Adult paratesticular leiomyosarcoma. A clinical and pathological case report

G. Pupca[1], L. Daminescu[1], S. David[1], Mihaela Iacob[2], V. Bucuraş[1]
[1] *Department of Urology*
[2] *Department of Pathology, Timisoara County Clinical Emergency Hospital, „Victor Babeş" University of Medicine and Pharmacy Timişoara*

Abstract

Purpose: Paratesticular leiomyosarcoma is a rare malignant tumor in an adult patient that must be known by the urologist in order to adopt the most appropriate treatment.
Materials and methods: We present the case of 71 years old man with a right malignant paratesticular tumor, without inguinal, pelvic or retroperitoneal adenopathy, diagnosed by clinical examination, blood tests, sonography and CT-scan examination. During surgery we performed paratesticular tumor excision.
Results: Histopathological examination showed a high grade leiomyosarcoma, pT_{2a}.
Conclusions: This type of paratesticular sarcoma (leiomyosarcoma) is very rarely quoted in the literature, especially at the age of our patient. The prognosis depends on a correct surgical treatment and a complete adjuvant oncological therapy (radio- +/- chemotherapy).

Keywords: adult patient, leiomyosarcoma, paratesticular tumor

Correspondence: Dr.Gheorghe Pupca
Clinica de Urologie, Spitalul Clinic Județean de Urgență Nr. 1, Timişoara
Bd. I. Bulbuca Nr. 10, Timişoara
Email : gpupca@yahoo.com

Introduction

The majority of the malignant extratesticular tumors arise from the spermatic cord. Sarcomas are the most common malignant tumor, with rhabdomyosarcoma being common in children while liposarcoma is common in adults. However, pleomorphic sarcoma, leiomyosarcoma, mesothelioma, and lymphoma also occur in the scrotum [1]. Leiomyosarcoma, the second most common paratesticular sarcoma in adults after liposarcoma, is represented in the literature primarily as isolated case reports.

Case report

A 71-year-old male presented in our Ambulatory Department with a painless swelling of the right hemiscrotum content, which had gradually enlarged during the past 12 months. No urologic or constitutional symptoms were present. He did not receive any radiation therapy or anabolic corticosteroids in the past.

Clinical examination revealed an 1.5 cm ulceration of the right hemiscrotum skin, corresponding to the upper side of a hard, relatively irregular, nontender scrotal mass, 5/4 cm in size, entirely separate from the right testicle, without associated inguinal adenopathy (Fig. 1).



Fig. 1. Ulceration of the right hemiscrotum

The routine hematologic profiles were all within the normal limits. Also the serum level of alpha-fetoprotein (AFP) and beta human chorionic gonadotropin (b-HCG) was normal.

Scrotal sonography (Fig. 2) and CT-scan (Fig. 3) showed a solid, heterogenous mass of 5/4.5/5 cm. Abdomen-pelvis CT-scan and bone scan were negative for pelvic or retroperitoneal adenopathy, visceral or bone metastasis. The left testis was normal.



Fig. 2. Right hemiscrotal sonography image



Fig. 3. Scrotal CT-scan image

Clinical examination and scrotal sonography showed a mass arising separate from the testis, which allowed exclusion of a testicular origin of the tumour. Clinical and sonographic findings were not specific to a tumour type. In all cases, all tumours of the paratesticular region are amenable to adequate surgical resection. Definite diagnosis is determined by histological evaluation.

During surgery, we have observed that the tumor had no connection with the testicle, epididymis or spermatic cord (Fig. 4). The tumor and a large part of the skin around the ulceration were excised.



Fig. 4. Intraoperative completely isolated tumor

This tumoral mass was located completely outside the testicle, well circumscribed and the cut surface was firm, white, solid mass with focal necrosis and hemorrhage (Fig. 5).

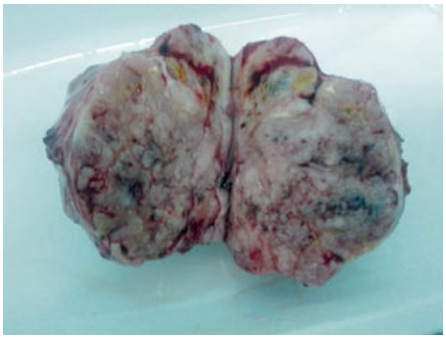


Fig. 5. Gross appearance of paratesticular leiomyosarcoma

Histological examination showed a lesion composed of spindle smooth muscle cells, arranged in interlacing fascicles. Cells showed focal nuclear pleomorphism. There were 2-10 mitoses per high power field and atypical mitoses and focal necrosis were noted. Immunohistochemistry of the elongated cells showed positive staining for smooth muscle actin and desmin and negative staining for CD-68. The combined histological and immunohistochemical findings were diagnostic for leiomyosarcoma grade pT2a (Fig. 6A, Fig. 6B).

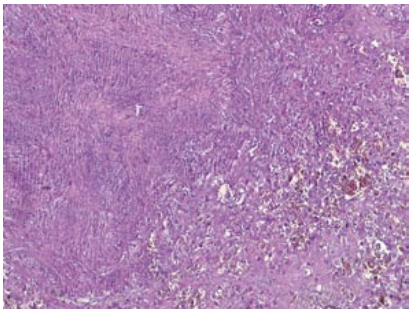


Fig. 6A Transition between leiomyosarcoma and malignant fibrous histyocitoma, HE x50

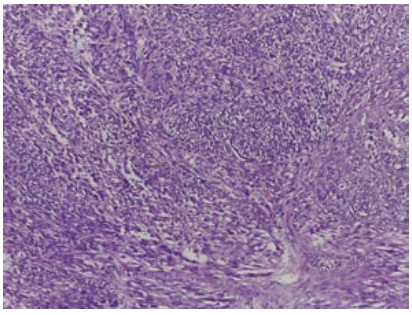


Fig. 6B Leiomyosarcoma tumoral contingent, HE x100

The patient left our Clinic with surgical healing wounds and recommendation to perform a complete adjuvant oncological radio- +/- chemotherapy.

Discussion

Leiomyosarcoma occurs commonly in the gastrointestinal tract, such as colon and stomach, but is rare in the genital region. The presence of such elements in the scrotal subcutis, dartos muscle, tunica albuginea, blood vessels and testicular parenchyma could provide a site of origin for these rare smooth muscle tumors. Paratesticular leiomyosarcoma is a rare and described as an indolent tumor with the potential for distant metastases. A few cases of primary paratesticular leiomyosarcoma had been reported, however, the age at diagnosis were most after fifth decade of life [2].

Scrotal leiomyosarcoma is thought to arise from the dartos layer of the scrotum [3]. Like other sarcomas, leiomyosarcoma tends to infiltrate local tissues. Lymphatic spread may involve the external iliac, hypogastric, common iliac, and retroperitoneal lymph nodes while haematogenous metastases are primarily pulmonary [4, 5]. The behavior of leiomyosarcoma is related to the site, size (particularly in areas where anatomical constraints limit adequacy of resection), histological grade, and presence of nodal or distant metastases [6, 7]. Typical clinical presentation is of a painless, firm, slow-growing, intrascrotal mass with palpation usually revealing the mass to be well defined, lobulated, mobile, and sometimes associated with a small hydrocele [5]. Work-up should include ultrasonography which is the primary imaging method for any cord or scrotal abnormality, with a sensitivity of 95-100% for differentiating intratesticular from extratesticular lesions [8]. A solid, heterogeneous mass is usually identified [9]. Currently there is no clear indication for prophylactic lymphadenectomy for paratesticular leiomyosarcoma. The general consensus in the literature is that paratesticular leiomyosarcomas rarely involve locoregional lymph nodes rather spreading most frequently by direct extension.

There is some evidence supporting the use of adjuvant radiotherapy for paratesticular sarcomas [10-12]. In a series of 21 cases, Catton and colleagues noted a 5-year disease-free survival of 58% with surgery alone and 100% with the addition of adjuvant radiotherapy [12]. Despite these findings, there have been no studies on the use of radiotherapy in leiomyosarcoma specifically. However, in the case presented, radiotherapy was recommended.

The role of adjuvant chemotherapy was observed as effective in pediatric rhabdomyosarcoma [10], but not in the leiomyosarcoma [12].

Conclusions

Leiomyosarcoma should be considered as a differential diagnosis in any elderly male presenting with an intrascrotal mass. These tumours may be clinically indistinguishable from testicular tumours.

All paratesticular tumours are amenable to adequate surgical resection.

Adjuvant radiation therapy can lead to improved locoregional control and has a role in patients with nonmetastatic paratesticular leiomyosarcoma.

The limited number of cases of this rare disease as well as the inconsistent management strategies utilised requires that further research be performed to formulate an ideal treatment protocol.

Conflict of interests: The authors declare that there is no conflict of interests regarding the publication of this paper.

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