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PAINFUL HEART: Pheochromocytoma / Paraganglioma - induced Takotsubo Cardiomyopathy

Zhimin Lin[1,2], Bennett Soh[2], James Lee[2,3], Kee Yuan Ngiam[2,3], Anil Dinkar Rao[1,2], Rajeev Parameswaran[2,3]

1 Khoo Teck Puat Hospital, Division of Endocrine Surgery, Singapore. 2 Yong Loo Lin School of Medicine, National University of Singapore, Singapore. 3 National University Hospital, Division of Endocrine Surgery, Singapore.

INTRODUCTION

Pheochromocytoma and paraganglioma (PPGL) can present with Takotsubo cardiomyopathy which has a presentation like acute coronary syndrome (ACS) and cardiogenic shock.

There are only 104 cases described in the literature[A], hence it is a very rare yet life-threatening disease that should be promptly recognised.

We present a local series of PPGL-induced Takotsubo cardiomyopathy and the challenges associated with the condition, for both diagnosis and management.

METHODOLOGY

Retrospective cohort review of patients who presented with PPGL-associated cardiomyopathy over 7 years (2017 – 2023) in 2 Singapore institutions with a dedicated Endocrine Surgery service.

DISCUSSION & CONCLUSION

- PPGL-induced cardiomyopathy usually precedes or leads to the diagnosis of PPGL[B]
- PPGL should be suspected in patients with acute heart failure without evidence of valvular or coronary artery disease[C], and with normal cardiac biomarkers
- These patients are associated with worse rates of morbidity and mortality compared to patients with only PPGL that were incidentally found[A,B,D]
- Metanephrines may be unreliable at the acute phase, especially with the use of catecholaminergic agents in ICU. This test his will also take time for processing (at least 2 weeks in our institution).[B]
- A CT-scan to look for PPGL should be performed[B]
- Removal of catecholamine exposure via PPGL resection was associated with an improvement (82%) in LV systolic function[E]

RESULTS

DEMOGRAPHICS	
Age (years)	Range 30 – 76, Mean 56
Gender	3 Male; 3 Female
Family history	1: Bilateral Pheochromocytomas (Mother) 5: None

PRESENTATION WITH TAKOTSUBO CARDIOMYOPATHY	
Cardiac symptoms	5: Angina type symptoms & Hypertensive crisis, breathless, requiring ICU stay 1: Hypotensive cardiogenic shock, requiring veno-venous ECMO. Initially diagnosed as a STEMI.
Troponins	All normal
Trans-thoracic echocardio-graphy	All had multiple regional wall motion abnormalities, severe systolic dysfunction, with global hypokinesia. Ejection fraction range 28% - 60%
Pre-op coronary angiogram	4: Normal 2: Triple vessel disease
Coronary Artery Bypass Graft Surgery (CABG)	Both patients with triple vessel disease underwent CABG, intra-op found <70% stenosis of all vessels

DIAGNOSIS AND MANAGEMENT OF PHEOCHROMOCYTOMA / PARAGANGLIOMA	
Metaneprhines + Catecholamines	All 4 fold higher than normal (Serum / Urine)
Screening for other adrenal hormones	All normal
Imaging for localisation	1: Bilateral pheochromocytomas (Left 9.9cm, Right 8.2cm) 3: Unilateral adrenal mass (size range 3.4 – 9.4cm) 1: Mediastinal paraganglioma (3.5cm) 1: Para-aortic paraganglioma (6.6cm)
Surgical approach (Adrenal masses)	Open for Bilateral Pheochromocytoma 1 Laparoscopic Retroperitoneal 2 Laparoscopic Transabdominal
Surgical approach (Paragangliomas)	Mediastinal: Open sternotomy Para-aortic: Open trans-abdominal
Pre-op α and β blockade	Used for all patients
Post-op cardiac function	All had normal echocardiography All had none or minimal anti-hypertensive medications

TUMOUR FEATURES & FOLLOW UP	
Genetic Mutations	SDH-B: 3 patients MEN 2A-634: 1 (Bilateral pheo) VUS: 1 patient
Follow up time	Range: 3 – 96 months Mean: 27 months
Metastasis development	Present in 2 patients with paragangliomas
Mortality	None

DEFINITIONS

Takotsubo Cardiomyopathy: Reversible cardiac failure. Characterised by left apical ventricular wall motion abnormalities, systolic dysfunction. Associated with normal cardiac biomarkers, and an absence of occlusive coronary artery disease.

Khoo Teck Puat Hospital
National Healthcare Group

National University Hospital

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