



Letter

Phaeochromocytoma of the urinary bladder presenting with malignant hypertension and hypertensive retinopathy



Dear Editor,

Phaeochromocytoma of the urinary bladder is a rare tumour that originates from chromaffin tissue of the sympathetic nervous system situated within the urinary bladder wall [1]. These are tumours of the sympathetic nervous tissue and the symptom profile will depend on the secretory function [1]. These account for less than 0.05% of all bladder tumours and less than 1% of all phaeochromocytoma [1]. As phaeochromocytoma of the urinary bladder is such a rare condition, only limited literature is available to direct clinical decision making.

The usual triad of symptoms are hypertension during micturition, haematuria, and systemic symptoms due to raised catecholamines such as headache, dizziness and palpitation. However, 27% of phaeochromocytoma of the urinary bladder do not feature any hormonal activity [1, 2]. We report a case of a 15-year-old male who presented with malignant hypertension and loss of vision. Ethical approval was obtained from the Ethical review committee of National Hospital of Sri Lanka (AAJ/ETH/COM/2017-13). Informed written consent was obtained from the patient and his mother prior to collecting information.

He presented with rapidly progressive bilateral loss of vision and headache for 2 weeks duration and was found to have elevated blood pressure of 200/120 mmHg. He denied any urinary symptoms including haematuria. Fundoscopy revealed bilateral papilloedema. Examination of the abdomen was unremarkable. Abdominal contrast enhanced computed tomography revealed a 5.3 cm×4.3 cm enhancing lesion in the right lateral bladder wall with no lymphadenopathy (Fig. 1). Both kidneys and adrenal glands looked normal. His serum

creatinine was 61.6 $\mu\text{mol/L}$ and blood urea was 2.6 mmol/L. Serum sodium levels and potassium levels were 135 mmol/L and 3.5 mmol/L, respectively; and his complete blood count was within normal limits. His urinary vanillylmandelic-acid level was 30 mg/24 h (normal: 2–7 mg/24 h). Therefore, aphaeochromocytoma of the bladder was suspected. Prior to the surgery, the patient was started on non-selective alpha blockade with prazosin and also on a calcium channel blocker to control the blood pressure. Beta-blockade was achieved using atenolol, a cardioselective beta-blocker. Intravenous phentolamine, a reversible nonselective alpha-adrenergic antagonist was used during the intraoperative period to control the blood pressure.

Cystoscopy revealed a slightly bulging lesion with normal looking overlying bladder mucosa on the right side of the dome of the bladder. The exploration of the bladder through a Pfannenstiel incision did not reveal a palpable lesion in the right paravesical area and the lateral bladder wall. A midline vesicotomy was done and a large (6.5 cm×4.5 cm) mobile, very firm, oval shaped lesion on the dome of the bladder with an exophytic component on the peritoneal surface was noted. Partial cystectomy was done with complete excision of the mass.

Macroscopic assessment of the specimen revealed a circumscribed, bisected, capsulated mass measuring 6.2 cm×4 cm×3.5 cm weighing 54.37 g. The cut surface was solid and yellow with small cavities (Fig. 2). There was no necrosis, haemorrhage or cyst formation. Histopathology revealed tumour cells arranged in well-defined nests (Zellballen pattern) bounded by a delicate fibrovascular stroma which contained brownish pigments. The cells varied in size and shape and had finely granular cytoplasm. Intracytoplasmic inclusions were not seen. Nuclei were moderately pleomorphic and vesicular with prominent nucleoli. There were scattered cells with hyperchromatic atypical nuclei. Mitotic figures were not seen. The resection margins were free of tumour. Thus, a neuroendocrine tumour with histological features suggestive of a phaeochromocytoma was detected.

Normalisation of blood pressure with improvement of the vision was noted in the immediate post-operative period and he was discharged without any anti-hypertensives. During 1 year follow-up his blood pressure was persistently normal with a visual acuity of 6/9.

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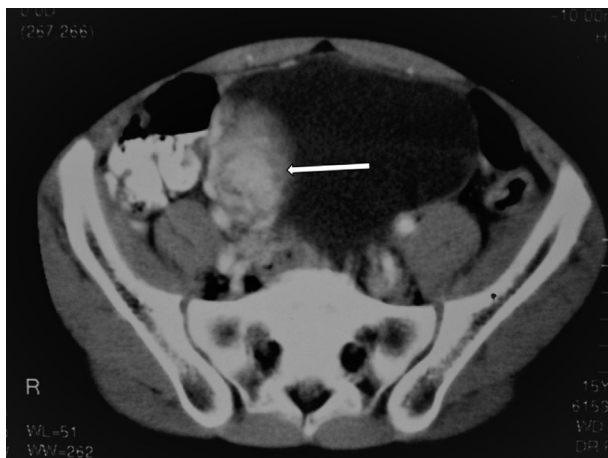


Figure 1 The abdominal contrast enhanced computed tomography scan revealed a 5.3 cm×4.3 cm enhancing lesion in the right lateral bladder wall with no lymphadenopathy. White arrow: Tumour.

Phaeochromocytomas of the urinary bladder are rare and mostly reported to occur in young adults but the age at presentation can range from 11 years to 84 years [1]. The most striking feature of this tumour is the vast variations in the initial presentation and thus a high degree of suspicion is required for early diagnosis. An extensive review of existing literature was done by Beilan et al. [1] and reported the largest analysis of phaeochromocytomas occurring in the urinary bladder. This included an analysis of 106 patients from many case reports and series and of those, 65 patients had biochemically functional tumours.

Based on the available literature, the typical symptoms include micturition attacks of headache and palpitations with haematuria. Some patients lacked more common presenting symptoms of bladder phaeochromocytoma, such as hypertension and had haematuria and lower urinary tract symptoms. Furthermore, the consequences of hypertension

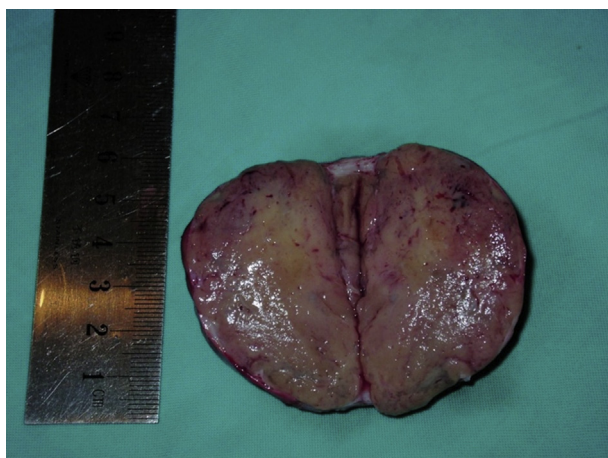


Figure 2 Macroscopic appearance of the specimen. The mass was circumscribed, bisected, and capsulated measuring 6.2 cm × 4.0 cm × 3.5 cm and weighing 54.37 g. The cut surface was solid and yellow with small cavities.

in the acute setting can mislead the initial diagnosis as noted in our patient [3]. Thus the presentation may vary considerably and the physicians must have a high degree of suspicion for an undiagnosed phaeochromocytoma in the setting of unexplained hypertension.

Our case is unusual in the patient presented with no obvious urinary symptoms suggestive of phaeochromocytoma of the bladder such as painless haematuria or symptoms of sympathetic over activity related to micturition, although the tumour was quite large. This patient initially presented to the eye surgeon due to the rapid onset of loss of vision and was diagnosed to have malignant hypertension and hypertensive retinopathy and was urgently referred to the physician. The possibility of a bladder phaeochromocytoma was suggested by the abdominal contrast enhanced computed tomography scan and was referred to the urology unit for urgent intervention.

There are several modalities for treatment of phaeochromocytoma which include catecholamine blockade, surgery and chemoradiation therapy. The accepted treatment modality for localized or locally advanced phaeochromocytoma of the bladder is surgery [2,4]. At least a partial cystectomy will be needed for adequate clearance as the sympathetic plexus is scattered between all layers of the bladder [4]. Transurethral resection will not be suitable to remove the entire tumour with adequate margins. Therefore, open surgery would be necessary. However, minimally invasive laparoscopic tumour resection has also been reported [2,4]. Cystoscopy examination is helpful to determine the exact location of the tumour in terms of the depth of invasion and involvement of the ureters. The role of biopsy for confirmation of this tumour is debatable [2].

Prognosis of the disease depends on the size of the lesion, presence of metastases and familial endocrinopathy [1,2]. At the cellular level, catecholamine-secreting bladder paragangliomas (PGLs) are indistinguishable from phaeochromocytomas. However, PGLs have stronger association with hereditary syndrome and higher risk for malignancy. Therefore plasma or urine metanephrine at 6 months after surgery and then annually is recommended, even in a patient with a probably benign tumour. Follow-up cystoscopy and imaging using computed tomography or metaiodobenzylguanidine (MIBG) scintigraphy should be considered particularly in patients with malignant tumours for the detection of metastases [2,5].

A high index of suspicion is required for the diagnose phaeochromocytoma of the urinary bladder due to its highly variable presentations. Phaeochromocytoma of the bladder can present with acute hypertensive emergency with hypertensive retinopathy without any urinary symptoms. A thorough investigation by imaging should clinch the diagnosis.

Author contributions

Study design: Umesh Jayarajah, Kasun Bandara Herath.

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Conflicts of interest

The authors declare no conflict of interest.

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