Case Report

Pheochromocytoma - An unusual cause of broken heart syndrome.

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Abstract

Takotsubo cardiomyopathy or stress induced cardiomyopathy is a recently emerged clinical entity though it was known as broken heart syndrome for sometime. Here we report a 47-year- old female with type 2 diabetes mellitus who presented with an episodic chronic headache and excessive sweating of face and head. She had an acute coronary syndrome leading to cardiogenic shock, respiratory failure and needed assisted ventilation during the hospital stay while being evaluated for headache. She had marked haemodynamic instability as well. A coronary angiogram confirmed normal epicardial vessels and the 2D echocardiogram revealed antero-septal and apical hypokinesia with poor systolic function. Considering the clinical picture, a presumptive diagnosis of pheochromocytoma was made and a right supra renal mass was detected by ultrasonography and subsequently confirmed by a CT scan. However, the urinary Vanillin Mandalic Acid (VMA) and metanephrine levels were repeatedly within normal limits. Once she recovered from the acute cardiac event, she under went a laparoscopic adrenalectomy and the histology of the tumour and subsequent immunohistochemical studies confirmed a pheochromocytoma. All her symptoms completely disappeared, and she was normotensive with stable blood pressure recordings after the surgery.

Key words

phaechromocytoma, takotsubocardiomyopathy, cardiogenic shock

Case Report

A 47-year-old woman from northern Sri Lanka presented with a history of intermittent headaches and excessive sweating of face and head of one-week duration. She was diagnosed with diabetes mellitus two years ago and had been under reasonable glycaemic control. Headaches were episodic in nature and were associated with palpitation, nausea and vomiting. Each episode of headache lasted around ten minutes and it was not associated with photophobia or phonophobia. On admission, her vitals were stable with a blood pressure of 90/60mmHg and a pulse rate of 78/minute, regular. No focal neurological signs were elicited. Rest of the systemic examination findings and initial investigations were normal.

She developed severe shortness of breath six hours following a contrast CT brain which was taken to rule out an intracranial lesion. During this episode, her blood pressure dropped to 80/60 mmHg and her heart rate was raised to 106 beats per minute. There were bilateral fine crackles on both lung fields more marked on mid and the lower chest. She also started to desaturate and her SpO_2 dropped to 76 % on ambient air. She was clinically diagnosed with an acute coronary syndrome complicated with acute left ventricular failure. Repeated ECGs taken at this point showed dynamic T inversions in the anterior leads. She was treated with anti-anginal medications along with other supportive care including Bi-PAP. Despite the initial treatment, her clinical condition deteriorated, and she went into refractory hypotension and respiratory failure. Thereafter she

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was managed with invasive ventilation and inotropes (needed four inotropes with maximum doses) along with other supportive care.

Troponin I levels 6h after this event became positive with the titre of 6.07ng/ml and 2D-ECHO done at bed side showed antero-septal and apical hypokinesia with an ejection fraction of 30%. With supportive care there was improvement in terms of physiological and clinical parameters and finally she was weaned off the ventilator and inotropes.

Subsequently she underwent a coronary angiogram which showed normal epicardial vessels. During her ICU stay, she experienced episodes of both hypotension and hypertension and erratic blood pressure recordings which required both anti-hypertensive medications and inotropes alternatively. These hypertensive episodes were associated with excessive sweating and palpitation.

Considering the whole clinical picture and course of the illness, pheochromocytoma was considered as a differential diagnosis. Initial ultrasound scan of the abdomen showed right sided suprarenal mass which was further confirmed by CECT abdomen which showed small enhancing mass of 4.3cm x 2 cm in the right suprarenal area (Figure 01). Her 24hours urinary excretion of VMA and metanephrines were repeatedly within normal range.

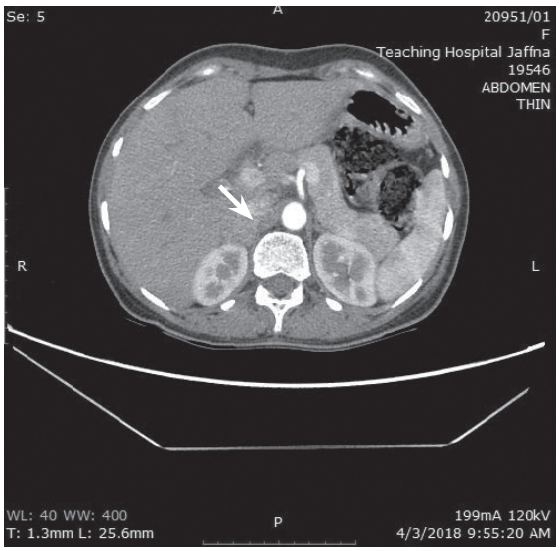

Figure 01: Contrast enhanced computerized tomographic image of Right side adrenal gland (Arrow)

Despite the biochemistry being normal, given the high degree of clinical suspicion, finally, she underwent a laparoscopic adrenalectomy. Surgery was performed after complete recovery from acute cardiac event and it was confirmed by a repeated 2 D Echocardiogram as well. She opted for a laparoscopic approach in a multidisciplinary team meeting when her cardiopulmonary function was back to normal after the acute cardiac event. Macroscopic appearance of tumour is shown in figure 2. Histology of tumour turned out to be a pheochromocytoma and it was further confirmed by immune histochemistry (Figure 03). All her symptoms completely disappeared after the surgery and she became normotensive with stable blood pressure.

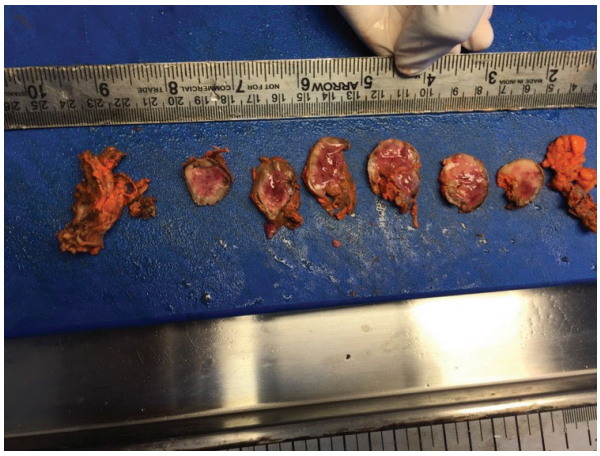

Figure 02: Macroscopic appearance of adrenal tumour after surgical removal

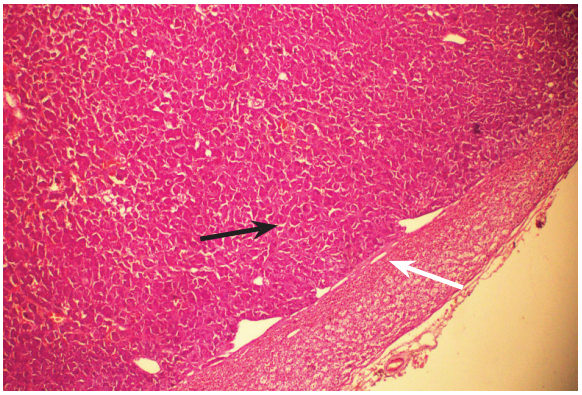

Figure 03: Histological appearance of normal tissue (white arrow) and the tumour (black arrow) interface of pheochromocytoma under Haematoxylin and eosin stain

Discussion

Pheochromocytoma is a tumoral disease that originates from chromaffin cells in the sympatho-adrenal system. These tumours secrete epinephrine and norepinephrine. However, they may also produce atrial natriuretic peptide, vasoactive intestinal peptide, endothelin, erythropoietin, dopamine, neuropeptide Y, and adrenomedullin(1). Pheochromocytoma is a rare disease; it is present in only 0.1% of hypertensive patients(2). Although there is no difference in age or gender, occurrence increases in the 4th and 5th decades. Pheochromocytoma is often called as a '10% tumour' because 10 percent of them are bilateral, malignant, extra adrenal, multiple, familial and occur in children(3). Our patient had a right sided, non-malignant and single adrenal tumour. The most common clinical sign of pheochromocytoma is a sustained or paroxysmal hypertension, and the most common symptoms are headache, excessive truncal sweating, and palpitation. Angina pectoris, abdominal pain, queasiness, anxiety, and paleness are other symptoms of it as well. Patients with tumours that predominantly secrete epinephrine can present with hypotension or even shock, caused by hypovolemia, desensitization of adrenergic receptors or abrupt cessation of catecholamine secretion owing to tumour necrosis. This might be the possibility in our patient who had persistent hypotension before surgery (4).

Acute coronary syndrome in the absence of atherothrombotic coronary artery disease has limited differential diagnosis. Takotsubo cardiomyopathy is one of the main causes for this kind of presentation. Relationship between catecholamine-induced shock and pheochromocytoma has been recognized for more than 40 years, but the link between takotsubo cardiomyopathy and a pheochromocytoma related crisis leading to acute cardiac failure or shock was not recognised until recently. It can be triggered by any stresses. Though benign, surgery is advocated for giant pheochromocytomas. As laparoscopy offers a better anatomical exposure, shorter length of stay, a decrease in postoperative pain, faster return to preoperative activity level, improved cosmetic, and reduced blood loss, early to resumption of oral feeding (5), we offered her a minimally invasive surgery option and which resulted in a favourable outcome. Early vascular control, minimal handling of the tumour and a multidisciplinary approach to combat potential intra-operative crisis are the cornerstones in managing such cases. Malignant counterparts need to be excluded histologically and our patient didn't have malignant component (6). The possibility of relapses of the tumour makes lifelong follow-up obligatory (7).

The authors would like to emphasize two key messages for the clinicians from this rare clinical presentation. The clinical picture of acute coronary syndrome and LV dysfunction with normal coronary epicardial vessels should raise the suspicion of stress induced cardiomyopathy and episodes of headache associated with palpitations and fluctuating blood pressure should warn us the possibility of pheochromocytoma being the cause for the cardiomyopathy.

Although abnormal biochemistry is considered the cornerstone of diagnosis for endocrine tumours however as illustrated by this case report a high degree of clinical vigilance should prompt the clinician to recommend surgical excision of the tumor for histological confirmation of diagnosis and for the cure of this potentially fatal condition.

Conflict of interest

The authors declare that there is no conflict of interest regarding the publication of this paper.

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