Casos Clínicos

Primary Testicular Leiomyosarcoma – Case Report

Rui Pedro Borges,[1] Fernando Vila,[1] Victor Cavadas,[1] João Queirós,[1] Filinto Marcelo,[1] Ana Encinas[2]

Departments of [1] Urology and [2] Pathology, Hospital Geral Santo António, Oporto, Portugal

Abstract

Primary leiomyosarcoma of the testis is extremely rare. We describe a 19-year-old man who complains of left scrotal swelling for 3 months, and underwent radical orchiectomy. The histological examination revealed intratesticular leiomyosarcoma.

Introduction

Leiomyosarcoma is a soft-tissue tumor arising from smooth muscle cell of mesenchymal origin. Although para-testicular leiomyosarcomas are reasonably common, primary testicular leiomyosarcoma is extremely rare with only nine cases reposted in literature.[1-9] We report a case of testicular leiomyosarcoma in a 19-years-old man who underwent left radical orchiectomy and adjuvant chemotherapy and radiotherapy.

Case Report

A 19-year-old man was referred to the Urology department with a 3-month history of left testicular pain and swelling. There was no history of local trauma, usual medication or associated pathology. On physical examination he had a large hard and firm left testis The right testis and epididymis were normal and there was no evidence of hidrocele or paratestiscular patalogy. Liver function and tumor markers, including a-fetoprotein, lactate dehydrogenase, and b human chronic gonadotrophin assays, were all within normal ranges.

Ultrasound scan showed a 7x5 cm mass on the left testis, with a multinodular heterogeneous pattern. Chest and abdominal computed tomography scans revealed no metastasis.

A left radical orchiectomy with high ligation of the spermatic cord was performed. The pathological examination revealed a mass weighing 152 g including testis and epididymis (8.5x6x4.5cm) and spermatic cord (3.5cm). The tumor (7x5cm) was yellowish-with, solid and existed only in the testis with focal invasion of tunica vaginalis and without any involvement of epididymis or spermatic cord.

Microscopy revealed a tumor with an winterweaving pattern of elongated cells arranjed in fascicles with high mitotic index and foci of necroses (fig. 1).

Immunohistochemistry was negative for CAM5.2, AE/AE3, sarcomeric actin and S100 protein, and positive for vimentin, smooth muscle actin and desmin.

These data supported the diagnosis of high grade intratesticular leiomyosarcoma.

The patient received adjuvant chemotherapy (Gencitabine plus Docetaxel) and radiotherapy (50Gy, in 25 fractions of 2Gy a day, five times a week) to prevent distant and local metastasis respectively.

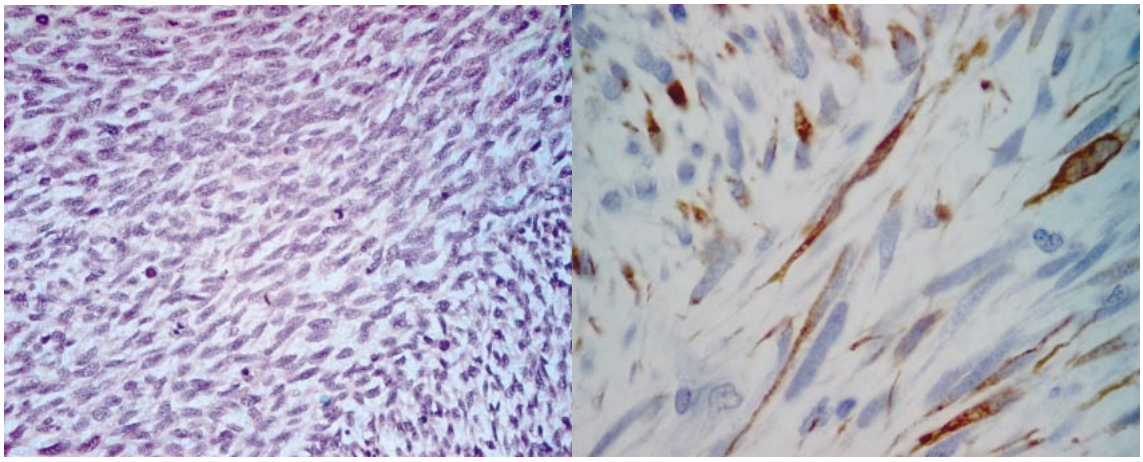

Fig 1 - Pattern of elongated cells arranjed in fascicles with high mitotic index and foci of necroses (Desmin 0x, 100x)

After 16 months of follow-up the patient remains asymptomatic but CT scan reveled a large bulky mass in the retroperitoneum (fig. 2) with central necrotic area and hidronephrosis of the left kidney. This mass, kidney and a segment of ileum where removed. Microscopy confirmed distant recurrence (with invasion of left kidney, and ileum peri-intestinal fat and serosa).

Patient starts a new salvage chemotherapy scheme, and a new CT scan not yet performed.

Discussion

Leiomyosarcoma is a soft-tissue tumor arising from smooth muscle cells of mesenchymal origin. The origin of leiomyosarcoma in the testis is controversial. Its origin has been attributed to contractile cells in the tunica propria of the seminiferous tubules, to the muscular layer of blood vessels, and to smooth muscle elements in the tunica albugínea. [1] High-doses of anabolic steroids [4] and chronic inflammation [5] are reposted to be risk factors for intratesticular leiomyosarcoma.

The clinical and biologic behavior of these tumors is very hard to predict. However, the high mitotic activity is considered an important criteria for malignancy. These tumors might spread via three routes: local invasion, lymphatic dissemination and hematogenous metastasis.[8]

In all reported cases of intratesticular leiomyosarcoma a radical orchiectomy performed[1-9]. Retroperitoneal lymph node dissection was additionally performed in one case and another one underwent chemotherapy. Only one patient developed pulmonary metastases post-surgery,[6] and there was no lymphatic invasion.

These tumors are exceedingly rare and there is no question that radical orchiectomy is the treatment of choice, however standard therapy is difficult to recommend.

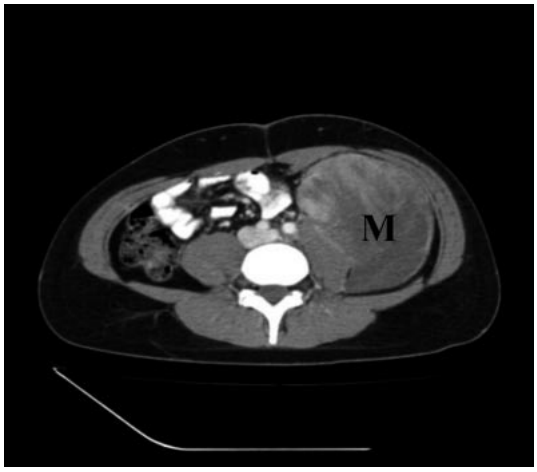

Fig 2 – M: Bulky mass in retroperitoneum

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