Catecholamine-induced Cardiomyopathy: Pitfalls in Diagnosis and Management

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Background

Cardiomyopathy as the initial presentation of phaeochromocytoma (PCA) is uncommon. Diagnostic work-up and peri-operative management may be challenging within this context. We report 3 cases of PCA presenting with cardiomyopathy to illustrate the pitfalls in diagnosis and management. None of the patients had typical adrenergic symptoms of catecholamine excess and all patients were well established on beta-blockers on presentation. All 3 patients had an adrenalectomy and a PCA was confirmed on histology.

Case Reports

Patient 1: A 66 year-old male was referred with a 3.1 cm heterogeneous right adrenal mass with an attenuation of 56 Hounsfield units (HU). Previous investigations revealed severe left ventricular impairment and non-flow limiting coronary disease. Plasma and urinary metadrenalines were raised and asymmetrical adrenal uptake was noted on an MIBG scan.

Patient 2: A normotensive 56 year old male with dilated cardiomyopathy, was found to have an incidental longstanding 3 cm right adrenal nodule with an attenuation of 48 HU. Plasma and urine metadrenaline levels were raised during decompensated heart failure and improved with heart failure treatment (figure 1 and 2). However, a subsequent increase catecholamine levels was noted despite clinical and echocardiographic improvement of cardiac function. MIBG and Octreotide scans were both negative but Computed Tomography (CT) scan of the right adrenal lesion with wash-out studies showed indeterminate characteristics. Adding alpha-blocking therapy to his beta-blocker resulted in troublesome hypotension prior to surgical intervention.

Patient 3: A 61 year-old female with Type 1 Neurofibromatosis presented with Takotsubo cardiomyopathy. Fractionated metadrenaline levels remained elevated despite normalisation of cardiac function on echocardiography. A 2.4 cm left adrenal mass with an attenuation of 43 HU was found on CT scan. An increased left adrenal uptake was demonstrated on subsequent MIBG scan.

Figure 1: Plasma metadrenaline levels prior to surgery
(reference value 80 – 510 pmol/L)

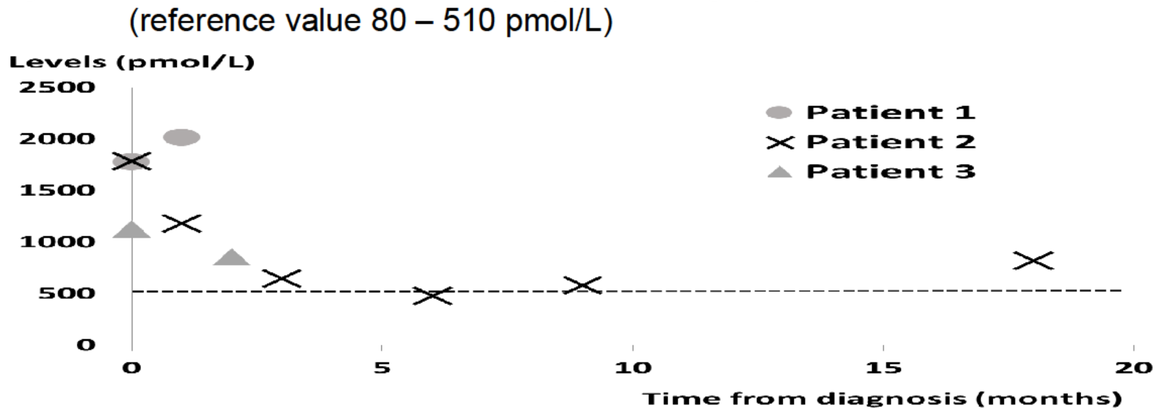
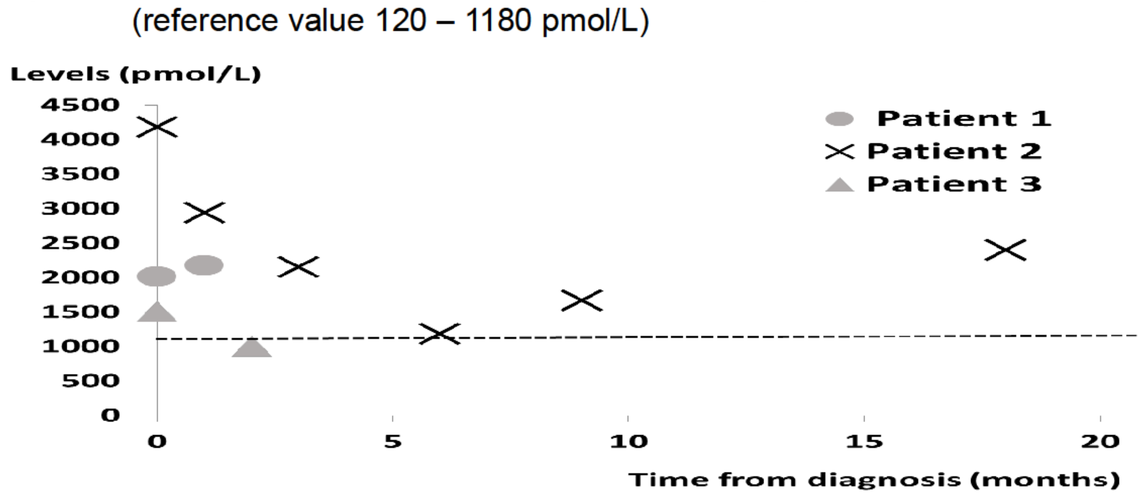


Figure 2: Plasma normetadrenaline levels prior to surgery
(reference value 120 – 1180 pmol/L)



Literature Review

We found 49 cases of PCA presenting primarily with cardiomyopathy between 2000 and 2013. Table 1 shows the demographic and presenting characteristics of these patients. Apart from a younger median age, the absence of adrenergic symptoms was notable in a significant proportion of patients. Takotsubo cardiomyopathy was relatively over-represented in this population. These adrenal lesions were over 2 cm in size when documented.

Table 1: Characteristics of 49 cases of PCA crises presenting with cardiomyopathy between 2000 and 2013

Parameter	N
Median Age (years)	39 (13 – 72)
Female Gender	25 (51%)
Adrenergic symptoms	20 (41%)
Blood Pressure anomalies	37 (76%)
Type of Cardiomyopathy	
Takotsubo	12 (24%)
Left ventricular dysfunction or unspecified	35 (71%)
Myocarditis	2 (5%)
Left adrenal PCA	17 (63%)
Mean size of PCA (cm)	5.9 (2.1 – 14)

Discussion

Screening for PCA in patients with cardiomyopathy is problematic as cardiac failure of any aetiology generates an adrenergic response. Hence interpretation of biochemical tests for catecholamine excess is difficult. The routinely used screening tests (plasma and urinary catecholamines) have not been validated in patients with cardiac failure.

Screening all patients with idiopathic cardiomyopathy for catecholamine excess is highly likely to generate false positive results, hence not a recommended strategy. In addition the incidence of adrenal incidentalomas is rising due to the increasing use of abdominal CT imaging studies over the last decade. With most of these CT scans being performed in the older patient group, a significant proportion of these incidentally discovered adrenal lesions is expected to coexist in patients with cardiac failure of known or unknown aetiology. Current endocrine guidelines on investigating adrenal incidentalomas do not differentiate this subpopulation group.

In the absence of peer-reviewed guidelines a high index of suspicion of occult phaeochromocytoma, in patients with idiopathic cardiomyopathy or with adrenal incidentalomas and cardiac failure of any aetiology, should prompt further diagnostic work-up.

Factors contributing to a high pre-test probability of PCA are: younger age, genetic predisposition to PCA, presence of adrenergic symptoms and blood pressure abnormality (labile blood pressure, hyper and/or hypotension), takotsubo cardiomyopathy and radiological characteristics of PCA. The latter includes adrenal lesions of heterogeneous composition, increased attenuation (>20 HU), delayed contrast medium washout and possibly greater than 2cm in size. The role of CT with washout studies as second line diagnostic imaging modality in this population requires special consideration given that one of our patients had negative MIBG and octreotide scans but the washout studies were the only abnormal investigations revealing indeterminate characteristics.

Conclusions

Investigating patients with cardiomyopathy for possible phaeochromocytoma remains challenging. Routine screening tests are difficult to interpret. A high index of suspicion should prompt further diagnostic work-up with judicious use of currently available imaging modalities as the second line investigative strategy.

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