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FIBROUS PSEUDOTUMOR OF THE TUNICA VAGINALIS: CONTRAST ENHANCED SONOGRAPHY WITH PATHOLOGIC CORRELATION

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We report the case of a 28-year-old man who presented at the emergency department with recent left painful scrotal swelling, without history of genitourinary infection or trauma. On physical examination, left scrotal swelling with nodular palpation was noted. Contrast enhanced sonography demonstrated nodular vascularized thickening of the tunica vaginalis. Surgical exploration revealed multiples solid nodules of the vaginal wall, with, at frozen section analysis, fibroblastic tissue, vessels and chronic inflammation without malignity, suggestive of fibrous pseudotumor of the tunica vaginalis. We discuss the sonographic aspect of this rare entity and the difficulty to establish a diagnosis of benignity without surgical exploration.

Key-word: Scrotum, US.

Scrotal fibrous pseudotumor is a rare tumor characterized by benign reactive fibroinflammatory tissue involving the paratesticular space, most frequently the tunica vaginalis (76%), less often the epididymis (10%), the tunica albuginea or the spermatic cord (1).

Different sonographic aspects have been described, generally non-specific and insufficient to avoid surgical exploration, required to rule out malignant processes.

Case report

A 28-year-old man presented at the emergency department with recent left painful scrotal swelling. No history of previous scrotal infection or trauma was noted.

General physical examination and blood tests were normal (no inflammatory syndrome, C-reactive protein = 0,7 mg/dl). Scrotal examination performed by a senior urologist revealed left scrotal painful swelling with nodular palpation.

Scrotal ultrasound (US) performed with a 9 MHz linear probe (Acuson S2000™ system from Siemens Medical Solutions) showed a multinodular, mild echoic, thickening of the tunica vaginalis (1,6 mm) associated with moderate anechoic hydrocele and normal testicle (Fig. 1A). No color Doppler (CD) signal was observed within the nodules (Fig. 1B). Epididymis had a normal US aspect as the right scrotum. Contrast enhanced sonography (CEUS) of the left scrotal wall, obtained after bolus intravenous injection of 4 ml

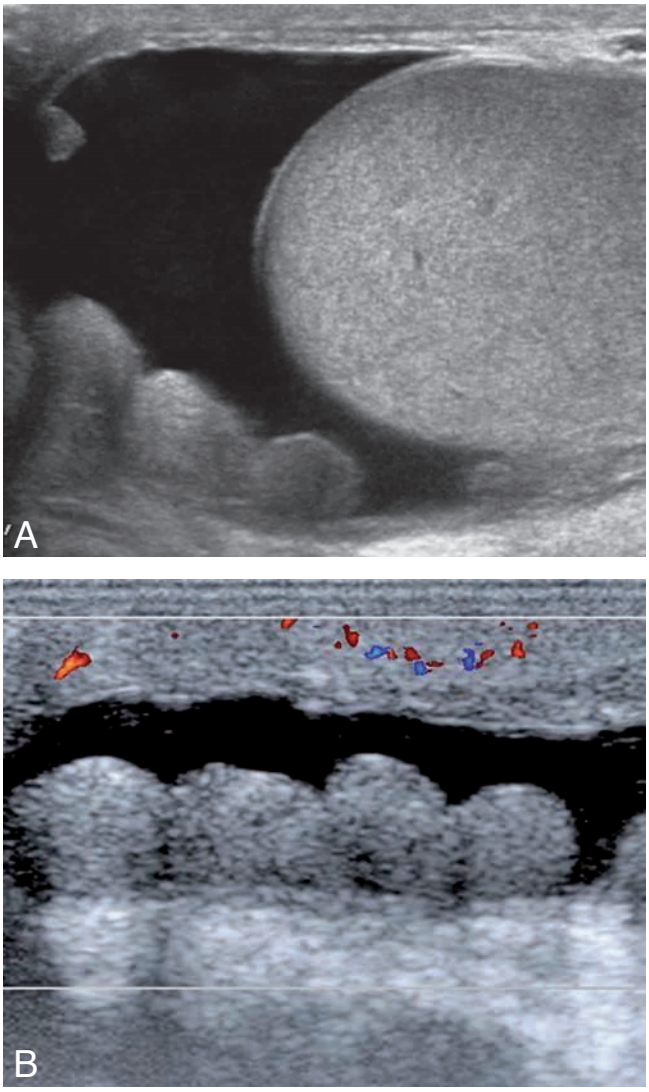

Fig. 1. — A. Bmode ultrasound of the left scrotum showing multinodular thickening of the tunica vaginalis, surrounded by anechoic hydrocèle and normal testicle. B. Color Doppler sonography of the nodules showing the absence of signal.

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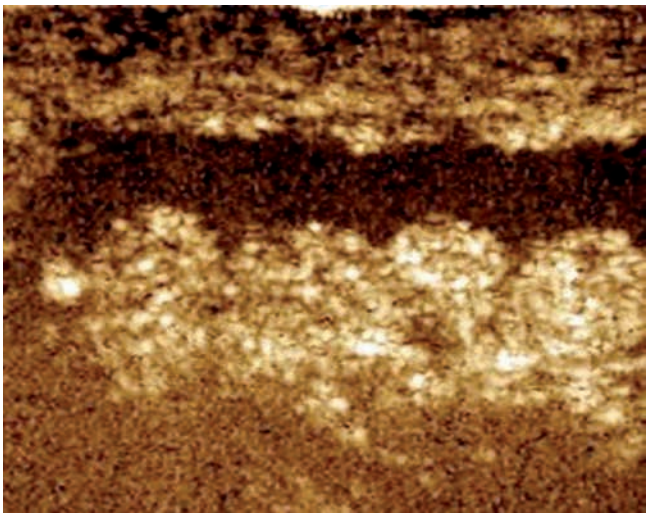

Fig. 2. — Contrast enhanced sonography showing homogeneous enhancement of the nodules.

Fig. 3. — Gross specimen before excision showing multiple solid nodules of the vaginal wall, arranged in bunch of grapes.

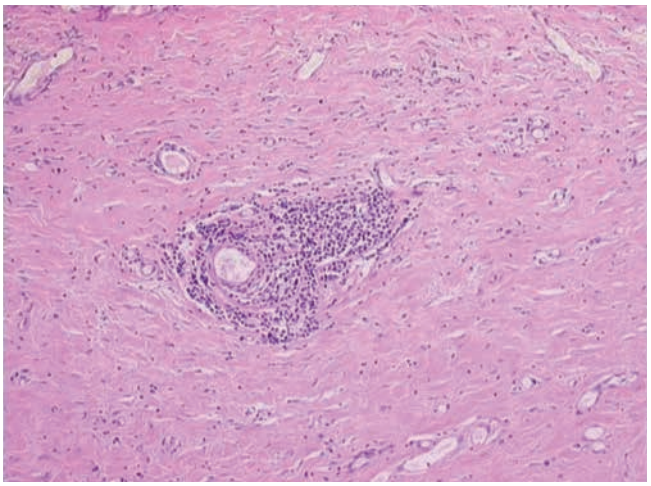

Fig. 4. — Microscopic section (hematoxylin and eosin x100) showing diffuse sclerous tissue with few fibroblast and cluster of inflammatory cells around a vessel.

contrast media (Sonovue®, Bracco), showed progressive, homogeneous, enhancement of the nodules, followed by a slow decrease of the signal intensity without washout aspect at the late time (Fig. 2).

Surgical exploration revealed multiple solid nodules of the vaginal wall, arranged in bunch of grapes (Fig. 3). A fragment of one nodule was submitted to frozen section analysis which showed fibroblastic tissue, vessels and chronic inflammation without malignity. All the visible nodules were removed by the surgeon without touching the testicle.

Diagnosis of fibrous pseudotumor was confirmed by histologic analysis showing normal mesothelial cells without atypia, proliferation of fibroblast cells mixed with collagen fibers surrounding vascular structures and focal inflammatory mononuclear cells (Fig. 4). Immunohistochemical staining on the spindle component was negative for calretinin, cytokeratin 5/6, cytokeratin AE1/AE3 and WT1.

Discussion

Reactive fibroinflammatory lesions of the paratesticular space, so called fibrous pseudotumor, is a rare entity, generally observed in the third decade (patients range from 16 to 75 years of age), very rarely in paediatric population (1, 2).

Fibrous pseudotumor is a generic term proposed by Mostophy that includes a series of different name like inflammatory pseudotumor, nodular and diffuse fibrous proliferation, proliferative funiculitis, chronic proliferative periorchitis, reactive periorchitis, fibromatous periorchitis, fibrous mesothelioma, benign fibrous paratesticular tumor, nonspecific peritesticular fibrosis (3).

The origin of this fibroinflammatory reaction is unknown and the pathogenesis remains obscure. It is frequently associated with history of prior infection or trauma but sometimes no apparent cause is found (3, 4). *Schistosomia haematobium* infection and HIV infection have also been associated with fibrous pseudotumor (5).

Macroscopically, nodules have gray-white and fibrogelatinous cut appearance with a whorled pattern (2).

Histologically, they contain hyalinized fibrous tissue with scattered collagen bundles and proliferative fibroblasts, some vascular channels and scattered inflammatory cells, rare focal calcification and ossification. Myxoid change may be present (2, 6).

Immunohistochemical staining of the fibrous pseudotumor is highly positive for vimentin, smooth muscle-specific actin, and common muscle actin and negative for calretinin, cytokeratin 5/6 and WT1, specific markers for mesothelioma. Necrosis and increased areas of mitosis and pleomorphism are absent (4).

Clinical examination (palpation) usually reveals single or multiple, painless, firm nodule, frequently associated with hydrocele or hematocele.

Sometimes, scrotal painful swelling is the only reason for consultation.

Different US aspects of scrotal fibrous pseudotumor have been described in the literature, depending on the localization.

The most frequent US aspect is, like in our case, multiple solid hyperechoic or hypoechoic small masses, attached to the inner part of the parietal layer of the tunica vaginalis, surrounded by hydrocele (7, 9, 10). The echogenicity of the nodules depends on the amount of collagen or fibroblast content. No spontaneous CD signal is visible in the nodules and testicle is normal. Sometimes, bigger solid heterogeneous masses unrelated to the epididymis are associated with shadowing and small amount of CD vascularity (8).

In our case, no CD signal was spontaneously visible in the nodules. Nevertheless, CEUS demonstrated a progressive, homogeneous, enhancement of the nodules that is consistent with the presence of vessels, followed by a slow decrease of the signal intensity, without washout aspect. This type of enhancement is highly suggestive of a benign enhancement. To our knowledge, this is the first description of such an enhancement.

Less frequently, fibrous pseudotumor of tunica vaginalis appears like a small solitary mass, containing eventual focal central or linear calcification, fixed or free within the vaginalis, also described then as scrotal pearl (2, 9, 10).

When the albuginea is involved (differentiating between albuginea and visceral layer of tunica vaginalis is impossible), a round, hypoechoic, mass of variable size, is attached to the testicle, simulating a testicular neoplasm (11). The extratesticular development of the mass within the vaginal cavity is typical with usual hydrocele.

One case of bilateral synchronous fibrous pseudotumor of the tunica albuginea has been described in a paediatric patient (12).

Diffuse fibrous pseudotumor, previously named nodular fibrous periorchitis or fibromatous periorchitis, is a very rare form of diffuse peritesticular thickening of the tunica vaginalis with focal linear calcifications (13).

Others very rare localization of fibrous pseudotumor have been described as hypoechoic mass in the epididymal tail or in paratesticular lymph nodes (14).

Sonographic differential diagnostic for fibrous pseudotumor of the paratesticular space includes, in the most frequent multinodular presentation, malignant mesothelioma, fibrosarcoma, leiomyosarcoma or rhabdomyosarcoma, in the solitary nodule presentation, fibrous mesothelioma, fibroma of the tunics, leiomyoma, neurofibroma, idiopathic fibromatosis and in the epididymal presentation, adenomatoid tumor or adenocarcinoma (8, 10, 14).

Few MRI descriptions of fibrous pseudotumor have been published-showing intermediate to low signal intensity on T1-weighted images, similar to the testis, low signal intensity on T2-weighted images and little or no gadolinium enhancement (15).

Despite the relative typical CD signal and MRI appearance of multinodular fibrous pseudotumor, imaging is generally not sufficient to affirm benignity and surgical exploration is required to rule out malignant processes. Only histological analysis with immunohistochemical staining may affirm the benign lesions and exclude malignancy.

However, because fibrous pseudotumor is a benign process, radical orchiectomy is not necessary and local excision of the mass is the treatment of choice for histologic study and correct diagnosis (7).

CEUS is an interesting new imaging modality which seems to give supplementary argument for the benign nature of the lesions. Unfortunately, for instance, no publication exists in the literature concerning the enhancement of scrotal fibrous pseudotumor and future CEUS explorations are needed to confirm this hypothesis.

In conclusion, we present CD signal and CEUS appearance of a rare paratesticular fibrous pseudotumor.

The radiologist should know this rare entity and its benign nature. Establish a correct preoperative diagnostic of benignity remains difficult but is important for correct surgical management.

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