



Pheochromocytoma: A Great Masquerader

A 30 y old female with hypertension presented with intermittent headaches, chest pain and an episodes of presyncope a month ago followed by an episode of syncope after 2 weeks. Her headaches and chest pain generally coincided with rise in blood pressure. Her BP on admission was 160/90 mm Hg on Long acting Prazosin (5 mg) and Metoprolol (50 mg). She was hospitalised for further work up. ECG was normal (Fig 1) and 2D echo revealed mild LVH with normal LVEF without any RWMA. Her Troponin-I and CPK-MB were markedly elevated (Fig 2) and hence she was subjected to coronary angiogram which revealed normal coronary arteries. Her ECGs taken during symptomatic episodes (chest pain and headache) revealed marked QTc prolongation (583 and 553 msec) in two tracings (Fig 3,4), ST depression in inferior and lateral leads in one (Fig 5) and marked sinus tachycardia at the rate of 140 per minute in one tracing (Fig 6). Her free and 24 hours urinary normetanephrines and metanephrines were normal (Fig 7,8), however a well circumscribed 4.1 cm x 3.9 cm right suprarenal mass was found on abdominal ultrasound examination which was later confirmed on CT scan. An I-131 MIBG nuclear scan clinched the diagnosis of pheochromocytoma (Fig 9). After adequate pre-operative alpha and beta receptor blockade, she underwent robotic (da Vinci, Intuitive surgicals, Sunnyvale, CA, USA) right adrenalectomy (Fig 10) uneventfully. She required noradrenaline support for about an hour after surgery.

Fig 1

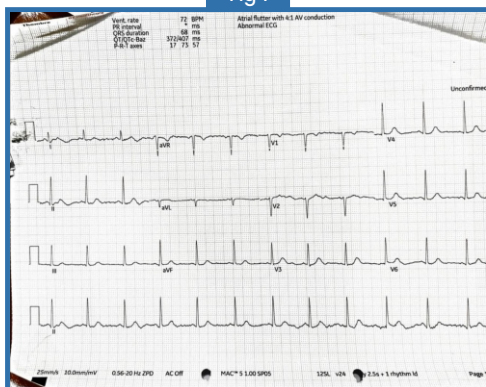


Fig 2



Fig 3

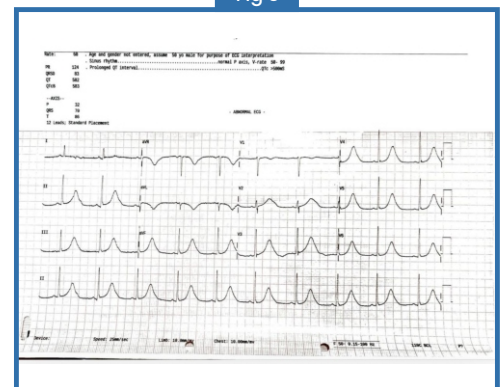


Fig 4

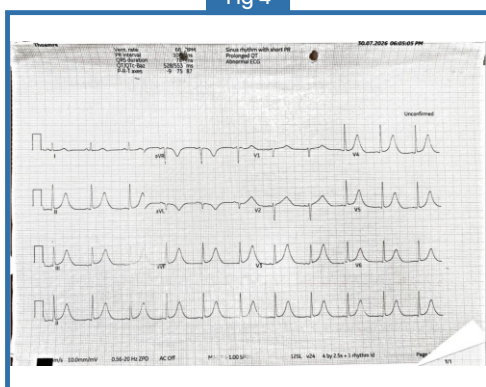


Fig 5

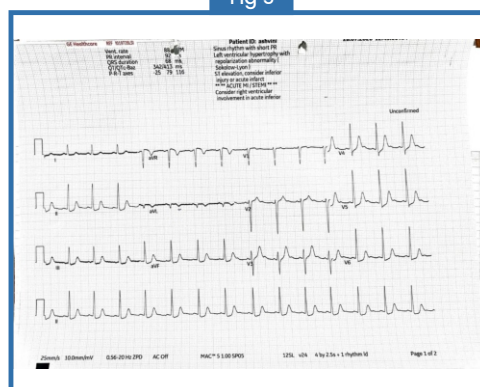


Fig 6



The Following Points Need Discussion

1. Her free and 24 hours urinary nor-metanephrines and metanephrines were normal and it was a surprising finding. However false-negative biochemical testing can occur with:

- small tumours
- intermittently secreting tumours (in this case this was the strongest possibility)
- non-functional/minimally functional tumours
- some genetically defined PPGLs
- tumours with unusual catecholamine biosynthesis.

2. MIBG is taken up by norepinephrine transport mechanisms in chromaffin tissue, so strong uptake in an adrenal mass substantially supports pheochromocytoma. However, MIBG is not an absolute biochemical confirmation. Normal adrenal tissue can show physiological uptake and false positives/false negatives are recognised. In this case, though, the combination of: Right adrenal mass on ultrasound & CT + compatible symptoms + hypertension + strongly positive MIBG on right side made pheochromocytoma considerably more plausible than an incidental non-functioning adrenal adenoma.

3. Intermittent marked QT prolongation was an interesting yet a well described finding. Catecholamines can produce dynamic myocardial electrical abnormalities, including:

- QT prolongation
- T-wave inversion
- ST-segment changes
- ventricular ectopy
- ventricular tachyarrhythmias.

Although the exact cause of her syncope could not be documented in this case, episodes of torsade-de-pointes secondary to prolonged QT remains a strong possibility.

4. Elevated Troponin and ST-T changes on ECG can mimic ACS as was evident in this case. Pheochromocytoma-associated cardiac disease can mimic ACS, myocarditis and Takotsubo syndrome. Importantly, ECG repolarisation abnormalities may occur with only modest troponin elevation and completely normal coronary arteries.

5. The major advantage of a robotic procedure is surgeon's ability to clip the adrenal vein/s before handling the tumour. This prevents catecholamine surges and hypertensive peaks during surgery.

Note: We thank our nuclear medicine chief Dr. Prathamesh Joshi, Robotic surgeon Dr. Rajesh Saoji for lending us the images.

Fig 7



Fig 8



Fig 9

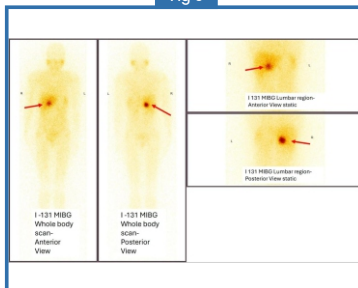


Fig 10





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