Pheochromocytoma in children: a case report

Nguyen Trong Thanh1, Vu Chi Dung1, Bui Phuong Thao1, Nguyen Ngoc Khanh1, Can Thi Bich Ngoc1, Nguyen Thu Ha1, Do Thi Thanh Mai1
1 Department of Endocrinology, Metabolism & Genetics; National children’s hospital (NCH), Hanoi, Vietnam

Background

• Severe prolonged hypertension occurs in 0.1% of the children’s population and only about 2% of these have underlying endocrine causes.
• Pheochromocytoma (PCC) is a rare tumor arising from the adrenal medulla as well as extra-adrenal paraganglion system and secreting catecholamines causing severe hypertension in children.
• The gold standard for diagnosis is the measurement of free plasma levels of metanephrines while imaging studies performed for the location and size of the tumor and possible metastatic lesions.
• Preoperative treatment with alpha blockers, beta blockers and tyrosine hydroxylase inhibitors improves safety of the surgical procedure.

Objectives

To describe the clinical characteristics, investigation data and outcome of patients with Pheochromocytoma

Methods

This is case study including clinical symptom descriptions, biochemical and imaging investigations, and management for the child with PCC.

Results

• The female patient was 7 years old, with no medical history. She presented with vomiting, headache, abdominal pain and convulsions. Blood pressure was 220/190 mmHg and heart rate 130/min on admission.
• Abdominal CT showed a left adrenal tumor of 24x25 mm in size. Plasma level of cortisol and noradrenaline were 1374 nmol / L and 971 pg / ml, respectively; and urinary HVA level was 7.8 µmol/mmol of creatinine. Cerebrospinal fluid testing is normal.
• Preoperative blood pressure was controlled within 30 days with nicardipine, doxazosin mesylate and amlodipine. A 20x30 mm tumour was removed through endoscopy. Histopathological tissue was Pheochromocytoma, benign.
• After surgery, patient was stable with normal blood pressure, without using antihypertensive drugs.

Conclusions

• PCC is the cause of treatable secondary hypertension. Stabilizing blood pressure prior to surgery, contributes to ensure surgical treatment safety.
• Multidisciplinary collaboration is warranted to optimize the management of patients: Pediatric endocrinology - Imaging diagnosis - Anesthesia - Surgery - Resuscitation after surgery.

References


E-mail address: dungvu@nch.org.vn