Leiomyosarcoma of urinary bladder with extensive osseous metaplasia

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ABSTRACT

Extensive ossification in a bladder tumor is extremely rare. A case of bladder leiomyosarcoma with extensive osseous metaplasia is reported in a 84 years old Libyan male. The patient underwent radical cystectomy and ureterosigmoid urinary diversion for muscle invasive bladder tumor. The patient died three months after surgery with local tumor recurrence, inter current sepsis and metabolic complication of urinary diversion.

Keywords: Leiomyosarcoma, osseous metaplasia, urinary bladder.


Genitourinary sarcomas are a group of malignancy arising in mesodermally derived extra osseous tissues. These tumors share prognostic characteristics of soft tissue sarcoma from other sites. Primary sarcomas of urinary bladder are uncommon. In adults, leiomyosarcomas are the most common histological type among urinary bladder sarcomas and they account for less than 1% of all urinary bladder tumors.1-3 Bone and cartilage formation within bladder neoplasms either as a neoplastic or metaplastic component is distinctly rare.4-5 We describe an unusual case of leiomyosarcoma of urinary bladder with extensive areas of metaplastic bone formation, observed among 340 (0.29%) cases of primary malignant bladder tumors managed during the last fourteen years (Jan. 1983-Dec.1996) at our institution.

Case Report. A 84 year old Libyan male was admitted on July 22nd 1989 to the Department of Urology in 7th October (Teaching) Hospital, Benghazi with the complaint of irritative voiding symptoms of two months duration. Gross hematuria was absent. Abdominal examination including external genitalia was unremarkable. Laboratory work up revealed a normal complete blood picture with raised ESR (17 mm/1st hour). Hepatic and renal functional profiles were normal. Urinary sediment showed pus cell 80-90/hpf, red blood cells 10-15/hpf. Urine was sterile on culture. Plain X-ray of abdomen showed specks of calcification in urinary bladder area.

Abdominal sonography. Showed a large strawberry like irregular mass measuring 4.5 x 5 cm with mixed echo, occupying right antero-lateral wall. Excretory urography suggested a spastic bladder with persistent filling defect in the right lateral wall with mild fullness of right pelvicalyceal system.

Cystoscopy. Revealed a markedly reduced structural bladder capacity < 250 ml and a solitary cauliflower mass encroaching bladder neck from right lateral wall. EUA (Examination Under Anesthesia) confirmed a mobile resectable tumor. Trans urethral biopsy from bladder lesion revealed an ulcerated tumor composed of spindle shaped cells suggesting the diagnosis of leiomyosarcoma. Patient

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underwent radical cystectomy and ureterosigmoid
urinary diversion as a definitive therapy for muscle -
invasive tumor.

Pathological findings. Cystectomy specimen
revealed a bony hard to firm cauliflower mass
measuring 8 x 6 x 6 cm infiltrating the right antero-
lateral wall. The areas of bone formation occupied
approximately 50-60% of tumor mass. Multiple
section from tumor showed the tumor is composed of
spindle shaped cells arranged in fascicles, the
cytoplasm is abundant and eosinophilic and nuclei
are plump with blunt ends (Figure 1). Masson
trichrome and phosphotungstic acid hematoxylin
stain (PTAH) showed longitudinal myofibrils in the
cytoplasm. Mitotic figure averaged 1-2/hpf. The
bony hard areas showed lamellar bone formation
surrounded by benign osteoblasts (Figure 2). There
was no evidence of direct woven bone formation by
the tumor cells. The diagnosis of leiomyosarcoma
with osseous metaplasia was made.

Follow up and fate. During the three months
following surgical therapy, patient had recurrent
episode of febrile right pyelonephritis, metabolic
acidosis, inter current intra-abdominal sepsis. Serial
abdominal sonographic evaluation revealed treatment
failure as a result of local recurrence without
evidence of systemic (metastatic) disease. A (7.2 x
6.5 cm) irregular hyper dense mass was revealed in
right half of pelvis. Patient succumbed due to local
recurrence, sepsis and metabolic complications.

Discussion. The limitation with soft tissue
sarcoma from urological organs is the relative rare
occurrence of the tumor and a lack of uniform staging
system as well as treatment strategy. Generally,
leiomyosarcoma is an uncommon tumor of urinary
bladder and occurs mainly in patients past the age of
40. Approximately 100 cases have been reported in
the literature since Gussenbauer described the first
case, a century before.6 Men are more frequently
involved than women. Rarely it is known to occur in
children.7 Earlier reports indicated the involvement
of lateral bladder wall yet, the involvement of trigone
with ureteral obstruction was noted8 as in our case.
Histologically, a typical leiomyosarcoma is
composed of interlacing fascicles of spindle shaped
smooth muscle fibres with high mitotic rate. Epitheloid
and myxoid variant have been described.
The myxoid variant, because of its bland histology,
could be mistaken for a benign or an inflammatory
pseudotumor.9

Our case is interesting from the point of view that
the tumor is associated with extensive areas of
metaplastic bone formation involving more than half
of the tumor. It is important to distinguish the
metaplastic bone formation from the neoplastic one.
The formation of mature lamellar bone surrounded
by benign osteoblast favors metatstatic process
whereas, in the neoplastic process, woven bone is
formed and closely intermixed with malignant tumor
cells.4 Curative treatment of choice is partial cystectomy
when the size and location of the tumor allows for
adequate surgical margin.7,9 While radical
cystectomy is indicated for more extensive lesions as it
seems to be an adequate form of monotherapy.3
The existing systemic chemotherapy is ineffective for
urological sarcomas other than rhabdomyosarcoma
and the definite role of radiotherapy in the treatment
of urological sarcoma is difficult to assess.10

Leiomyosarcoma is a rare tumor with poor
prognosis. Using Memorial Sloan Kettering Cancer
Center (MSKCC), clinical and pathological
prognostic criteria for staging, our case was of stage
3 (high grade, >5cm in size, and deeply invasive)
with an anticipated poor prognosis and survival
despite radical cystectomy.10

Local recurrence in high stage tumor is associated
with metastatic disease with its attendant mortality.10
We suggest that the treatment success or failure and
the prognosis depends upon the clinical stage (superficial/muscle-invasive) and intrinsic character (histopathological grading) of the tumor as evident by local and systemic failure in our case even following radical cystectomy.

References


