Paratesticular fibrous pseudotumour: Intraoperative frozen section analysis can help prevent unnecessary orchiectomy

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Abstract

Paratesticular fibrous pseudotumours are rare intrascrotal lesions, most frequently affecting the testicular tunics. They are benign in nature; however, their pathogenesis is not completely understood. Presenting features are similar to testicular malignancy, which may result in unnecessary radical surgery. It has been suggested that additional diagnostic imaging combined with frozen section analysis may help prevent orchiectomy in these patients. We describe a case of paratesticular fibrous pseudotumour in a 40-year-old male treated with testicle-sparing surgery aided by intraoperative frozen section analysis.

Introduction

Paratesticular fibrous pseudotumours (PFPs) are rare, benign lesions believed to result from fibroinflammatory processes. They present most commonly as painless scrotal masses and occur as single or disseminated nodules. They frequently involve the tunica vaginalis, with rarer cases arising from the tunica albuginea, epididymis and spermatic cord.[1-6] Peak incidence occurs in the third decade of life, but reports exist in all age groups.[7] Although patients may report a history of testicular trauma, infection or hydrocele, the similarity in presentation to malignancy often results in radical surgery with diagnosis made on the resected specimen.[2-4]

Case report

Clinical findings

A 40-year-old man presented with a history of right hemiscrotal fullness and palpable nodules. The nodules grew tender over time. Physical examination revealed several small, right-sided scrotal lesions. Interpretation of scrotal ultrasound described microlithiases, and an enlarged, lobulated and echogenic right epididymis. There was one small epididymal cyst measuring 3 mm. The patient was booked for surgical exploration, which revealed solid nodules lining much of the tunica vaginalis. A specimen sent for frozen section revealed heavily collagenized, hypocellular tissue with chronic inflammatory infiltrate. No malignancy was identified. About one-third of the tunica vaginalis was removed and the remainder of the testicle was spared (Fig. 1).

Pathological findings

Gross examination revealed tan-grey tissue measuring 7.0 × 3.5 × 1.2 cm, with a roughened surface and opposing smooth, pearly grey tunica/mesothelial surface containing many polypoid projections (Fig. 2). Nodules measured up to 1.1 cm in greatest dimension. Microscopic examination revealed hypocellular tissue composed of thick collagen bundles with areas of hyalinization, thick and thin walled vessels and a background mixed inflammatory infiltrate consisting primarily of plasma cells with admixed lymphocytes and occasional mast cells (Fig. 3). There were occasional lymphoid aggregates and plump fibroblasts. Several vessels demonstrated concentric perivascular fibrosis. Immunohistochemistry revealed monokeratin- and vimentin-positive cells between the collagen bands. These cells also stained weakly for D2-40 and CD34, consistent with fibroblasts/myofibroblasts. LCA stain highlighted the background inflammatory cells and lymphoid aggregates. Vessel walls stained with CD31 and actin.

Discussion

Early reports of PFPs include publications by Cooper in 1830 and Balloch in 1904.[8,9] These lesions comprise about 6% of paratesticular masses, second to adenomatoid tumours.[1,10] Still, their relatively rare occurrence has meant that most available information comes from case reports and series.

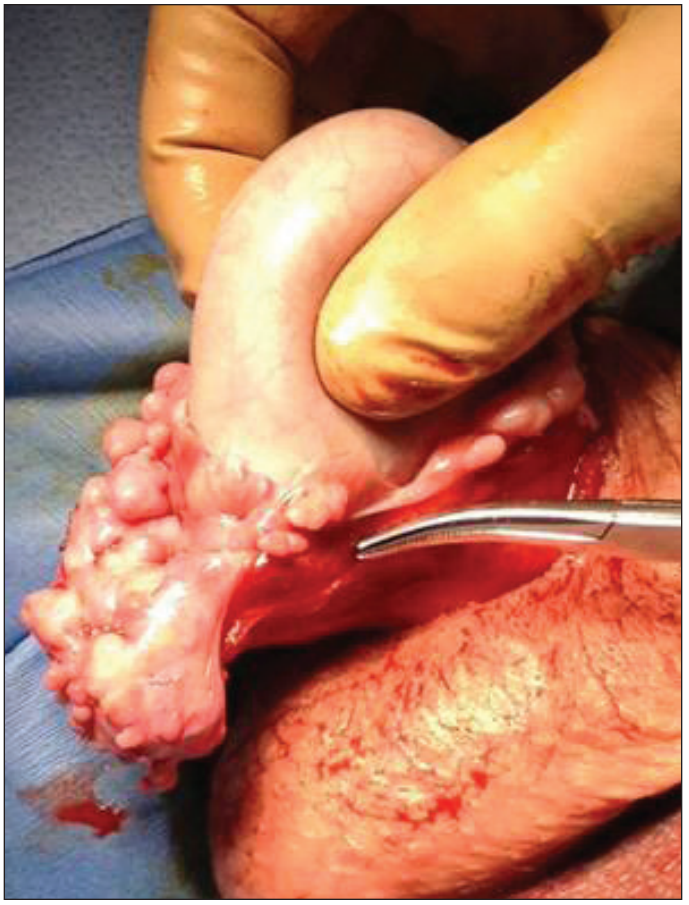

Fig. 1. Intraoperative photograph of prominent lobulated right epididymis.

Fig. 2. Gross photograph of excised polypoid right epididymis.

The pathogenesis of PFPs remains poorly understood and is controversial.[1,3,8] Mostafi and Price endorsed the term "fibrous pseudotumour" due to the belief that these lesions are the result of benign reactive processes in the testicular tunics. This description encompassed several variants and synonyms used to describe inflammatory and fibrotic paratesticular lesions,[2,11] and has since been included in most textbooks and guidelines.[6] Jones and colleagues, who studied a series of 9 cases, proposed further categorizing benign fibromatous testicular and paratesticular lesions into fibromas of testicular tunics or of gonadal stromal origin, angiomyofibroblastomas and fibroblastic/myofibroblastic pseudotumours.[12] Recent publications have questioned whether pseudotumours may belong to a growing list of IgG4-related sclerosing diseases, noting the presence of high numbers of IgG4-positive plasma cells.[5,6] Characteristics of these diseases include a high proportion of IgG4-positive plasma cells, predominantly T-lymphocytic infiltrate, storiform fibrosis, and venulitis.[13] These features were not seen in our case.

The macroscopic appearance of PFPs involves nodular, firm, often white, well-defined lesions.[2,8] In general, the lesions are comprised primarily of hyaline cartilage with collagen bundles, spindle cells, plasma cells, and lymphocytes;[8] however, they remain a pathological challenge due to histologic variability.[6] This variability may represent lesions at various stages, including early lesions with greater inflammatory infiltrate and myofibroblastic proliferation, as well as mature hypocellular lesions appearing more collagenized and nodular.[5] The cell of origin for these lesions is believed to be the fibroblast or myofibroblast.[14] Lesions tend to lack features suspicious for malignancy, such as increased mitotic activity, necrosis and pleomorphism. Based on a case series of 13 patients, Miyamota and colleagues have suggested subdividing the spectrum of histologic appearance into myofibroblastic, inflammatory sclerotic, and plaque-like.[3]

The differential diagnosis for the presentation of PFPs includes both testicular and paratesticular lesions. Testicular lesions to consider include malignant tumours, cysts, intratesticular vericocele, adrenal rest tumours and splenogonadal fusion. Benign paratesticular lesions include adenomatoid tumours, spermatoceles, cystadenomas, hydroceles, hernias, vericoceles, calculi, polyorchidism, neurofibroma, tunic fibroma, and leiomyoma, while malignant lesions include liposarcoma, rhabdomyosarcoma, leiomyosarcoma, mesothelioma, and papillary serous tumours.[2,15] These differential diagnoses may be distinguished based on a combination of clinical findings, gross and microscopic pathology, radiologic findings and immunohistochemistry. Fibrous pseudotumours are often positive for vimentin, smooth muscle actin and muscle-specific actin, and negative for S-100, keratin and desmin.[10,12] Focal discrepancies in these staining patterns may occur, and variability exists between lesions with significant inflammatory infiltrate versus relatively hypocellular lesions.[3]

Because a large proportion of palpable testicular masses are diagnosed as germ-cell malignancies, orchiectomy

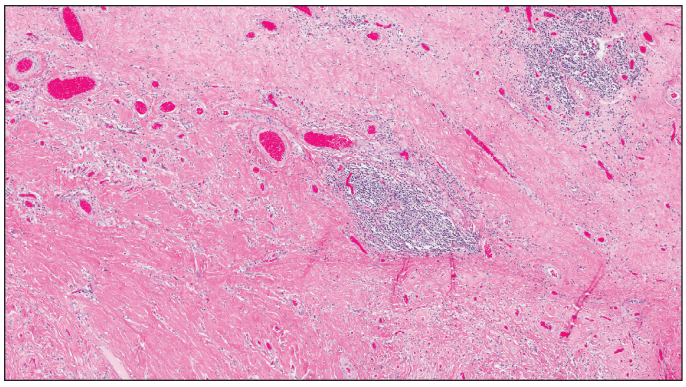

Fig. 3. Light microscopy of resected tissue showing thick collagen bundles with areas of hyalinization, thick- and thin-walled vessels, and background chronic inflammatory infiltrate (hematoxylin and eosin, 40× magnification).

remains the standard of care. With increasing use of ultrasound for infertility or trauma, the incidental detection of small masses is increasing. Fibrous pseudotumours may be hyper- or hypoechoic on ultrasound, and may be accompanied by microcalcifications. A hydrocele is present in nearly half of cases.[1,2] Magnetic resonance imaging has been suggested as a potentially useful imaging modality for PFP; however, data to support radiographic diagnosis is lacking.[6,8,16] Intra-operative frozen section analysis (FSA) may provide a means for testicle-sparing surgery in patients with PFP, although it may be difficult to definitively rule out malignancy with a limited sample.[4,16] In some cases, diffuse tunica involvement complicates surgery, which may preclude sparing the testicle.[8]

Conclusion

PFP is a rare, benign lesion with presentation similar to testicular malignancy. It most commonly affects the tunica vaginalis, and may occur as a single or multiple nodules. The histologic appearance can vary, which may be the result of disease progression/chronicity. Recent literature has revealed a potential association with IgG4-related diseases. Because of the presentation overlap with testicular malignancy, recent investigations have shown the potential for diagnostic imaging and FSA to allow for testicle-sparing surgery. However, more research is required. In our case, FSA allowed for testicle-sparing surgery, with PFP diagnosed later through microscopic and immunohistochemical investigation.

Competing interests: The authors declare no competing financial or personal interests.

This paper has been peer-reviewed.

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