Malignant Sertoli Cell Tumor of the Testis With a Large Retroperitoneal Mass in an Elderly Man

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INTRODUCTION
We report a malignant Sertoli cell tumor distinguished clinically by its rapid enlargement, early metastasis, and age of presentation.

CASE REPORT
A 78-year-old man presented with a new growth in his right testis which was uncomfortable, but not painful. He reported a progression in growth over the past few months, and stated that it was doubled in size in the previous month. His past medical history included mumps as a child, hypertension, and recurrent urolithiasis with reported spontaneous passage of calcium oxalate calculi several months prior to this presentation. Physical exam revealed a moderately enlarged right hydrocele and a firm right testicular mass as well as right inguinal adenopathy consisting of non-tender, small nodes. There was no evidence of gynecomastia.

Complete blood cell count, blood chemistry, and urine analysis were within normal limits, and serum tumor markers, including human chorionic gonadotropin and alpha-fetoprotein were negative. Serum levels of estrogen and testosterone were not initially measured. Scrotal ultrasonography revealed an area of mixed echogenicity in the mid-lower portion of the right testis measuring 4.0 × 2.1 × 2.3 cm with increased flow on color Doppler ultrasonography consistent with a testicular neoplasm, as well as a large right hydrocele. Computed tomography scan revealed multiple enlarged retroperitoneal lymph nodes and a large aortocaval lymph node measuring 4.2 × 2.1 cm.

Right inguinal orchiectomy was performed without complication and the patient was discharged from the hospital the following day. Serum level of testosterone was within the normal range (287.13 ng/dL) at 11 months after the orchiectomy. On gross inspection, the specimen included the testis with attached spermatic cord and surrounding soft tissue, measuring 5.3 × 4.5 × 4 cm. Sectioning revealed the testis that weighed 53 gr and a 4 × 4 × 3.5-cm firm mass, with solid white to yellow cut surfaces replacing 95% of the testis tissue. The tumor focally invaded the surrounding tunica albuginea, and 3 small solid yellow nodules were identified on the surface of the parietal tunica vaginalis. Furthermore, one solid nodule measuring 0.9 × 0.7 × 0.5 cm was identified in the spermatic cord.

Microscopic examination revealed a Sertoli cell tumor with focal necrosis, vascular invasion, and...
direct extension into the spermatic cord and
tunica vaginalis. A high mitotic count of 15 per
10 high-power fields was noted. The tumor cells
were positive for inhibin and focally positive for
neuron specific enolase. Computed tomography
guided biopsy of the retroperitoneal lymph nodes
was offered to the patient on multiple occasions,
but he refused any further diagnostic tests. The
patient was counseled extensively regarding
the risks and benefits of retroperitoneal lymph
node dissection. He decided not to undergo the
procedure. At the time of the 1-year follow-up
visit, the patient’s aortocaval lymph node had
increased in size to 4.3 × 9.6 cm and there was
an additional new lymph node in the pelvis with
central necrosis measuring 3.3 × 2.6 cm. The
patient again elected to continue observation and
refused a diagnostic biopsy or retroperitoneal
lymph node dissection.

DISCUSSION

Sex cord-stromal tumors account for
approximately 4% of the testicular neoplasms,
with Sertoli cell tumors comprising 0.4% to 1.5%
of all primary testicular tumors.(1,2) In spite of
their rarity, Sertoli cell tumors have been rather
heterogeneous and much debate has revolved
around their distinguishing characteristics, clinical
course, and potential for malignancy.(3)

Two distinct subtypes have emerged from
careful study, a large cell calcifying Sertoli cell
tumor, first described in 1980, exhibits diffuse
intratubular and extratubular calcification, and
has been known to exhibit virilization and
extragonadal manifestations.(4) The second variety,
a sclerosing Sertoli cell tumor, was first described
by Zuckerberg and colleagues in 1991.(3) This
subtype, distinguished by extensive hypocellular,
collagenous stroma separating clusters of Sertoli’s
cell, is the least reported one and has questionable
malignant potential, with only one reported
tumor showing evidence of malignant features
pathologically, but with no evidence of metastasis
before the patient suffered from a cardiac related
death.

Even after distinct subtype classification had been
established, the low incidence of these tumors
makes appreciation for histological variability
difficult and subtyping a challenge, as factors such
as the degree of sclerosis weigh heavily upon the
categorization. For example, microscopic and
histological analysis of the tumor in our patient
originally yielded a diagnosis of a sclerosing
Sertoli cell tumor. However, consultation with
Armed Forces Institute of Pathology confirmed
the diagnosis of a malignant Sertoli cell tumor,
but the degree of hyalinization of the stroma
was not enough to categorize this as a sclerosing
Sertoli cell tumor. Furthermore, the largest series
of Sertoli cell testicular tumors in the literature,
consisting of 60 subjects with Sertoli cell tumors,
not otherwise specified, found a tendency in
the older literature to report the neoplasm now

![Figure 1. Abdominal computed tomography scan showing the
largest of multiple retroperitoneal lymph nodes, an aortocaval
node measuring 4.2 × 2.1 cm.](image1)

![Figure 2. Histology of tumor showing trabecular/acinar
arrangement of malignant Sertoli cells. Focal necrosis can be
seen in the upper left corner of figure 2a.](image2)
recognized as a juvenile granulosa cell tumor as a Sertoli cell tumor, and a similar tendency to label other neoplasms as Sertoli cell tumors that would be best placed in other categories, including Sertoli-Leydig cell tumors. They believed that this inconsistency weighs heavily on reported clinical features, including the age of presentation and frequency of gynecomastia.(5)

With this in mind, we feel it is important to accurately document the clinical manifestations and pathologic features of these tumors, especially those that are malignant. Although the large size (> 5 cm), poor tumor demarcation, invasion to adjacent structures, blood vessel and lymphatic invasion, and increased mitotic activity (> 5 mitotic figures per 10 high-power fields) are all suggestive of malignant potential, the designation of malignancy can be made certainty only in the presence of metastasis.(6) The first and most common site of metastatic disease of patients with sex cord-stromal tumors is the retroperitoneal lymph nodes,(7) and according to the literature, about 10% to 12% of these tumors have evidence of metastasis.(1,4)

The malignant tumor in our patient appears to be aggressive when compared to the 60 tumors analyzed by Young and associates. In his study, the mean age of presentation and the average duration of a “slowly enlarging” mass were 45 and 3.7 years, respectively. Only four patients had metastatic disease at the time of presentation. Of whom, in only two, vascular invasion, necrosis, nuclear pleomorphism, and mitotic rate >5 were all present.(5) Our patient presented with an enlarging mass over only months (doubling in size over the last month), had 4.2 × 2.1 cm retroperitoneal mass, and while slightly missing the large size criterion for pathologic malignancy, was significant for the remainder of the afore-mentioned standards. Furthermore, this patient’s age of 78 years is a rarity amongst this variety of tumor, as approximately one-third of the recorded patients with Sertoli cell tumors have been 12 years or younger.(6) While it has been observed that in Sertoli cell tumors, benign neoplasms occurred at a younger mean age than those proven to be malignant,(8) the oldest patient reported in the Young and colleagues’ study was 80 years old (a benign tumor),(5) and in a study by Lindegaard and Merck on metastasizing Sertoli cell tumors, only one patient, a 79-year-old man, exceeded the age of our patient.(1)

**CONFLICT OF INTEREST**
None declared.

**REFERENCES**