Reminder of important clinical lesson

Unsuspected pheochromocytoma of the urinary bladder: reminder of an important clinical lesson

Harvinder Singh Pahwa, Awanish Kumar, Rohit Srivastava, Anurag Rai

Department of Surgery, CSMMU Lucknow, Lucknow, Uttar Pradesh, India

Correspondence to Dr Harvinder Singh Pahwa, pahwakgmu@yahoo.co.in

Summary

Bladder pheochromocytoma creates an extremely rare situation (0.06% of all bladder tumours). We came across a case with a complaint of intermittent episodes of haematuria. Cystoscopy revealed a solid, bluish submucosal growth with increased vascularity. Immediately after taking biopsy, the patient developed a sharp headache, chest heaviness and marked rise in blood pressure. The procedure was abandoned. Later, serum and urinary markers for pheochromocytoma were within normal limits. The histopathology report came out to be pheochromocytoma of the bladder. Later, partial cystectomy was carried out under general anaesthesia and histopathology confirmed the same. Thus, a surgeon/urologist should have a high index of suspicion for bladder pheochromocytoma while dealing with such cases if he comes across the characteristic symptoms of sharp headache, hypertension, palpitation, sweating, fainting or blurring of vision immediately after voiding or during the cystoscopic manipulation of tumour or cystoscopic findings of a submucosal supratrigonal vascular tumour with a bluish hue.

BACKGROUND

Pheochromocytoma is a catecholamine-secreting tumour of chromaffin cells and is a rare cause of hypertension. Unsuspected pheochromocytoma greatly increases the per operative mortality in patients undergoing even relatively minor surgical procedures.

However, most surgeons and anaesthesiologists are unfamiliar with pheochromocytoma of the urinary bladder. We report one such case of unsuspected pheochromocytoma of the urinary bladder with severe hypertensive episode during cystoscopic biopsy.

CASE PRESENTATION

A forty-year-old man presented in district hospital with a history of intermittent episodes of haematuria. Ultrasonography suggested an echogenic lesion in the urinary bladder that was misinterpreted as bladder stone and was planned for cystolithotomy. Per-operatively, a urinary bladder growth was seen and the procedure was abandoned without biopsy due to excessive bleeding. Blood transfusion was done and the patient was referred to our institute for definitive management.

INVESTIGATIONS

At our institute, a fresh ultrasound was done which was suggestive of a solid mass with mixed echogenicity. The patient was subjected to cystoscopy under spinal anaesthesia that revealed a submucosal solid, broad-based growth with increased vascularity and a bluish hue of about 2×3 cm size near the dome of the urinary bladder (figure 1). As soon as biopsy was taken, the patient started crying with severe headache and heaviness in the chest. Blood pressure rose to 220/160 mm Hg (preoperatively 128/76 mm Hg) in spite of proper anaesthesia. The procedure was abandoned and the irrigating fluid was

released. Blood pressure settled to normal and the patient did not require any antihypertensive in the postoperative period. In view of fluctuation in blood pressure, we clinically suspected this growth to be pheochromocytoma. The biopsy report revealed extra-adrenal pheochromocytoma. Later, estimation of serum levels of epinephrine and norepinephrine and 24 h urinary vanillyl mandelic acid came out to be normal. This patient has no complaints of headache or diaphoresis, during micturition with no hypertensive episodes either in the past or in follow-up. We planned for methyl iodo benzyl guanidine (MIBG) scan to rule out other sites of pheochromocytoma but since this facility was not available at our institute and the patient being unaffordable, he declined this study; we had to go further without MIBG scan. A CT scan was done preoperatively for staging, which showed an intensely enhanced lesion at the dome of the urinary bladder (figure 2). There was no extra vesicle extension or pelvic lymphadenopathy.

TREATMENT

The patient was given adequate fluids preoperatively for expansion of extracellular volume. Phenoxybenzamine and intravenous infusions of phentolamine, nitroprusside and metoprolol were kept ready to control hypertensive crisis and any unwarranted tachycardia/arrhythmia during surgery but were not needed in this case. Partial cystectomy was carried out under general anaesthesia. The bladder was approached through a midline infraumbilical incision; a growth (2×3 cm) was identified at the dome of the urinary bladder after anterior vesicotomy. The tumor was isolated without manipulation by using intestinal clamps and resected with adequate (2 cm) margins. The intraoperative and postoperative periods were uneventful. The histopathological examination of

Figure 1 Cystoscopic view: sub-mucosal solid vascular tumour with a bluish hue in the supratrigonal area.

resected specimen confirmed the previous biopsy report of extra-adrenal pheochromocytoma.

OUTCOME AND FOLLOW-UP

The patient made an uneventful recovery. In follow-up, we are sequentially monitoring the urinary and serum markers along with cystoscopy and ultrasonography. There are no standard set guidelines for the follow-up of bladder pheochromocytoma, although some have suggested MIBG scan on an annual basis.

DISCUSSION

Pheochromocytoma of the urinary bladder is rare, accounting for less than 0.06% of all bladder tumours.¹

Figure 2 CT scan showing an intensely enhanced lesion at the dome of the urinary bladder.

Extra-adrenal pheochromocytomas are rare tumours that are found in about 10% in adults and up to 30% in children suffering from phaeochromocytomas.²

Characteristic symptoms are sharp headache, hypertension, palpitation, sweating, fainting or blurring of vision immediately after voiding due to increased catecholamine release in association with bladder contraction during micturition.³ However, approximately half of the patients, such as in this case, lack these typical symptoms, a reason responsible for a large number of such cases being incidentally discovered (these unsuspected cases may be termed as incidentalomas). A common cause of intraoperative hypertension is insufficient depth of anaesthesia. The patient, in this case, noticed an oppressive sensation in the chest and severe headache during cystoscopic biopsy as the cause. The range or density of anaesthesia was sufficient for surgery. We could not find appropriate reasons to explain the hypertensive crisis at the time of cystoscopy and biopsy and the procedure had to be abandoned. Catecholamine release may have been facilitated when the bladder was distended or when the tumour was manipulated. The cause of the hypertension would have been more convincing if catecholamine levels had been measured from the blood samples obtained during the episodes.

The clinician must thus be aware of the adreno-sympathetic syndrome of headache, hypertension or blurring of vision with detrusor activity.³ Extracellular volume is contracted from the chronic sustained effect of the elevated catecholamine levels and should be re-expanded prior to removal of the neoplasm; otherwise, profound postoperative hypotension may result. Blood pressure is first controlled with α -blockade using phenoxybenzamine. β -Blockade may be added to counteract the rebound tachycardia of the former. Certain patients may require calcium channel blockers such as nifedipine to maintain adequate control. Intraoperative intravenous infusions of phentolamine and nitroprusside are kept ready for use in the case of a hypertensive crisis.

Strict control of blood pressure is mandatory prior to and during any surgery to prevent the potentially fatal

hypertensive crisis. Preoperative localisation of extra-adrenal pheochromocytoma and metastasis with I¹³¹-MIBG is essential.^{4 5}

Urinary bladder pheochromocytoma are located in the dome and above the trigone of the urinary bladder in up to 40% of the cases.³ Although transurethral resection has been reported, such resection rarely excises all tumour deposits as the tumour is intramural in location and sympathetic nerves run through the entire thickness of the bladder wall. Partial cystectomy is the most common procedure performed for this condition.^{5 6} For advanced disease, radical cystectomy with pelvic lymph nodal dissection has been advocated. Surgery provides the best chance of cure for the patient. There have been reports of laparoscopic excision of urinary bladder pheochromocytoma.^{6 7} Since histology cannot differentiate between benign and malignant pheochromocytoma, follow-up assumes prime importance. Sequential monitoring of blood pressure, urinary catecholamine studies and clinical observation of the persistence or recurrence of headaches may alert the clinician to the possibility of recurrent pheochromocytoma.⁵ These studies should be repeated on an annual basis with others recommending the addition of annual I¹³¹-MIBG scinti scanning.⁴

Competing interests None.

Patient consent Obtained.

REFERENCES

1. **Leestma JE**, Price EB Jr. Paraganglionoma of the urinary bladder. *Cancer* 1971;**23**:1063.
2. **Salanitri J**, Smith P, Schlicht S. Multifocal malignant extra-adrenal paragangliomas of the Organ of Zuckerkandl and urinary bladder. *Australas Radiol* 2001;**45**:229–32.
3. **Das S**, Bulusu NV, Lowe P. Primary vesical pheochromocytoma. *Urology* 1983;**21**:20–5.
4. **Lindsey CM**, DeHart HS, Glenn JF. Pheochromocytoma of the urinary bladder. *Urology* 1976;**7**:210–11.
5. **Thrasher JB**, Humphrey PA, Rajan RR, *et al.* Pheochromocytoma of urinary bladder: contemporary methods of diagnosis and treatment options. *Urology* 1993;**41**:435–9.
6. **Kozlowski PM**, Mihm FG, Winfield HM. Laparoscopic management of bladder pheochromocytoma. *Urology* 2001;**57**:365v–vii.
7. **Sweetser PM**, Ohl DA, Thompson NW. Pheochromocytoma of the urinary bladder. *Surgery* 1991;**109**:77.

Learning points

- ▶ Pheochromocytoma of the urinary bladder can surprise both the urologist and the anaesthesiologist during any invasive procedure such as cystoscopy or open surgery.
- ▶ It is prudent to have in mind that any patient developing hypertension or its symptoms thereof such as sharp headache, dizziness, blurring of vision, palpitation during micturition or during cystoscopic procedure or any submucosal solid supratrigonal growth in the urinary bladder with increased vascularity and bluish hue, might herald a diagnosis of pheochromocytoma and should be managed cautiously.
- ▶ Cystoscopic biopsy without proper preparation should be avoided in patients in whom pheochromocytoma is suspected on the basis of symptoms or cystoscopic findings.
- ▶ Whenever suspected, such patients must undergo preoperative MIBG scanning for confirmation of diagnosis and localisation of other sites of extra-adrenal pheochromocytoma.

This pdf has been created automatically from the final edited text and images.

Copyright 2012 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.

BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Pahwa HS, Kumar A, Srivastava R, Rai A. Unsuspected pheochromocytoma of the urinary bladder: reminder of an important clinical lesson. *BMJ Case Reports* 2012;10.1136/bcr-2012-006377, Published XXX

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow