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Case Report

Atypical presentation of adrenal pheochromocytoma with central tumour necrosis in a paediatric patient: a case report

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ABSTRACT

Pheochromocytoma is a rare catecholamine producing neuroendocrine tumour in children. It is usually presented with a triad of palpitation, headache, and diaphoresis. It is one of the causes of secondary hypertension in children. We report a case of a girl who presented with atypical symptoms of pheochromocytoma. A 10-year-old girl who presented with chest discomfort, shortness of breath and labile blood pressure. A young hypertension workup revealed a non-functioning left adrenal mass, making the dilemma in the initial clinical diagnosis. Confirmative diagnosis of pheochromocytoma was obtained via histopathological examination of the specimen through laparoscopic left adrenalectomy, which offered both diagnostic and therapeutic values. She was successfully treated via minimally access surgery. Diagnosing a pheochromocytoma in paediatric age group can be challenging due to its scarcity and possible atypical initial presentation. It requires high index of suspicion for a timely diagnosis and prompt treatment.

Keywords: Adrenal, Pheochromocytoma, Large necrosis, Paediatric

INTRODUCTION

Pheochromocytoma is a rare catecholamine producing neuroendocrine tumour in children. It is usually presented with a triad of palpitation, headache, and diaphoresis.[1] It is one of the causes of secondary hypertension in children.

It may be presented with normotension in children or atypical symptoms such as shortness of breath and chest discomfort. The diagnosis is via measurement of 24-hour urine catecholamines, imaging study for localization and identifying any metastatic lesions, and surgical resection for the definitive treatment.

Here we presented a girl who was delayed in diagnosis due to atypical presentation and alternating blood pressure. She was successfully treated with minimally access surgery.

CASE REPORT

A 10-year-old girl with a background history of bronchial asthma and allergic rhinitis, presented with a chief complaint of chest discomfort and shortness of breath one day before admission. She also had history of vomiting two days prior to the presentation. Otherwise, she denied any episodes of headache or blurring of vision. She visited the clinic on the second day of illnesses and was documented to have the blood pressure reading of 140/90 mmHg with the electrolyte imbalances: sodium 129 mmol/l, potassium 3.1 mmol/l. She was treated as an acute exacerbation of bronchial asthma and was discharged home after the symptoms resolved. Unfortunately, her symptoms recured, requiring her to visit the hospital for recurrent similar symptoms. Upon evaluation, patient was alert with the systolic and diastolic blood pressure ranging from 138-162 mmHg and 94-105 mmHg, respectively. She was also

tachycardic with heart rate of 120-140 beats per minute; regular and normal rhythm. There was no radio-radial or radio-femoral delay. Auscultation of the heart revealed a systolic murmur and a heaving apex beat. The fundoscopy assessment was normal. A young hypertension workout was carried out due to persistent serial high blood pressure readings. Laboratory findings indicated that a complete blood count, hepatic enzyme and thyroid function test were within the normal limit. Urinalysis was positive for protein (+1), erythrocyte (+1) and leukocyte 2/hpf. Echocardiography revealed apical hypokinesia with left ventricular hypertrophy. The ejection fraction near apex was 34%. Ultrasound renal doppler did not show any evidence of renal artery stenosis. However, abdominal ultrasound revealed a well-defined solid lesion measuring 2.6x3.2x3.9 cm at the left renal hilar region, displacing the left renal hilum vessels (Figure 1).

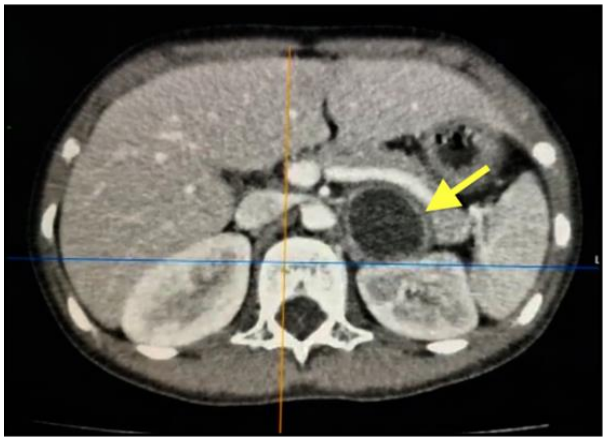

Figure 1: Encapsulated non-enhancing left adrenal lesion measuring 2.9×3×4 cm.

The patient was subjected for contrasted abdominal computed tomography which showed encapsulated non-enhancing left adrenal lesion with soft tissue and minimal internal fat attenuation, measuring 2.9×3×4 cm. No intra-lesion calcification was seen. Hormonal studies were sent to assess the functionality of the adrenal mass. A 24-hour urine catecholamine, serum renin and aldosterone level were normal in this patient. During the hospital stays, the blood pressure readings were labile with episodes of hypertension and hypotension as well. Acceptable control of blood pressures was achieved after patient was started on alpha-blocker medication (tablet Prazosin 0.5 mg TDS).

Surgical procedure

In view of no apparent causes of young hypertension except for a non-functioning left adrenal mass, she was arranged for laparoscopic transperitoneal left adrenalectomy after six months of pre-operative workup. The transperitoneal approach was chosen as the imaging showed tumour with a size of 4 cm, which was relatively bigger comparing to her body size. During the induction, the patient was given IV hydrocortisone 50mg. The left adrenal tumour was tightly adherent to the left renal vein, however, was able to dissect successfully. A 4x2x2 cm left adrenal mass was removed and the cut section showed small nodule measuring approximately 1 cm within the mass, which was smaller compared to the lesion in the initial imaging (Figure 2). The patient was fully conscious after the surgery and anti-hypertensive drug was able to wean off during the post-operative period. Her blood pressure remained within the normal range throughout the stays, and she was discharged on the fourth day following the operation.

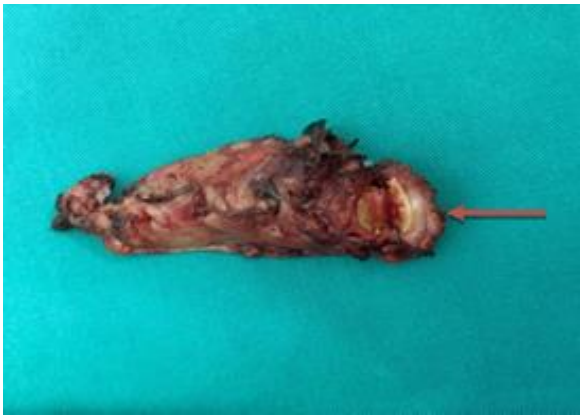

Figure 2: Left adrenal mass with small nodule (cut section, arrow).

Outcomes

The final histopathology examination showed left adrenal tumour; pheochromocytoma with substantial large central necrosis and granulomatous reaction. The necrosis and inflammation are bordered by some viable tumour cells. Ki-67 proliferative index is about 5%. No peri-adrenal adipose tissue invasion or vascular tissue invasion seen. The surgical margin is clear. The tumour cells are positive for chromogranin, synaptophysin, and S100 stain. The patient remained normotensive and well during subsequent follow up.

DISCUSSION

Pheochromocytoma is a neuroendocrine tumour that arises from the adrenal medulla, leading to catecholamine hypersecretion.[1] The diagnosis of pheochromocytoma in paediatric age group is rather challenging. The differential diagnoses include neuroblastoma and adrenocortical tumour. The pre-operative evaluation of adrenal mass must be done correctly using accurate analysis of clinical features, hormonal markers and imaging selection. Adrenal mass that arising in children also need a pre-operative workup of genetic counselling and testing, as one-third of patients may have a germ-line mutation.[1,2] Germ-line mutation of either of six known genes: RET (MEN 2A, 2B), NF1 (Von Recklinghausen neurofibromatosis type 1), Von Hippel-Lindau syndrome, SDHD, SDHC, and SDHB (mitochondrial supinate

dehydrogenase gene mutation) is associated with young age group and familial disease. Hereditary disease, extra-adrenal, metastasis, multifocal, and recurrent tumour are higher in children than in adults.[2] The patient in this case study presented with alternating hypertension and hypotension with episodic chest pain and shortness of breath. Her earlier presentation was treated as an acute exacerbation bronchial asthma, which leads to a delay in the detection and treatment of pheochromocytoma. In addition to her atypical symptoms, the functional studies for the left adrenal lesion were normal, which making the diagnosis of pheochromocytoma even more difficult. The patient developed left ventricular hypertrophy and apical hypokinesia due to uncontrolled hypertension and high afterload attributed to stunned myocardium.

The confirmatory diagnosis of pheochromocytoma was obtained via histopathological examination of the left adrenal mass which was retrieved through laparoscopic left adrenalectomy. The resected specimen showed extensive central necrosis with granulomatous reaction. This substantial necrosis of the adrenal gland can be explained due to a few factors such as infarction secondary to embolism to the vascular structures, shock or vasoconstriction, which induced by catecholamine. Pheochromocytoma's necrosis process could have explained the alternating release of massive catecholamine into the systemic circulation that leads to episodic hypertension and hypotension, which manifest as chest discomfort, palpitation, and shortness of breath.[3] Necrosis of the adrenal gland may contribute to the normal hormonal level during the pre-operative workup. It may also cause the size of the tumour to become smaller. The necessity to repeat scans such as ultrasound to assess the extent of the adrenal tumour before the operation can help the surgeon choose what approach they may use during the surgery. There are few case reports that described patient with extensive necrosis in adrenal pheochromocytoma has resolved symptom while receiving their treatment and waiting for the surgery.[3] It causes a real challenge and dilemma to obtain an early clinical diagnosis, hence the prompt treatment. The surgical approach for pheochromocytoma should be planned according to the functional status, presence of a bilateral lesion, and surgeon's experiences. Minimally access adrenal surgery has become the gold standard in the adult patient, and it was first described in 1992. As in children, the adrenal tumour has more prevalent malignant in nature, massive size during the presentation, and smaller body proportion, which contribute to difficulty in performing minimally access surgery. The transperitoneal approach becomes a primary choice in paediatric patients as it gives ample working space and excellent exposure as well as the familiarity of an intraperitoneal space to most surgeons.[4] It is also useful in dealing with bilateral adrenal tumour and abdominal lymph node sampling. Minimal access adrenal surgery proven to be safe from the oncological viewpoint for selected cases and giving the benefit of a smaller wound and early recovery.[5] Thus, the decision for transperitoneal approach was adopted in this patient because of the size of the lesion as well as post-operative benefits of minimally access surgery.

CONCLUSION

Pheochromocytoma represents a group of neoplasms that carry several issues and strongly requires a multidisciplinary approach. The incidents in paediatric patients are rare and may present with atypical symptoms. High clinical suspicion is needed to achieve an accurate timely diagnosis. Surgical therapy is a mainstay of its definitive treatment. Minimally access adrenal surgery is safe and effective in a selected paediatric patient with benign or malignant adrenal pathology.

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REFERENCES

1. Virgone C. Pheochromocytomas and paragangliomas in children: data from the Italian cooperative study (TREP). Pediatr Blood Cancer. 2020;4:e28332.
2. Pamporaki C. Characteristics of pediatric vs. adult pheochromocytomas and paragangliomas. J Clin Endocrinol Metab. 2017;102(4):1122-32.
3. Ohara N. Histopathological analysis of spontaneous large necrosis of adrenal pheochromocytoma manifested as acute attacks of alternating hypertension and hypotension: a case report. J Med Case Rep. 2016; 10(1):279.
4. Fascetti-Leon F. Minimally invasive resection of adrenal masses in infants and children: results of a European multi-center survey. Surg Endoscop. 2017; 31(11):4505-12.
5. Dokumcu Z. Laparoscopic adrenalectomy in children: A 25-case series and review of the literature. J Pediatr Surg. 2018;53(9):1800-5.

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