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Research Letter

Adrenal and extra-adrenal pheochromocytomas presenting as life-threatening ventricular arrhythmias: Report of three cases

ABSTRACT

Pheochromocytoma patients can rarely have prolonged QT interval in the ECG. We report three cases of pheochromocytoma in females presenting with ventricular arrhythmia; two had torsades de pointes and a third patient had frequent VPCs and nonsustained ventricular tachycardia. All the patients were treated with surgical removal of the tumor with complete relief of symptoms and normalization of QT interval.

Patients with pheochromocytoma are known to have various cardiac complications, including arrhythmias, heart failure, myocardial infarction, and cardiomyopathy. QT prolongation has been reported in these patients and can rarely predispose to serious ventricular arrhythmias resulting in syncope and cardiac death in the worst cases. There are however only few cases of torsades de pointes (TDP) associated with pheochromocytoma reported in the literature.[1,2]

In the last 4 years, we noted three cases of pheochromocytoma presenting with ventricular arrhythmia; two had TDP and a third patient had frequent VPCs and nonsustained ventricular tachycardia (NSVT). All the patients were treated with surgical removal of the tumor with complete relief of symptoms.

1. Case 1

A 35-year-old female, not a known hypertensive, presented with episodes of giddiness, palpitations, and sweating. Nearly 5 symptomatic episodes were seen over a period of 2 months. During an episode, her blood pressure (BP) was recorded as 240/120 mmHg. ECG monitor showed polymorphic VT (Fig. 1), which was cardioverted with 200 J DC shock. Her serum electrolytes, liver, and thyroid function tests were within normal limits. She was investigated for the labile hypertension with polymorphic VT. Baseline ECG showed a long QTc of 540 ms (Fig. 2). 2D echo showed structurally normal heart and coronary arteries were normal on coronary angiography (CAG). The Holter study showed no VT but there was evidence of persistent long QT of 598 ms. 24 h urinary metanephrines was 600 mcg (normal range: 30–180 mcg). Ultrasound and contrast CT-abdomen showed 7 cm × 4.6 cm echogenic mass in the right adrenal gland region (Fig. 3A).

After control of BP with prazocin 5 mg twice daily and metoprolol 50 mg twice daily, the patient underwent surgical excision of the adrenal tumor. Histopathology of the tumor showed round to polygonal cells with abundant granular amphophilic cytoplasm arranged in well-defined nests (zell-balen appearance) suggesting pheochromocytoma (Fig. 4). Postoperative period was uneventful. The antihypertensive drugs were weaned off gradually after the surgery. Follow-up Holter showed normal QT interval. The patient remained asymptomatic and normotensive over 3 years of follow-up and there was no evidence of ventricular arrhythmia.

2. Case

A 49-year-old female, a known hypertensive and hypothyroid patient, presented with recurrent syncopal attacks for the last 2 months, which were preceded by headache, sweating, and

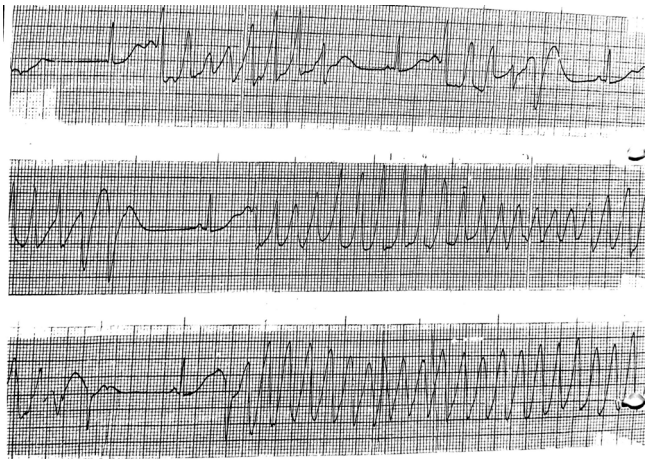

Fig. 1 – Cardiac monitor tracing showing polymorphic ventricular tachycardia.

palpitations. During the hospital admissions, she was found to have polymorphic VT requiring DC version three times in a span of 2 months. She was referred to our hospital for automatic implantable cardioverter defibrillator (AICD) implantation for recurrent TDP. During the index hospital admission, her BP was recorded as 230/120 mmHg. Her electrolytes, renal, liver, and thyroid status were normal.

Her baseline ECG was showing long QT of 580 ms. 2D echo and CAG were normal. Holter trace showed TDP (Fig. 5). Her plasma metanephrine was 200 nmol/L (normal range <0.5 nmol/L). Ultrasound and contrast CT-abdomen revealed a well-defined 6.4 cm × 4.3 cm mass in the right paracaval region (Fig. 3C) and a provisional diagnosis of extra-adrenal pheochromocytoma was made.

After titration of antihypertensive drugs (prazocin 10 mg twice daily and metoprolol 50 mg twice daily), she was subjected to surgical removal of tumor. Histopathology of the tumor confirmed the diagnosis of pheochromocytoma. She remained asymptomatic and her BP was normal during follow-up. Holter showed normal QT interval with no evidence of ventricular arrhythmia in the follow-up of 2 years.

3. Case

A 48-year-old female presented with episodic headache, sweating, and right loin pain of 2 months duration. She was found to have BP of 180/100 mmHg during these episodes of headache and sweating. She was investigated and contrast CT-abdomen revealed right adrenal mass of 10.4 cm × 6.2 cm (Fig. 3B). Her ECG, 2D ECHO, and CAG were normal. Plasma metanephrine was 280 nmol/L. She was started on prazocin

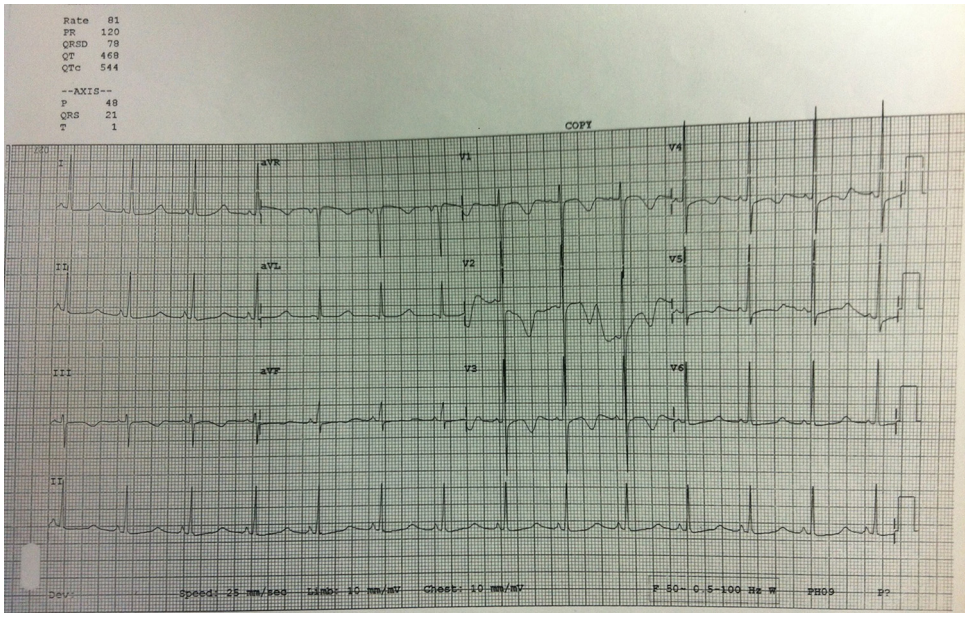

Fig. 2 – Baseline ECG showing long QTc of 540 ms.