Case Report

ANNALS OF MEDICINE & SURGERY

OPEN

Sacral chordoma: an unusual cause of neurogenic bladder – a case report

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Introduction and Importance: Sacral chordoma is a rare tumor arising from notochord remnants. Sacral pain is the most common sign. The authors report the case of a sacral chordoma diagnosed in a urology department. It was a rare cause of erectile dysfunction and it was unusually revealed by lower urinary tract symptoms (LUTSs).

Case Presentation: A 67-year-old man had severe dysuria associated with erectile dysfunction. On digital rectal examination, we palpated a mass protruding through the posterior wall of the rectum. The gluteal region is infiltrated by a firm mass. MRI revealed an osteolytic sacral tumor. A percutaneous biopsy of the tumor was performed. A pathological examination confirmed the diagnosis of sacral chordoma. The patient had palliative radiotherapy. One year after radiotherapy, he had rectal stenosis associated with severe constipation.

Clinical Discussion: Sacral chordoma is a rare malignant tumor. Sacralgia is the most common sign. LUTS and erectile dysfunction were rarely reported in patients affected by this bone tumor. MRI is very suggestive of this pathology. Diagnostic confirmation requires pathological examination. The curative management consists of a large resection of the tumor. For inextricable tumors, palliative radiotherapy can be carried out. The prognosis of sacral chordoma is defined by a high recurrence potential and a low metastatic risk.

Conclusions: Sacral chordoma is a rare bone tumor. It is an unusual cause for LUTS and erectile dysfunction. Complete tumor excision is the only curative treatment. Recurrence after treatment is the main challenge for surgeons.

Keywords: biopsy, case report, erectile dysfunction, neurogenic bladder, radiotherapy, sacral chordoma

Introduction

Chordoma is a bone tumor arising from embryonic notochord remnants. It is a rare malignant tumor with an annual incidence estimated between 0.5 and 8 cases per 1,000,000 inhabitants[1]. The sacrococcygeal region is the most common site of chordoma[2,3]. The mitotic activity of tumor cells is very low[4]. Consequently, the clinical expression of this tumoral disease is insidious and the diagnosis is often made tardily. However, it is a locally aggressive tumor.

HIGHLIGHTS

- Chordoma is a rare malignant tumor.
- Sacral chordoma is a rare cause of neurogenic bladder and erectile dysfunction.
- On MRI, chordoma is typically lobulated and has a T2 hyperintense signal.
- Complete tumor resection is the only curative treatment for chordoma.
- Recurrence after treatment is the main challenge for surgeons.

We report an unusual case of the neurogenic bladder caused by locally advanced sacral chordoma. Erection dysfunction was a rare clinical sign of this sacral tumor.

This case report has been reported in line with the SCARE Criteria[5].

Case presentation

A 67-year-old man, who had no pathological antecedent, was referred to our urology consultation for the management of lower urinary tract symptoms (LUTSs).

The disease history dates back to a year when he had severe dysuria complicated with acute urinary retention. He also had a recent severe erectile dysfunction (International Index of Erectile Function score = 6) (Table 1). Furthermore, he reported mild sacral pain that worsened when sitting (visual analog scale = 2/10).

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Sponsorships or competing interests that may be relevant to content are disclosed at the end of this article.

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Annals of Medicine & Surgery (2023) 85: 41–45

Received 15 October 2022; Accepted 25 December 2022

Published online 18 January 2023

http://dx.doi.org/10.1097/MS9.0000000000000209

Table 1

The International Index of Erectile Function (IIEF-5) Questionnaire.

Over the past 6 months						
How do you rate your *confidence* that you could get and keep an erection?	Very low 1	Low 2	Moderate 3	High 4	Very high 5
When you had erections with sexual stimulation, *how often* were your erections hard enough for penetration	Almost never/never 1	Much less than half the time 2	About half the time 3	Much more than half the time 4	Almost always/always 5
During sexual intercourse, *how often* were you able to maintain your erection after you had penetrated (entered) your partner?					
During sexual intercourse, *how difficult* was it to maintain your erection to completion of intercourse	Extremely difficult 1	Very difficult 2	difficult 3	Slightly difficult 4	Not difficult 5
When you attempted sexual intercourse, *how often* was it satisfactory for you?	Almost never/never 1	Much less than half the time 2	About half the time 3	Much more than half the time 4	Almost always/always 5

IIEF-5 scoring, the IIEF-5 score is the sum of the ordinal responses to the five items:
22–25: no erectile dysfunction.
17–21: mild erectile dysfunction.
12–16: mild to moderate erectile dysfunction.
8–11: moderate erectile dysfunction.
5–7: severe erectile dysfunction.

Table 2

WHO performance status.

Grade	Definition
0	Asymptomatic: fully active, able to carry on all predisease activities without restriction
1	Symptomatic but completely ambulatory: restricted in physically strenuous activity but ambulatory and able to carry out work of a light or sedentary nature. For example, light housework and office work
2	Symptomatic, <50% in bed during the day: ambulatory and capable of all self-care but unable to carry out any work activities. Up and about more than 50% of waking hours
3	Symptomatic, > 50% in bed, but not bedbound: capable of only limited self-care, confined to bed or chair 50% or more of waking hours
4	Bedbound: completely disabled. Cannot carry on any self-care. Totally confined to bed or chair
5	Death

On physical examination, the patient had a good health status (WHO performance status = 0) (Table 2).

On digital rectal examination, we palpated a painless mass protruding through the posterior wall of the rectum. The prostate was enlarged without suspicious lesions. In the prone position, we noticed the presence of an excrescence slightly deforming the left hemisacrum (Fig. 1). On palpation, the tumor was noninflammatory and firm. No neurological deficit was revealed in the lower limbs.

For a better exploration of the LUTS, urodynamic testing was performed. The cystometry objectified a low-compliant and overactive bladder (Fig. 2).

MRI revealed a large osteolytic tumor centered on the sacrum and associated with intracanal extension (Fig. 3). As part of the extension assessment, a thoracoabdominopelvic computed tomodensitometry did not reveal distant metastasis.

A junior trainee assisted by a senior performed, in our urological department, a percutaneous fine-needle biopsy of the gluteal portion of the sacral tumor. No radiological control was necessary for this prominent tumor.

Four biopsy fragments were addressed for the pathology department. On histological examination, the tumor consists of cell masses and cords with a basophilic chondromyxoid matrix. The tumor cells display an 'epithelioid' appearance. The nuclei are rounded and monomorphic with focal cytoplasmic pseu-

Figure 1. Clinical photography of the sacral mass.

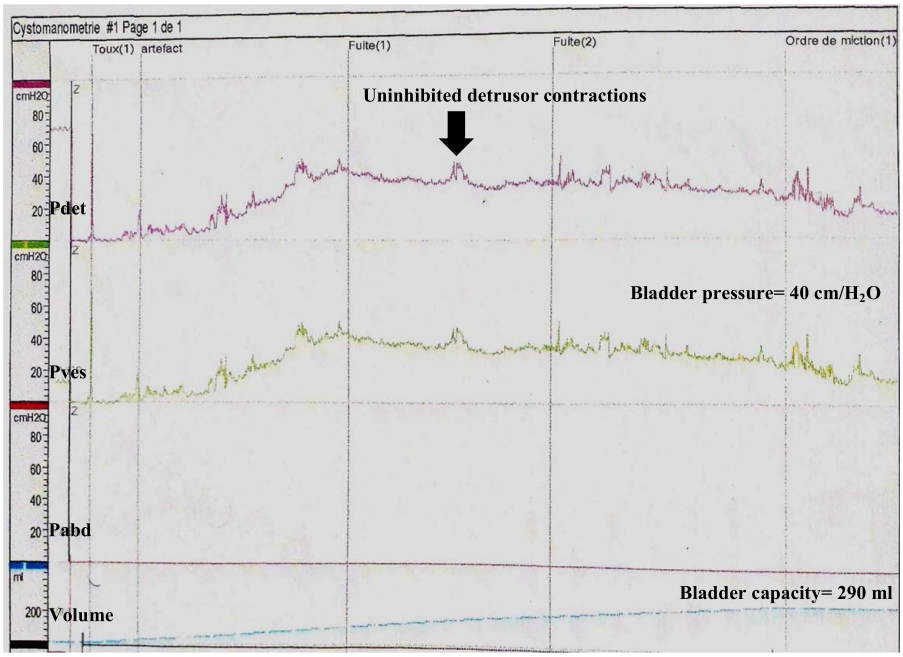

Figure 2. Cystomanometric study. *During the filling phase, uninhibited detrusor contractions were* observed. The intravesical pressure rised to 40 cm H_2O. This lower bladder compliance (Compliance= 8.33 ml /cm H_2O) was associated with a reduced capacity.

doinclusions (Fig. 4). No mitosis was identified. On immunohistochemical study, chordoma is positive for pan cytokeratin and S100 protein.

Considering the clinical data, radiological, and pathological characteristics of the tumor, the diagnosis of locally advanced sacral chordoma was made.

The patient had palliative radiotherapy. For the management of LUTS related to neurogenic bladder, we have indicated intermittent urinary self-catheterization combined with anticholinergic.

At the last follow-up dating back to one year after radiotherapy, the patient had an alteration in health status (WHO performance status = 2). Sacral pain had increased (visual analog scale = 6/10). He also reported severe constipation. However, LUTS had been significantly improved.

A digital rectal examination revealed rectal stenosis. The gluteal portion of the sacral chordoma had slightly increased in size.

Discussion

Chordoma is a rare bone tumor that arises from embryonic remnants of the notochord[3,6]. The annual incidence of this pathology varies between 0.5 and 8 cases per 1,000,000 inhabitants[1]. The sacrum represents the main site for the

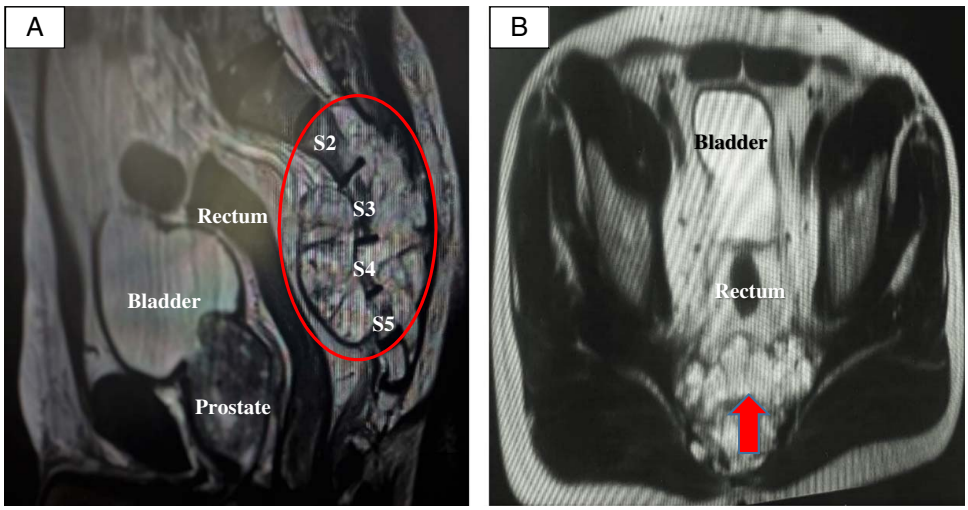

Figure 3. MRI showing a tumor mass (red circle) centered on the sacrum, which is lobulated, well-circumscribed (a: sagittal MRI) with hyperintense T2 signal (b: axial MRI).

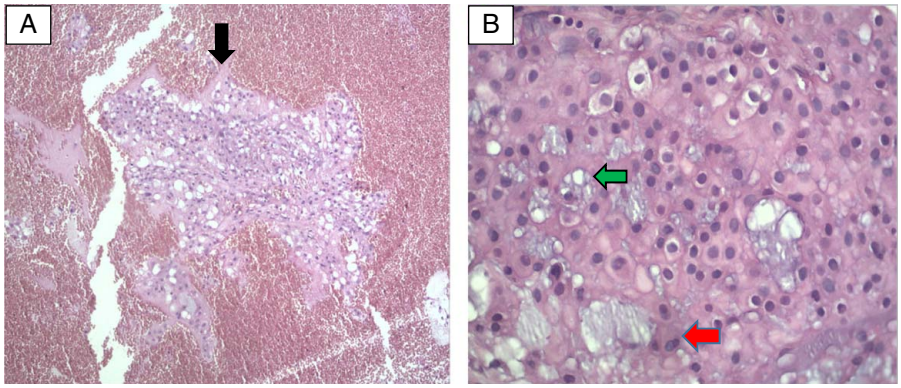

Figure 4. Pathologic aspect of sacral chordoma. (A) Tumor cells are organized in masses and cords. They are separated from chondromyxoid matrix by fibrous septa (black arrow). (Hematoxylin & eosin, HE × 40). (B) Epithelioid cell (red arrow): the cytoplasm is eosinophilic and contains more than one nucleus. Physaliphore cell (green arrow) has multivacuolar cystoplasm (Hematoxylin & eosin, HE × 200).

development of the chordoma[3,7]. In our case, four sacral vertebrae (S2, S3, S4, and S5) were affected by the tumor. This pathology occurs preferentially in men with a sex ratio of 3 : 1[8]. It can be observed at any age, with a peak around the fifth decade[9]. Several theories have been formulated regarding the etiopathogenesis of this neoplasm[1].

Patients with sacral chordoma often consult with a locally advanced disease. In our case, the patient had a large sacral tumor with invasion of the gluteal muscles. The clinical expression of this neoplasm is varied and equivocal. Low back pain is the most frequently reported sign[3]. In the series of Ozaki *et al.*[10], 11 out of 12 patients with sacral chordoma had pain in the sacrococcygeal region.

LUTS can rarely reveal sacral chordoma. They are related to an underactive bladder. This detrusor underactivity is secondary to the invasion of the nerve plexuses involved in the neural control of micturition. Clinically, patients may report voiding and storage symptoms. The urodynamic study shows hypocontractile and hyposensitive detrusor. In our case, the bladder demonstrated a mixed functional profile. Urodynamic signs of an overactive bladder are explained by autonomic nervous system disorders[11].

Sacral chordoma can rarely affect the patient's sexuality by infiltrating the sacral center of erection (S2, S4). Consequently, reflex erection is impaired. Few studies have evaluated erectile function in these cancer patients[12]. In our case, the patient had a low International Index of Erectile Function-5 score.

Bowel dysfunction can also be associated with LUTS[13]. Constipation was the only digestive sign reported by our patient.

Furthermore, rare cases of sensorimotor disorder of the S1–L1 dermatomes have been described in patients with proximal sacral chordoma (S1, S2)[14]. Up to 9 months of follow-up after radiotherapy, no neurological disorder affecting the lower limbs was observed in our patient.

In the majority of cases, physical examination reveals a gluteal or posterior perineal mass[3]. The digital rectal examination assesses the endopelvic extension of the sacral tumor. In our case, invasion of the left gluteal region was evident on palpation. On digital rectal examination, we palpated the endopelvic part of the tumor.

MRI is the reference radiological examination for chordoma. In accordance with our case, it is typically a lobulated osteolytic mass with a T2 hyperintense signal[15].

Macroscopically, the tumor is lobulated and dotted with gelatinous nodules. It can include foci of hemorrhagic necrosis. Calcifications can rarely be observed[4,16]. Histologically, tumor cells can be organized in cords, spans, or layers. They are separated from a myxoid stroma by fibrous septa[4]. Sacral chordoma consists of two types of cells[4]:

(1) Physaliphore cells: the cystoplasm is multivacuolar. The nucleus has a spider web appearance.
(2) Epithelioid cells: the cytoplasm is eosinophilic and can contain more than one nucleus.

In the case of our patient, a percutaneous biopsy was the diagnostic tool allowing anatomopathological examination of tissue samples. The tumor was composed mainly of epithelioid cells. Rare physaliphore cells were observed.

Sacral chordoma can pose a differential diagnosis with mucus-secreting adenocarcinomas, myxoid chondrosarcomas, and metastases of a clear cell carcinoma[4].

The immunohistochemical profile of chordoma associates epithelial differentiation antigens (cytokeratin) with mesenchymal differentiation antigens (S100 protein)[16].

The mainstay of treatment is surgical resection of the tumor. In 30–40% of cases, the excision is incomplete, indicating adjuvant radiotherapy[8,16]. For inextricable tumors, as is the case in our observation, palliative radiotherapy can be temporarily beneficial[17].

The natural history of chordoma is characterized by slow tumor growth and low metastatic potential[18]. However, these tumors are locally aggressive and can cause major functional sequelae[3]. In our case, sexuality and micturition were severely impaired. After complete excision, the main challenge for surgeons is the high risk of local recurrence[19]. The mean survival of patients who underwent radiotherapy alone was 5.4 years. At 9 months after radiotherapy, our patient had clinical tumor progression causing symptomatic rectal stenosis.

Our study highlights the urological (neurogenic bladder) and sexual (erectile dysfunction) complications revealing a rare bone tumor, which was unusually diagnosed in a urology department.

The short follow-up period is the main limitation of our case.

Conclusions

Sacral chordoma is a rare, locally aggressive tumor. It can be an unusual cause of LUTS and erectile dysfunction.

MRI is the gold standard in imaging. A pathological examination is necessary to establish the diagnosis.

Complete tumor excision is the only curative treatment. For locally advanced tumors, palliative radiotherapy can be temporarily effective.

The prognosis of chordoma is defined by a high recurrence rate and a low metastatic risk.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Ethical approval

Given the nature of the article, a case report, no ethical approval is required.

Patient consent

Written informed consent was obtained from the patient for the publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Sources of funding

This study has not received any funding.

Author contribution

N.J., A.M., and A.S. participated in clinical data collection and interpretation. M.S. participated in the interpretation of the urodynamic study. S.F. performed the histological examination of the vertebral biopsy and is a major contributor in writing the manuscript. N.J., I.C., B.J., and S.C. conducted a literature review. N.J., A.S., and S.F. developed the study methodology and participated in the article writing process. A.M. and M.Y.B. reviewed and supervised the study. N.J., I.C., B.J., and S.C. carried out the edition of the study. All authors read and approved the final manuscript.

Conflicts of interest disclosure

The authors declare that they have no conflict of interest.

Research registration unique identifying number (UIN)

No registration is needed.

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Availability of data and materials

All data sets generated for this study are included in the article.

Acknowledgments

None.

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