Case Report

AN UNUSUAL ENCOUNTER OF PSEUDOTUMOUR OF PERITONEUM IN HERNIAL SAC- CASE REPORT

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ABSTRACT

A fibrous pseudotumor is a relatively rare nonneoplastic condition characterized by usually nodular, probably reactive proliferation of fibrous tissue and inflammatory cells, most often involving the lungs and liver, other sites have been reported with most common site as the tunica vaginalis. Although benign, this often clinically mimics testicular malignancy and is usually not diagnosed preoperatively, with radical orchidectomy often performed. We present a case of a fibrous pseudotumor involving the peritoneum in hernial sac.

CASE

A 65 yrs old male was admitted in MIMSR Medical Collegefor right sided inguinal hernia. He was investigated thoroughly and was advised surgery for the same.

He underwent a herniorapphy. After identifying and separating the sac, the sac was opened. An index finger was passed into the sac to reduce the contents; on reaching the peritoneal cavity at the neck of the sac, a hard structure was felt. It was slippery and difficult to catch hold of. So the incision was extended and surprisingly a whitish, rounded and glistening mass was taken out of the peritoneal cavity (Figure 1) The specimen was sent to histopathological examination. On gross the mass was of size 8x7x4 cm. The cut surface was greywhite firm (Figure 2). The microscopy revealed storiform pattern of spindle shaped cells (Figure 3) Hence we came to a surprising diagnosis of pseudotumour of peritoneum.

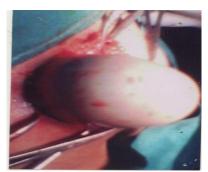


Figure 1: Intraoperative GlisteningMass



Figure 2: Gross cut section of tumor

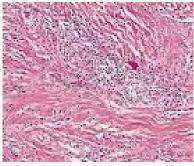


Figure 3: Charecteristic fibrous Pseudotumor

Postoperatively recovery of the patient was uneventful and patient was discharged on day 8th.Patient is regularly attending the follow-ups and at present having no complaints.

DISCUSSION

A fibrous pseudotumor of the scrotum is an uncommon lesion with an incompletely understood etiology. Since an early reference to a case of a peritesticular fibromatous mass by Sir Astley Cooper in 1830 and a case report by Balloch in 1904 various case reports and small series have been published in the urology, pathology, and radiology literature(Tobias-Machado,2000). Multiple other names have been applied to this and related entities, including chronic proliferative periorchitis, fibromatous periorchitis, pseudofibromatous periorchitis, reactive periorchitis, an inflammatory pseudotumor, nodular and diffuse fibrous proliferation of the tunica, a benign fibrous paratesticular tumor, granulomatous periorchitis,

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nonspecific peritesticular fibrosis, a nodular fibropseudotumor, and proliferative funiculitis, (Seethala, 2003) partly reflecting the variable and overlapping spectrum of pathologic findings and various etiologic theories. Although the terminology, classification, and proposed pathogenesis have been confusing and controversial, these lesions are generally accepted to represent a benign reactive proliferation of inflammatory and fibrous tissue, likely in response to trauma, surgery, infection, or inflammation, as distinct from true benign fibrous neoplasms, which may also rarely occur in the testis and testiculartunics (Jones, 1997). The pathology literature separates these reactive lesions into inflammatory and fibrous pseudotumors depending on the degree of cellularity, possibly representing early and later phases of a temporal spectrum of the same underlying process. Fibrous pseudotumors have been reported over a very wide age range, with the most common occurrence variably described as in the third decade or third through sixth decades, but is very rare before 18 years (Seethala 1997, Bostwick 1997, Mostifi 1973). Although uncommon, reactive fibrous proliferative lesions apparently represent the third most common benign tumor-forming paratesticular condition, after spermatic cord lipomas and epididymal adenomatoid tumors (Ulbright 1997, Mostifi 1973). Patients typically present with a painless lump of widely varying size. There is a reported association with a hydrocele in nearly 50% of cases and with prior trauma or epididymoorchitis in about 30%, (Akbar, 2003) leaving a majority of clinically idiopathic cases, as in our case. A rare or isolated association has also been reported with other conditions, including testicular infarction, Schistosoma haematobium infection, retroperitoneal fibrosis, and Gorlin (nevoid basal cell carcinoma) syndrome (Seethala, 2003). At least 2 cases of inflammatory pseudotumors have been reported in association with human immunodeficiency virus infection (Navai, 2005). Fibrous pseudotumors usually present as 1 or more (sometimes numerous) discrete or confluent hard unilateral extra testicular nodules or, less often, plaques, typically ranging from 0.5 to 8 cm, with 1 reported case measuring 25 cm (Al Otaibi, 1997). A less common form involves poorly defined fibrous lesions with sometimes diffuse involvement of the tunica, which may partly or completely encase the testis, for which some authors favor the term fibromatous periorchitis (Ulbright, 1997). About three fourths of cases involve the tunica vaginalis, with about 10% involving the epididymis and the remainder involving the spermatic cord or the tunica albuginea (which may not be possible to differentiate from tunica vaginalis involvement) (Akbar,2003). Inflammatory pseudotumors have been said to most commonly involve the spermatic cord.1Rarely, there is extension into the testis by an inflammatory pseudotumor or fibromatous periorchitis (Orosz, 1995). Microscopically, fibrous pseudo-tumors are composed of dense fibrous tissue consisting of hyalinized collagen and fibroblasts/myofibroblasts in varying proportions but usually paucicellular, sometimes with mixed inflammatory cells or granulation tissue, with orwithout varying amounts of calcification or even ossification. About half of spermatic cord lesions have been said to be histologically atypical (Mostofi, 1973). The differential diagnosis for a fibrous pseudotumorincludes fibrous mesothelioma (solitaryfibrous tumor), fibroma of the tunics, leiomyoma, neurofibroma, and idiopathic fibromatosis (Bostwick, 1997). Analysis of morphologic, histologic, and immunohistochemical features should allow differentiation of these entities in most cases. The sonographic appearance of fibrous pseudotumorsis widely variable, typically showing 1 or more solid paratesticular or tunica nodules or masses with variable echogenicity, with the characteristics depending on the amount of contributing fibrous and cellular tissue components, presence or absence of calcification, gross morphologic characteristics (e.g., single or multiplenodules, size, and confluence), and the structures involved (Dogra,2003). As noted above, ahydrocele is a frequent but nonspecific finding. Although case reports with MRI are limited, a more specific appearance of the nodular lesions on MRI has been suggested, with intermediate to low signal intensity on T1-weighted images, similar to the testis, and low signal intensity onT2-weighted images, secondary to the fibrous nature of these lesions, with little or no gadolinium enhancement (Woodward, 2003). Thus, MRI may be the preferred modality for the preoperative diagnosis and possibly also the follow-up of patients with fibrous pseudotumors. Most cases described in the literature report a benign course of this disease with cure after resection, which has tended to be radical orchidectomy, because the correct diagnosis has seldom been made preoperatively due to the nonspecific

International Journal of Basic and Applied Medical Sciences ISSN: 2277-2103 (Online) An Online International Journal Available at <u>http://www.cibtech.org/jms.htm</u> 2012 Vol. 2 (2) May-August, pp.132-135/Kale et al.

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clinical and sonographic findings. However, because this is a benign condition, complete local resection of the abnormal tissue is ideally the treatment of choice. Incomplete excision of the pseudotumor might be expected to result in eventual clinical recurrence if there is continued inflammatory or fibrous tissue proliferation, although a review of the English literature using Ovid and PubMed search engines (as of June 2006) found no reports of pseudotumor recurrence; 1 case report with pelvic recurrence was asserted to represent a scrotal desmoidtumor or aggressive fibromatosis rather than a fibrous pseudotumor (Lai, 1995). No metastatic potential has been reported with this entity. Because it is postulated that prior trauma or surgery may play a causative role in the development of a fibrous pseudotumor, it is tempting to speculate that less than complete resection could potentially initiate a vicious cycle, inducing reactive inflammation, which might incite or evolve into further inflammatory or fibrous pseudotumor formation. However, this remains to be shown on follow-up of cases treated with local resection. The inconclusive initial frozen section findings suggest the difficulty in establishing the correct diagnosis by that means. It is important to consider the possibility of a fibrous pseudotumor and related conditions in the differential diagnosis of sonographically detected paratesticular lesions, particularly if there is tunica vaginalis involvement and hydrocele and if a history of prior trauma or inflammation is available. This should prompt consideration of preoperative MRI, which may suggest the correct diagnosis in cases of fibrous pseudotumor. This may allow the surgeon to avoid unnecessary orchidectomy in at least some cases of this benign entity.

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International Journal of Basic and Applied Medical Sciences ISSN: 2277-2103 (Online) An Online International Journal Available at <u>http://www.cibtech.org/jms.htm</u> 2012 Vol. 2 (2) May-August, pp.132-135/Kale et al.

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