

An Interesting Presentation of Pheochromocytoma

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Abstract

Pheochromocytoma is a rare cause of hypertension, but it could have severe consequences if not recognized and treated appropriately. Pheochromocytoma classically presents with paroxysms of hypertension and adrenergic symptoms including classic triad of episodic headache, sweating, and tachycardia. The clinical presentation of pheochromocytoma can mimic a number of other medical conditions including migraine, cardiac arrhythmias, myocardial infarction, and stroke, thus making the diagnosis of pheochromocytoma difficult, and treatment is directed toward presenting issue rather than underlying problem in such patients. We present a case of a 41-year-old male patient who presented with cerebellar infarct and found to have aortic thrombi and later developed acute myocardial infarction during same hospitalization. To the best of our knowledge, this is the first reported case of this kind.

Keywords: Acute cerebellar infarct, acute myocardial infarction, aortic thrombi, pheochromocytoma

INTRODUCTION

Pheochromocytoma is rare catecholamine-secreting neuroendocrine tumors arising from chromaffin tissue in the adrenal medulla or extraadrenal paraganglia. It occurs with an annual incidence of approximately 2–8 million person-years and rare cause of hypertension in the general population, representing <0.2% of cases.^[1,2]

Pheochromocytoma has classic triad of symptoms, but most do not present with this. It has atypical presentations in 9%–10% which often delay diagnosis.^[1] Even with classical presentations, the diagnosis is often missed for a number of years unless the patient is evaluated in a center experienced in this disease.^[3]

CASE REPORT

A 41-year-old male presented with 6 h history of sudden onset dizziness, vomiting, and difficulty in walking. His BMI was 28.3. On evaluation, he was conscious, oriented with heart rate 81/min regular, blood pressure 220/120 mmHg, and normal temperature. Examination was relevant for the presence of gaze-evoked nystagmus, impaired finger-nose test, truncal, and gait ataxia. MRI brain showed acute left cerebellar infarct. He was shifted to Intensive Care Unit on labetalol infusion. His past medical history was not significant except the diagnosis

of hypertension 1 year back for which he was not taking any treatment. His admission laboratory values were significant for leukocytosis and high fasting sugar but normal glycosylated hemoglobin and lipid profile.

On the next day, he complained of severe chest pain with radiation to upper back, so clinical suspicion of dissection was made and computed tomography (CT) aortogram done which revealed no evidence of dissection, but four small floating thrombi in the ascending aorta near junction with arch measuring 10–11 mm along with well-defined round-to-ovoid soft-tissue density lesion measuring 4.1/3.1 cm in the left suprarenal region [Figures 1 and 2]. He was initiated on enoxaparin and warfarin in addition to oral labetalol, perindopril, aspirin, atorvastatin, and nitroglycerin. At this time, there was no evidence of active coronary ischemia based on echocardiography, electrocardiogram, and troponin results and CT neck and cerebral angiography were unremarkable.

He was investigated for hormonal excess in view of adrenal mass and hypertension. On 5th day while awaiting hormonal report, he developed chest pain and neck pain again. This time electrocardiogram revealed fresh changes in precordial

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Access this article online

Quick Response Code:



Website:
www.ijccm.org

DOI:
10.4103/ijccm.IJCCM_407_17

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How to cite this article: Garg SK, Garg P, Urumdas M. An interesting presentation of pheochromocytoma. *Indian J Crit Care Med* 2018;22:40-2.



Figure 1: Filling defect due to thrombi in ascending aorta

leads (biphasic T-waves in V2–V4), positive and rising troponin, and new onset of regional wall motion abnormality in anterior wall and septum confirming non-ST elevation myocardial infarction. He was started on bisoprolol, and labetalol was stopped. The patient refused for coronary angiography and requested to go back to his home country for further management. The patient left for his home country on the same day and lost to follow-up. On 6th day, laboratory investigation reports confirmed pheochromocytoma as his plasma metanephrines, and urinary catecholamines were significantly elevated. His plasma-free metanephrine level was 76 pg/ml and urinary dopamine and norepinephrine 512 ug/g creat and 126 ug/g creat, respectively.

DISCUSSION

Pheochromocytoma is known to have varied clinical presentation. However, the presence of stroke, aortic thrombi, and myocardial infarction in one patient during single hospitalization has not been reported in medical literature. We also suspected thrombophilia initially; however, there was no evidence of venous thrombosis and presence of adrenal mass in hypertensive patient lead us to look for pheochromocytoma.

The clinical presentation of pheochromocytoma is mainly attributable to the excessive secretion of catecholamines.^[4] Approximately 91% of pheochromocytoma patients present with typical symptoms. The remaining 9%–10% present with atypical symptoms such as weakness, chest pain, paralysis, or claudication.

Atypical central nervous system symptoms are mainly due to ischemia.^[1] Cerebral ischemia leading to neurological features is a rare manifestation of pheochromocytoma. Various factors considered responsible for cerebral ischemia include severe hypertension with deranged cerebrovascular autoregulation, cerebral vasospasm, thrombosis *in situ*, or embolization from a left ventricular thrombus.^[1] Since a CT cerebral angiographic study was normal in our case, we cannot precisely pinpoint the cause as there was presence of aortic thrombi also. The precise

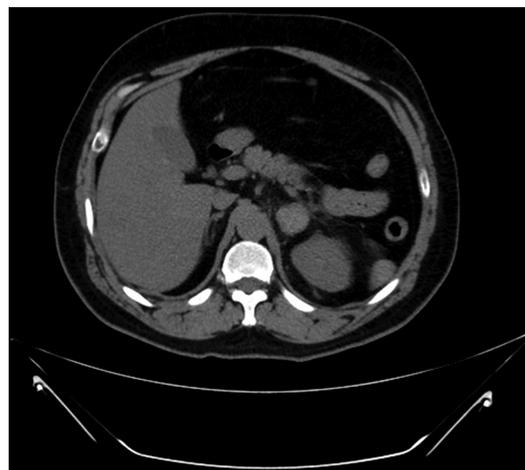


Figure 2: Left adrenal mass

vascular cause of such a stroke can seldom be determined with certainty. Our patient showed disappearance of neurological signs within 24 h.

Pheochromocytoma can result in various cardiac manifestations including ventricular hypertrophy, myocarditis, dilated cardiomyopathy, and congestive heart failure or occasionally, as in our case, acute coronary ischemia.^[5] During a pheochromocytoma crisis, a myocardial oxygen demand-supply mismatch can occur due to an increased afterload (vasoconstriction), catecholamine driven tachycardia, and catecholamine-driven coronary vasospasms.^[5] This can precipitate myocardial ischemia with concomitant electrocardiographic and cardiac enzyme abnormalities, even in the absence of coronary atherosclerosis.^[5] In general, patients presenting with symptoms and signs of an acute myocardial infarction will be treated with β -adrenergic blockage. However, in the presence of a pheochromocytoma, inhibition of β -adrenergic receptors will cause paradoxically increased blood pressure due to unopposed α -adrenergic receptor stimulation, thus further aggravating the condition of the patient as it happened in our patient. When our patient developed acute coronary ischemia, we changed his labetalol to bisoprolol which led to marked rise in his blood pressure again. This rise in blood pressure was due to initiation of bisoprolol, or merely a manifestation of episodic nature of pheochromocytoma cannot be ascertained definitely. Our patient had recurrent chest pain and which occurred even when the patient was on full medical management including aspirin, atorvastatin, and enoxaparin and it always improved spontaneously.

Numerous published reports have documented the association of pheochromocytoma with the development of systemic and intracardiac thrombi.^[6] It is likely that catecholamines and other hormones secreted by pheochromocytoma may play an important role in the pathogenesis of thrombosis.^[3] It has been reported that platelet aggregation is increased in patients with a pheochromocytoma.^[7,8] Of the 28 patients reported in the literature, a left ventricle thrombus was identified in eight. Inferior vena cava thrombi were present in 13 patients.^[6]

Only one case report has documented an association between pheochromocytoma and an isolated acute arterial thrombosis.^[9]

CONCLUSION

The diagnosis of pheochromocytoma is challenging because the symptoms can be episodic and the overall incidence and index of suspicion are low. Whenever, a patient who presents with unusual clinical manifestations or course of organ ischemia along with hypertension which is difficult to control with conventional oral or intravenous antihypertensives, it should be suspected. Failure to diagnose and treat the underlying disorder has its own consequences including wrong management and mortality. Management of its all manifestations is primarily based on the control of excessive catecholamine release.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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