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A Huge Testicular Fibrous Inflammatory Pseudotumor - A Rare Case Report and Literature Review

Chin Yiun Lee*, Syahril Anuar Salauddin, Hamid Ghazali

Department Of Urology, Hospital Tengku Ampuan Afzan (HTAA)

*Corresponding author: lcyiun@hotmail.com

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Abstract

Testicular fibrous inflammatory pseudotumors represent a rare pathological entity characterized by diagnostic challenges and histopathological complexity. These lesions, composed of myelofibroblastic spindle cells amidst a backdrop of chronic inflammation, pose difficulties in the pre-operative evaluation due to their resemblance to malignant testicular neoplasms. A middle-aged man, Mr. A, with a history of right inguinal hernioplasty, presented with a two-month history of progressively enlarging right scrotal swelling. Physical examination revealed a sizable, semi-solid testicular mass measuring 20x20 cm, with serum tumor markers within normal limits. Scrotal ultrasound depicted extensive cystic lesions with sedimentation, obscuring the normal testicular architecture. Subsequent thoracoabdominal pelvis (TAP) CT imaging delineated a right scrotal cystic lesion measuring 11x9 cm superiorly and 11x11 cm inferiorly, indicative of an encysted hydrocele with concurrent hydrocele of the right testis. Surgical intervention via high inguinal ligation orchidectomy revealed a complex histopathological composition comprising myelofibroblastic cells, fibrin, necrotic materials, and hematoma within the excised specimen. The anatomic location of paratesticular tumors, normal testicular tumor markers, elevated IgG4 levels, and ultrasound findings help distinguish benign pseudotumors from malignancies. Intraoperative frozen section biopsy is crucial before definitive testicular sparing surgery. Testicular sparing surgery emerges as the standard of care in managing testicular pseudotumors.

Keywords:

Fibrous Inflammatory Pseudotumor, Testicular Neoplasm, High Inguinal Orchidectomy, Testicular Sparing Surgery

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Introduction

Testicular pseudotumors pose a clinicopathological challenge as they are often misdiagnosed as malignant testicular tumors, despite radiological assessments, leading to unnecessary high inguinal orchidectomy[1]. Clinically, they commonly present as painless, rapidly growing scrotal masses. The term "fibrous inflammatory pseudotumor" was first described by Balloch et al. in 1904[2]. Pathologically, pseudotumors are characterized by fibroproliferative and chronic inflammation features. Due to their benign nature, they are not listed in the World Health Organization (WHO) classification of testicular tumours as of 2022. We aim to report a rare case of a large testicular pseudotumor and review the latest literature to enhance understanding and management of this condition.

Case Description:

Mr. A, a 42-year-old gentleman with no significant medical history, presented to the outpatient Urology clinic with right scrotal swelling persisting for 2 months. He noticed the swelling grew rapidly in size over time. Physical examination revealed a right semi-solid testicular swelling measuring 20x20 cm. His serum testicular tumor markers were within normal limits. Ultrasound of the scrotum showed large right cystic scrotal lesions with microcalcifications, obscuring the normal right testicular structure [Figure 1]. Further imaging with a thoracoabdominal pelvis (TAP) CT scan revealed a right scrotal cystic lesion measuring 11x9 cm for the superior pole with an enhancing cystic wall and 11x11 cm for the inferior pole [Figure 2]. He underwent a right high inguinal ligation orchidectomy. Intra-operatively, a large right testicular tumor weighing 2.8 kg was removed en bloc via high inguinal ligation technique. He recovered well and was discharged home on day 2 after the operation. Macroscopically, the surgical specimen showed two spherical cystic tumors attached, as depicted in [Figure 3]. Histologically, myelofibroblastic cells, fibrin, necrotic materials, and hematoma were observed, with no features of testicular neoplasm.

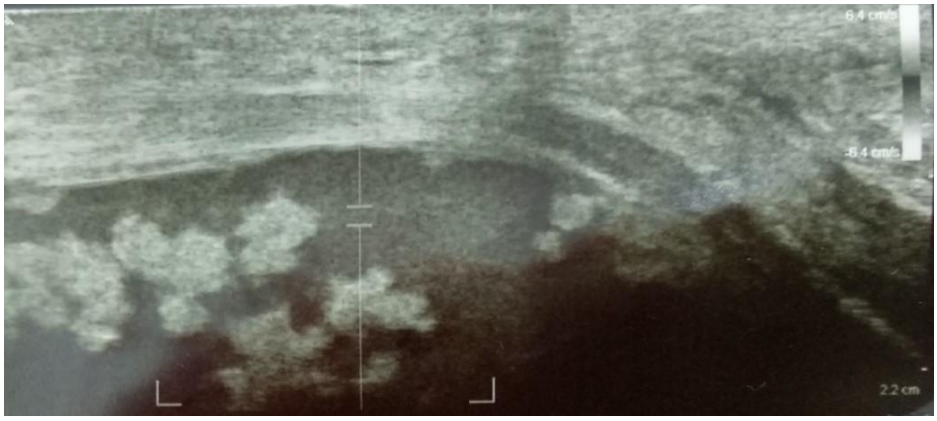

Figure 1: Ultrasound of the right intra-scrotal mass revealed microcalcifications.

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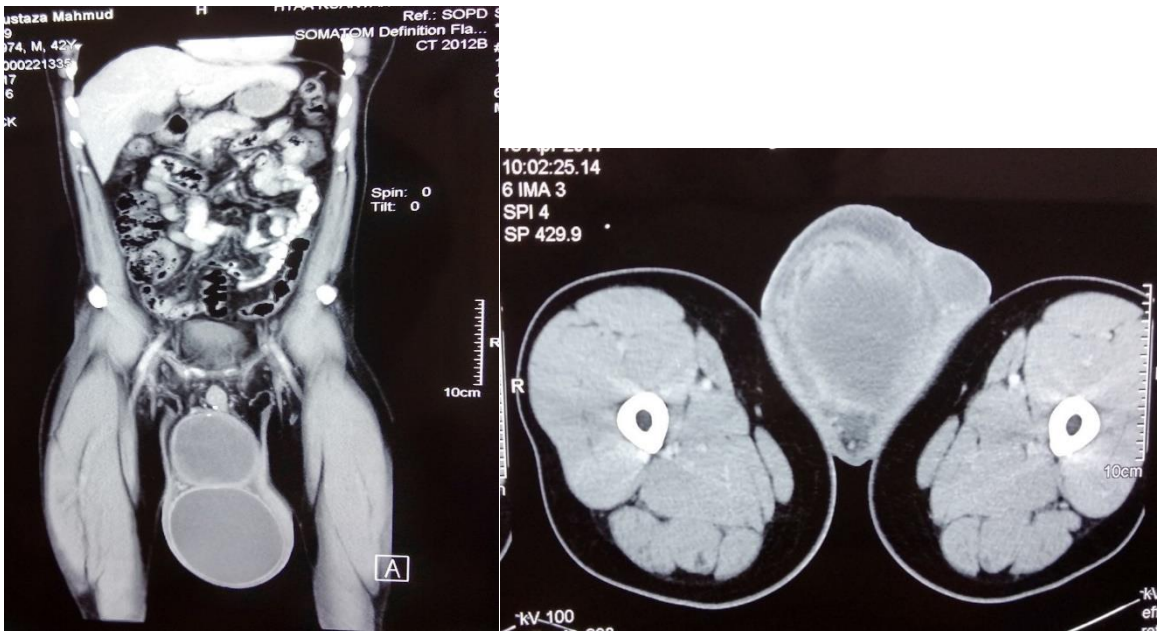

Figure 2: CT showed enhancing cystic wall and heterogeneity of the right testicular tumor.

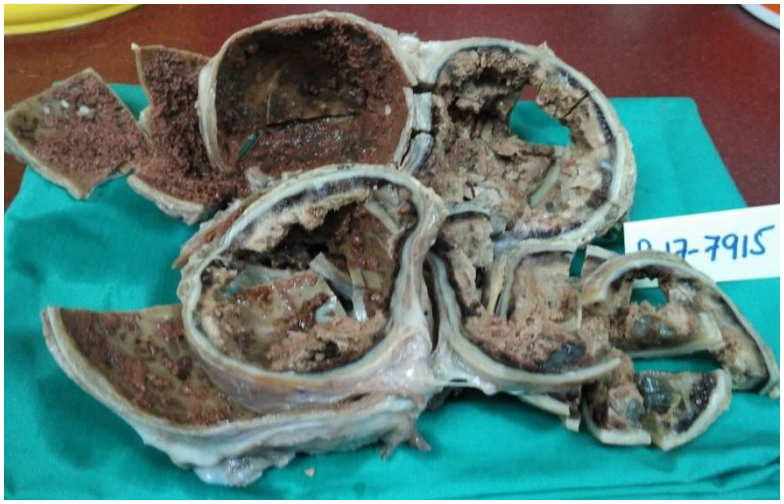

Figure 3: The right testicular pseudotumor comprised of hard and spherical wall.

Discussion

The reported incidence of fibrous inflammatory pseudotumor is estimated to be up to 200 cases to date. Fibrous inflammatory pseudotumors (FIP) of the testis present a diagnostic challenge due to their resemblance to malignant testicular tumors. Clinically, patients often present with painless, rapidly growing scrotal masses, which can be mistaken for testicular neoplasms. The lack of specific clinical features makes it difficult to differentiate pseudotumors from malignant tumors based on presentation alone. Moreover, the rarity of pseudotumors contributes to the challenge of diagnosing them accurately. As demonstrated in our case, the patient's scrotal swelling rapidly increased in size, prompting concern for malignancy and leading to surgical intervention.

The pathogenesis of fibrous inflammatory pseudotumors involves fibroproliferative and chronic inflammatory processes. Chronic inflammation leads to the accumulation of plasma cells and lymphocytic

cell infiltration within the testicular tissue, resulting in the formation of pseudotumors. Elevated levels of serum IgG4 have been proposed as a common pathogenic mechanism, suggesting a potential link to IgG4-related diseases[3]. Histologically, pseudotumors exhibit characteristic features such as bundles of spindle cells, fibrin, necrotic materials, and hematoma, often with prominent lymphocytic and plasma cell backgrounds. These histological features, although nonspecific, are helpful in distinguishing pseudotumors from malignant neoplasms.

Understanding the role of IgG4 in FIP may have implications for treatment strategies. While surgical excision is often necessary, particularly in cases where malignancy cannot be ruled out, consideration of immunomodulatory therapies targeting IgG4-related pathways should be explored in selected cases. Ongoing research is needed to elucidate the exact role of IgG4 in the pathogenesis of FIP and its significance in clinical management. Larger studies are required to establish the prevalence of the IgG4 immune-mediated mechanism in FIP and to evaluate the response to anti-inflammatory therapies.

Imaging modalities play a crucial role in the diagnosis and management of testicular pseudotumors. Ultrasonography is often the initial imaging modality used to evaluate scrotal masses, revealing hypoechoic lesions with microcalcifications, although these findings may not always be present. Computed tomography (CT) and magnetic resonance imaging (MRI) can provide additional information about the extent and characteristics of the lesion, aiding in surgical planning. However, distinguishing pseudotumors from malignant tumors based on imaging findings alone can be challenging, as demonstrated in our case. Surgical management typically involves radical orchiectomy, although testicular sparing surgery may be considered for small tumors or cases where pseudotumor is suspected preoperatively. Intraoperative frozen section biopsy can be valuable in confirming the diagnosis and guiding surgical decision-making.

In a case series by Jones et al, radical orchidectomy was performed in 8 out of 9 patients with fibrous inflammatory pseudotumor[4]. However, recent trends favour testicular sparing surgery for small tumors or cases suggestive of pseudotumor[5]. Testicular sparing surgery (TSS) is indicated for lesions that are suspected to be FIP based on imaging and biopsy findings, which do not show aggressive features suggestive of malignancy.

By offering testicular sparing surgery, we can preserve the testicular function of fertility and reduce the risk of hypogonadism. This technique also provides a more cosmetically favorable outcome compared to radical orchidectomy, particularly in younger patients concerned about body image.

In all testicular sparing surgery cases, intraoperative frozen section analysis should be performed to confirm benign pathology and ensure complete excision while ensuring a clear surgical margin. Minimally invasive techniques such as partial orchidectomy or enucleation of the lesion are employed to achieve good oncological safety and functional preservation. Unfortunately, radical inguinal orchidectomy was performed for our case given the absence of normal testis architecture on pre-operative radiological assessment.

Emerging evidence supports the feasibility and safety of TSS in selected cases of FIP, with studies reporting favorable outcomes in terms of oncological control and preservation of testicular function[6]. Long-term data on recurrence rates and functional outcomes are still evolving, emphasizing the need for continued research and prospective studies to validate the efficacy of TSS in managing FIP.

The prognosis of fibrous inflammatory pseudotumors of the testis is generally favorable, as these lesions are benign and do not metastasize. However, misdiagnosis and inappropriate surgical management can lead to unnecessary morbidity for patients. Future research directions may include further elucidating the pathogenesis of pseudotumors, exploring novel diagnostic modalities to improve preoperative diagnosis, and investigating the optimal surgical approach for managing these lesions. Collaboration among urologists, radiologists, and pathologists is essential to ensure accurate diagnosis and appropriate management of testicular pseudotumors.

Conclusion

The anatomic location of paratesticular tumors, normal testicular tumor markers, elevated IgG4 levels, and ultrasound findings help distinguish benign pseudotumors from malignancies. Intraoperative frozen section biopsy is crucial during definitive testicular sparing surgery. Morphological and immunohistochemical profiles aid in confirming the diagnosis of benign pseudotumors. Testicular sparing surgery emerges as the standard of care in managing testicular pseudotumors.

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Conflict of Interest

The authors declare that they have no conflict of interest.

Consent for Publication

The authors declare that they consented to the publication of this study.

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