A. Pridjian,
Department of Urology, Thomas Jefferson University, 1025
Walnut Street, Suite 1100, Philadelphia, PA 19107 USA

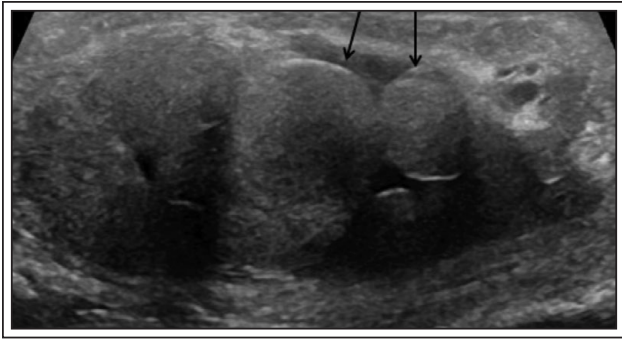


Figure 1. Scrotal ultrasound. Multiple extratesticular hypochoic mass-like lesions (arrows) with peripheral vascularity along the right epididymis.

vascularity along the right epididymis, measuring together at 4.4 cm x 2.3 cm x 2.7 cm. Bilateral testicles and the left epididymis were sonographically normal, Figure 1. Magnetic resonance imaging (MRI) further characterized these lesions as circumferential contrast-enhancing nodules arising from the tunica vaginalis with no regional lymphadenopathy, Figure 2.

We proceeded with inguinal exploration. After delivering the testis, we discovered over 40 fingerlike nodular projections dispersed throughout the tunica, Figure 3. Frozen section confirmed our suspicion for fibrous pseudotumor. We strongly considered resecting each lesion individually in order to spare the testis. However, due to the sheer volume of nodules and concern for recurrence due to incomplete resection,⁴ we proceeded with radical orchiectomy.

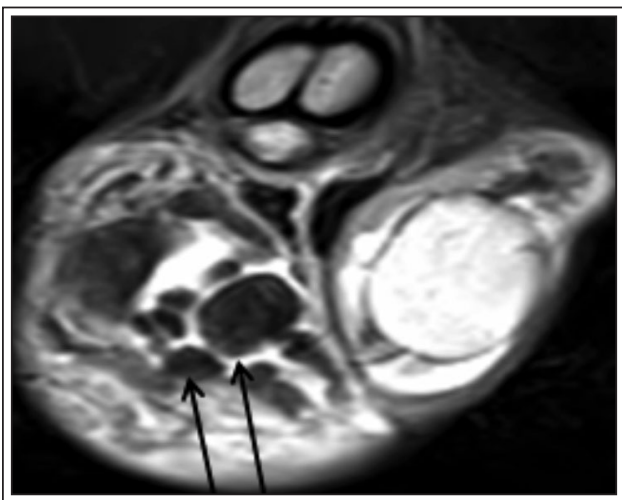


Figure 2. T1-weighted MR image. Contrast-enhancing nodules (arrows) found to be arising from the tunica vaginalis.



Figure 3. Gross specimen. Numerous pseudotumors seen intimately involved with the tunica vaginalis.

Histologic analysis revealed nodular fibrous thickening of the tunica vaginalis association with numerous plasma cells and lymphocytes. Of note, IgG4 plasma cells were focally increased with a IgG4/IgG ratio of nearly 50%.

Discussion

Fibrous pseudotumor of the paratesticular tissue is extremely rare. Over a 37 year period, the Canadian Reference Center for Cancer Pathology reported only 6 of 111 paratesticular tumors as fibrous pseudotumors.⁵ These pseudotumors usually present as one or more painless scrotal masses; they are not easily distinguishable from malignancy, often requiring surgical exploration with intraoperative frozen section. Organ-sparing local excision is the treatment of choice whenever possible.⁶

Scrotal ultrasound and MRI can help preoperatively identify these lesions. Sonographic appearance typically shows one or more solid paratesticular or tunical nodules or masses with variable echogenicity, at times with internal calcifications, and often associated with a hydrocele. MRI more specifically delineates the fibrosis in the nodules; as intermediate to low signal intensity on T1-weighted images, and low signal intensity on T2-weighted images. Lesion enhancement is slow but persistent, which is typical of fibrous tissue.⁷

Recent case reports have suspected paratesticular fibrous pseudotumors to be a manifestation of IgG4-related sclerosing disease.^{4,8} This is a newly recognized multi-organ system condition in which certain pathological features remain consistent across a wide range of organ systems. Diagnosis is made with characteristic histopathological findings and an elevated IgG4/IgG ratio of at least 40%. Follow up serum testing showing IgG4 > 135 mg/dL is 83% sensitive and 87% specific for diagnosis of IgG4-related systemic sclerosing

disease.⁹ Our patient's histologic IgG4/IgG ratio was nearly 50, making him a good candidate for serum IgG4 testing. Steroids (glucocorticoids) are typically the first line of therapy for systemic disease,¹⁰ however, this modality has yet to be explored as a treatment option for scrotal pseudotumors.

Radical orchiectomy is not the treatment of choice for this disease, and we highlight our surgical management of this case as an exception. There were several factors that swayed us against local excision – preoperative discussions with the patient, advanced age, extent of disease, and concern for recurrence due to incomplete resection. We still encourage organ-sparing local excision whenever possible. □

References

1. Mostofi FK, Price EB. Tumors of the male genital system. In: AFIP Fascicle 8;1973:151-154.
2. Ulbright TM, Amin MB, Young RH. Tumors of the testis, adnexa, spermatic cord, and scrotum. Atlas of tumor pathology. *Amer Registry Pathol* 1999;25:217-218.
3. Bösmüller H, von Weyhern CH, Adam P et al. Paratesticular fibrous pseudotumor—an IgG4-related disorder? *Virchows Arch* 2011;458(1):109-113.
4. Germaine P, Simerman LP. Fibrous pseudotumor of the scrotum. *J Ultrasound Med* 2007;26(1):133-138.
5. Strigley JR, Hartwick RW. Tumors and cysts of the paratesticular region. *Patho Annu* 1990;25(Pt 2):51-108.
6. Grebenc ML, Gorman JD, Sumida FK. Fibrous pseudotumor of the tunica vaginalis testis: imaging appearance. *Abdom Imaging* 1995;20(4):379-380.
7. Cassidy FH, Ishioka KM, McMagon CJ et al. MR Imaging of scrotal tumors and pseudotumors. *Radiographics* 2010;30(3):665-683.
8. Dieckmann KP, Strauss WJ, Frey U et al. Paratesticular fibrous pseudotumor in young males presenting with histological features of IgG4-related disease: two case reports. *J Med Case Rep* 2013;7:225.
9. Hao M, Liu M, Fan G et al. Diagnostic value of serum IgG4 for IgG4-related disease: a PRISMA-compliant systematic review and meta-analysis. *Medicine (Baltimore)* 2016;95(21):e3785.
10. Deshpande V, Zen Y, Chan JK et al. Consensus statement on the pathology of IgG4-related disease. *Mod Pathol* 2012;25(9):1181-1192.