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HYPERTENSION

CASE REPORT: CLINICAL CASE

Micturition Hypertension in an Adolescent

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ABSTRACT

Pheochromocytomas of the urinary bladder are rare tumors that represent <1% of all pheochromocytomas, and their main symptom is hypertension. In children, the evaluation of hypertension should focus on secondary causes, which involves ruling out catecholamine-secreting adrenal and extra-adrenal pheochromocytomas. (JACC Case Rep 2024;29:102435)

HISTORY OF PRESENTATION

An 11-year-old boy presented to the emergency department with a week-long history of paroxysmal headaches and sustained elevated blood pressure. He brought his cerebral magnetic resonance imaging (MRI), which was normal, and reported the ineffectiveness of pain medication. His medical history revealed only mild persistent asthma treated with inhaled corticosteroids, and he had no significant family history.

Observations showed a blood pressure of 129/87 mm Hg (98th percentile) with no difference in 4 limbs, a heart rate of 85 beats/min, normal oxygen saturation, and a body temperature of 37.2°C. On examination, he appeared healthy, was not experiencing symptoms at the time, and showed no signs of neurological disorder. He had good peripheral pulses, normal heart sounds, and no murmurs. His chest was clear. The electrocardiogram revealed a sinus rhythm with no sign of ventricle hypertrophy.

Still in the emergency room, 5 minutes after using the toilet, he complained of terrible headaches and palpitations. Blood pressure taken at this precise moment revealed a hypertensive peak of 155/97 mm Hg. Subsequently, the patient reported a relationship between micturition and headache attacks. He had no hematuria.

LEARNING OBJECTIVES

- To list secondary causes of arterial hypertension in adolescents and set up investigations to determine the etiology.
- To perform systematic and stepwise imaging in the case of a paraganglioma to avoid missing rare locations or synchronous metastases.
- To advocate routine genetic testing for paraganglioma or pheochromocytoma-predisposing germline mutations.

DIFFERENTIAL DIAGNOSIS

Hypertension secondary to renal parenchymal disease, renal artery stenosis, coarctation of the aorta, thyroid disorders, and adrenal disorders were considered.

INVESTIGATIONS

Initial blood results showed normal complete blood count, thyroid-stimulating hormone (1.2 μU/mL),

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ABBREVIATIONS AND ACRONYMS

CT = computed tomography

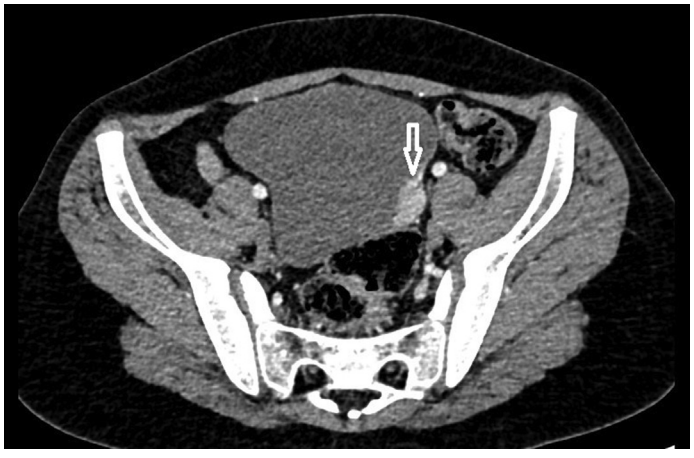
MRI = magnetic resonance imaging

creatinine (50 μmol/L), and electrolytes including level of potassium (4.5 mmol/L). Echocardiography showed normal left ventricle diameter, thickness, and ejection fraction. There was no valvulopathy or coarctation. We performed an abdominopelvic contrast-enhanced computed tomography (CT) scan that revealed a 3-cm mass located in the bladder (Figure 1). Strongly suspecting a paraganglioma, a treatment with α- and β-blockers was initiated, and urinary catheterization alleviated the headache attacks. Screening to confirm an extra-adrenal pheochromocytoma showed abnormal 24-h urine metanephrines: total metanephrine 0.31 mg (normal <0.27 mg/24 h) and normetanephrine of 1.41 mg (normal <0.46 mg/24 h). Plasma normetanephrine was also abnormal at 420 ng/L (normal <196 ng/L) but not total metanephrine, which was normal at 39 ng/L (<65 ng/L). 123I-labeled meta-iodobenzylguanidine imaging was performed and showed no synchronous metastases.

MANAGEMENT

The patient underwent surgery on day 6, with removal of the entire mass. The urinary catheter was kept in place for 10 days after the operation, with progressive weaning of the hypertension treatment.

FIGURE 1 Pelvic CT Scan



A mass located in the left posterior wall of the bladder suggesting a paraganglioma. CT = computed tomography.

DISCUSSION

Paraganglioma are rare tumors in the general population and even more uncommon in the pediatric population. The extra-adrenal location in the bladder is called bladder pheochromocytoma. It originates from the chromaffin cells of the sympathetic nervous system in the urinary bladder wall and account for <1% of all pheochromocytomas.[1]

The 3 most common symptoms in children are hypertension, palpitation, and headache, the same as in adults. Unlike other symptoms, hypertension has a sustained pattern and should be investigated further, particularly in children, in search of a secondary cause.

The screening test is based on the standardized urinary assays. Fractionated metanephrines and normetanephrines in 24-hour urine have a slightly lower sensitivity 97%, but a better specificity 98%, than serum-free metanephrines and normetanephrines, with a sensitivity of (97%-100%) and a specificity of (82%-85%).[2]

MRI and CT scan have around 95% sensitivity and 70% specificity, for detecting adrenal pheochromocytomas. However, MRI is better than CT scan as it has higher 90% sensitivity for localizing extra-adrenal pheochromocytomas.[3]

Most bladder pheochromocytomas are benign, but some can be malignant.[1] In the World Health Organization's updated classification of endocrine tumors, the latter is referred to as metastatic pheochromocytoma. Metastasis at initial presentation was found in 14% of children with pheochromocytoma or paraganglioma.[4] The 5-year survival rate of pediatric patients with nonmetastatic pheochromocytoma was 90% and ranged from 30%-60% in patients with metastatic disease for whom the 10-year survival rate was 31%.[5] This is why functional imaging with 123I-labeled meta-iodobenzylguanidine is indicated in the search for metastatic disease or identification of multiple chromaffin tumors.

Routine genetic testing must be performed in all cases. A germline mutation in one pheochromocytoma or paraganglioma susceptibility gene was identified in 77% of affected children who underwent genetic testing.[4] Inheritance is autosomal-dominant for pheochromocytoma-predisposing germline mutations. This means that the offspring of a carrier has a 50% likelihood of inheriting the parent's mutation. Sporadic cases may represent index cases for their families.

The treatment is usually surgical removal of the tumor covered by preoperative treatment with α- and β-blocking agents, sometimes with adjuvant therapies such as chemotherapy or radiotherapy.

FOLLOW-UP

At 1-month follow-up, blood pressure was normal for his age, less than the 90th percentile. The patient's parents were advised about genetic testing for their son.

CONCLUSIONS

Pheochromocytomas of the urinary bladder are exceedingly rare tumors that represent <1% of all pheochromocytomas. Arterial hypertension could be the only symptom detected. In children, the evaluation of hypertension should focus on secondary causes, which involves ruling out catecholamine-secreting adrenal and extra-adrenal pheochromocytomas.

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