Paratesticular well-differentiated liposarcoma initially diagnosed as fibrous pseudotumour

Svitlana Y. Bachurska, Petar A. Antonov¹, Ivan Y. Dechev¹, Antonio Lopez-Beltran²
Departments of General and Clinical Pathology and ¹Urology, Plovdiv Medical University, Plovdiv, Bulgaria, ²Department of Surgery, Unit of Anatomical Pathology, Faculty of Medicine, Cordoba, Spain

Address for correspondence:
Dr. Svitlana Y. Bachurska, Department of General and Clinical Pathology, Plovdiv Medical University, Plovdiv, Bulgaria.
E-mail: svitba@gmail.com

ABSTRACT

Paratesticular tumours are relatively rare and mostly of the mesenchymal origin. Due to its rarity, general surgical pathologists might have limited experience on the diagnostic entities and relevant differential diagnoses related to mesenchymal paratesticular tumours. This may likely cause diagnostic difficulties in a daily pathology practice. Paratesticular liposarcoma is a highly heterogeneous tumour and may be misdiagnosed as a benign fibromatous lesion. Herein we present a case of well-differentiated paratesticular liposarcoma of the sclerosing type initially diagnosed as a fibrous pseudotumour. Main differential diagnostic considerations are highlighted.

KEY WORDS: Fibrous pseudotumour, liposarcoma, paratesticular

INTRODUCTION

Paratesticular tumours are relatively rare and in the most cases are of a mesenchymal origin.[1] Often in a daily pathology practice mesenchymal tumours cause diagnostic difficulties regarding their biologic behaviour due to the wide variation of the histological features and overlapping clinical presentation.[2] Herein we present a case of paratesticular well-differentiated liposarcoma (sclerosing type) initially diagnosed as a fibrous pseudotumour. Relevant literature review and differential diagnosis considerations are included.

CASE REPORT

An 83-year-old male was admitted due to a painless mass above the left testis, which had a progressive, slow growth lasting 5 months. Serum tumour markers were negative; chest CT and X-ray study revealed no abnormalities; CT-scan of the scrotal zone showed testiciles and epididymis with homogenous structure and without abnormalities as well as heterogeneous mass above the left testicle with areas of calcification with diameter 65mm [Figure 1a]. The patient underwent radical orchectomy.

Pathology Findings: The encapsulated, well-defined soft mass measure 6 × 3 × 2cm in the area of spermatic cord was identified. The tumour was yellow on the cut surface with extremely heterogeneous structure presenting soft, gelatinous-like zones admixed with foci of bone-like structure [Figure 1b]. Histologically the tumour consisted of the dense fibrous tissue [Figure 1c], scattered lymphocytic infiltrates and areas of ossification [Figure 1d]. Cells which composed the tumour resembled well-differentiated fibrocytes and fibroblasts with some degree of the nuclear atypism. Immunohistochemical analysis demonstrated positive expression of actin, desmin and CD68. The patient was given a diagnosis of benign tumour-like condition: Fibrous pseudotumour.

Three months later he developed recurrence of the disease and underwent a second resection. Gross and microscopic features of the lesion were close to the initial one demonstrating fibrous tissue with ossification [Figure 2a]. Although histologically tumour demonstrated mostly well differentiated appearance with presence of mature adipocytes and fibroblasts, there were areas composed of vacuolated lipoblasts and atypical spindle cells embedded in a loose myxoid fibrous stroma [Figure 2b]. On a high magnification atypical mitoses could be seen [Figure 2c]. Therefore the slides were sent for a second opinion. S100 and MDM2 staining showed...
positive immunoreactivity in the suspicious cells (Figure 2d). CD34 and CDK4 were not performed. Final Diagnosis: Well-differentiated (low grade) liposarcoma (sclerosing type). The patient recovered without any complications. He did not receive any chemotherapy and radiotherapy following surgery due to concomitant illness-severe heart failure. The patient died due to a heart attack 26 month after initial surgery.

**DISCUSSION**

The paratesticular area represents a sophisticated union of diverse anatomical structures as testis, spermatic cord, testicular tunics, epididymis and its appendices. Histogenic origin of the components of this area has epithelial, mesothelial and mesenchymal character. Thus tumours arising in this region form a heterogeneous group with different behavioural and morphological patterns.[3]

Paratesticular tumours are relatively rare and account for 7% to 10% of all intrascrotal tumours. The majority of paratesticular tumours are benign (~70%). Among the remainder of ~30% of malignant tumours, the most frequently diagnosed are liposarcoma and leiomyosarcoma.[4] First case of paratesticular liposarcoma (PLS) was described in 1952 and since then around 100 cases were reported, representing 3% to 7% of all scrotal sarcomas.[3] Majority of the authors claimed that this neoplasm originates from spermatic cord tissue; however, others believe that it may developed based on the pre-existing benign lipomas.[5] Patients with PLS represent a broad age range (from 16 to 87).[3] Clinically, most recent reports of PLS showed slow-growing tumours, with a usually painless palpable scrotal or inguinal mass (sized between 3.5 to 20 cm in diameter) (Table 1).[6–11] In our case the patient is relatively older compared to the others, but with very similar clinical picture of slowly growing painless scrotal mass with diameter 6 cm. Due to

**Table 1: Recent studies that have reported paratesticular well-differentiated sarcoma**

| Author, reference | Year | n | Age | Size | Presentation | Treatment | Histology | Follow up |
|-------------------|------|---|-----|------|--------------|-----------|-----------|-----------|
| Montgomery and Fisher[6] | 2003 | 19 (33) | 63 | 3, 5-18 | Scrotal mass | Radical orchidectomy | 11-exclusively lipoma-like, 8 - varying sclerosing and inflammatory components | 6 cases with rec in 4 to 21 y 2 rec in 15 m |
| Küçük et al.[7] | 2013 | 1 (6) | 35 | 6 | Scrotal swelling | Radical orchidectomy | Well- differentiated Liposarcoma, sclerosing subtype | 18 m, no rec |
| Omidvari et al.[8] | 2014 | 1 | 55 | 4, 5 | Mass in left scrotum | Local excision | Lipoma, after 1 year rec - benign spindle cell tumor after 3 m - low grade malignant fibrous histiocytoma Radical orchidectomy - well- differentiated liposarcoma | 1 rec in 30 m |
| Gabriele et al.[9] | 2014 | 1 | 50 | 5 | Mass in right hemiscrotum | Local excision | Well- differentiated ‘lipoma-like’ Liposarcoma | 1 rec in 30 m |
| Vinayagam et al.[10] | 2014 | 1 | 50 | 20 | Swelling in the right hemiscrotum of 20 years | High orchidectomy | Well- differentiated Liposarcoma, sclerosing subtype | 2m, no rec |
| Pănuş et al.[11] | 2015 | 1 | 62 | 8 | Right scrotal mass | Radical right orchidectomy | Well- differentiated Liposarcoma | - |
| Our case | 2017 | 1 | 83 | 6 | Painless mass above the left testis | Radical orchidectomy | Well- differentiated Liposarcoma, sclerosing subtype | 1 rec in 26 m |
this clinical presentation, differential diagnosis should include a broad spectrum of benign conditions as scrotal lipoma, groin hernia, hydrocele and epididymitis.[9]

The World Health Organization (WHO) classification of the liposarcomas recognizes five categories of this malignancy: well differentiated, dedifferentiated, myxoid, round cell and pleomorphic. Most PLS represent well differentiated, low-grade malignancies with minimal tendency to metastasize but with some degree of local invasive potential.[10] Most of the published cases as well as the present case demonstrated recurrence of the disease in the period from 3 months to 21 years [Table 1].

Well differentiated PLS demonstrate mature and atypical lipoblasts with pleomorphic, hyperchromic nuclei, and variable areas of pleomorphic spindle cells and sclerosis.[9] Therefore, they are further subdivided into adipocytic (lipoma-like) liposarcoma, sclerosing liposarcoma, and into the two more rare types of inflammatory and spindle cell liposarcoma.[8] The sclerosing form is the more commonly seen in the retroperitoneum and spermatid cord.[10] Histological diversity may lead to the frequent misdiagnosing of these tumours as benign or tumour-like conditions (lipoma, leiomyoma, solitary fibrous tumour). Abundance of dense fibrous tissue, ossification and presence of the inflammatory cells in the initial histological picture of our case mask neoplasm as a benign tumour-like condition as fibromatosis or as in this case was the rationale for the diagnosis of fibrous pseudotumour. Additional difficulty in the morphological analysis of our case was scarce amount of atypical lipoblasts. In these controversial cases immunohistochemistry may be extremely helpful.

The other “hot” issue is differentiation of the low-grade sclerosing liposarcoma from the dedifferentiated liposarcoma which has worse prognosis and outcome. Histologically former presents as a paucicellular fibrotic lesion with single mitoses, whereas later appears as a densely cellular area with numerous mitoses resembling high grade mesenchymal tumours as malignant fibrous histiocytoma.[6] In present case the overall appearance is of well-differentiated fibrous, adipose and osseous tissue and the dedifferentiated areas occupied less than 5% of the tumour. These histologic characteristics support diagnosis of the well-differentiated (low grade) sclerosing type liposarcoma.

The important clinical and pathological findings which pathologists should be aware of, signing-out paratesticular tumours showing proliferation of fibrous tissue include the followings:
1. Fibrous pseudotumour is associated with hydrocele in 50% and involves tunica vaginalis in 75% of the cases[4]
2. Liposarcomas are usually presented as the largest masses (up to 1.2 kg)
3. The most frequent histological subtype of PLS is a sclerosing subtype[6]
4. Immunohistochemistry:
   - Fibrous pseudotumour is positive for vimentin and cytokeratin
   - Liposarcoma is positive for S-100 and CD34
5. Molecular assays for MDM2, p16 and CDK4 are also useful not only to define the histological subtype but also to outline the malignant character of the tumor.[3]

CONCLUSION
It is of a great value a wide sampling of paratesticular tumours and tumour-like conditions to avoid missing representative areas that might support the diagnosis. The use of molecular techniques and second opinion consultation should be of help in this type of uncommon paratesticular lesions.

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Conflicts of interest
There are no conflicts of interest.

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