CASE REPORT

Pheochromocytoma of urinary bladder - a case report

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Abstract

Pheochromocytomas are catecholamine producing neuro-endocrine tumours. Bladder involvement with pheochromocytoma is rare that usually presents with hypertension, hematuria and syncope often related to voiding. Clinical manifestation may be inconsistent and high index of suspicion is required for accurate diagnosis. We report a case, who presented with gross intermittent hematuria and mild elevation of pulse rate and blood pressure. Patient was adequately treated with partial cystectomy.

Keywords Bladder neoplasms · Pheochromocytomas · Partial cystectomy

Introduction

Pheochromocytoma of the bladder is a rare disease that accounts for less than 0.05% of all bladder neoplasms. Embryonic rests of chromaffin tissue arising from sympathetic plexus within bladder wall are believed

to be the source of these tumours [1]. The symptomatology consists of gross hematuria, paroxysmal or sustained hypertension, palpitation, profuse perspiration, pallor, weakness and headache often associated with micturition [2]. Due to variable clinical presentation and non-specific physical findings a high index of judgement is required for proper diagnosis. The unusual location of these tumours and in a way their interesting feature undoubtedly accounts for the low incidence of reported cases. We present a case, which presented with gross hematuria.

Case presentation and management

A 16-year-old girl was referred to our hospital for intermittent gross hematuria after suffering for 4 weeks. Physical examination was unremarkable, her pulse rate and blood pressure were 90/minute and 140/90 mmHg respectively. Urinalysis revealed full field RBC and urine culture was negative. Ultrasound showed an intravesical mass. Cystoscopy revealed 4 × 3 cm solid mass on the posterior wall above left ureteric orifice, biopsy was taken. The pathological picture of resected tumour was characterized by nest of fairly uniform cells having a pale, partially clear cytoplasm with eosinophilic granules throughout and relatively uniform, dark staining, oval or spherical nuclei. The tumour was partially surrounded by transitional mucosa from the bladder. Chromagranin staining was shown to be positive. A diagnosis of pheochromocytoma of the bladder was made. The 24-hour urine study revealed elevated levels of nor epinephrine (240 µg), epinephrine (150 µg) and vanillyl mandelic acid (VMA) (15 mg). Computerized tomography (CT) revealed a mass confined to bladder without any obvious multicentricity (Fig. 1). In preparation for removal of the tumour patient was treated with a 10 day course of α

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blockade and adequate hydration. On exploration a solid, submucosal tumour located above left ureteric orifice confined to bladder was found (Fig. 2). Partial cystectomy was done. Histopathological examination confirmed Pheochromocytoma. Postoperatively patient's pulse rate and blood pressure were 80/min and 120/80 mmHg respectively. In twenty months follow-up, patient's blood pressure, urinary VMA, nor epinephrine remained normal.

Discussion

Pheochromocytoma of the bladder is a rare occurrence. There are more than 80 reported manifestation of pheochromocytoma in general and bladder involvement often causes micturitional attacks that involve headache, fainting, palpitation, blurred vision, sweating or hypertension.

Fig. 1 Computerized tomography (CT) of bladder

Fig. 2 Solid submucosal tumor above the left ureteric orifice

These attacks may also be initiated by bladder distension, abdominal palpation, defecation or sexual intercourse. Painless hematuria has been noted in 50–65% of cases and hypertension in 65–80%. But not all patients, like the one reported here, show typical signs and symptoms even though their catecholamine levels are high. Reduction of the sensitivity of catecholamine receptors is considered the cause of this phenomenon [3]. The most helpful diagnostic laboratory test available is the determination of blood or urinary levels of epinephrine and nor epinephrine and their metabolic byproducts. Urinary VMA >9.0 mg/24 hours, nor epinephrine > 80 µg /24 hours and epinephrine > 20 µg/24 hours have been found in most of the patients. Diagnostic imaging is important for localizing the tumour and helping to exclude multifocal and metastatic disease. In addition to CT and magnetic resonance imaging, functional imaging methods such as metaiodobenzylguanidine scintigraphy and indium ¹¹¹pentetreotide scintigraphy have been used in the detection of pheochromocytoma. Positron emission tomography (PET) using 6 – [¹⁸F] flurodopamine is a promising new technique for functional imaging of pheochromocytoma and may aid in precise localization of all the tissues involved [4]. Alpha blockers and adequate hydration to allow intravascular volume reexpansion is essential in preoperative preparation. Beta blockers may be started if cardiovascular conditions, such as tachycardia and arrhythmias warrant. The majority of pheochromocytomas of bladder are intramural, therefore transurethral resection may not eradicate the entire lesion and is not recommended and may precipitate hypertensive crises or arrhythmias. The tumour can usually be adequately excised by partial cystectomy. A thorough examination of the surrounding structures and the regional lymph nodes should be done. Malignant potential is not predictable by histology but by clinical behaviour. Bladder pheochromocytoma seems to have a slightly better prognosis than other extra adrenal Pheochromocytoma [5]. Extra-adrenal pheochromocytomas are more likely to recur and metastasise making lifelong follow up of these patients mandatory. Biochemical evidence of excess catecholamine production usually precedes the clinical manifestations therefore annual determination of urinary catecholamine and / or their metabolites is recommended. Our case serves as a reminder that although bladder pheochromocytomas have a classic presentation, it is by no means exclusive. The case we encountered presented only with hematuria and mild elevation of pulse rate and blood pressure in spite of increased urinary concentrations of metabolic by-products of catecholamine. A high clinical judgement is important in evaluating bladder masses in patients of all ages.

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