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A Case Report

DILATED CARDIOMYOPATHY WITH HIDDEN PHEOCHROMOCYTOMA

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Abstract:

This report describes the case of a 38-year-old male with a no significant past medical history. The patient presented with the complaints of shortness of breath and episodic headache. In ER, his BP was 138/88 and physical examination was significant for lower lung field crackles and 1+ edema on dorsum of feet. His labs showed increased pro-NT BNP for 1328, CXR with mild to moderate pulmonary edema and cardiomegaly. Transthoracic echocardiogram showed EF of 25% with global hypokinesis. Administration of metoprolol succinate and lisinoprol, increased the systolic BP to 190 and secondary workup for hypertension showed increased plasma metanephrines which was subsequently confirmed by elevated 24-hour urine metanephrines. He had abdominal CT and MRI which showed mass in left adrenal gland. Elective left adrenalectomy yielded an adrenal gland mass which on biopsy confirmed pheochromocytoma.

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INTRODUCTION:

Pheochromocytoma is an important but sometimes untraceable cause of secondary hypertension. The pulsatile secretion of catechol- amine not only leads to hypertension, but also leads to a series of cardiac complications including cardiomyopathy and arrhythmia, [1-4] which sometimes are the initial manifestations of pheochromocytoma, and also make the diagnosis more intriguing

CASE:

This report describes the case of a 38-year-old male presenting with shortness of breath. In ER, his BP was 138/88 and physical examination was significant for lower lung field crackles and 1+ edema on dorsum of feet. His labs showed increased pro-NT BNP for 1328, CXR with mild to moderate pulmonary edema and cardiomegaly. He was given Lasix and his symptoms improved.

Transthoracic echocardiogram was done which showed EF of 25% with global hypokinesis. His cardiac stress test was normal. He was started on metoprolol succinate and lisinoprol, following which his systolic BP during the hospital stay was found to increase to 190s which was initially thought to be secondary to anxiety. But secondary workup for hypertension showed increased plasma metanephrines which was subsequently confirmed by elevated 24 hour urine metanephrines. He had abdominal CT and MRI which showed mass in left adrenal gland. He was subsequently started on phenoxybenzamine and discharged home with plan for followup after 2 weeks for elective left adrenalectomy.

2 weeks later on he had abdominal surgery for the resection of adrenal gland mass which on biopsy confirmed pheochromocytoma. Post operatively he followed with outpatient cardiology and was continued on metoprolol and lisinoprol. 4 months after the surgery, followup echocardiogram showed improved ejection fraction to 55-60%.

Histopathology Findings: Well circumscribed lesion with polygonal cells with granular cytoplasm (highlighted with yellow arrow head) with round to oval nuclei arranged in nested/trabecular pattern. A typical histopathological pattern of pheochromocytoma

CXR: Inc pulmonary vascular congestion and increased interstitial marking consistent with pulmonary edema. Moderate cardiomegaly, Transthoracic echocardiogram with apical 4 chamber view: dilated left ventricular cavity with normal thickness. LV ejection fraction of 20-25%

Advanced Radiological Findings: CT abdomen showing right adrenal adenoma 36 x 23 mm and MRI right adrenal adenoma 36x24 mm. In this condition on MRI patient had non-enhanced lesion with lipid features. However, in clinical context of elevated plasma metanephrine and 24 urine metanephrines. Patient underwent left adrenalectomy and subsequent biopsy result proved that patient had pheochromocytoma.

Lab findings:

Free plasma metanephrine 186 pg/ml (normal is less than 57 pg/ml)

Free plasma normetanephrine 391 pg/ml (normal is less than 148 pg/ml)

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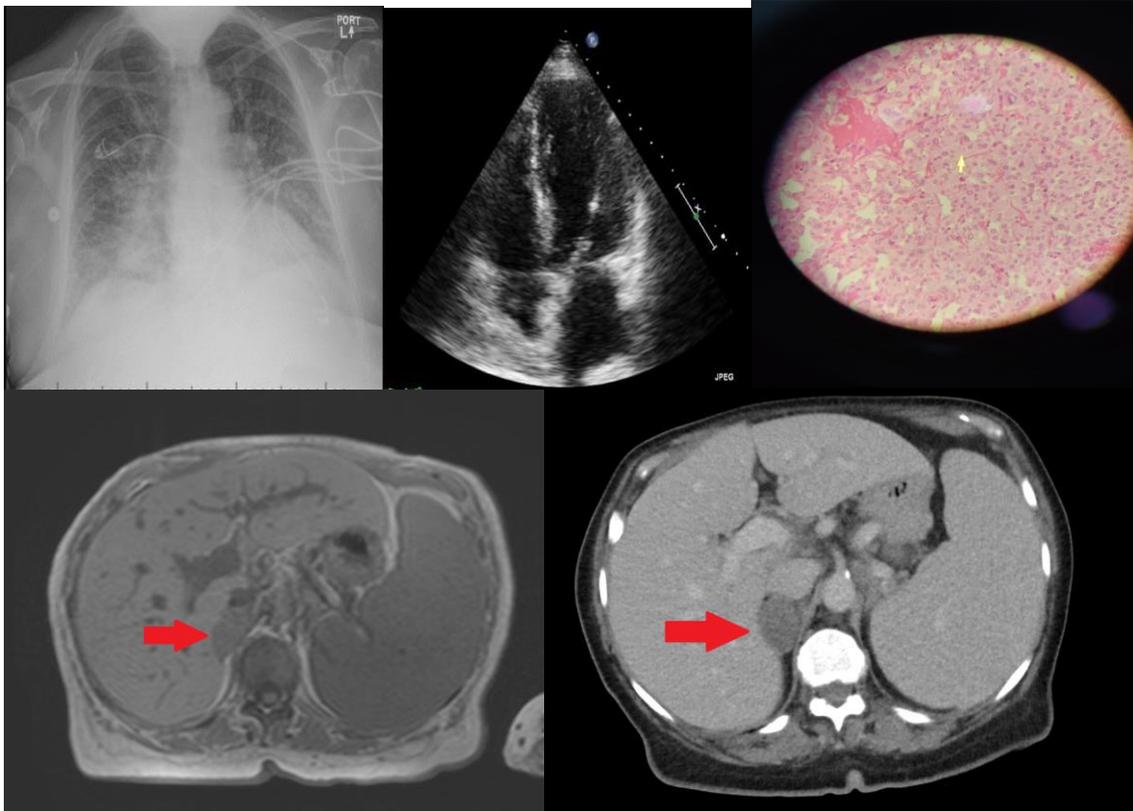
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HISTORY:

There was no significant past medical history, except shortness of breath (3 months). He also admitted to experiencing episodic headaches which were diagnosed as migraines and he was taking acetaminophen and naproxen for it, which helped decreasing pain. He denied smoking or using any illicit drugs.

DISCUSSION:

The intraoperative presentation of pheochromocytoma is rare but has been previously reported.^[5-8] The peak

blood pressure and catechol- amine levels measured were very high compared with those reported in other series.^[9] Predominantly epinephrine-secreting tumors make up 10% to 17% of all pheochromocytomas and are more likely to manifest with paroxysmal symptoms of beta-adrenoreceptor hyperstimulation, namely, palpitations, sweating, blanch- ing, anxiety, and hyperpyrexia.^[9] Apart from headaches, this patient had no symptoms, possibly because of beta-adrenoreceptor antagonism with atenolol.

The patient demonstrated both typical and atypical features of pheochromocytoma. The development of pulmonary edema and dilated cardiomyopathy are rare but recognized associations of pheochromocytoma. The pulmonary edema is secondary to postcapillary pulmonary vasoconstriction and catecholamine-induced myocardial dysfunction. It is usually fatal unless identified and managed promptly. ^[10] The myocardial damage results from the direct cardiotoxicity of high concentrations of catecholamines, increased cellular oxygen demand, and decreased coronary perfusion caused by adrenergic-mediated vasospasm. ^[11, 12]

The ECG changes together with the raised troponin T level and low cardiac output in this patient support the diagnosis of catecholamine-induced myocardial necrosis. Echocardiograms demonstrated dilated cardiomyopathy. This is the commonest pheochromocytoma-associated cardiomyopathy, and normally reverses over a period of months after tumor resection or aggressive medical treatment. ^[13]

Furosemide was used initially to treat the acute hypertensive crisis because the use of labetalol in pheochromocytoma has also been reported to precipitate hypertensive crisis and elevated vascular resistance. ^[14]

CONCLUSION:

In summary, this case highlights some interesting points. Pheochromocytomas can occur unexpectedly in the perioperative period, and the mortality rate is quoted as 33% to 50%. ^[5, 15, 16] A pheochromocytoma that manifested as a hypertensive crisis and cardiovascular instability following external palpation of the anesthetized patient's abdomen is detailed. Pulmonary edema and reversible catecholamine-induced cardiomyopathy developed. The cardiovascular instability and cardiomyopathy were reversed with medicinal treatment and successful surgical removal of the tumor at a later date.

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