A LUMP TOO MANY – A CASE OF MULTIPLE PEDIATRIC TUMOURS

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Abstract

We report the case of a twelve year old male who presented with abdominal pain, vomiting, night sweats and weight loss. Examination revealed hypertension and imaging showed multiple abdominal tumours due to Von Hippel-Lindau disease. Surgical excision of adrenal and pancreatic lesions with management of endocrine dysfunction was facilitated by a multi-disciplinary team approach.

Keywords: Von Hippel-Lindau, phaeochromocytoma, pancreas, neuroendocrine, pediatric

Introduction

Von Hippel-Lindau disease is an autosomal dominant condition that is associated with a germline mutation in the VHL tumour suppressor gene on the short arm of chromosome 3 [1, 2]. It has perpetuated the name of two prestigious European physicians: Eugen von Hippel and Arvid Lindau [3]. It has an internationally variable incidence of 1 in 36000-91000 live births [4]. The mutation leads to the formation of benign or malignant tumours in multiple organ systems [1-3]. Before screening programmes became available the prognosis was poor with less than half of patients reaching the age of 50 but with appropriate medical and surgical treatment after early detection this has improved considerably [1]. It is a rare condition in children but we present our experience of a case in a pediatric patient [5]. Our case presents an uncommon and challenging problem for the pediatric surgeon.

Case History

A 12 year old Pakistani male presented to his local hospital with 5 months of night sweats and weight loss on a background of 3 years of intermittent abdominal pain and vomiting. At clinical examination he was found to be hypertensive but an otherwise normal child. Computed tomography (CT) and magnetic resonance imaging (MRI) was obtained which revealed a pancreatic lesion and bilateral adrenal lesions (Fig. 1 and 2). He underwent 24h urine analysis which revealed elevated levels of catecholamine consistent with a suspected diagnosis of phaeochromocytoma for the adrenal lesions. Southern blotting and DNA sequence examination confirmed Von Hippel Lindau disease. He therefore went on to have central nervous system imaging for other lesions but this along with ophthalmological assessment was normal. His case was discussed at the oncology multi-disciplinary team meeting (MDT) where a decision was made to proceed to surgical resection of the adrenal lesions. Although there were no radiological suspicious features of the pancreatic lesion for malignancy the decision was made to resect it at the same time in keeping with current recommendations [6, 7]. Preoperative preparation was planned in conjunction with the pediatric endocrinology team and involved alpha blockade with phenoxybenzamine 8 weeks before the operation. The aim of this was to achieve a normal blood pressure and postural hypotension. Beta blockade with propanolol was introduced after a few days, with absence of postural tachycardia indicating a sufficient blockade. The operation was performed jointly with pediatric, endocrine and hepatobiliary surgeons. The anaesthetic was performed with joint participation with pediatric anaesthetist with a shared interest in adult endocrine surgery ideally suited to this case. Invasive monitoring is essential for these patients and central and arterial lines were placed at induction of anaesthesia. An epidural was sited for analgesia and hydrocortisone was administered intravenously at induction and post adre-
nal resection. Sodium nitroprusside and magnesium were used intravenously to maintain vasodilation and prevent hypertensive crisis intra-operatively. A noradrenaline infusion was commenced after removal of the second adrenal gland to maintain blood pressure. Surgically, laparotomy was performed via a rooftop incision and the patient underwent first right and then left adrenalectomy. Following this, a subtotal pancreatectomy was performed for the pancreatic lesion. Splenectomy was performed at the same time, as the lesion was encasing the vessels. The specimens were sent for urgent histological analysis (Fig. 3) and the patient went to the pediatric intensive care unit post operatively. He was commenced on lifelong hydrocortisone, fludrocortisone and phenoxybenzamine. The noradrenaline infusion was stopped after 24 hours and he made a good recovery. Histological analysis confirmed bilateral phaeochromocytomas for the adrenal lesions. The pancreatic lesion was a beta cell tumour with pleomorphic features indicating malignant potential.

Discussion
This case illustrates the significant challenge that such rare diseases in childhood present for the pediatric surgeon. The endocrine disturbance from phaeochromocytomas is not frequently encountered with only 2% occurring in children. However this is still the most common pediatric endocrine tumour and 40% will be associated with genetic syndromes making cases such as this a significant possibility for the paediatric surgeon [8, 9]. Bilateral tumours as in this case have also been reported to occur with greater frequency in children compared to adults (20% versus 5-10%) [10]. Early consideration of the diagnosis will lead to the best outcomes therefore it is important for paediatric clinicians to consider the diagnosis particularly in hypertensive children which, unlike in adults, is usually due to underlying pathology [5]. In addition, phaeochromocytoma in children is almost always associated with sustained hypertension unlike in adults and so this is a sensitive diagnostic feature [10]. We have shown that such cases can be managed successfully with a multi-disciplinary team approach combining adult expertise with pediatric specialists.
REFERENCES