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Oncology

Neurofibroma of bladder: A case report and literature review



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ABSTRACT

To improve the understanding and treatment level of urogenital nerve fibroma by sharing the clinical manifestations, imaging features, and pathological characteristics of a case of urogenital nerve fibroma. The patient was a middle-aged male with initial symptoms of painless gross hematuria, bladder irritation, and pelvic mass. Imaging examination showed a bladder mass, and transurethral bladder tumor resection was performed to reduce the tumor. Pathology was confirmed to be bladder neurofibroma. After 6 months of surgery, follow-up examination showed a significant reduction in tumor volume and no recurrence. The successful diagnostic and therapeutic experience of this case can be referenced for clinical practice.

1. Introduction

Neurofibroma of the genitourinary system is very rare in clinical practice, and neurofibroma of the bladder is the most common organ. Patients often experience clinical symptoms at a young age, with a much higher proportion of male patients than female patients. We report a case of primary neurofibroma of the bladder in a 36-year-old male whose clinical symptoms were clinical symptoms of painless gross hematuria, bladder irritation, and pelvic mass. Pelvic magnetic resonance imaging shows thickening of the right posterior wall of the bladder, with irregular soft tissue signal shadows protruding into the cavity. The prostate and bilateral seminal vesicles are also affected. Through cystoscopy and pathological biopsy, it was found to be consistent with neurofibroma of the bladder. Immunohistochemical examination further confirmed the diagnosis.

2. Case presentation

The patient, a 36-year-old male, presented to our hospital for 2 days due to painless gross hematuria accompanied by lower abdominal pain; The patient has a family history of neurofibroma (his grandmother and father); Physical examination: A cystic mass can be palpated in the bladder area of the lower abdomen, and multiple cystic masses can be palpated along the spine in the lower back (Fig. 1 A).Multiple coffee-

latte spots and subcutaneous soft tissue masses are found on the skin of the whole body (Fig. 1B). Urinary tract ultrasound shows a solid cystic mass in the pelvic cavity. Further magnetic resonance imaging of the lumbosacral vertebrae and pelvic cavity revealed enlargement of the sacral canal and some sacral foramen, with a local cystic protrusion towards the pelvic cavity (Fig. 2A and B). Nodular shadows were observed in the subcutaneous fat layer of the lumbosacral region, with larger ones ranging from approximately 1.9cm–1.3cm. Thickening of the trigone and right posterior wall of the bladder, with irregular soft tissue signal shadows protruding into the cavity, with a maximum cross-section of approximately 4.6cm \times 3.3cm(Fig. 2C). A cystic shadow can be seen between the posterior wall of the bladder and the sacrum, with a size of approximately 4.9cmX2.9cm. The prostate and bilateral seminal vesicles are enlarged with uneven signals(Fig. 2 D).

3. Treatment and outcome

The patient has persistent symptoms of hematuria and bladder irritation. After admission, the patient underwent transurethral cystoscopy and bladder tumor biopsy; During the operation, a bladder diverticulum was visible on the lateral wall of the bladder, and a solid mass with a wide base was observed on the right posterior wall of the bladder, measuring about 5cmX3cm in size. The surface showed mucosal redness, swelling, and easy bleeding; Clamp the tissue and send it for

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Fig. 1. Preoperative physical examination pictures. A:Back physical examination picture; B:Abdominal physical examination picture.

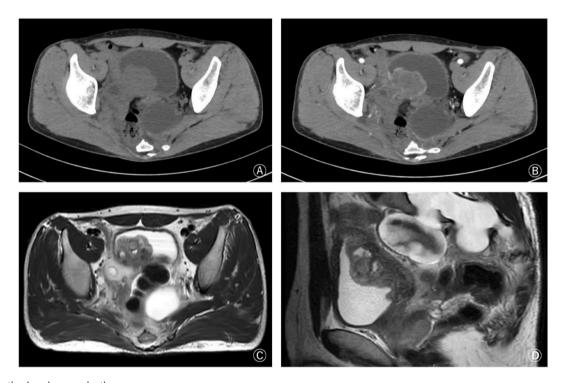


Fig. 2. Preoperative imaging examination.

A:Preoperative non-enhanced Bladder CT image; B:Preoperative enhanced CT; C:Coronal magnetic resonance imaging; D:Sagittal magnetic resonance imaging.

pathological examination. The pathological results indicate that there is spindle cell proliferation in the stroma (biopsy of bladder tumor), combined with immunohistochemical results, it is considered as neurofibroma. If necessary, NF gene testing can be performed to further clarify neurofibromatosis. Vimentin (+), SOX-10 (+), S100 (+), CK (-), EMA (-), CD21 (-), CD35 (-), CD117 (-), DOG-1 (-), CD34 (vascular+), Bcl-2 (weak+), CD31 (vascular+), CD68 (histiocyte+), Desmin (-), SMA (-), B-catenin (plasma+), Ki67 (3–5%+)(Fig. 3A–F).

After being diagnosed with neurofibroma of the bladder, considering the patient's younger age and medical needs, transurethral resection of the bladder tumor was performed on the patient; Postoperative pathology was still considered as bladder neurofibroma. After surgery, the patient's symptoms of hematuria and bladder irritation disappeared, and a follow-up urological enhanced CT scan at the 6th month after surgery showed a significant reduction in bladder tumor burden (Fig. 4A and B). Re-examination of cystoscopy 6 months after surgery showed that the tumor volume was significantly reduced, and the patient refused to undergo a second transurethral resection of the bladder tumor (Fig. 4C and D). The work has been reported in line with the SCARE

criteria.

4. Discussion

Type I neurofibromatosis (NF1) is an autosomal inherited disease.² Mutations in the NFI gene can develop into neurofibroma.^{3,4} Patients often experience symptoms in the early stages or even adolescence, and neurofibromatosis lesions can occur in the prostate, urethra, testes, spermatic cord, and ureter of the urinary system, but the bladder is the most common site.^{5,6} As reported in this case, bladder neurofibroma typically originates from the pelvic cavity, bladder nerves, and prostate plexus.⁷ Common clinical features include gross hematuria, difficulty urinating, recurrent urinary tract infections, bladder irritation symptoms, and pelvic masses. In isolated cases of bladder neurofibroma, differential diagnosis of leiomyosarcoma, ganglioneuroma, and paraganglioma should be considered.⁸ In addition to clinical manifestations, imaging examinations can help evaluate lesions in the abdominal, pelvic, and spinal regions, especially in cases where neurofibromatosis is not yet clear in the initial patient.⁹ Imaging examinations include

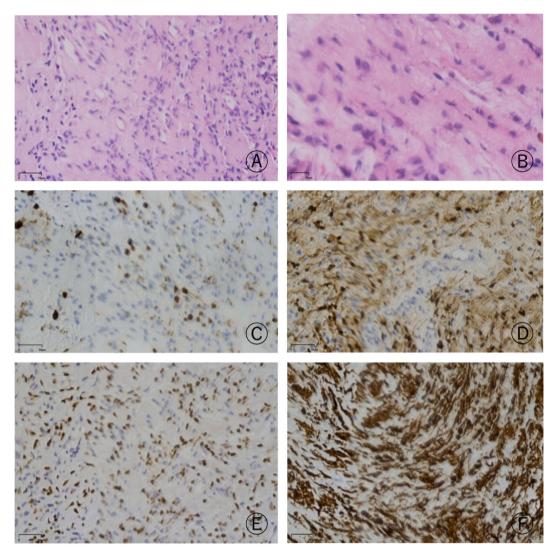


Fig. 3. Microscope Images (HE) and Pictures of immunohistochemical pathology results. A:HE(X100); B:HE(X200); C:Ki67(+); D:S100(+); E:SOX-10(+); F:Vimentin(+).

abdominal enhanced CT, MRI, cystoscopy, and biopsy pathology. Enhanced CT can evaluate the depth and metastasis of bladder neurofibroma invasion, but the CT manifestations of bladder wall ganglioneuroma or other tumors may be similar. 10,11 In this case, contrast-enhanced CT of the urinary system showed diffuse thickening of the bladder wall, irregular soft tissue mass shadow on the right posterior wall of the bladder, with lobulated edges and no significant enhancement. Enhanced CT with unclear enhancement cannot be used to completely clarify bladder neurofibroma, but it can be used to distinguish the possibility of malignancy or metastasis of the tumor. ¹² In our case, imaging examinations did not detect the possibility of malignant tumor metastasis. So further pathological examination is needed. In terms of histopathology, the histological manifestations of neurofibroma may be similar to low-grade malignant schwannoma, leiomyosarcoma, and rhabdomyosarcoma. Bladder neurofibroma usually presents with proliferation of spindle shaped cells in the stroma. Neurofibroma cells are composed of medium-sized spindle shaped cells with slender nuclei, and spindle shaped cells are mostly S-100 protein positive in immunostaining.8 The unique clinical manifestations, imaging, and immunohistochemical features of neurofibroma of the bladder can usually provide a clear diagnosis. 13 Due to the benign nature of bladder neurofibroma and its good prognosis, asymptomatic patients can choose to undergo regular follow-up monitoring 14 or medication treatment.

Studies have shown that regular use of selumetinib can lead to the reduction of neurofibroma and prevent the development of related complications. ¹⁵ For patients who continue to experience bladder irritation symptoms, anticholinergic drugs can be used for treatment. If the disease progresses during conservative treatment, such as obstructive urination symptoms or persistent hematuria due to lesion occupancy, surgical intervention becomes the preferred treatment method for symptomatic patients. Transurethral bladder tumor resection or partial cystectomy is suitable for patients without upper urinary tract obstruction. ¹⁶ If ureteral obstruction and hydronephrosis occur, ureteral orifice replantation can be considered as a surgical option. At the same time, rapid frozen histological examination of the distal ureter is required during surgery to avoid residual affected tissue. ¹⁴

5. Conclusion

The unique clinical manifestations, imaging, and immunohistochemical features of bladder neurofibroma can usually be clearly diagnosed. As bladder neurofibroma is a benign tumor with a good prognosis, regular follow-up monitoring can be chosen for asymptomatic patients. If patients experience obstructive urination symptoms or persistent hematuria due to the occupying space of the lesion, surgical intervention becomes the preferred treatment method for symptomatic

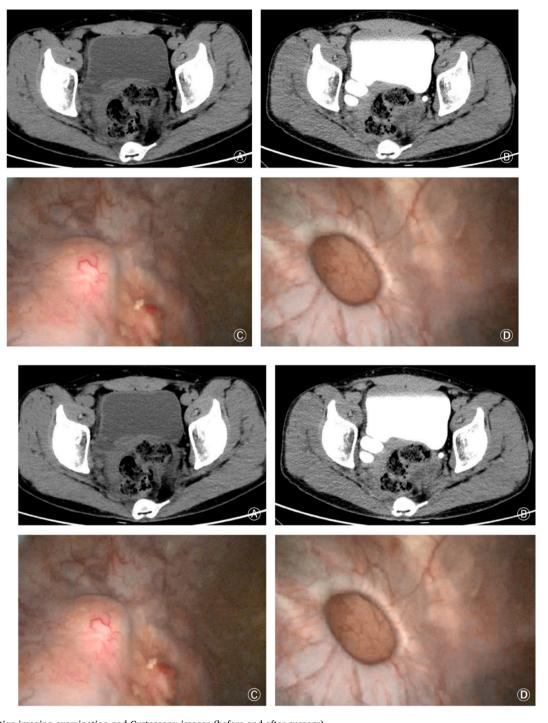


Fig. 4. Postoperative imaging examination and Cystoscopy images (before and after surgery).

A:Postoperative non-enhanced Bladder CT image; B:Postoperative enhanced CT; C and D:Postoperative Cytocopy images.

patients, while transurethral bladder tumor resection or partial cystectomy is suitable for patients without upper urinary tract obstruction.

CRediT authorship contribution statement

Zhipeng Yan: Writing – original draft. Baoqun Xu: Data curation. Maokun Sun: Data curation, Investigation. Heng Zhang: Data curation. Xiaolu Sun: Data curation. Lihua Li: Data curation, Investigation. Yulan Gao: Formal analysis, Investigation, Supervision. Yunji Sun: Writing – review & editing, Supervision, Validation.

Patient consent for publication

Signed informed consent was obtained from the patient for publication of this case report and any accompanying images.

Availability of data and materials

All data generated or analyzed during this study are included in this published article.

Ethics approval and consent to participate

The study is approved by the Ethical Committee of Shandong Provincial Third Hospital.

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Declaration of competing interest

The authors declare that they have no competing interests.

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