Case Report

Pheochromocytoma with Uncommon ECG Presentation

Osama Tariq, Aftab Siddiqui, Adil Al Riami, Fatima Amir, Yasser W. Mula-Abed, Mohammed Hammed Gillani

ABSTRACT

Pheochromocytoma is a rare neuroendocrine tumour with a highly variable clinical presentation. It may present with unusual presentation and these tend to mislead the physician into a delayed diagnosis. Typical cardiac manifestations include ST-segment deviations mimicking myocardial infarction, QT interval prolongation, blood pressure fluctuations, arrythmias mostly tachycardia but rarely may present with bradyarrythmia. We describe a case of pheochromocytoma with rare findings of Intermittent AV dissociation associated with Junctional and ventricular ectopic rhythm.

KEYWORDS: Pheochromocytoma, Myocardial Infarction, QT interval prolongation, AV dissociation, Bradyarrythmia

INTRODUCTION

Pheochromocytoma is a neuroendocrine tumour of the medulla of the adrenal glands, originating in the chromaffin cells, or extra adrenal chromaffin tissue that failed to involute after birth[1]. It secretes excess amounts of catecholamines, usually noradrenaline, and adrenaline to a lesser extent[1]. The clinical manifestations of a pheochromocytoma result from excessive catecholamine secretion by the tumour. Cardiovascular manifestations are hypertensive emergencies, reversible dilated or hypertrophic cardiomyopathy[2] .ST segment changes mimicking MI[3,4] and sinus arrhythmias, namely sinus tachycardia with palpitations as the presenting symptoms, ventricular tachycardia [5] atrial fibrillation , and AV block [6].

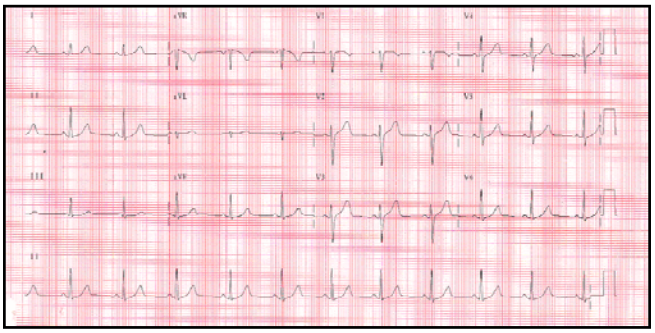
CASE PRESENTATION

A 37 year old married woman, mother of two children, presented with two years history of paroxysmal headache, palpitations and dry mouth. There was also a history of associated chest tightness lasting for few minutes. She was found to have high blood pressures during these occasions, with systolic >200 mm of Hg, and diastolic >100 mm of Hg mostly. She was on a combination antihypertensive (ARB with hydrochlorothiazide), despite which she had paroxysmal readings of high blood pressure. She had a family history of hypertension and her past medical history was not significant except for two caesarean sections and an appendicectomy..

She was seen initially in the medical OPD and was then admitted under the medical team, for work up of pheochromocytoma as a cause of her hypertension. Her antihypertensive medications were modified to labetalol 200mg BD and amlodipine 10 mg OD. On admission, a routine ECG was done which showed AV dissociation with junctional ectopics. A 24 hour holter monitor, 24 hour BP monitoring, Echo, 24 hour urinary metanephrines, CT abdomen and MIBG scan were ordered.

CT abdomen showed a right adrenal tumour 3.9 X 3.5 cm, and the MIBG scan showed abnormal focal area of increased radiotracer uptake in the right suprarenal region suggestive of pheochromocytoma. Echo showed mildly thickened AV cusps, but otherwise was a normal study. She was then discharged with plan to review the results in the clinic and was also referred to the surgeons. Her 24 hours holter monitor showed intermittent AV dissociation with ectopic junction and ventricular rhythm.

ECG I: NORMAL SINUS RHYTHM



ECG II: INTERMITTENT AV DISSOCIATION WITH JUNCTIONAL RHYTHM

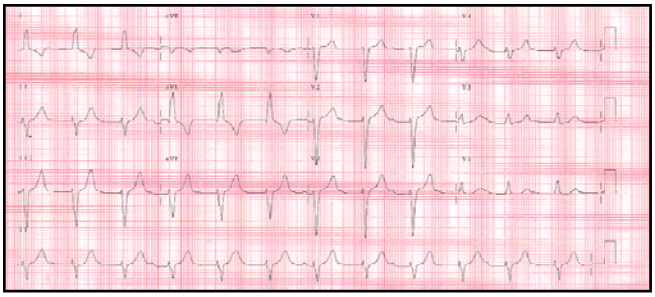
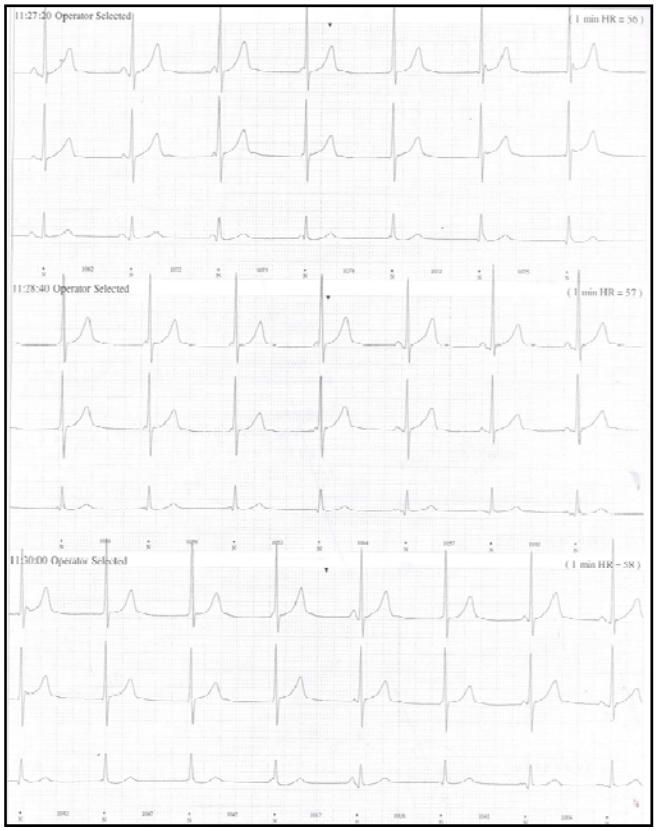
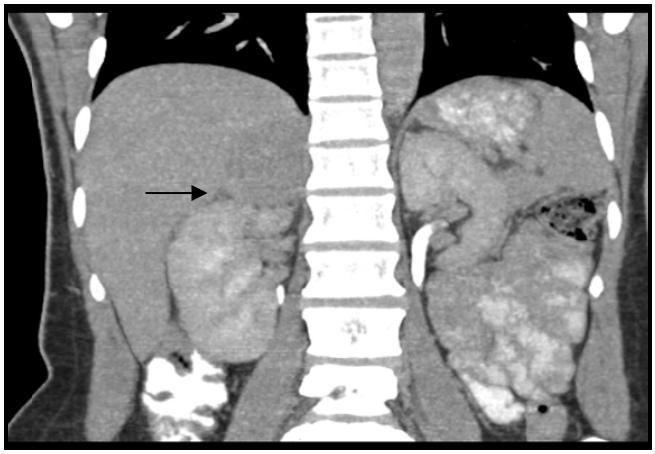


FIGURE A: 24 HOURS ECG HOLTER MONITORING TRACING



CT SCAN:

FIGURE B: RIGHT SUPERARENAL PHEOCHRO-MOCYTOMA



A week later, she presented to the emergency department with headache, palpitation associated with chest tightness. Her BP was found to be 210/140 mm of Hg, and it came down to 138/80 spontaneously after ten minutes. Her labetalol dose was increased to 200mg TID daily and amlodipine increased to 20mg OD.

Her biochemical results were reviewed in the endocrine OPD, and showed high catecholamine levels,

FIGURE C: RIGHT SUPERARENAL PHEOCHRO-MOCYTOMA

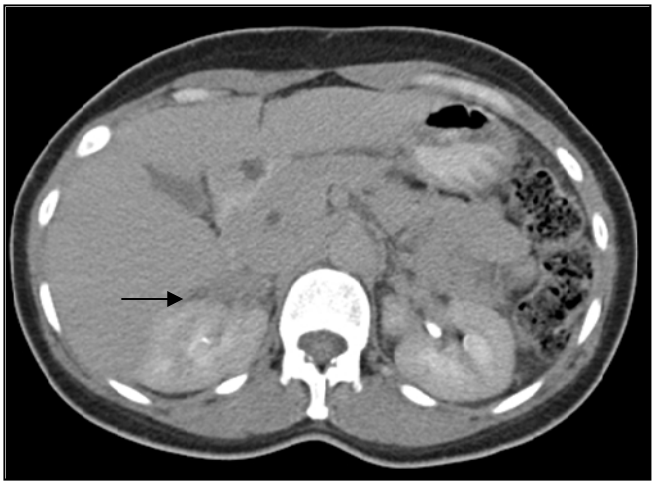
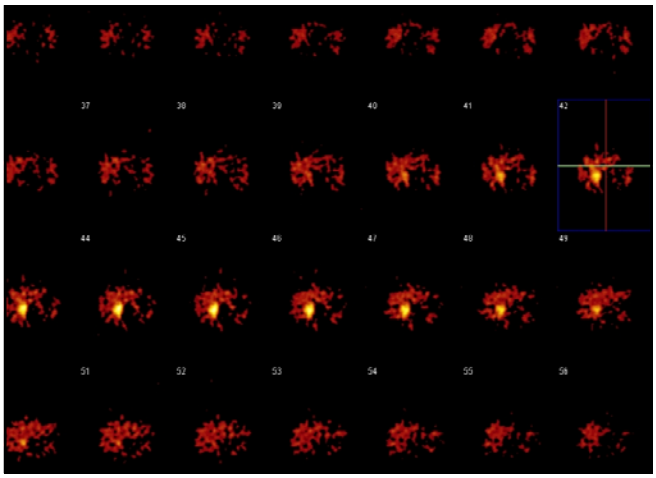


FIGURE D: MIBG SCAN SHOWING UPTAKE IN THE AFFECTED RIGHT ADRENAL GLAND



with plasma normetanephrine 16.2 n mol/L (n < 0.94), plasma metanephrines 1.49 n mol/L (< 0.37) . 24 Hour urine normetadrenaline 18900 n mol/L/24 Hrs (N-300-2200), urine metadrenaline 3891 n mol/L (N-200-1500) and orthmethyldopaine 2355 n mol/L (N-200-1400).The tumour was surgically removed, and her histopathology revealed pheochromocytoma. She was followed up in the OPD subsequently with no manifestation of her disease and her cardiac rhythm and ECG remained normal throughout the follow up period of one and a half year.

DISCUSSION

The serious and potentially lethal cardiovascular complications of pheochromocytoma are due to the potent effects of secreted catecholamines[1]. Many cardiac arrythmias have been described occurring with this catecholamine releasing tumour [3]. Catecholamine release is usually associated with tachycardia, although

up to 10% of the patients show bradycardia during the florid phases of their illness [4]. Some patients who have pheochromocytoma present with brady arrhythmia or asystolic arrest [5]. These situations are the result of a reflex mechanism in which sinus slowing occurs at the onset of a sudden rise in blood pressure during a paroxysm rarely, atrioventricular dissociation and bigeminy [3, 6] right bundles branch block and sick sinus syndrome occur in patients with pheochromocytoma. The diverse ECG abnormalities observed in patients with pheochromocytoma result from excessive adrenergic stimulation of the myocardium by the high plasma catecholamine levels.

Sinus arrest due to catecholamine release, most often with nodal escape rhythm has been published in several case reports [7, 8]. However occurence of AV block in pheochromocytoma is rare [5, 6]. There are very few case reports available in literature [7]. The mechanism of these brady arrhythmia postulated in previous studies [2, 3] is a rise in arterial pressure that stimulates the baroceptors and evokes reflex vagal discharge which slows the sinus node. They found out that when hypertension was most severe, nodal escape and atrioventricular dissociation was seen. The occurrence of sinus depression and AV dissociation with or without ectopic ventricular beats in association with the paroxysmal hypertension of pheochromocytoma has been documented previously mostly in adrenaline secreting pheochromocytoma, with only minimal noradrenaline secretion. [4, 5]

This case underlines intermittent AV dissociation with AV-nodal escape rhythm as a potential early and unusual presentation of pheochromocytoma. These changes disappeared after successful removal of the tumour.

The diagnosis of pheochromocytoma as a cause of different tachy or brady arrythmias is frequently delayed, resulting in more invasive procedures like pacemaker implantation in some cases and even ablation of conduction pathway but usually in such cases after resection of the tumor the arrhythmias resolved in almost all cases.[8,10]

CONCLUSION

Patients with pheochromocytoma can have diverse manifestations that must be recognized in the context of other clinical symptoms as soon as possible to allow early diagnosis and appropriate treatment.

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AUTHOR AFFILIATION:

Dr. Osama Tariq
Registrar Cardiology, Department of Medicine
Sultan Qaboos University Hospital, (SQUH), Oman.

Dr. Aftab Siddiqui *(Corresponding Author)*
Consultant, Department of Medicine, SQUH, Oman.
Email: aftabsidpk@hotmail.com

Dr. Adil Al Riami
Consultant Cardiologist, Dept. of Medicine, SQUH, Oman.

Dr. Fatima Amir
Acting Registrar, Department of Medicine, SQUH, Oman.

Dr. Yasser W. Mula-Abed
Registrar, Department of Medicine, SQUH, Oman.

Dr. Mohammed Hammed Gillani
Acting Consultant, Department of Medicine, SQUH, Oman.