A Rare Abdominal Tumour in a Paediatric Age Group: Pheochromocytoma

Sah R, Sah RG, Mahmood S

Abstract

A 10 year boy presented with history of abdominal pain and palpitation. Abdominal ultrasound and CT oriented towards the diagnosis. After preoperative optimization, laparotomy with complete excision of an unusual pheochromocytoma tumour of about 8x10 cm adherent to liver and IVC performed. Histopathology confirmed the diagnosis without evidence of malignancy.

Key words: Adrenalectomy, Laparotomy, Pheochromocytoma

Introduction

Pheochromocytoma is a rare tumour of endocrine origin and a cause of secondary hypertension in children. It accounts for only about 1% of paediatric hypertension and often is associated with a variety of genetic syndromes. The National Registry of Childhood Cancers reports an incidence of 0.11 benign and 0.02 malignant pheochromocytomas per 1 million children. It is a catecholamine secreting tumour that arises from chromaffin cells of the sympathetic nervous system. Early diagnosis is vital because the tumour may be fatal if undiagnosed. The diagnosis benefited from the advancement of imaging, including Computed Tomography (CT), Magnetic Resonance Imaging (MRI) and meta-iodobenzylguanidine scintigraphy (MIBG). Its therapeutic management is multidisciplinary. Confirmatory diagnosis is based on the histopathological study of the surgical specimen. We report the case of a 10 year old boy with a unilateral pheochromocytoma revealed by short history abdominal pain and palpitation that has evolved well after surgical treatment.

The Case

A 10 year old boy, weighing 18kg, with two month's history of intermittent periumbilical abdominal pain. Pain was non radiating and severe in intensity but non-radiating, reported in our paediatric OPD. The pain increased with meals and was not associated with nausea and vomiting. He also had reduced weight of about 6 kg. Before coming to our department, the patient was taken to the local doctor where medication was prescribed. Pain was relieved for few days, however, after 2-3 days, he started having abdominal pain again with off and on palpitation which was not relieved by the medication. So the patient was brought to the Paediatric Medicine OPD, Mayo hospital.
A unusual large tumour of about 8 × 10 cm adherent to liver and IVC was excised [Figure 2]. Intraoperative blood pressure was kept 105-120/60-85 mmHg during the surgery. Histopathology confirmed the diagnosis of pheochromocytoma without any evidence of malignancy. He made an uneventful recovery with normalization of blood pressure and was discharged after 10th day of surgery. BP readings were normal on follow up on many occasions.

**Discussion**

Pheochromocytoma is a neuroendocrine tumour derived from the chromaffin cells of the sympathetic nervous system. It originates in the adrenal gland, but can arise in any part of the body that contains chromaffin cells. In children, pheochromocytoma is more frequently familial, extra-adrenal, bilateral and multifocal than in adults. Patients having pheochromocytoma may present not only with classical signs and symptoms, but also with various non-specific symptoms including abdominal pain. Keisuke Takeda et al. reported in their case that non-specific abdominal pain was most probably due to hypercalcemia, but in our case report the patient had a short history of abdominal pain which was probably due to the tumour itself. Presence of hypertension in children is uncommon and secondary to underlying pathology.

Our patient’s BP was not recorded during his initial visit to the general practitioner and was blindly treated for abdominal pain. BP measurement is not a routine check-up in paediatric age group. However, during paediatric assessment BP measurement should be considered an essential component. Elevated levels of catecholamine in blood and urine examination are useful in diagnosing pheochromocytoma. Radical excision is the best treatment of adrenal pheochromocytoma. Minimal handling of the tumour and early venous control are the best safeguards against intraoperative hypertensive crises. The blood pressure may not return to normal immediately in long standing cases and may require medication for some time.  

![Fig 1: The representative axial images of contrast enhanced CT scan showing mass of soft tissue attenuation in the right adrenal gland pushing IVC upward.](image)

![Fig 2: Shows an excised tumour (pheochromocytoma) of size 8 × 10 cm after laparotomy in a ten year child.](image)
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Conclusion

This case highlights the importance of children blood pressure, tumour size and presence of metastatic disease. Lack of good medical practice that has made patient suffered from unexplained abdominal pain and palpitation which was brought to normal after tumour resection. Thus, the overall prognosis in patient with pheochromocytoma appears to be related to early diagnosis, meticulous planning and tumour resection.

References


