

## CASE REPORT

# Intrapelvic malignant nerve sheath tumors presenting as acute urinary retention: A case report and literature review

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## Abstract

Malignant nerve sheath tumors (MPNSTs) are rare sarcomas tumors which rarely present as intrapelvic mass and are hard to diagnose clinically. We present a 29-year-old male patient presented with acute urinary retention and was diagnosed with large intrapelvic mass. After complete surgical resection, the histopathology confirmed the diagnosis of low MPNST.

## KEYWORDS

acute urinary retention, case report, malignant nerve sheath tumors, pelvic mass

## 1 | INTRODUCTION

A malignant peripheral nerve sheath tumor (MPNST) is a sarcoma that develops from the central or peripheral autonomic nervous systems.<sup>1</sup> MPNST is a benign tumor that can be seen sporadically in 0.001% of the general population; however, patients with neurofibromatosis type 1 (NF1) have an incidence of 0.16%.<sup>2</sup> The posterior thoracic cavity, retroperitoneal area, adrenal gland, and soft tissue of the head and neck are the mostly affected anatomical

sites.<sup>3</sup> MPNST pelvic retroperitoneum cases are rare and may arise from the sacral plexus.<sup>4</sup>

The clinical characteristics of MPNST, such as symptoms, malignancy transformation pathway, and attributes of metastasis are widely undefined and are still under investigation.<sup>5</sup> To date, there are few cases of MPNST originating from the pelvic area and causing urinary retention in the literature.<sup>5</sup> Hence, we describe our patients' clinicopathologic characteristics, including clinical follow-up, focusing on the disease diagnosis, treatment option, and outcome.

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## 2 | CASE REPORT

A 29-year-old man was referred to the urology outpatient clinic in May 2021 with a history of abdominal pain and acute urinary retention in the last week. The pain was mild and located in the suprapubic area. There was no history of malignancy in his family members or neurological problem. An abdominal examination revealed palpated suprapubic mobile mass with mild tenderness. Another physical examination, including a neurological examination, was normal. No symptoms of muscle wasting, weakness, or functional disability were observed. No skin manifestations for NFI such as cafe-au-lait were observed.

The blood investigation, including liver and renal function tests, was within normal limits. Urine analysis showed microscopic hematuria (15–20 RBCs/HPF). The abdomen and pelvis computed tomography (CT) scan revealed a large, well-defined, and smooth border soft tissue mass of about  $8.5 \times 6.5 \times 8.5$  cm in the midline and left paramedian pelvic region. The mass appeared capsulated with a smooth border without calcification and associated with a soft pressure effect on the adjacent structures, including the rectum and bladder. The bladder was displaced antero-superiorly and toward the right side. There was also a displacement and pressure effect on the prostate and seminal vesicles (Figure 1A). A sonography-guided True-cut biopsy from the mass revealed spindle cell fascicular mass with myofibroblastic differentiation and low proliferative activities.

The patient was admitted for surgery. After general anesthesia and low midline incision, the solid mass was identified and wholly excised without complication. The mass was a capsulated, yellowish color with a smooth surface measuring about  $11.5 \times 9 \times 7$  cm (Figure 1B). At the microscopic level, the tumor showed a low-grade tumor with moderate cellularity, mild increased nuclear to cytoplasmic ratio, and mild hyperchromasia with a neurofibroma-like pattern of growth without tumor necrosis or lymphatic invasion. Immunohistochemical (IHC) results were Ki-67: 1%–2%, CD56: positive, and SMA, S100, CD34, Staat 6, C-kit, Desmin, SOX 10: negative (Figures 2 and 3). On the third postoperative day, the

patient was discharged from the hospital without complications. After a 5-month follow-up, the patient was fine and had no signs of recurrence.

## 3 | DISCUSSION

MPNSTs are a rare soft tissue sarcoma that develops from the peripheral nerve or its sheath from Schwann cells and accounts for 30% of all soft tissue sarcomas. The prevalence of MPNSTs in the general population is 0.001%.<sup>5</sup>

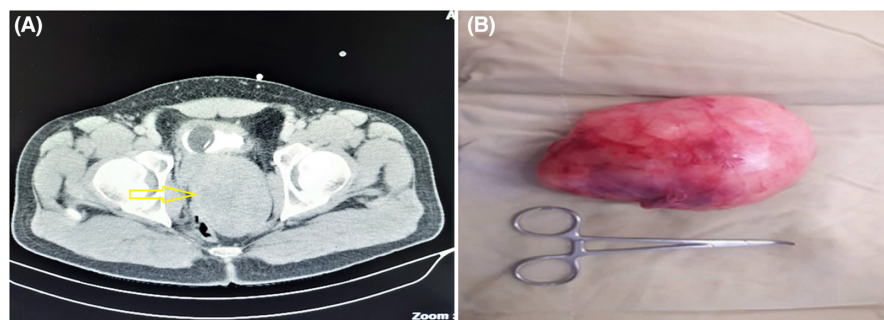
NF1, existing benign plexiform neurofibromas, and a history of radiotherapy are the most critical risk factors for MPNST development.<sup>4,5</sup> The sporadic MPNST affects both men and women equally, with peak incidence occurring in the seventh decade of life.<sup>6</sup> Our patient was a young man with no symptoms of NF 1.

MPNSTs can occur anywhere in the deep tissues along the peripheral nerves. The importance of intrapelvic MPNST lies in its diagnostic challenges in clinical practice. The lungs are the most commonly affected by systemic spread, and pulmonary metastasis is the leading cause of death in MPNST cases.<sup>4</sup>

Pelvic tumors grow slowly and cannot be noticed along the nerve path. They are usually harder to predict until symptoms appear.<sup>4</sup> Pain, neurological signs, and numbness are the most frequent clinical complaints in these patients. They can occasionally present with back or suprapubic pain, and symptoms caused by compression of adjacent tissue such as the bladder.<sup>7</sup> The location of the tumor in the posterior bladder wall, as seen in this patient, suggests an origin from one of the branches of the inferior hypogastric plexus. Ajani and associations reported a similar case with similar location.<sup>13</sup>

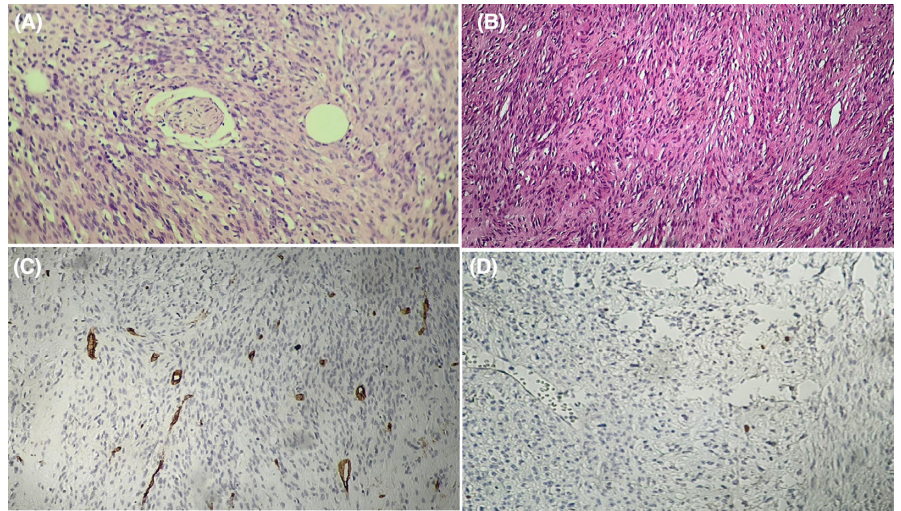
The intrapelvic area is an extremely rare location for MPNST, and only a few cases have been reported in the literature. Table 1 summarizes a recently reported intrapelvic MNST arising in the bladder wall.<sup>4,5,8–12</sup> Despite having a large tumor, our patient had no neurological complaints but had urinary retention due to mass compression.

Ultrasonography, CT scan, and magnetic resonance imaging (MRI) are helpful radiologic investigation methods.<sup>13</sup>

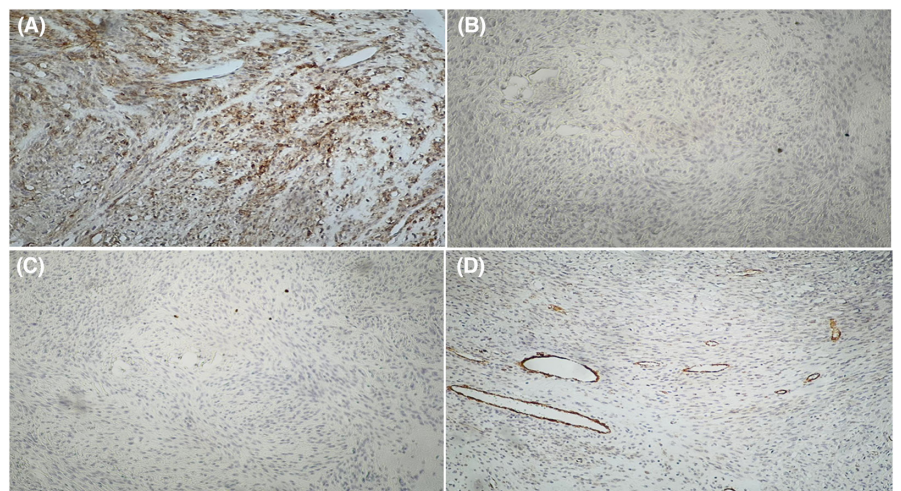


**FIGURE 1** Show (A); A computed tomography scan showed the mass with pressure on the bladder, rectum, and prostate. (B); Photography of the mass after surgical removal

**FIGURE 2** Show (A); Low-power view showing aggregates of foamy histiocytes between the tumor cells. (B); SD 1. (C); CD34. (D); Ki67



**FIGURE 3** Show (A); CD 56. (B); C Kit. (C); S100. (D); SMA



In our patient, only a CT scan showed a large, well-defined, smooth border soft tissue mass in the pelvic area. A similar protocol was performed by Benz et al.<sup>14</sup> However, MRI is still the most useful radiologic tool, which allows for assessing the associations of the tumor with adjacent structures.<sup>15</sup>

Because there is no definitive IHC or unique chromosomal anomaly, unique general histopathology, or clinical criteria, diagnosis of MPNST is incredibly difficult. MPNST is diagnosed in a combination of its clinical presentations, radiologic findings, and histologic representations. The S-100 protein and the Ki-67 index, on the contrary, are frequently used as IHC markers for MPNST.<sup>16</sup>

In our patient, the fine needle aspiration (FNA) biopsy was not conclusive and the final IHC showed to be negative for S-100 protein, and the Ki-67 index was 1%–2% in favor of low MPNST. Similarly, Ogose et al. reported that in MPNSTs around the pelvis, fine needle aspiration just gave a correct diagnosis in 4 out of 11 patients and core needle biopsy gave a correct diagnosis in all patients. In this study, all benign tumors were diffusely positive for S-100 protein in IHC analysis. In contrast, malignant tumors

were negative or focally positive for S-100 protein. The Ki-67 index was less than 4% in all benign tumors, while malignant tumors ranged from 7% to 36%.<sup>7</sup>

Complete surgical excision of intrapelvic mass is the standard gold treatment.<sup>4,17</sup> The goal of surgery is to eradicate the mass with tumor-free margins and relieve the symptoms. When tumors extend beyond the pelvis via the sciatic notch, a combined surgical approach should be considered, such as a transabdominal plus gluteal approach.<sup>17</sup>

As in our case, most benign pelvic tumors can be dissected bluntly after incising the epineurium over the mass. If the tumor involves the sciatic nerve or the pelvic plexus, complete resection may be difficult and may result in nerve damage.<sup>17</sup> Additional treatments, such as adjunctive radiotherapy and chemotherapy, have improved survival outcomes.<sup>17</sup> However, radical surgical removal is the optimal choice with a good prognosis.<sup>17</sup>

Huge size, high-grade tumor, proximal location, surgical margin with tumor invasion, NF1, and Ki-67 index greater than 7% are adverse prognostic factors associated with poor prognosis.<sup>18</sup> While tumor diameter of less than



TABLE 1 Summary of a recently reported intrapelvic MNST arising in the bladder wall

Author	Age (year) / Gender	Main symptoms	NF	Treatment	Status
Eltoum <sup>8</sup>	39/M	Hematuria	(-)	Cystoprostatectomy and adjuvant RT	Recurrence 2 months later
Petracco <sup>9</sup>	50/M	Urinary retention	(+)	Cystoprostatectomy	No recurrence after 18 months
Kalafatis <sup>10</sup>	57/M	Hematuria	(+)	Cystoprostatectomy with pelvic LND	No recurrence after 12 months
Ajani <sup>5</sup>	58/M	Abdominal swelling	(-)	-	Died after 12 days
Umakanthan <sup>11</sup>	52/M	Hematuria and pelvic mass	(-)	-	Lost during follow-up
Wang <sup>4</sup>	46/F	Abdominal pain	(-)	Mass resection	Pulmonary metastases after 3 years
Shen <sup>12</sup>	20/M	Urinary retention	(+)	Mass resection	Not mentioned

Abbreviations: M, male; F, female; NF, neurofibromatosis; RT, radiation therapy; LND, lymph node dissection.

5 cm, gross total resection of the tumor, and younger age were favorable prognostic variables.<sup>17</sup>

The 5 years survival rates are linked to NF1. The 5 years survival rate for patients without NF1 vs. those with NF was 50% and 10%, respectively.<sup>19</sup> Cai et al. reported in another study that the risk of recurrence is up to 40%, commonly with subsequent hematogenous metastasis, and the 5 years survival has varied from 15% to 66%.<sup>20</sup>

We present a rare case of substantial intrapelvic MPNST who presented with acute urinary retention. We aimed to highlight that while intrapelvic MPNST is uncommon, it should be considered in the differential diagnosis of pelvic masses because its symptoms can mimic other urologic conditions. To promote a better prognosis, we recommend open biopsy, surgical intervention, and a multimodal approach for diagnosing MPNST adequately.

## 4 | CONCLUSION

MPNSTs can occur in unusual locations such as the pelvic area without neurofibromatosis manifestation. They should be considered in the differential diagnosis of pelvic masses because their symptoms can mimic other urologic conditions, especially in patients with obscure urinary retention. Performing radiological imaging studies, open biopsy, and expert IHC of the mass help early diagnosis of these tumors. The gold standard treatment for these tumors is complete surgical resection.

## AUTHOR CONTRIBUTIONS

A A designed the study and were involved in the record collection. FA and S H wrote the manuscript. S T and A K edited the manuscript and provided guidance. FA conceptualized the study, designed the study, edited the manuscript, provided guidance, and approved the final version of the manuscript.

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## CONFLICT OF INTEREST

None.

## ETHICAL APPROVAL

None.


## CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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