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Leiomyosarcoma Of Urinary Bladder

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Abstract

Mesenchymal tumours represent a small number of bladder malignancies. Leiomyosarcoma is the most common histological variant with 0.1% of bladder cancer. This tumour has been historically considered as highly aggressive and shows a poor prognosis. Despite very low survival rates showed in older reports, some authors indicate that some patients could have a better outcome. Immunohistochemical studies play an important role in the diagnosis of this tumour. Wide surgical excision with adjuvant chemotherapy is the mainstay of treatment. We report a case of 50 year old female presented with high grade leiomyosarcoma of urinary bladder.

Key Words: Leiomyosarcoma, urinary bladder, immunohistochemistry

INTRODUCTION

The urinary bladder cancer are rare neoplsma. The 95% of bladder tumour are epithelial in origin, while mesenchymal tumours accounting less than 5% in adults.[1] Essentially any mesenchymal tumour can occur in adult bladder. Smooth muscle neoplasms (leiomyoma, leiomyosarcoma) accounting for over 75% of bladder mesenchymal tumours. Out of which 0.1% are liomyosarcoma. About 100 cases of liomyosarcoma reported in world medical literature till date.[2] There is a lack of consensus about a standard treatment, and little is known about natural history and prognosis of the tumour, due to a very low incidence.

As reported, most cases present with gross urinary symptoms, more often with an earlier onset. This may, eventually, lead to earlier diagnosis, allowing a safer therapeutic approach
The radiological evaluation leads to a final surgical approach to the disease. The high-grade of the tumour and involvement of one of the margin, indicates mostly a poor prognosis in these cases. We report a case of an adult female presented with abdominal mass and gross urinary symptoms including burning micturition, urinary incontinence and hematuria.

CASE PRESENTATION

A 50 yr old female presented with complaints of burning micturition, urinary incontinence, constipation and loss of appetite since 2-3 months. General condition of patient was fair, she was afebrile and vitals were normal. Per abdominal examination was not contributory. The patient had undergone hysterectomy 02 yrs back. No history of drug intake and addiction. Abdominal USG revealed a pelvic mass. CT scan confirmed the diagnosis of bladder tumour. Bilateral iliac node dissection with partial cystectomy with wide excision of bladder tumour was done. On the gross examination tumour was grey white in color covered with thin capsule, measuring 8 x 6.5 x 6.5 cms. At one place the tumour appears polyploidal, infiltrating the capsule (Fig. 1A). Cut surfaces appeared nodular with some necrotic areas (Fig. 1B). Six right and six left iliac nodes were recovered from the fatty tissue separately. Small node measured 0.75 x 0.5 x 0.5 cms and large one was 2.5 x 1.0 x 1.0 cms. Microscopic examination from tumour and lymph nodes revealed tumour partially lined by urothelial epithelium. The subepithelium and lamina propria showed necrosis, oedema and inflammatory infiltrate. The muscular layer showed high grade spindle cell tumour. The cells are arranged in fascicles and bundles showing storiform pattern, whorly pattern and criss-crossing each other. The cells have pink cytoplasm with spindle shaped nuclei with blunt ends and irregular chromatin. Mitotic activity is >10/HPF(Fig. 2A & 2B). Iliac nodes did not show any metastasis. The diagnosis of high grade spindle cell tumour (leiomyosarcoma) was given and immunohistochemistry (IHC) was advised for confirmation of histogenesis of tumour. IHC panel showed immunopositivity with desmin and vimentin (Fig. 3A & 3B) confirming the diagnosis of Leiomyosarcoma.

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DISCUSSION

Sarcoma is the most frequent mesenchymal malignancy of the bladder with leiomyosarcoma as most common histologic variant, according to two retrospective reviews of mesenchymal genitourinary tumors.[1] They occur at extremes of life, with rhabdomyosarcomas being the more common variety in children and leiyomyoma more common in adults. Most of the bladder tumors are located at the base of the bladder in adults aged 40 to 60 years.[3]

Histopathologically, leiomyosarcoma is composed of interlaced bundles of spindle cells with nuclear abnormalities. They differ from leiomyoma primarily in their infiltrative growth pattern, although the increased mitotic rate and nuclear pleomorphism are often helpful clues. The designation of leiomyoma has been reserved for circumscribed, noninfiltrating lesions with little or no mitotic activity and minimal cellular atypia. The immunohistochemical findings are important in differentiating leiomyosarcoma from other spindle cell-containing tumors. Ikegami et al. reported 14 cases of sarcomatoid carcinoma of bladder and found that the sarcomatoid component showed a positive immunoreaction to at least one of the epithelial markers, pankeratin or EMA.[4] Mills el al. reported 15 cases of bladder leiomyosarcoma and found that all of those cases were uniformly negative for cytokeratin and EMA, while all were positive for muscle-specific actin, and 8 cases were positive for desmin.[5] In this report the case exhibited immunopositivity

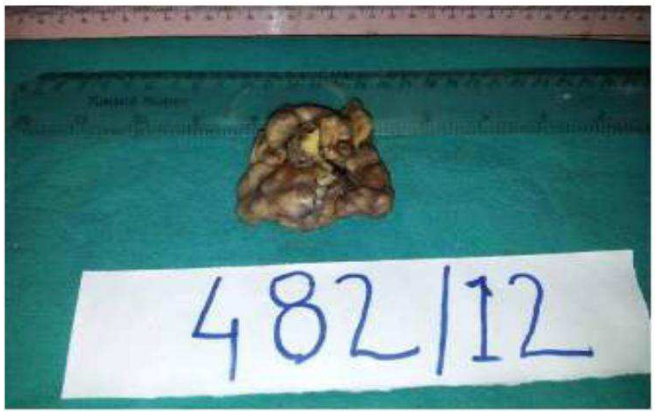

Figure -1A Gross photograph of tumour.

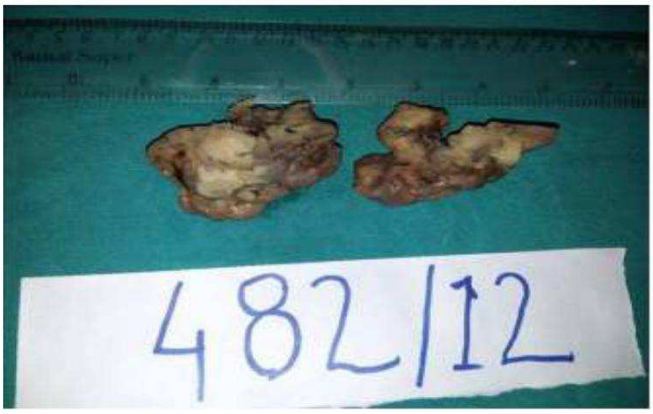

Figure 1B Cut section of the tumour showing nodularity along with necrotic areas.

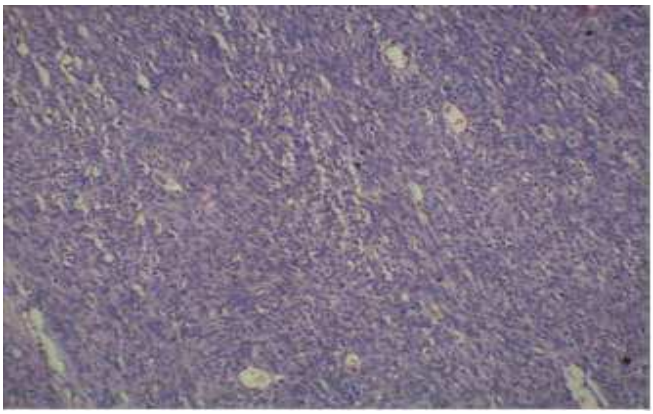

Fig- 2A: Microscopic photograph of tumour (100x) Showing proliferating tumour cells with eosinophilic cytoplasm.

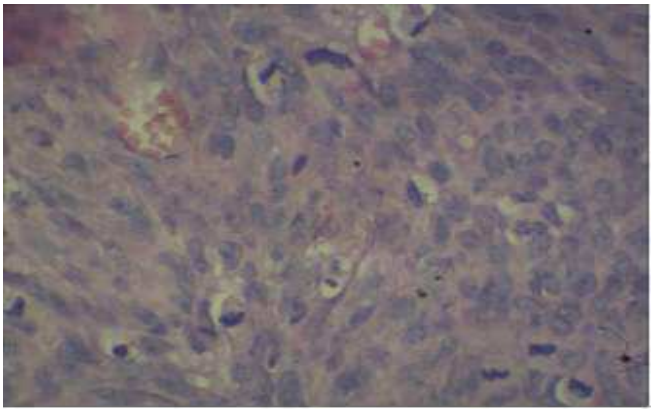

Fig-2B: Microscopic photograph of tumour(400x) showing spindle-shaped nuclei with irregular chromatin and numerous mitotic figures.

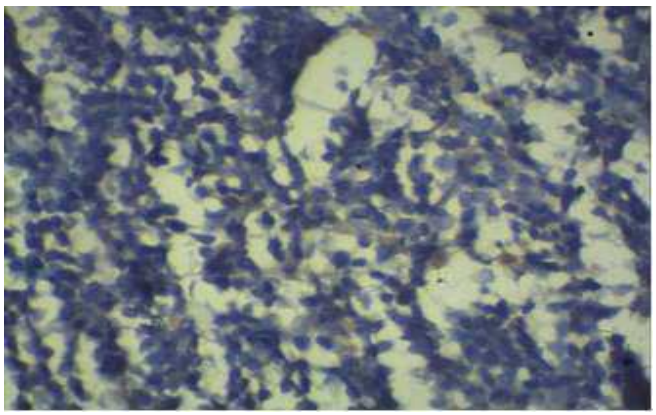

Fig-3A: Vimentin positivity (IHC) (400x)

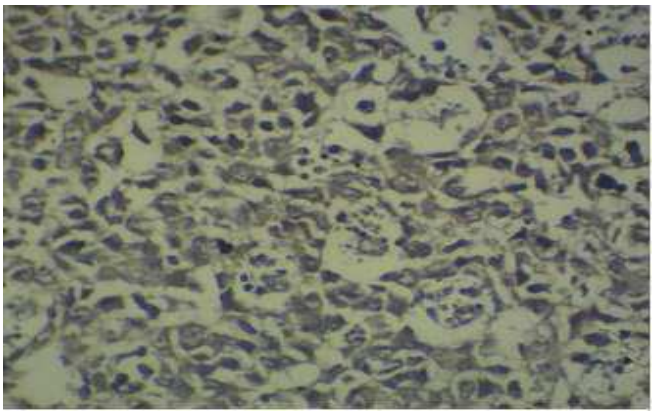

Fig-3B: Weak Desmin positivity (IHC) (400x)

for desmin and vimentin both.

There is no universally accepted grading system or staging system for leiomyosarcoma of urinary bladder. Mills et al used mitotic activity alone to distinguish low and high grade tumours. Tumours with <5 Mitotic figures/HPF had an excellent outcome whereas tumours with >5 Mitotic figures/HPF metastasized.[5] Martin et al considered tumours as low grade those showing mild to moderate nuclear atypia, <5 Mitotic figures/HPF and <25% necrosis. Where as those showing moderate to high nuclear atypia, >5 Mitotic figures/HPF and >25% necrosis were considered as high grade. By definition all tumours classified as leiomyosarcoma were infiltrative.[5,6]

Painless frank hematuria is the most common presenting symptom. Urinary frequency, dysuria, nocturia, urgency, urinary retention and recurrent urinary tract infections are less common presentations.[7] In our case burning micturition and urinary incontinence were the presenting complaints. Like other soft tissue sarcoma, wide surgical excision is the mainstay of primary treatment. Swartz et al. suggested that the curative treatment of choice is a partial cystectomy when the size and location of the tumour allow for adequate surgical margins. Radical cystectomy is indicated for more-extensive lesions.[8]

Multimodal treatment with pre and post operative chemotherapy setting targeting mesenchymal cancer (as sarcoma

therapy protocol using doxorubicin, ifosfamide, cisplatinum and docetaxel) should be mandatory for metastatic tumor.[2]

The prognosis was thought to be very poor traditionally. Ahlering et al reported 11 patients with leiomyosarcoma of the bladder and prostate. The authors achieved a 5-year survival rate with aggressive combination treatment involving radical surgery, preoperative and postoperative chemotherapy, and radiotherapy in 90% of the patients.[9] However, the best prognostic factor seems to be the presence of free margins. In addition to margin status, local invasiveness and size, as well as tumour grade, seem to play an important role in determining the final outcome.[1] According to one of the greatest series available to date, overall local recurrence of bladder leiomyosarcomas is about 16%, with most recurrences occurring in the pelvis. Overall recurrence of distant metastases is about 53%, with the most common sites of metastases being the lungs, liver, bone, and brain.[2]

CONCLUSION

Leiomyosarcomas of the urinary bladder are rare malignancy. Immunohistochemical studies are important to differentiate leiomyosarcomas from epithelial and other spindle cell proliferations. Surgical resection along with chemotherapy with doxorubicin and cis-platinum must be used for better prognosis.

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