CASE REPORT
Urooncology

Doi: 10.4274/jus.1652
Journal of Urological Surgery, 2018;5(4):208-210

Spermatocytic Seminoma with Sarcomatous Transformation: The First Brazilian Case Report

Sarkomatöz Dönüşüm ile Spermatositik Seminom: Brezilya'ya Özgü İlk Olgu Sunumu

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Abstract

The cases of spermatocytic seminoma with sarcomatoid transformation are rare. When there is sarcomatoid transformation, the tumor takes an aggressive pattern, with high rates of metastasis and a worse prognosis. We present the case of a 42-year-old man with a slow enlargement of the left testicle over more than 18 months, which progressed to a pronounced and painful growth in the last six months. Computed tomography showed a left retroperitoneal mass in the renal area affecting the stomach. Total orchiectomy with partial scrotectomy was performed in the left testicle and the patient was referred to the oncology department for chemotherapy.
Keywords: Spermatocytic seminoma, Sarcoma, Testicular neoplasms, Orchiectomy

Öz

Sarkomatöz dönüşüm gösteren spermatositik seminom olguları nadirdir. Sarkomatöz dönüşüm olduğunda, tümör yüksek oranda metastaz ve daha kötü bir prognoz şeklinde agresif bir yol izler. On sekiz aydan fazla bir süredir sol testiste yavaş bir büyüme olan, son altı ayda ise bu büyüme belirgin ve ağrılı bir hale gelen, 42 yaşında bir erkek hastayı sunuyoruz. Bilgisayarlı tomografi, renal bölgede mideye etki eden sol retroperitoneal kitleyi gösterdi. Sol testiste parsiyel skrotektomi ile total orşiektomi yapıldı ve hasta kemoterapiye yönlendirildi.
Anahtar Kelimeler: Spermatositik seminom, Sarkoma, Testiküler neoplazi, Orşiektomi

Introduction

Testicular tumors account for approximately 1% of all neoplasms in men, with solid neoplasms more frequent in young adults (1,2). Primary testicular tumors are divided into germinal and non-germinal tumors. Germ cells comprise 90% of testicular neoplasms. Most are curable and have good prognosis. With various therapeutic approaches including surgery, chemotherapy and radiotherapy, 5-year survival rate can be increased to as much as 90%. However, spermatocytic seminoma (SS) with sarcomatous transformation, a rare testicular tumor, has a poor prognosis (1,3).

Case Presentation

A 42-year-old man presented to the Emergency Department at Antônio Pedro University Hospital on December 11th, 2016, with a slow enlargement of left testicle over the previous 18 months, which had progressed to a pronounced and painful growth in the last six months. The patient also complained of postprandial fullness. No other comorbidities were present.

Physical examination was unremarkable except for emaciation and hypocorous (+/++++). The left testicle presented as an enlarging testicular mass with a ten-fold increase in size, mobile, with stony consistency and pain. There was a palpable mass in the abdomen at the epigastric region, the umbilical region and the left flank. No other abnormalities were found on the exam.

Computed tomography of the abdomen and pelvis showed a left retroperitoneal mass in the renal area, which pushed the stomach and a heterogeneous testicular mass on the left side (Figure 1). Tumor markers were as follows: alpha-fetoprotein: 2.6 ng/mL; human chorionic beta-gonadotrophin: 7.36 mIU/mL; lactate dehydrogenase: 416 U/L and stadium: S1.

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Received: 20.09.2017 **Accepted:** 10.12.2017

Cite this article as: Carvalho D, Bozzi R, Coltro A, Reis B, Scheinkman J. Spermatocytic Seminoma with Sarcomatous Transformation: The First Brazilian Case Report. J Urol Surg 2018;5(4):208-210.

Total orchiectomy and a partial scrotectomy were done in the left testicle due to the intimate and indissociable contact with the skin, with staging T3N3. Macroscobically, the tumor was measuring 13.5x8.0x7.0 cm in size, weighing 350 grams, capped with areas of necrosis and showed albuginea involvement and tunica vaginalis, and replacement of the entire testicular parenchyma. Histopathological analysis revealed SS in small focus and extensive high grade sarcomatoid component (Figure 2). The immunohistochemical stains S100 were positive in sarcomatous component. The surgical limit of the spermatic cord had neoplastic emboli. The patient was referred to the department of oncology for chemotherapy.

Written informed consent was taken from patient.

Discussion

SS is a rare germ cell neoplasm, accounting for about 1% of all testicular tumors, and mainly affects men in around the fifth decade of life (1,3). It was first described by Pierre Masson in 1946 as a variant of seminoma. However, SS and classical seminoma have distinct pathogens (1,2). For this reason, some authors prefer to use the term spermatocytoma, a term not widely accepted in the scientific community (2). SS also differs from seminoma by having a higher rate of cell differentiation, to the point of performing spermatogenesis. It has no association with cryptorchidism or other testicular neoplasm, as opposed to the classical seminoma (1,4).

The clinical presentation is in the form of an insidious and painless enlargement of testis (1). The possibility of metastasis is rare and the prognosis is favorable. Orchiectomy and surveillance of the contralateral testis is the appropriate therapy, without the need for adjuvant chemotherapy in most cases (2,3). The main differential diagnoses are classical seminoma and testicular lymphoma (1).

Approximately 6% of SS cases can develop from an anaplastic transformation of tumor cells into a sarcomatous differentiation (1,2). The risk of sarcomatous transformation increases mainly in cases of long-standing SS (3). It is a neoplasm that shows no ovarian equivalence and no association with intraepithelial testicular neoplasms (5). Possible differential diagnoses are: seminoma, lymphoma, teratoma, and primary testis sarcoma (1,4). There are less than 20 reports of SS with sarcomatous transformation in the literature, this being the first reported case of SS with sarcomatous differentiation from Brazil (3,4,6).

When there is sarcomatoid transformation, the tumor takes an aggressive pattern, with high rates of metastasis and worse prognosis (1,2,3). Thus, SSs that have metastasized normally present sarcomatous transformation (2). In these cases, distant metastasis or lymph node metastasis occurs.

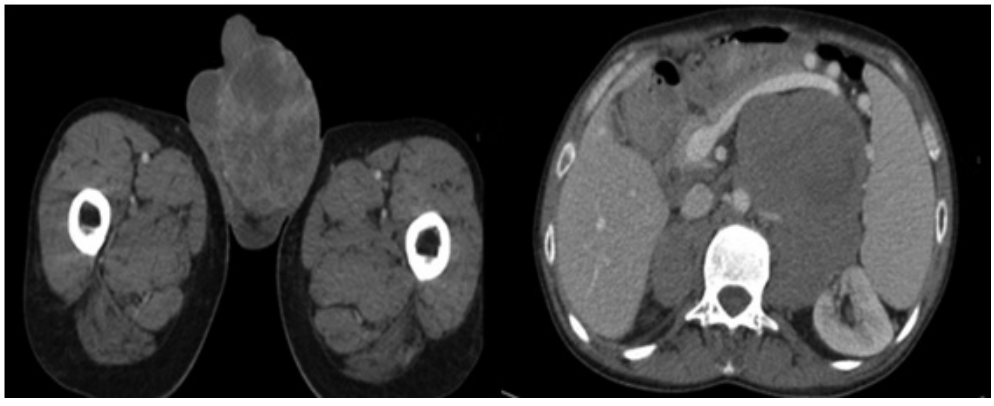

Figure 1. Computed tomography, which evidences tumor in the left testicle and voluminous retroperitoneal mass in the left that pushes away the adjacent structures

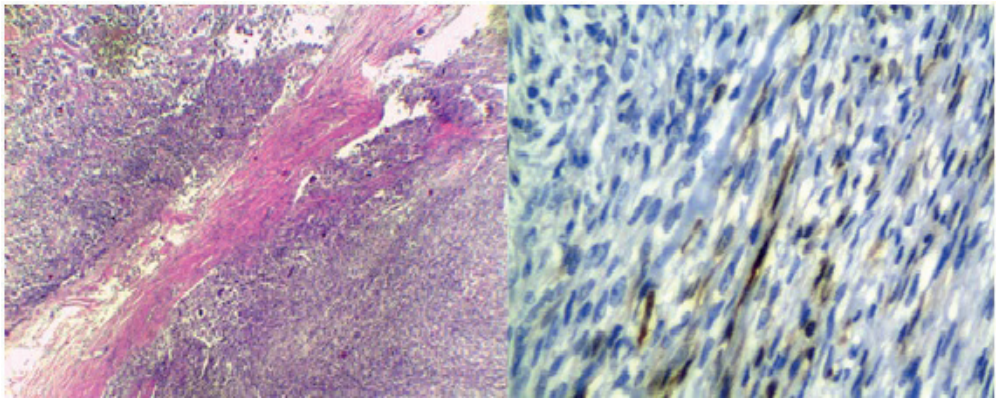

Figure 2. Spermatocytic seminoma histopathology (upper left) and sarcomatous transformation (lower right). On the right, immunohistochemistry stains S100 was positive for the sarcomatous component

In the literature, there are records of lung, bone and liver metastases, as well as retroperitoneal lymph node metastasis up to the level of the ipsilateral renal vein (2,6). The sarcomatoid components reported in the literature are rhabdomyosarcoma, chondrosarcoma, undifferentiated sarcoma or high-grade sarcoma (2,4). Undifferentiated sarcoma is the most commonly found component (4). All reported cases of rhabdomyosarcoma showed distant metastasis, although there is no significant evidence of difference in prognosis between histological types of sarcomatous transformation (6,7). Some studies have shown that only the sarcomatous elements of SS metastasize (2).

Histopathologically, SS is characterized by cells poor in cytoplasmic glycogen and with spherical nuclei (5). It is common for it to present a heterogeneous population of cells: small cells (6-8 µm) with dense and basophilic nuclei, medium cells (15-20 µm) with fine chromatin filaments and large cells (50-100 µm) which may be multinucleated (4). Unlike classic seminoma, SS has a sparse or simply absent lymphocytic infiltrate (5). It is common to find mitoses, including atypical forms (4).

The cases of SS with sarcomatous transformation are clinically characterized by a rapid increase in the size of pre-existing tumor, and may present with pain (3,4). It commonly affects only one testicle, being bilateral in about 10% of cases (3). Most patients have metastasis at the time of diagnosis (2). Differently from classic SS, where a sarcomatous component is present; greater benefits are obtained if the treatment includes radiotherapy and adjuvant chemotherapy in addition to radical inguinal orchiectomy (3,6). Thus, aggressive treatment with the combination of surgical approach and adjuvant chemotherapy is recommended in all cases, although the prognosis remains reserved despite chemotherapy (1,6). For some authors, the need for radiotherapy or chemotherapy is not clear in cases without radiological evidence of metastasis (4).

Acknowledgments

We thank Antônio Pedro University Hospital for its support.

Ethics

Informed Consent: The informed consent was obtained from the patient.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: D.C., B.R., Concept: D.C., R.B., Design: D.C., R.B., Data Collection or Processing: D.C., R.B., Analysis or Interpretation: D.C., J.S., Literature Search: A.C., R.B., Writing: A.C., D.C., R.B.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study received no financial support.

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