A Rare Abdominal Tumour in a Paediatric Age Group: Pheochromocytoma

Sah R[1], Sah RG[2], Mahmood S[3]

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 8×10 cm adherent to liver and IVC performed. Histopathology confirmed the diagnosis without evidence of malignancy.

Key words: Adrenalectomy, Laparotomy, Pheochromocytoma

[1]Dr. Rajesh Prasad Sah, Department of Paediatric Surgery, Kanti Children Hospital, Maharajgunj, Kathmandu, Nepal, [2]Dr. RG Sah, Department of Clinical Neurosciences, University of Calgary, Canada, [3]Dr. Shaukat Mahmood, Department of Paediatric Surgery, King Edward Medical University, Mayo Hospital, Lahore, Pakistan.

Introduction

Pheochromocytoma is a rare tumour of endocrine origin and a cause of secondary hypertension in children[1]. 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.

Address for correspondence
Dr. Rajesh Prasad Sah
E-mail: drrajeshsah@gmail.com

How to cite
Sah R, Sah RG, Mahmood S. A Rare Abdominal Tumour in a Paediatric Age Group: Pheochromocytoma. J Nepal Paediatr Soc 2017;37(1):86-88.

doi: http://dx.doi.org/10.3126/jnps.v37i1.16885

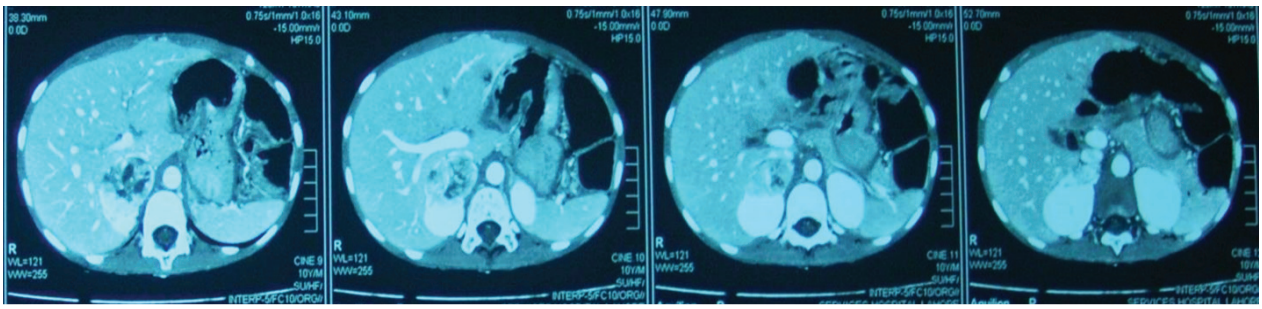

Fig 1: The representative axial images of contrast enhanced CT scan showing mass of soft tissue attenuation in the right adrenal gland pushing IVC upward.

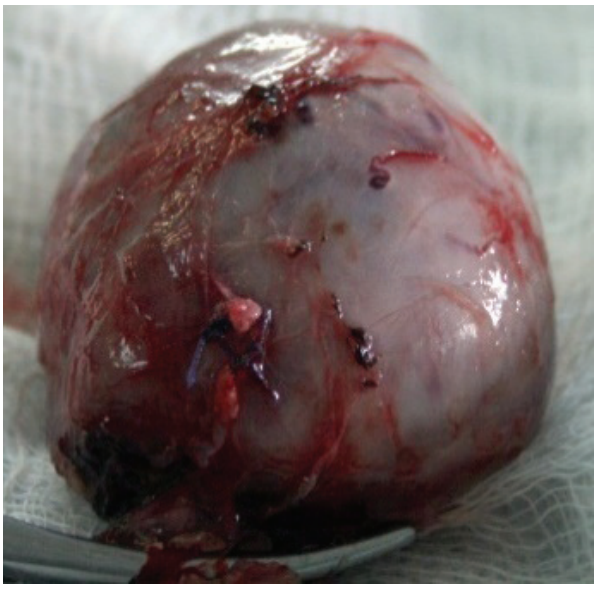

Fig. 2: Shows an excised tumour (pheochromocytoma) of size 8 × 10 cm after laparotomy in a ten year child.

During the nine days of admission he was investigated for abdominal pain and palpitation. Blood Pressure(BP) recorded was up to 160/100 mmHg, which was controlled with medication. Urinary VMA (vanillymandelic acid) was 18.0 mg/24hr. The level of calcium was 6.9mg/dl. Ultrasonography showed a solid well capsulated lesion measuring 3.9 × 4.0 cm in the area of right adrenal gland displacing inferior vena cava (IVC). An abdominal CT revealed a heterogeneous mixed density lesion measuring about 4cm in size with solid and cystic components with enhancement both in arterial and venous phases. The lesion was found adjacent to segment VI of the liver and was separable from it. The mass displaced the IVC. Right adrenal gland was not visualized separate from the mass; hence the mass was diagnosed to be arising from the right adrenal gland [Figure 1]. The left adrenal gland was normal.

He was treated with alpha receptor blocking drugs prior to surgery. After pre-operative optimization,general anaesthesia was induced with fentanyl and propofol and maintained with sevoflurane, dexmeditomidine and epidural infusion. Laparotomy was done and an unusually 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[2,3]. It originates in the adrenal gland, but can arise in any part of the body that contains chromaffin cells[4]. In children, pheochromocytoma is more frequently familial, extra-adrenal, bilateral and multifocal than in adults[5]. Patients having pheochromocytoma may present not only with classical signs and symptoms, but also with various non-specific symptoms including abdominal pain[6,7]. 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[8]. 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[9]. Elevated levels of catecholamine in blood and urine examination are useful in diagnosing pheochromocytoma. Radical excision is the best treatment of adrenal pheochromocytoma[10]. 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.

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

1. Silverman MA, Walker AR, Nicolau DD, Bono MJ. The frequency of blood pressure measurements in children in four EDs.*Am J Emerg Med* 2000;18(7):784-88.DOI: 10.1053/ajem.2000.16311.
2. Y, Yasukawa M, Kido T. Pheochromocytoma of the posterior mediastinum undiagnosed until the onset of intraoperative hypertension. *Gen ThoracCardiovascSurg* 2008;56:509-11. DOI: 10.1007/s11748-008-0282-3.
3. Young WF Jr. Paragangliomas: clinical overview. Ann N Y Acad Sci. 2006; 1073:21–29.
4. Ross J. Pheochromocytoma. Special considerations in children. *UrolClin North Am* 2000;27:393-402.DOI: 10.1196/annals.1353.002.
5. Havekes B, Romijn JA, Eisenhofer G, Adams K, Pacak K. Update on pediatricpheochromocytoma. *PediatrNephrol* 2009;24:943-50. DOI: 10.1007/s00467-008-0888-9.
6. Kopetschke R, Slisko M, Kilisli A, Tuschy U, Wallaschofski H, Fassnacht M, *et al.* Frequent incidental discovery of phaeochromocytoma: data from a German cohort of 201 phaeochromocytoma. *Eur J Endocrinol* 2009;161:355-61.DOI: 10.1530/EJE-09-0384.
7. Neumann HP, Eng C. The approach to the patient with paraganglioma. *J ClinEndocrinolMetab* 2009;94:2677-83.doi: 10.1210/jc.2009-0496.
8. Takeda K, Hara N, Kawaguchi M, Nishiyama T, Takahashi K. Parathyroid hormone-related peptide-producing non-familial pheochromocytoma in a child. *Int J Urol* 2010;10:1442-2042.DOI: 10.1111/j.1442-2042.2010.02555.
9. National High Blood Pressure Education Program Working Group on High Blood Pressure in Children and Adolescents. The fourth report on the diagnosis, evaluation and treatment of high blood pressure in children and adolescents. *Pediatrics* 2004;114:555-76.
10. Bissada N, Safwat A, Seyam R, Al Sobhi S, Hanash K, Jackson R, et al. Pheochromocytoma in children and adolescents: a clinical spectrum. *J PediatrSurg* 2008;43:540-543. DOI: 10.1016/j.jpedsurg.2007.10.038.