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CASE REPORT

Urinary bladder paraganglioma with a family history of neurofibromatosis type 1 treated with partial cystectomy

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Introduction

Paraganglioma is a rare tumour of the sympathetic nervous system often causing excessive production of catecholamines. These catecholamines, such as noradrenaline or adrenaline, can cause disruptive symptoms such as headaches and hyperhydrosis. Urinary bladder paraganglioma is exceedingly rare and therefore, if not managed correctly, can cause intraoperative complications secondary to excessive catecholamine production.

Discussion

Paraganglioma is a rare, extra-adrenal, catecholamine-secreting tumour, originating from the neuroectoderma of the sympathetic nervous system (Kurose et al, 2020). Paraganglioma tumours are more commonly situated within the mediastinum or upper abdomen, but they can rarely be located in the urinary bladder. These urinary bladder tumours account for 1% of extra-adrenal paraganglioma and 0.06% of all bladder tumours (Li et al, 2013).

Traditionally, paragangliomas are categorised into functional (83%) and non-functional (17%) modalities depending on their catecholamine secretion status, with common symptoms being paroxysmal hypertension, tachycardia and hyperhydrosis (Male et al, 2019). Specifically to urinary bladder paraganglioma, of which 27% are functional, episodes of dysuria, haematuria and hypertension upon micturition, triggered by bladder contraction or pressure, can be profound (Li et al, 2013). As a result of the potential intraoperative

Case report

A fit and well 34-year-old man, presenting initially with visible haematuria, and eventual acute urinary retention secondary to blood clots, was admitted to hospital. He had no significant urological past medical or family history of urological disease. However, he did have a strong positive family history of neurofibromatosis type 1, including his grandmother, mother and cousin. He reported no symptoms of hypertension, headaches or palpitations during his day-to-day life or while micturating.

Given the patient's age and unusual findings, he had a contrast computed tomography urogram, which showed evidence of a bladder tumour (Figure 1). Flexible cystoscopy confirmed a solitary papillary lesion on the right bladder wall. Subsequently, he underwent a transurethral resection to diagnose the specific type of tumour. Chippings from this transurethral resection were analysed by histopathology and confirmed to be paraganglioma of the bladder with muscular invasion (Figure 2).

Endocrine investigations, which included measurement of levels of serum and urine metadrenalines (a metabolite of catecholamine breakdown), were normal.

The patient's case was discussed in the local urological department multidisciplinary team meeting to plan his management. He was offered a partial cystectomy using bladder mapping to ensure precise removal of the tumour. The patient recovered well postoperatively and returned to his normal level of function quickly.

The patient's recovery was uneventful, and he was discharged 4 days post operation. He is scheduled to have a flexible cystoscopy in a year's time.

Scheduled follow up was a repeat computed tomography scan 6 months post procedure and a minimum of 10 years of annual flexible cystoscopies, with consideration for this to be lifelong. He has been referred to the regional genetic team for investigations for neurofibromatosis type 1.

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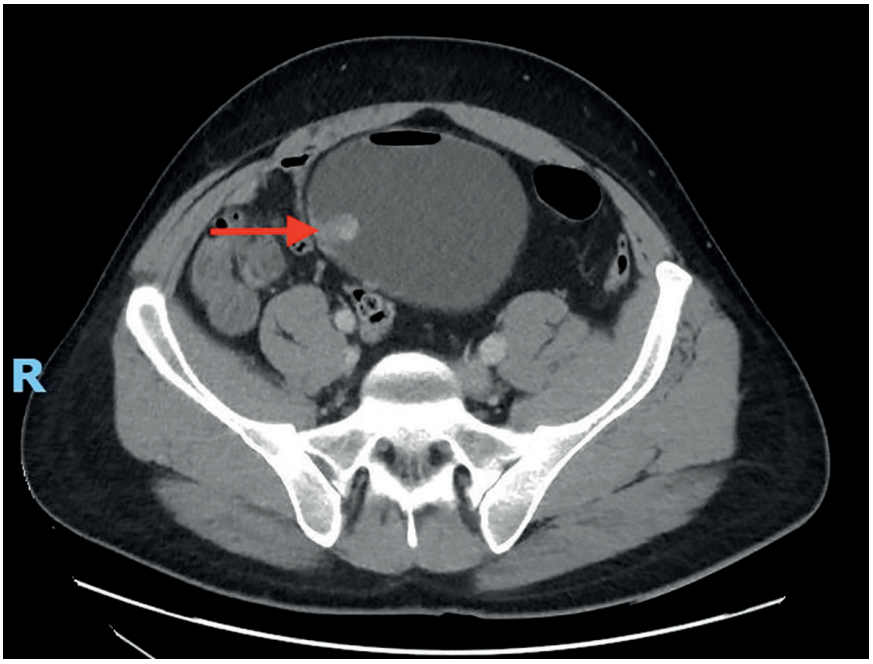

Figure 1. Computed tomography image showing right-sided bladder mass (red arrow).

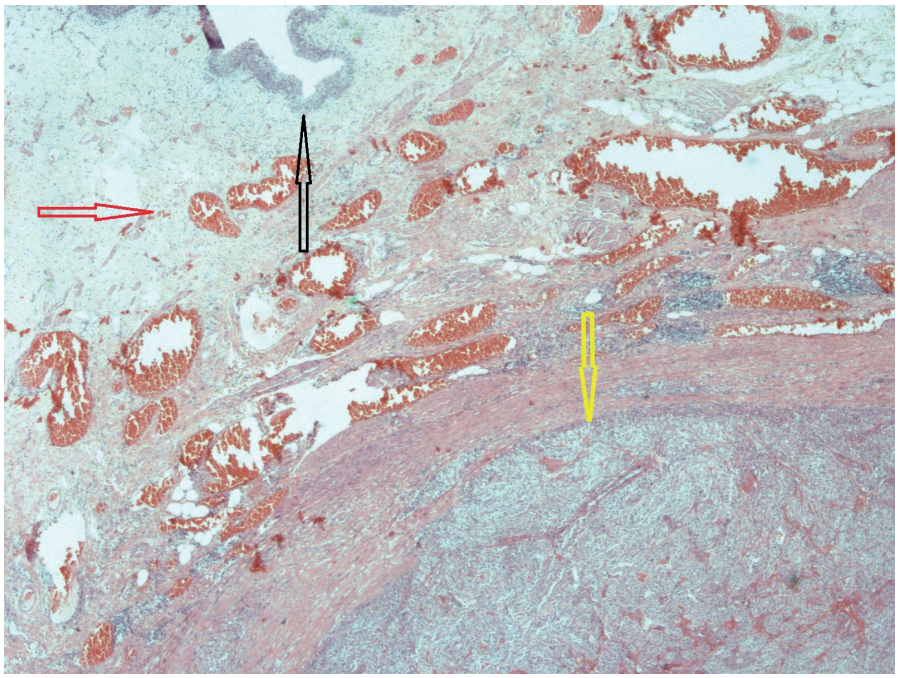

Figure 2. Chippings from the bladder tumour obtained via transurethral resection under haematoxylin and eosin stain. 20x magnification. Black arrow: normal surface endothelium. Red arrow: submucosa. Yellow arrow: paraganglioma.

secretion of catecholamines, some patients may require full androgenic blockade and therefore preoperative preparations are essential to reduce the risk of intraoperative crises.

Urinary bladder paragangliomas are usually treated with surgical intervention, typically transurethral resection of bladder tumour, partial cystectomy or radical cystectomy (Peng et al, 2015). The choice of surgical intervention is not guideline-driven at present in the UK, but multiple studies have been published arguing the specific merits for each of the three interventions. Both transurethral resection of bladder tumour and partial cystectomy, robotically or laparoscopically, appear to have widespread published merits compared to open cystectomy (Ren et al, 2017; Zhai et al, 2017). However, it is essential that treatment is tailored to each patient's specific requirements for best possible outcomes. In this specific

Learning points

- It is vital to explore the family history of cancer in patients presenting with haematuria, especially if the patient is young.
- Consider urinary bladder paraganglioma as a differential diagnosis in patients with a family history of neurofibromatosis type 1 and haematuria.
- If neurofibromatosis type 1 is present in the medical or family history, consider functional testing via the measurement of metadrenalines on micturition before surgical intervention to avoid a potential hypertensive crisis intraoperatively.
- Surgical intervention for urinary bladder paraganglioma is not guideline driven, but transurethral resection of bladder tumour and partial cystectomy appear to be preferable. A multidisciplinary approach is advisable.

case, the patient was young, and the tumour was relatively small. After a multidisciplinary team discussion, it was agreed, with patient involvement, that a partial rather than complete cystectomy would be the best surgical option.

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