



Spermatocytic seminoma of testis associated with undifferentiated sarcoma revealed in metastatic disease: A review and case report analysis

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ABSTRACT

Spermatocytic seminoma is a relatively rare testicular tumor and is characterized by a good prognosis. The discovery of a sarcomatous contingent modifies the prognosis of the indolent neoplasm. Only 20 cases being reported in English literature. We present the case of a 66-year-old man with a two-year history of left-sided scrotal pain and swelling. Tumor markers were normal. Ultrasound demonstrated a very large solid-cystic testicular mass. Orchidectomy was performed. Further imaging investigations revealed lung, vertebra, and retroperitoneal lymph node metastases. Histological examination and immunohistochemistry of the orchidectomy specimen concluded on spermatocytic seminoma associated with undifferentiated sarcoma component.

Introduction

The spermatocytic seminoma (SS) was first described by Pierre Masson in 1946. This is a relatively uncommon testicular cancer, usually of good prognosis, which never associates with another germ component.¹ The development of a sarcomatous component transforms the prognosis of the usually innocuous spermatocytic seminoma into a highly aggressive neoplasm.² We report a case of SS of testis associated with undifferentiated sarcoma and a review of the literature.

Case report

A 66-year-old man presented with a 2-year history of left-sided scrotal pain and swelling. Clinical examination revealed a massive left testicular mass, which was solid and adherent to the scrotal skin. Inguinal lymph nodes were not palpable. The serum tumor markers alpha-fetoprotein (α FP) and beta-human chorionic gonadotropin (β -HCG) were without abnormality. A scrotal ultrasound has revealed a voluminous solid-cystic testicular mass measuring 15 × 87 mm (Fig. 1). The thoracic abdominopelvic scan revealed multiple left retroperitoneal lymph nodes (largest 20 × 22 cm at the left para-aortic area), multiple lung nodules (diameters 6 mm–12 mm), and lytic lesions in the thoracic and lumbar vertebra D7, L5 (Fig. 1: A, B).

The patient underwent a left orchidectomy via an inguinal approach. Macroscopically, the specimen measured 17 × 17 × 6 cm and weighed 1700 g (Fig. 2: A, B). Testicular parenchyma was destroyed by a

voluminous tumor of heterogeneous appearance, solid, including cystic and necrotic-hemorrhagic remodelings. The spermatic cord and epididymis were involved.

Histopathological examination revealed tumor proliferation with a dual germinal and sarcomatous component. The first germinal component was formed of 3 different cell types of variable size of small, intermediate, and large cells with spherical nuclei and absent lymphocytic infiltrate, characteristic of a spermatocytic seminoma. The second component, sarcomatous, was composed of fascicles of spindle-shaped cells with fusiform nuclei (Fig. 2: C, D).

Immunohistochemical analysis revealed positivity focal staining of antibody to CD117 (c-KIT) and vimentin. Immunohistochemical stains of antibodies to PLAP, HCG, AFP, and the myeloid markers CD34 and myoglobin were all negative.

A final diagnosis of spermatocytic seminoma with undifferentiated sarcoma metastatic was established. Immediately after the orchidectomy, the patient received poly-chemotherapy with a combination of etoposide, ifosfamide, and cisplatin (VIP). The patient died after 7 months of follow-up.

Discussion

The spermatocytic seminoma (SS) is an uncommon variety of germinal cell tumors, exclusively observed in the testis, and represents 3–7% of seminomas and 1–2% of all testicular germinal tumors. SS is an indolent neoplasm, with a long duration of symptoms, extremely low

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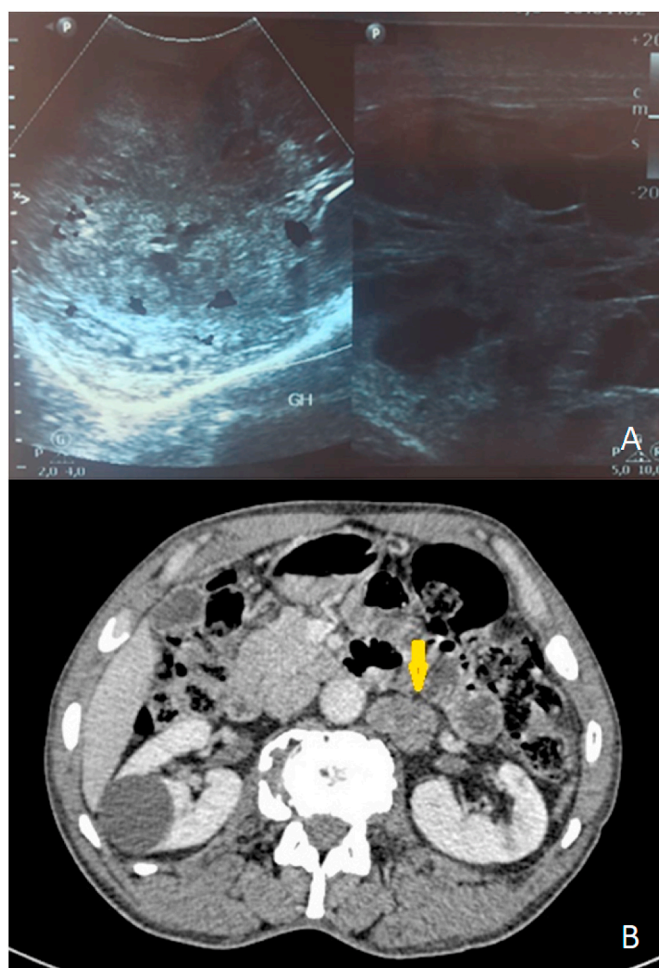


Fig. 1. A: Scrotal ultrasound demonstrating a large solid-cystic mass, B: The para-aortic lymph node size was about 2 cm (arrow).

metastatic potential and is associated with an excellent prognosis.¹ It may derive from spermatocytes and spermatogonia and not be associated with other germinal cell components. It usually occurs in older men with a mean age of 54 years (range 25–87 years).²

SS with sarcoma is an extremely rare diagnosis with only 20 cases previously reported in the literature (Table 1).^{2–4} This sarcomatous transformation, which occurs in approximately 6% of cases, induces a sudden rapid growth in the size of the tumor and is associated with aggressive behavior, presence of metastasis, and poor prognosis. The SS with a sarcomatous component has the same age incidence as the pure SS, the mean age of 51 years (range 29–68).^{2,3}

The sarcomatous component is usually undifferentiated spindle cell, but specialized differentiation, including rhabdomyosarcoma. Only one case showed both rhabdomyosarcoma and chondroid differentiation, and one case shows a focal area of chondrosarcomatous differentiation within a largely undifferentiated sarcomatoid component.⁵

The immunohistochemical analysis helps to identify and categorize the sarcomatous element. Immunohistochemically, the spermatocytic component shows negativity for CD30, smooth muscle actin, c-kit, placental alkaline phosphatase, and alpha-fetoprotein. On the other hand, the rhabdomyosarcomatous element shows strong positivity for desmin and weak positivity for myoglobin. Furthermore, the undifferentiated spindle cell component is positive only for vimentin.⁴

The differential diagnosis includes other tumors in the testis with a sarcomatous appearance: teratoma with a secondary somatic type malignant component and primary sarcoma of the paratestis or testis.⁵

Eight cases of metastatic SS with sarcoma are documented in the

literature and are highly resistant to cytotoxic chemotherapy with a median survival of 5 months.³

The theory of sarcomatous transformation can be explained by an anaplastic transformation or dedifferentiation of a well-differentiated SS.⁴

The treatment of choice for non-metastatic SS is orchidectomy, followed by surveillance. Adjuvant chemotherapy or radiotherapy is rarely recommended in SS cases due to low risks of relapse and metastasis.²

However, the non-metastatic sarcomatous component of the SS gives a very high risk of metastasis, and the guideline of treatment has yet to be established in these cases.³

Some authors suggest orchidectomy followed by regular monitoring with computed tomography,⁵ for another the adjuvant chemotherapy following orchidectomy is the choice of treatment for these highly malignant tumors.²

In metastatic SS cases, the prognosis is very poor, even with the aggressive multimodal treatment.⁴ In the literature, five of these patients received platinum-based chemotherapy, and all responded poorly and died shortly (within 3–14 months) after diagnosis. However, only one case of metastatic SS responded well to the VIP combination chemotherapy after radical orchidectomy.³

Conclusion

SS with sarcomatous differentiation is an uncommon tumor. It should be considered when evaluating testicular tumors in older men. The development of this sarcomatous element transforms the prognosis

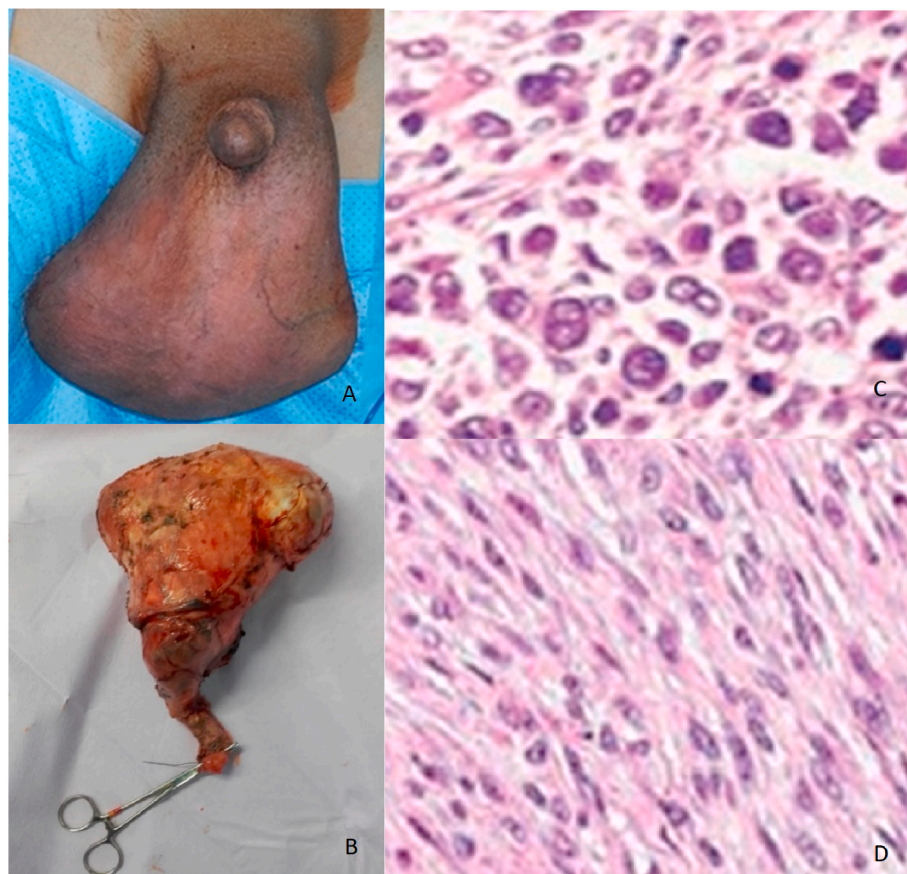


Fig. 2. A: Preoperative picture, B: surgical specimen.: C: High-power image showing: 3 types of cells (small, intermediate and large), typical of spermatocytic seminoma. D: fascicles of spindle shaped cells, typical of undifferentiated sarcoma.

Table 1
: Reported cases of Spermatocytic Seminoma associated with Sarcoma.

N°	year	Authors	Age (years)	Metastatic disease	Tumor Size (cm)	Treatment	Histology of Sarcoma	Follow-up
	2020	Our case	66	Lung + Bone + RL	17 × 17 × 6	OE + CT	Undifferentiated spindle cell	Died 7 months
1	2017	Stueck et al. ⁵	52	Nil	9,5	OE	chondrosarcoma	Alive 41 months
2	2015	Jeong et al. ³	52	RL + neck and iliac nodes	9	OE + CT + RPLND + RT	Rhabdomyosarcoma	N/A
3	2013	Wetherell et al. ²	29	Nil	6 × 5.5 × 5	OE	Undifferentiated spindle cell	Survived
4	2012	Narang et al. ¹	38	Nil	7 × 6 × 4	OE + CT + RT	Rhabdomyosarcoma	Survived
5	2011	Trivedi et al. ⁴	43	Lung	18 × 10 × 10	OE + CT	Undifferentiated spindle cell	Died 10 months
6	2009	Menon et al.	55	N/A	15 × 9.5 × 8	OE + RT	Rhabdomyosarcoma	n.a
7	2007	Robinson et al.	44	Bone	17	OE + CT	Rhabdomyosarcoma	Died 5 months
8	2006	Chelly et al.	50	Liver	14	OE + CT	Rhabdomyosarcoma	Died 3 months
9	1993	Burke and Mostofi	68	Present (site n.s.)	N/A	OE	Undifferentiated spindle cell	Died 9 months
10	1993	Burke and Mostofi	43	N/A	N/A	OE	Undifferentiated spindle cell	N/A
11	1993	Burke and Mostofi	68	Present (site n.s.)	N/A	OE	Undifferentiated spindle cell	Died 11 months
12	1993	Burke and Mostofi	39	N/A	N/A	OE	Undifferentiated spindle cell	N/A
13	1994	Sabater and Martorel	42	Nil	8 × 5 × 4	OE + CT	Rhabdomyosarcoma + chondrosarcoma	Alive 18 months
14	1990	Matoska and Talerman	51	Lung + Liver + RL	18 × 12 × 12	OE	Rhabdomyosarcoma	Died 2 months
15	1988	True et al.	55	Nil	6 × 5 × 5	OE	Undifferentiated spindle cell	Alive 36 months
16	1988	True et al.	66	Nil	17 × 11 × 10	OE + RT	Undifferentiated spindle cell	Died 16 years (metastatic prostate cancer)
17	1988	True et al.	40	Nil	7.5	OE	Undifferentiated spindle cell	Alive 9 months
18	1988	True et al.	60	lung + thyroid + heart	25 × 20 × 15	None (Tumor found at autopsy)	Rhabdomyosarcoma	Died 1 month
19	1980	Floyd et al.	42	Lung	9.5 × 7.5 × 5.5	OE + CT	Undifferentiated spindle cell	Died 1 year
20	1980	Floyd et al.	56	Lung + liver	9 × 6 × 3	OE + RPLND + CT	Rhabdomyosarcoma	Died 14 months

Abbreviations: RL = retroperitoneal lymph nodes; OE = orchiectomy; CT = chemotherapy.

of the relatively indolent SS tumor into a highly aggressive malignant neoplasm. These tumors often metastatic at the time of diagnosis and therefore treatment with adjuvant chemotherapy after radical inguinal orchidectomy should be considered in all cases.

Declaration of competing interest

None of the contributing authors have any conflict of interest.

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