CASE REPORT

Recurrent Pheochromocytoma After Initial Debulking Surgery

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ABSTRACT

A 14-year-old male with a history of intermittent headaches since age 8 was diagnosed with right adrenal pheochromocytoma at 11 years after presenting with severe headaches and sustained hypertension up to 200 mmHg. Imaging revealed a right suprarenal mass, and debulking surgery was performed in 2021, with histopathology confirming pheochromocytoma. Three years later, he presented with recurrent headaches, palpitations, syncope, and systolic hypertension of 180 mmHg. Imaging demonstrated a large hypervascular solid–cystic mass in the left suprarenal region, without residual tumor on the right. The patient underwent laparotomy with complete excision of a well-encapsulated left adrenal mass, and histopathology again confirmed pheochromocytoma measuring 10 cm. This case highlights metachronous contralateral recurrence following initial debulking surgery in a pediatric patient. It underscores the limitations of incomplete resection, the importance of early recognition of recurrent symptoms, and the need for lifelong structured follow-up to prevent severe cardiovascular complications and long-term morbidity.
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INTRODUCTION

Pheochromocytoma is a rare catecholamine-secreting neuroendocrine tumor arising from chromaffin cells of the adrenal medulla, with an estimated incidence of 2–8 cases per million population (1). Although uncommon in children, it represents an important cause of secondary hypertension and may lead to life-threatening cardiovascular complications if not recognized early (1,2). Excess catecholamine secretion typically manifests as hypertension, headache, palpitations, and diaphoresis, and may progress to hypertensive crisis, arrhythmia, or multiorgan dysfunction (1,2).

Complete surgical resection is the definitive treatment; however, recurrence remains a significant clinical concern, with reported rates ranging from 10% to 30% (3). Recurrence may result from incomplete tumor removal, multifocal disease, genetic predisposition, or the development of metachronous tumors in the contralateral adrenal gland (4). Pediatric patients are particularly at risk due to a higher prevalence of hereditary syndromes and bilateral disease (5).

Current guidelines strongly recommend lifelong follow-up after surgery, including periodic biochemical and imaging evaluation, as recurrence may occur years after initial treatment (3,4). Early recognition of recurrent symptoms is essential to prevent severe cardiovascular and cerebrovascular complications associated with catecholamine excess (1,2).

We report a case of metachronous contralateral pheochromocytoma occurring three years after initial debulking surgery in a pediatric patient, highlighting the limitations of incomplete resection and the importance of long-term surveillance.

CASE REPORT

A 14-year-old boy with a history of recurrent intermittent headaches since the age of 8 years was initially suspected of having a brain tumor. A head CT scan was performed and revealed no evidence of intracranial mass. The patient did not undergo further evaluation at that time.

At the age of 11 years, he presented again with severe headaches accompanied by intermittent abdominal pain. On examination, his systolic blood pressure reached 200 mmHg. Bilateral renal vascular Doppler ultrasonography was performed due to suspicion of renal artery stenosis as a cause of hypertension, which demonstrated bilateral diffuse renal parenchymal disease.

Subsequently, contrast-enhanced multislice CT (MSCT) angiography of the aorta revealed a right suprarenal

gland mass suggestive of pheochromocytoma (Fig. 1). The patient underwent tumor debulking surgery in 2021, and histopathological examination confirmed the diagnosis of pheochromocytoma.

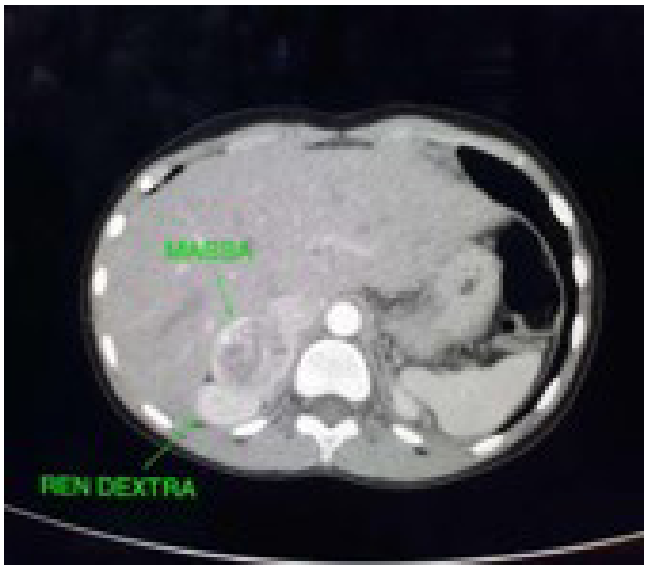

Figure 1: First Abdominal CT scan showed right side pheochromocytoma

Three years after the initial surgery, the patient again complained of headaches associated with palpitations and experienced a syncopal episode at school. On evaluation, his systolic blood pressure was 180 mmHg. He was referred back to the pediatric endocrinology clinic. Abdominal ultrasonography demonstrated a solid mass with cystic components located anterior to the left kidney. Contrast-enhanced abdominal CT scan confirmed a solid–cystic suprarenal mass with areas of necrosis and hypervascularity in the left adrenal region, measuring approximately 5.61 x 5.80 x 9.38 cm (anteroposterior x laterolateral x craniocaudal dimensions) (Fig. 2). The mass displaced the pancreatic tail anteriorly and the left kidney posteriorly. No residual pheochromocytoma was identified in the right suprarenal region.

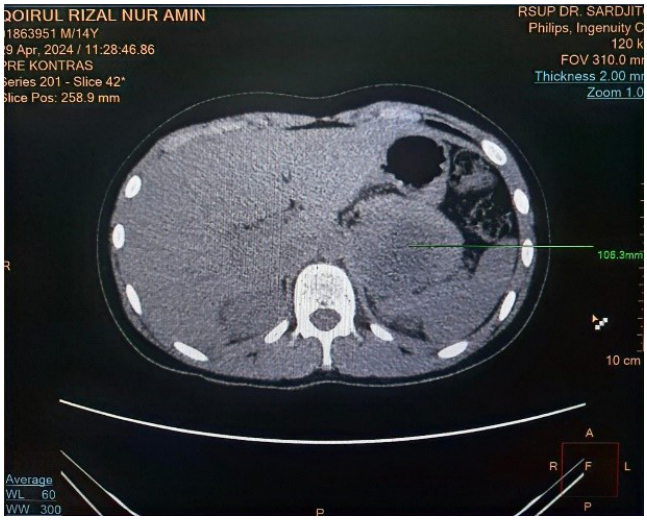

Figure 2: Abdominal CT scan carried out three years after initial debulking surgery

The patient subsequently underwent laparotomy with mass excision and a drain placement (Fig. 3). Histopathological examination confirmed pheochromocytoma measuring 10 cm in greatest dimension.

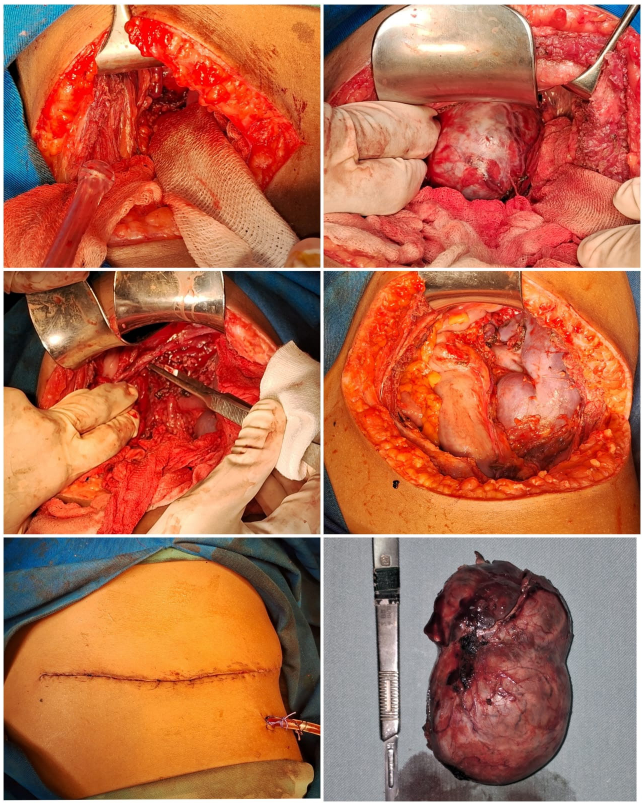

Figure 3: Left adrenalectomy and drain insertion

DISCUSSION

Pheochromocytoma, although rare in the pediatric population, represents a clinically significant and potentially life-threatening cause of secondary hypertension. Excess catecholamine secretion can lead to paroxysmal symptoms such as headache, palpitations, and diaphoresis, as well as sustained hypertension with severe cardiovascular and cerebrovascular consequences if left untreated (1,2). In children and adolescents, delayed recognition is particularly concerning, as prolonged catecholamine excess may result in irreversible target-organ damage, including cardiomyopathy, arrhythmia, and stroke.

This case is clinically important because it illustrates metachronous contralateral recurrence following initial surgical debulking, underscoring the persistent and unpredictable nature of pheochromocytoma. Although surgical resection is considered curative, recurrence rates of 10–30% have been reported even after apparently adequate treatment (3). Recurrence may result from residual disease, multifocal tumorigenesis, hereditary predisposition, or development of new tumors in previously uninvolved adrenal tissue (4). Pediatric patients have a higher likelihood of hereditary syndromes and

bilateral or recurrent disease compared with adults (5).

The implications of tumor recurrence extend beyond the presence of a new adrenal mass. Recurrent catecholamine excess re-exposes patients to hemodynamic instability and increases cumulative cardiovascular risk. Persistent or episodic hypertension may predispose to left ventricular hypertrophy, heart failure, arrhythmia, and cerebrovascular events (1,2). In addition, undiagnosed recurrence may pose substantial perioperative risk due to the potential for hypertensive crisis under anesthesia.

This case also underscores the importance of long-term structured surveillance following initial treatment. Current clinical guidelines recommend lifelong follow-up, including periodic biochemical and imaging evaluation, due to the persistent risk of recurrence (3,4). Pediatric patients require particular vigilance given their higher risk of hereditary disease and bilateral involvement (5). The three-year interval between initial surgery and recurrence in this patient highlights the necessity of continued monitoring even in asymptomatic periods.

Importantly, the initial management in this case involved tumor debulking rather than complete adrenalectomy. While debulking may be necessary in selected cases, incomplete resection may increase the risk of persistent or recurrent disease. This reinforces the principle that complete surgical excision, when feasible, remains the cornerstone of definitive management. Multidisciplinary collaboration involving pediatric surgeons, endocrinologists, radiologists, and genetic specialists is essential to optimize outcomes.

In summary, pheochromocytoma in children should be regarded as a chronic condition requiring lifelong vigilance. Recognition of recurrence risk, adherence to follow-up guidelines, and early intervention upon symptom recurrence are critical to reducing morbidity and preventing life-threatening complications (1–5).

CONCLUSION

This case demonstrates that pheochromocytoma can still occur on the other side even though debulking surgery has been carried out. Since recurrence happens in 10-30% of cases, long-term follow-up is crucial. Early detection and prompt treatment are crucial to prevent serious health issues and potentially life-threatening complications.

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