Phaeochromocytoma of the urinary bladder

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ABSTRACT

The occurrence of urinary bladder paragangliomas is rare. A 12-year-old Chinese girl who presented with history of blurring of vision was found to have grade IV hypertensive retinopathy. Investigations revealed a phaeochromocytoma on the posterior wall of the urinary bladder. A partial cystectomy with right ureter reimplantation was undertaken and her hypertension was promptly controlled. The diagnosis and management of this rare tumour is discussed.

Keywords: bladder neoplasm, extra-adrenal tumour, hypertension, phaeochromocytoma, urinary bladder tumour

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INTRODUCTION

Phaeochromocytoma of the urinary bladder is a rare neoplasm that accounts for less than 0.06% of all bladder tumours and less than 1% of all phaeochromocytomas[1-3]. It arises from the chromaffin tissues of the symphathetic nervous system within all the layers of the bladder wall. Haematuria and intermittent hypertension during micturition are among the usual clinical signs along with generalised symptoms due to raised catecholamines such as headache, blurred vision, heart palpitation and flushing. In addition, symptoms and signs of urethral obstruction may occur when the tumour is within the vicinity of the urethral opening[4]. Treatment strategies for these tumours are not well-defined because of their rare incidence. We present a rare case of phaeochromocytoma of the urinary bladder where a partial cystectomy and right ureter reimplantation was performed due to its involvement at the trigone of the urinary bladder.

CASE REPORT

A 12-year-old Chinese girl presented to an optometrist with sudden onset of blurring of vision and was then referred to a paediatrician for further assessment. It was subsequently found that she had hypertension of 180/110 mmHg that was complicated by grade IV retinopathy. There were no other symptoms and signs to suggest phaeochromocytoma. There was no family history of hypertension. Biochemical assessment revealed an elevated 24-hour noradrenaline of 2357.6 μg/24 hours (12.1-85.5). Initial localisation of a phaeochromocytoma was through computed tomography (CT) of the abdomen and pelvis which showed a suspicious mass lesion arising from the uterus. Magnetic resonance (MR) imaging showed an isointense lesion within the posterior wall of the trigone of the urinary bladder (Fig. 1).

After an extensive effort to control her blood pressure with prazosin, labetolol and nifedipine, excision of the tumour was performed under general anaesthesia, with prior cystoscopical examination. During cystoscopy, a 5 x 5cm tumour with superficial ulceration was found around the trigone area and which displaced the right ureteric orifice (Fig. 2). A partial cystectomy with reimplantation of the right ureter was succesfully performed with minimal fluctuation of her blood pressure. No other tumour or para-aortic lymph node enlargement was found. The post-operative period was uneventful and histopathological examination confirmed the diagnosis of a phaeochromocytoma of the urinary bladder. All anti-hypertensive medications were discontinued immediately after the operation.

DISCUSSION

Extra-adrenal phaeochromocytomas are rare tumours that are found in about 10% in adults and up to 30% in children[3] suffering from phaeochromocyomas. They are described to be potentially located between the clivus and the anus, although more than 85% of these tumours occur below the diaphragm[2]. Commonly, these urinary bladder phaeochromocytomas are hormonally active with elevated catecholamine metabolites, which help in their diagnosis. While MR imaging aids in the anatomical localisation, iodine-131 (I-131) – methyliodobenzylguanidine (MIBG) scintigraphy serves as a complimentary

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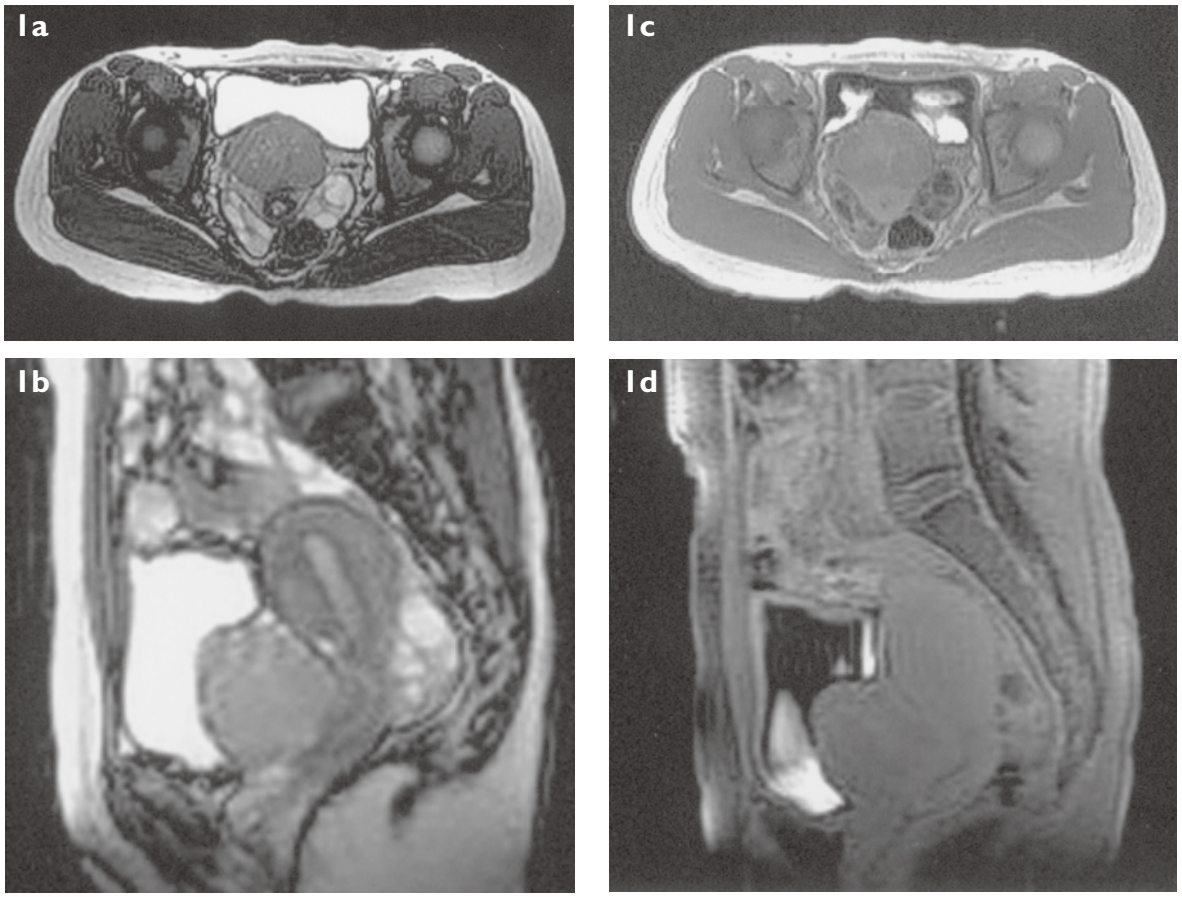

Fig. 1 (a) Axial and (b) sagittal T2-W MR images show a well-circumscribed isointense mass located between the bladder anteriorly and the cervix posteriorly. Enhanced (c) axial and (d) sagittal T1-W MR images show minimal enhancement of the mass.

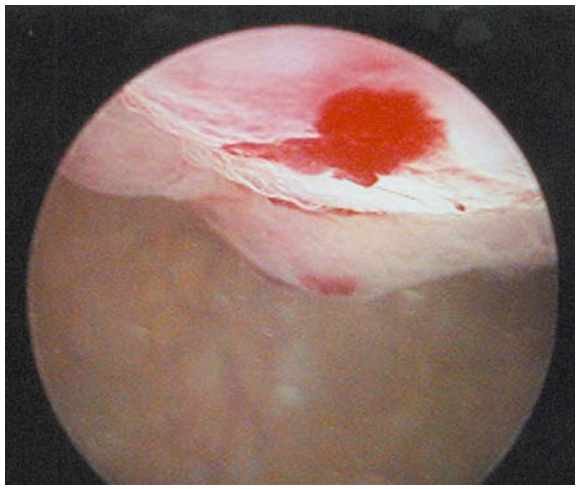

Fig. 2 Photograph taken during cystoscopy shows an ulcerated tumour on the posterior bladder wall.

functional diagnostic tool and together, they have 85-100% sensitivity in localising these paragangliomas[5,6].

Nakatani et al showed that using I-123-MIBG scintigraphy, tumours can be detected in cases where I-131-MIBG scintigraphy failed[6]. It is indeed well known that excision of these tumours requires the similar preoperative stabilisation of hypertension strategy as other phaeochromocytomas. This will involve the use of alpha-blocking agents for two to three weeks prior to the operation to counteract the effect of catecholamines, followed by beta-blockers. Adding nifedipine is beneficial if the blood pressure is not controlled.

As part of the intra-operative strategy, the use of cystoscopic examination prior to the excision helps to delineate the exact location of the lesion, especially with regard to the depth of invasion and the involvement of the ureters. Through cystoscopy, phaeochromocytomas appear as granulated and lobulated lesions with or without ulceration[2], as seen in our patient. Since the sympathetic plexus of the bladder is scattered between all the layers of the bladder, involvement of the entire layer of the bladder wall will require at least a partial cystectomy to achieve satisfactory results. For this, open surgery has been recommended[2,4,6] although a laparoscopic approach, as reported by Kozlowski et al, has recently been shown to be feasible[7].

Hwang et al has recommended laparoscopic ultrasonography for precise localisation and evaluation of tumour resection for laparoscopic management of extra-adrenal phaeochromocytoma[8]. In our case,

open partial cystectomy and right ureter reimplantation was performed. MR imaging of the pelvis together with pre-operative cystoscopy helped us in planning the type and extent of the surgery. Another approach to this tumour includes a transurethral resection as reported by Onishi et al[1] and Doran et al[2]. However, in both cases, the diagnosis of bladder phaeochromocytoma was not made pre-operatively.

From our literature review, the long-term outcome for each procedure, especially the recurrence rate, has not been well documented. This is perhaps due to its rarity and lack of long-term follow-up of these cases. It has however been reported that the prognosis is dependent on whether there is evidence of familial endocrinopathy or the presence of metastases[4]. Since histological features are usually unable to confirm malignancy, life-long follow-up is recommended for these patients with yearly endocrine evaluations, I-MIBG scintigraphy and MR imaging to detect recurrences or metastases[1,5]. In addition, these patients should also be monitored for post-operative complications such as stricture or obstruction at urethral reimplantation site, as hydronephrosis may result.

In summary, urinary bladder phaeochromocytoma is a rare tumour of the bladder which needs to be fully investigated before embarking on any surgical procedure. We strongly recommend prior cystoscopy and partial cystectomy in order to achieve complete resection due to its multilayer involvement of the bladder wall.

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