Case Report

Bilateral pheochromcytomas presenting as shock: A rare case report

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Abstract

Pheochromocytomas are rare chromaffin cell tumors of adrenal medulla (90%) that secrete catecholamines. Among children, the average age of presentation is 11–13 years, with a male preponderance of 2:1. Symptoms may be caused by catecholamine overproduction, local pressure, or metastasis. Sustained hypertension is the most common symptom. Elevated circulating catecholamines can cause cardiovascular alterations such as coronary vasospasm, ventricular and supraventricular arrhythmias, and dilated cardiomyopathy, precipitating in cardiogenic shock. We present a rare case of cardiogenic shock as the initial presentation of a bilateral pheochromocytoma in a child.

Keywords: Cardiogenic shock, children, pheochromocytomas

Introduction

Pheochromocytomas are rare chromaffin cell tumors of adrenal medulla.[1] They usually present a benign course; however, approximately 10% present signs of malignancy and 10% are bilateral.[1,2] Symptoms may be caused by catecholamine overproduction, local pressure, or metastasis.[1] Although presentation of pheochromocytoma is variable, 60% present with hypertension; other symptoms include palpitations, hyperthermia, diaphoresis, headache, and abdominal pain.[3]

To suspect, confirm, localize, and treat them is important due to associated cardiovascular morbidity and mortality.[4] It is thus important for primary care providers to refer children presenting with hypertension for appropriate referral and investigation.

Case Report

A 14-year-old boy presented with drowsiness, yellow discoloration of skin, and constipation for 4 days. On physical examination, his peripheries were cool, brachial pulses were feeble, and radial pulse and blood pressure were not recordable. Respiratory examination showed tachypnea, nasal flaring, and subcostal retraction. On cardiovascular examination, S3 gallops were heard. Central nervous examination concluded that he was drowsy but arousable, with pupils bilaterally reactive to light, and Glasgow coma scale was 13/15. Echocardiography showed mild concentric left ventricular hypertrophy, low left ventricular ejection fraction (45%), mild global hypokinesia, and grade 2 diastolic dysfunction.

He was treated with ionotropic support for cardiogenic shock, which was gradually tapered and stopped in view of

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hemodynamic improvement. He then showed persistent high blood pressure for which antihypertensives were administered.

Laboratory investigation revealed elevated total leukocyte count (16.7 x 10^3/μL) with 80% neutrophils, prothrombin = 20.3 s, and international normalized ratio (INR) = 1.83. Inflammatory markers such as C-reactive protein (64.7 mg/L), D-DIMER (>10000.00 ng/ml), and serum lactate dehydrogenase (9340 U/L) were elevated. Procalcitonin and creatine kinase were elevated, measuring 13.95 ng/ml and 3398 U/L, respectively.

Serum sodium was low (125 mmol/L), and serum potassium was normal. Kidney function and liver function showed progressive deterioration (serum creatinine: 2.53 mg/dl, serum urea: 140 mg/dl, alanine aminotransferase (ALT): 593 U/L), aspartate aminotransferase (AST): 880 U/L), serum total bilirubin (2.26 mg/dl), and direct bilirubin (1.17 mg/dl), but returned back to baseline after 2 weeks [Graphs 1 and 2]. No organism was isolated on blood or urine culture. The urine metanephrine was elevated (310 μg/24 h). Inflammatory markers also showed a significant reduction within 1 week [Graph 3]. The serum sodium also normalized on the third day of admission.

Ultrasound showed a fairly defined hypoechoic lesion in both suprarenal regions [Figure 1].

MRI of both adrenals showed well-defined bilateral supra renal lesions appearing isointense to hyperintense on T2W images with fluid [Figure 2], hemorrhage, and calcific foci [Figure 3]. Areas of calcific foci were confirmed on plain CT scan [Figure 4]. No intralesional fat components on in-phase and out-of-phase sequences or restricted diffusion on diffusion-weighted imaging were seen. These masses showed intense heterogeneous enhancement with minimal wash out in late post-contrast phases and showed central nonenhancing necrotic areas [Figures 5 and 6].

Positron emission tomography/computed tomography (PET/CT) was done to rule out multiplicity and/or metastasis, which showed bilateral adrenal, high-grade ^{18}F –fluorodeoxyglucose (FDG) avid lesions without metastasis.

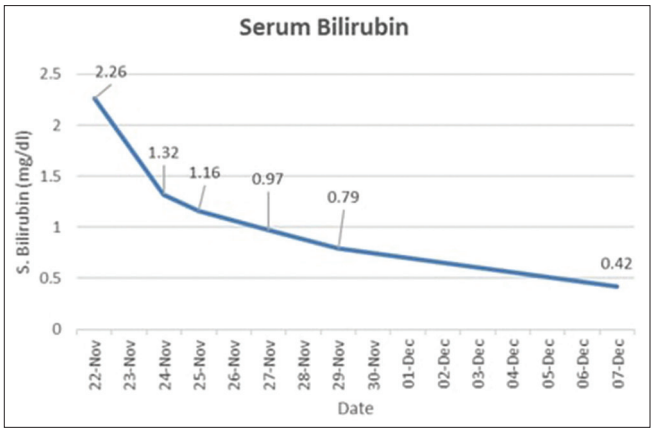
At 4 weeks, laparoscopy-guided bilateral cortical sparing adrenalectomy was done under general anesthesia. His vitals were stable throughout surgery as well as post-surgery. The patient was then discharged on oral hypertensives. Histopathological examination revealed an adrenal gland scale score of 8 (concerning for malignancy) on the left and 3 on the right (benign).

On follow-up after 1 month, the patient was asymptomatic with normal blood pressure.

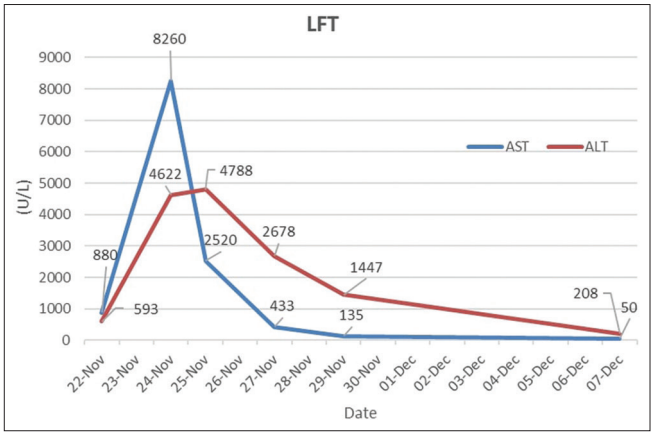
Discussion

The persistent high catecholamine levels secreted by pheochromocytomas have been associated with dysregulation of beta-adrenergic receptors, myofibril dysfunction, and reduction of contractile units. In addition, long-standing adrenergic stimulation generates an intense vasoconstriction and coronary spasm, which aggravates myocardial damage. This may lead to nonischemic, nonvalvular cardiomyopathy, or cardiogenic shock.[5-8]

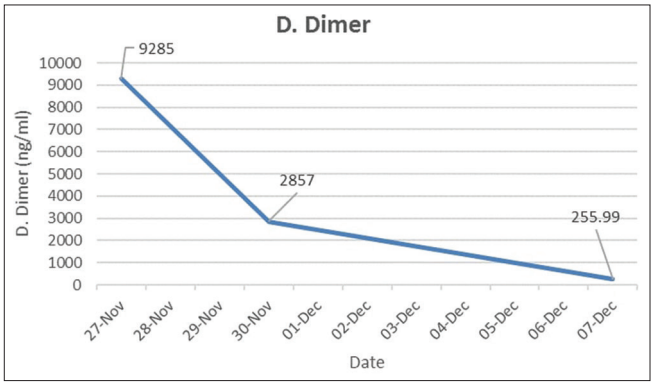
The recommended biochemical testing for the diagnosis of pheochromocytoma includes measurements of plasma-free metanephrines or urinary fractioned metanephrines.[4]



Graph 1: Reducing trend of serum bilirubin levels over 2 weeks



Graph 2: Reducing trend of liver function tests over 2 weeks



Graph 3: Reducing trend to D-Dimer levels over 1 week

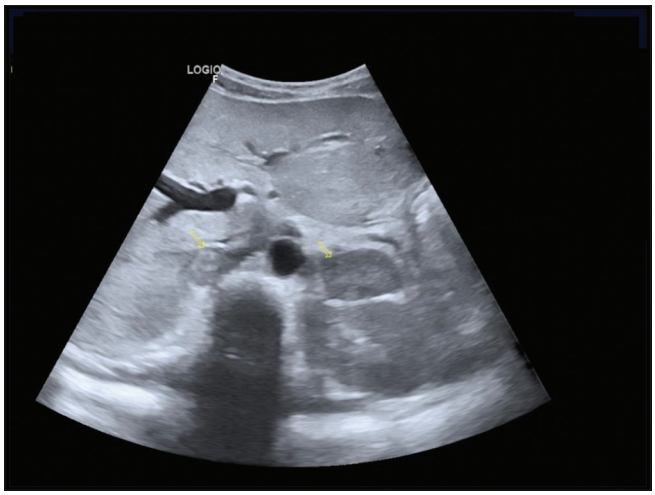


Figure 1: Ultrasound image shows well-defined hypoechoic lesions (yellow arrow) in bilateral suprarenal regions

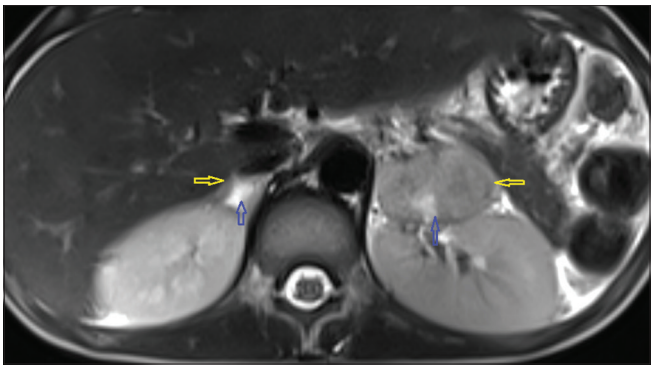


Figure 2: Isointense to hyperintense lesions (yellow arrow) on T2-weighted images with few T2 fluid signal foci within (blue arrow) in bilateral adrenals. Left suprarenal mass measured 2.8 cm (Anteroposterior) × 4.4.2 cm (Transverse) × 4.5 cm (Supero-inferior), and right suprarenal mass measured 1.9 cm (Anteroposterior) × 1.8 cm (Transverse) × 2.5 cm (Supero-inferior)

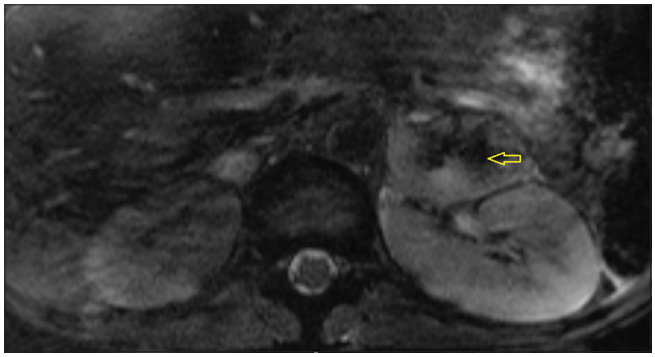


Figure 3: Areas of blooming (yellow arrow) in the left adrenal lesion on GRE, MRI images

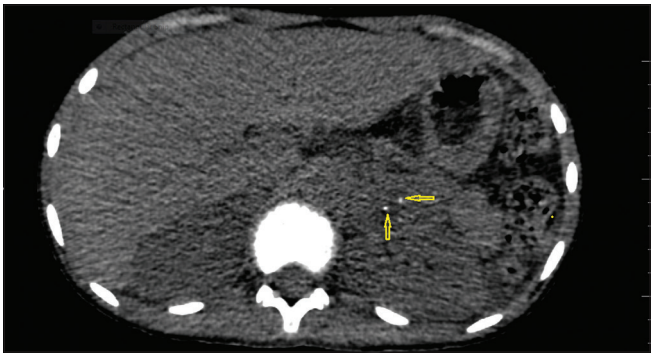


Figure 4: Foci of calcifications (yellow arrow) noted in the left adrenal lesion on axial unenhanced CT scan

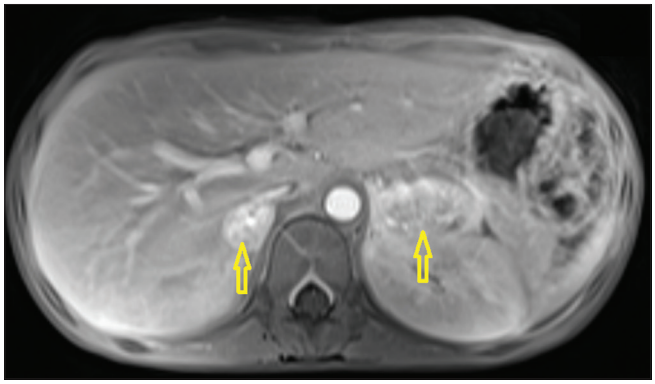


Figure 5: Intense heterogenous enhancement in bilateral adrenal lesions (yellow arrow) on post-contrast T1-weighted MRI images

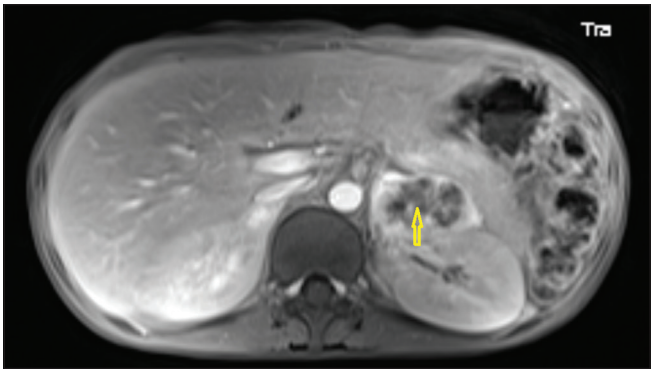


Figure 6: Nonenhancing necrotic areas (yellow arrow) on post-contrast T1 weighted MRI images in left adrenal lesion

The role of imaging lies in the localization and extent of tumors, diagnosing multiplicity, and metastatic lesions.

Computed tomography is considered as the first line of investigation owing to excellent spatial resolution.[4] On CT, these are well-defined masses with unenhanced attenuation greater than 10 HU. They show avid enhancement and delayed washout. Cystic areas, calcifications, necrosis, and hemorrhage may also be seen in atypical cases as it was seen in ours.

Magnetic resonance imaging has improved tissue contrast. Pheochromocytomas commonly demonstrate T2 prolongation and variable but intense contrast enhancement, more often resulting in the light bulb sign.[9] However, this appearance may vary, with 30% of lesions showing low signal intensity.[10]

Pheochromocytomas may sometimes demonstrate rapid contrast washout or contain sufficient fat to mimic adenomas. They may also show variable washout on occasions owing to varied pathological degeneration and are therefore confused with adenomas or metastases.[10]

^{18}F-FDG -PET/CT scanning is preferred over ^{123}I-MIBG scintigraphy in patients with metastatic pheochromocytoma.[4]

It has been suggested that patients should undergo preoperative catecholamine blockage in functional tumors to prevent perioperative cardiovascular complications. Preoperative evaluation should include, complete blood count, metabolic profile, plasm metanephrine, ECG, and echocardiography.[1]

Three critical perioperative instances that are associated with hypertensive episodes peri-operatively are endotracheal intubation, creation of pneumoperitoneum, and manipulation of the adrenal gland.[1] Laparoscopic adrenalectomy is recommended for most adrenal pheochromocytomas. Invasive tumors may require open resection to ensure complete tumor resection, prevent tumor rupture, and avoid local recurrence.[4]

Cortical-sparing adrenalectomy can be performed in patients with bilateral pheochromocytomas to avoid chronic steroid hormone replacement and the risk of Addisonian crisis.[11]

The reversibility of the myocardial affection after adrenalectomy has been described in cases of mild myocardial damage, but it is not possible in case of massive necrosis or extensive myocardial fibrosis.[12,13]

This condition requires long-term follow-up; the physician must evaluate blood pressure, heart rate and blood glucose, and plasma and urine levels of metanephrines annually to diagnose persistent disease and recurrence. MRI is the preferred imaging method for follow-up to minimize radiation exposure, but it should be noted that it may miss tumors in unusual places. Imaging may be performed every 1–2 years in patients with biochemically inactive pheochromocytomas to screen for local or metastatic recurrence.[14]

Conclusion

Timely referral and early diagnosis and prompt intervention can influence prognosis and prevent catastrophic complications caused by pheochromocytomas. Child presentation with shock is rare in pheochromocytoma; thus, imaging findings of suprarenal mass must alert the clinician to suspect pheochromocytoma.

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Conflicts of interest

There are no conflicts of interest.

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