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Non-cardiogenic Pulmonary Edema as the First Manifestation of Extra-adrenal Pheochromocytoma

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Abstract:

Pheochromocytomas are neuroendocrine, catecholamine-secreting tumors which are usually found in adrenal glands though they are occasionally seen in extra-adrenal locations. Classically the patients present with symptoms of headache, diaphoresis and tachycardia with hypertension. It is rarely associated with cardio-vascular manifestations, such as angina pectoris, acute myocardial infarction, myocarditis, acute heart failure, and cardiogenic shock among others. We present here a case of extra-adrenal pheochromocytoma presenting with flash pulmonary edema secondary to myocarditis.

Key words: Adrenal Gland Neoplasms, Catecholamines, Heart Failure, Myocarditis, Pheochromocytoma.

Introduction

Pheochromocytomas are well vascularized catecholamine producing tumors that arise from cells derived from sympathetic or para-sympathetic paraganglia. They are known to arise sporadically or may be associated with multiendocrine system disorder like multiple endocrine neoplasia type 2. It commonly arises from adrenal medulla or extra-adrenal abdominal paraganglion tissue. The catecolamines secreted by the tumor cause persistent hypertension [1].

Among these various symptoms, it is commonly associated with the triad of palpitations headache and sweating. Pheochromocytomas do not always produce symptoms and can be an incidental finding during imaging of abdomen for other causes and hence has been described as incidentaloma. The other end of the spectrum includes medical emergencies like myocardial infarction, myocarditis, and hypertensive crises with cerebrovascular accidents like intracranial bleed [2].

We present here a case of 16 year old boy who presented with pulmonary edema secondary to accelerated hypertension. Further evaluation revealed extra-adrenal pheochromocytoma with myocarditis. The patient underwent successful excision of the tumor and is presently normotensive on follow up.

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Case Report

A 16 year old boy presented to the emergency room with an eight hour history of sudden onset breathlessness. His blood pressure was 170/130 mm Hg and a high volume pulse. On physical examination the patient had pallor with raised jugular venous pressure. Respiratory system examination revealed bilateral basal crepitations with scattered rhonchi. Other systemic examinations revealed no abnormalities. The patient was started on intravenous diuretics and supportive treatment. A complete hematological examination revealed anemia with hemoglobin of 7.5 gm% with leucocytosis. Chest roentogram showed pulmonary edema, T wave inversions with left axis deviation was present on electrocardiogram. Echocardiography showed concentric left ventricular hypertrophy with global hypokinesia of left ventricle, mild diastolic dysfunction and ejection fraction of 45%. Serum creatinine phosphokinase was raised. A diagnosis of acute pulmonary edema with myocarditis probably of viral origin was made.

In view of accelerated hypertension in such a young patient, ultrasonography of abdomen and renal Doppler studies were performed. Ultrasonography revealed normal echotexture of renal parenchyma and normal renal artery Doppler studies. A mass with heterogeneous echotexture of the size 48x45x30 mm was noted in the right pre and para-vertebral region just above the umbilicus.

An abdominal contrast enhanced computed tomography revealed a heterogeneous enhancing retroperitoneal mass at the level of bifurcation of abdominal aorta. A diagnosis of paraganglioma arising from the organ of Zukerkandle suggestive of extra-adrenal pheochromocytoma was made. Twenty four hour urine metanephrines level were assessed and found to be raised thus confirming the diagnosis of extra-adrenal pheochromocytoma.

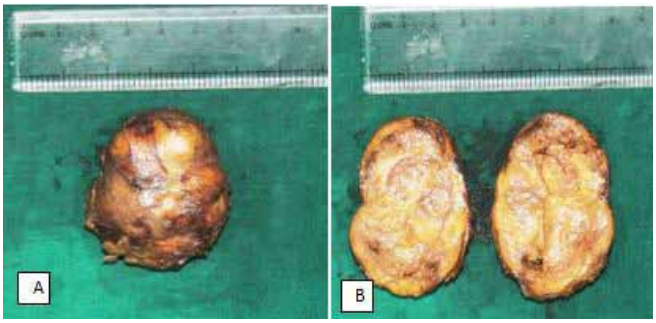

Fig.1:(A) Gross section measuring 4.5x4x4 cms. **(B)** Cut section showing gross areas of necrosis.

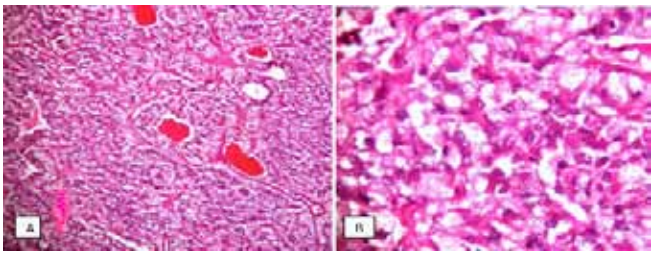

Fig.2: Large pink cells arranged in nests.

The patient was started on oral alpha blocker prazosin and scheduled for surgical excision of the tumor. An open excision of the tumor was done [Fig.1,2], intraoperative spike in blood pressure was managed by intravenous nitroprusside and esmolol. The patient was discharged after seven days. On further follow up visits the patient remained normotensive. Echocardiography after three months revealed no regional wall motion abnormalities and an ejection fraction of 55%.

Discussion

The word "Pheo chromo cytoma" is derived from Greek word phaios dark, chroma color, kytos cell; oma tumor [3]. It is derived from the chromaffin cells of the medulla of the adrenal gland which take up black colored staining secondary to chromaffin oxidation of catecolamines, hence the name.

Pheochromocytoma has been described as "the great masquerader" as it can present with a constellation of symptoms and signs misleading the diagnosis for years. The usual symptoms of headache and palpitations may be absent, as in this case. Pheochromocytoma can present with unusual presentations like dilated cardiomyopathy, myocarditis and sudden death [4]. Although hypertension is the hallmark of pheochromocytomas the patient can present with hypotension and even cardiogenic shock [5]. Excess norepinephrine can induce myocardial injury and desensitization of the blood vessels to adrenergic stimulation, which all result in cardiogenic shock [6].

It has been observed from previous studies that catecholamine-induced myocardial injury is reversible with complete recovery of cardiac dysfunction once the epinephrine secreting tumor is removed [7]. In the present case the patient was normotensive the next day of surgery with complete recovery of cardiac dysfunction at the end of three months.

Acute myocarditis as primary manifestation of pheochromocytoma occurs in less than 5% of the cases [8]. In younger age group as in our patient once the diagnosis of myocarditis is made, it is immediately suspected to be of viral etiology. This leads to a completely different line of investigations and treatment hence pheochromocytoma should always be considered as a differential even in normotensive patients. A screening abdominal sonogram must always be advised in such group of patients.

Conclusion

A diagnosis of pheochromocytoma should be considered in patients with hypertension and unexplained symptoms. Its recognition requires a high index of suspicion and failure to diagnose can result in life threatening complications.

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