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SURGICAL MANAGEMENT OF LARGE LIVER METASTASES FROM URINARY BLADDER LEIOMYOSARCOMA

Mohamed Hajri[1], Sahir Omrani[1], Skander Zouari[2], Seifeddine Baccouche[1], Wael Ferjaoui[1], Sana ben Slama[3], Ahmed Saadi[2], Riadh Ben slama[2], Ghofrane Talbi[1], Dhouha bacha[3], Hafedh Mestiri[1], Rached Bayar[1]

[1] Department of General surgery, Mongi Slim University Hospital, Faculty of medicine of Tunis, University of Tunis El Manar
[2] Department of Urology, Charle Nicolle University Hospital, Faculty of medicine of Tunis, University of Tunis El Manar
[3] Department of Pathology, Mongi Slim University Hospital, Faculty of medicine of Tunis,University of Tunis El Manar

Ferjaoui Wael

email: farjaouiwael4@gmail.com
phone: +216 52430099

ABSTRACT

Leiomyosarcoma is a malignant tumor developed from the smooth muscle. It can arise in different sites especiallyin gastrointestinal tract (stomach, small bowel and colon), genitourinary tract (uterus, bladder and kidney) and the retroperitoneum (inferior vena cava) (1,2). It's localization in the bladder is rare, representing 0.5% of all urinary bladder malignancies. There are over 210 cases reported in literature(3).Ten to 30% of the patients willpossibly develop metastases and the most common sites are lungs, liver and bone(4,5). There is no concensus on a standard treatment due to its very low incidence.

We report here a rare case of a surgical management of large liver metastases from urinary bladder leiomyosarcoma.

Keywords: leiomyosarcoma, urinary bladder, liver metastases

CASE PRESENTATION:

A 51year-old man with no known comorbidities presented to our hospital with macro hematuria and pelvic pain. His clinical status was good and no other symptoms were present. Physical exam found a hypogastric tenderness.Computed tomography found a urinary bladder mass of 7x8 cm with irregular contours. No distant metastases were reported in the abdominal cavity. Trans-urethral resection of bladder tumor was performed and pathological report concluded to a bladder localization of low grade leiomyosarcoma. Chest and abdomino pelvic scan were negative for metastases. Patient underwent radical cystectomy finding a confined tumor into the bladder wall. Urinary diversion was performed using Bricker ileal conduit. The post-operative course was uneventful and patient was discharged two weeks after the surgery. Final pathological report led to the diagnosis of urinary bladder localization of low grade leiomyosarcoma confirmed by immunohistochemistry study (Figures 1, 2, 3).

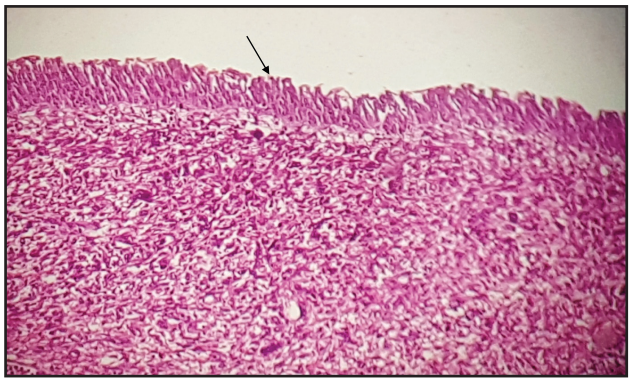


Figure 1: Microscopic aspects of Leiomyosarcoma of urinary bladder (arrow: urothelium) (x40)

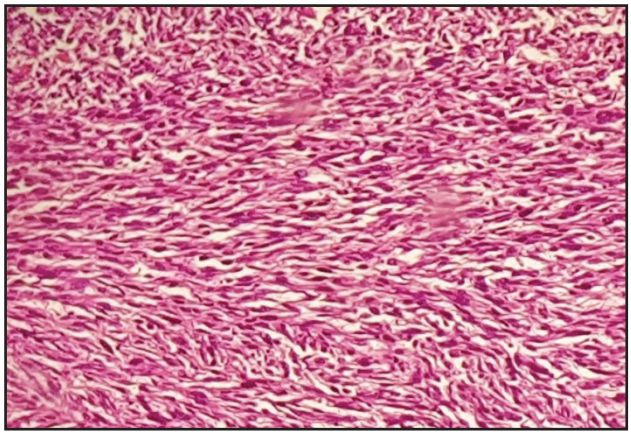


Figure 2: Microscopic examination: Tumor composed of interlacing fascicles of eosinophilic spindled cells (x 40).

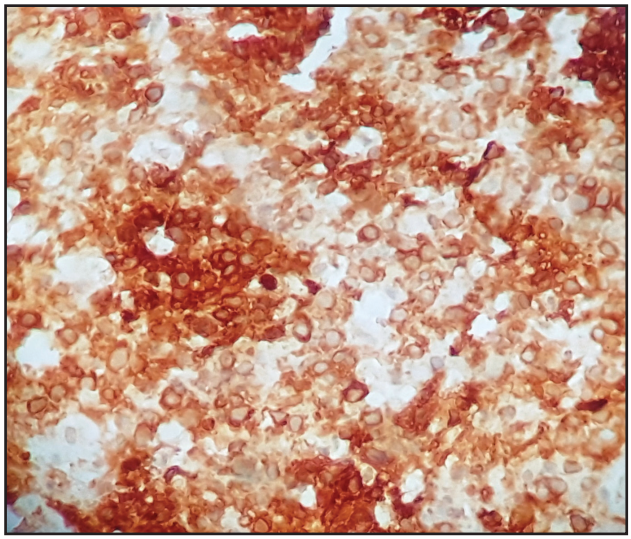


Figure 3: Immunohistochemistry: Positive immunostaining of tumor cells for desmin (x 400)

Patient was seen at three and six months post operatively with no signs of recurrence on imaging. Then, he was lost on follow up. He presented three years after to the hospital with upper abdominal pain. The physical exam showed a firm and fixed epigastric mass measuring 10 cm. Abdominal CT scan found a huge epigastric tissular mass developed in the left liver measuring 15x12x11 cm (figure 4).

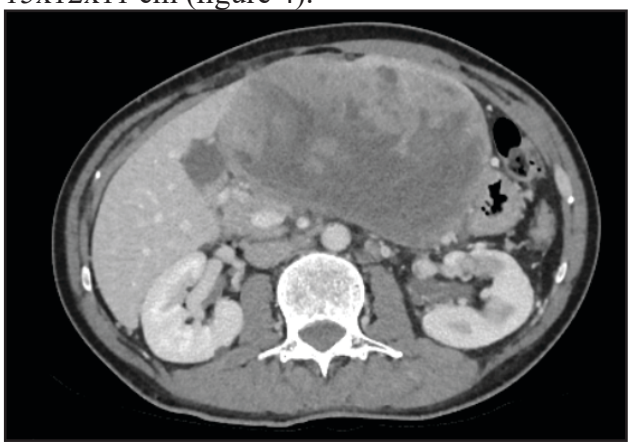


Figure 4: Abdominal CT scan showing the 15x12x11 tissular mass developed in the left liver

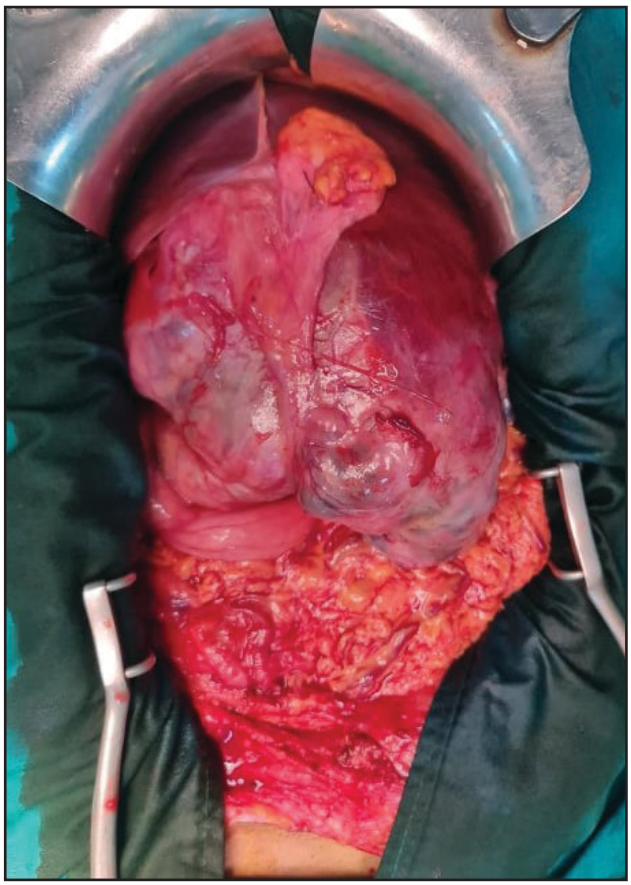


Figure 5: Intraoperative aspects of the liver leiomyosarcoma.

Percutaneous biopsy was performed and histology concluded to a leiomyosarcoma. The surgical approach was decided. Intra operative findings revealed a 15 cm left hepatic mass adhering to the stomach and the pancreas (figure 5), but showing a cleavage with these organs.

An en-bloc resection of the mass, with the segment III and the lower parts of the segment II and IV was performed.On cut surface, the tumour measured 16x11 cm and not well-defined (Figure6). The histological examination showed aspects of leiomyosarcoma (Figure 7).

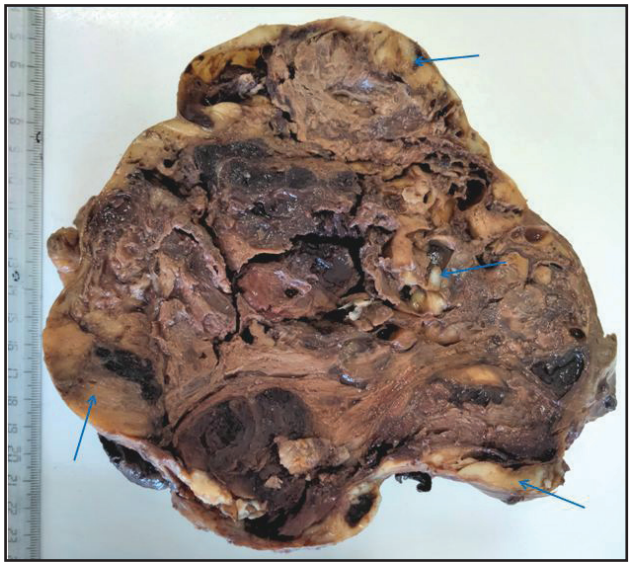


Figure 6: Gross examination: Large tumor with necrosis, hemorrhage and cystic degeneration, minimal fleshy territory (blue arrows).

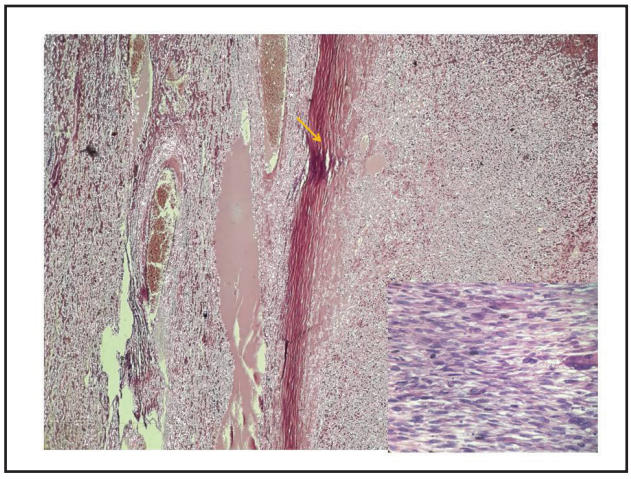


Figure 7: Microscopic examination: Proliferation of atypical spindle cells (on the right) separated of peripheral liver tissue on the left by a fibrous capsule (yellow arrow) (hematoxylin-eosin: HEx100). Cartridge: Fascicles of spindled cells eosinophilic cytoplasm with marked cytologicatypia (hematoxylin-eosin: HEx200).

Post-operative course was free from complications and the patient was discharged from the hospital six days after surgery. He was referred for chemotherapy.

Discussion

Most bladder tumors are urothelial cancers and other types of tumors are rare (less than 5%).

Altough leiomyosarcoma are presents the most common urinary bladder sarcoma, itaccounts less than 0.7 % of all primary bladder tumors(6). Leiomyosarcoma arise from normal urinary bladder smooth muscle.The most frequent clinical presentation is gross hematuria, followed by dysuria and pollakuria. Otherwise, the tumor may present as a hypogastric mass with abdominal pain(7). In our case, gross hematuria with abdominal hypogastric pain was observed.

Since clinical and radiographic aspects are not suggestive, such as for our patient, pathological examinations with immunohistological study are essential to confirm the diagnosis. Histologically, leiomyosarcoma is characterized by thepresence of a spindle cell proliferation forming bundles and fascicles. Cells present cigar shaped nuclei with cytologic atypia and mitotic figures(8). Immunohistology study confirms the smooth muscular origin and eliminates differential diagnosis. So leiomyosarcoma is immunoreactive for Smooth Muscle Actin (SMA), h-caldesmon and Desmin. Tumors cells are negative for other markers (epithelial, vasuclar and skeletal muscle). Such as sarcomas in other sites, urinary bladder leiomyosarcoma is graded into grade 1, 2 or 3 (for high)(9). Increasing grade, as well as age, stage and not curative resection, get worse survival (8,9).

Urinary bladder leiomyosarcomas are potentially aggressive neoplasms that require aggressive surgical resection, and radical cystectomy with wide margins should be performed whenever possible (10,11)

In a systematic review of the literature reported by Zieschang et al. (3) and information regarding treatment and outcome is scattered in the scientific literature. Interested clinicians and patients have to undertake troublesome search for treatment and outcome information.

Material and methods

We performed a systematic review of the literature using the PubMed and Web of Science

databases and included all identified cases published in English language between 1970 and June 2018 into a meta-analysis. Prior to the literature search, key questions were formulated and with the data obtained, answers to these questions should be derived.

Results

We analyzed clinical data of 210 cases of urinary bladder leiomyosarcoma revealed by this review and seen in our institution. The mean age of patients was 52 years. The majority (75% in 2019, 210 cases of urinary bladder leiomyosarcoma were analyzed. The most promising treatment option seemed to be surgery (radical or partial cystectomy) with negative resection margins, possibly supplemented by chemotherapy or radiation.

Adjuvant radiotherapy may be used for patients with positive surgical margins. Patients with local recurrence or metastatic bladder sarcomas should receive systemic chemotherapy based on doxorubicin and cisplatin, and/or radiotherapy (10–12).In our case, radical cystectomy was performed. The margins were negative and the tumor was grade 2, therefore, no adjuvant treatment was received. Unfortunately, a metachronous liver metastasis was diagnosed three years after surgery.

Other factors were associated with the development of distant metastasis including large size, deep location and recurrent disease The most common metastatic sites are lungs, liver and bone(4,5,13).

There have been few case reports of leiomyosarcomas with liver metastasis.

Faraj et al. (14) reported five cases of liver resection for metastatic colorectal leiomyosarcoma, with a median survival of 47 months.

Lang et al. (15) reviewed a total of 26 cases, who underwent 34 liver resections. Median survival was 32 months after R0 resection and 20.5 months after R1/2 resections. The 5-year survival rate was 13% for all patients and 20% after R0 resection.

Kim et al. (1) reported 10 cases with liver metastasis of intra-abdominal leiomyosarcoma which underwent surgical resection. The primary lesion was a retroperitoneal mass in 5 cases. All patients had resection surgery for the primary lesion and two had surgery for the primary lesion and liver metastasis at the same time. Eight patients had a partial hepatectomy, and the others had right hepatectomy and left lateral sectionectomy. The median survival period was 34.2 months.

Van cann et al. (16) reported 122 cases of metastatic leiomyosarcoma. The primary lesion was intra-abdominal in 23 cases. Liver metastases were present in 33 cases. Ninety eight patients were treated by systemic therapy, 28 by metastasectomy and 46 by radiotherapy.The median survival from diagnosis of metastasis was 20.5 months. On multivariate analysis, metachronous disease, no progressive disease as best response to first line treatment, the possibility of metastasectomy with curative intent and use of palliative radiotherapy were indicators for better survival.

In our case, surgical resection of the liver metastasis was performed and adjuvant chemotherapy was initiated.

It has been established that hepatic metastasectomy with R0 resection can improme survival in sarcoma patients. The main problem is that many patients are not surgical candidates (17–19). Leiomyosarcoma is known to be relatively chemoresistant, and chemotherapy response rates are poor. Doxorubicin is the first-line chemotherapy for patients with unresecable metastatic leiomyosarcoma. Liver-directed therapies (transarterial chemoembolization, radioembolisation and microwave ablation) are promising alternatives to systemic chemotherapy (4,20).

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