Cardiac calcification in acute intermittent porphyria

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ABSTRACT

Aetiology of pericardial calcifications can be multifactorial. Tuberculosis has been reported as the most common cause. Other known causes include uraemia, asbestosis, post-traumatic or postoperative. We report a rare case of pericardial calcification seen in a patient with established acute intermittent porphyria. A direct causal relationship cannot be established between porphyria and pericardial calcification, but it may be due to deposition of the porphyrin in the pericardium.

Keywords: Calcification, pericardium, porphyria

INTRODUCTION

Pericardial calcification is usually associated with previous pericardial insult such as post-tuberculous or pyogenic pericarditis, uraemia, asbestosis, post-traumatic or postoperative cases.[1,2] Pericardial calcification may be associated with constrictive pericarditis[1,3] or may present as an isolated finding. However, acute intermittent porphyria (AIP) as a cause of pericardial calcification has not been reported.

CASE REPORT

A 17-year-old female patient weighing 35 Kg presented to emergency department with history of chronic intermittent abdominal pain, vomiting, and anorexia for three months, history of recurrent generalized tonic clonic seizures for 2 days and history of rapidly progressive ascending type of limb weakness with respiratory distress for one day. She had no history of snakebite, pain chest, poison intake, or blunt injury to neck or abdomen.

She initially presented to a private nursing home where she was intubated and transferred to our hospital on portable ventilator. On examination she was conscious (E_4 V_T M_1) with power of 1/5 in all limbs, plantars were mute, and deep tendon reflexes were absent. Her heart rate was 120/min and blood pressure was 100/60 mm Hg. She was breathing at a set rate given by ventilator. Systemic examination was insignificant. She was transferred to intensive care unit with a diagnosis of acute inflammatory demyelinating polyneuropathy. She also had features suggestive of autonomic neuropathy in the form of postural hypotension, tachycardia, and sweating.

Routine baseline workups including hemogram, blood sugars, liver and kidney function, chest X-ray, electrocardiography, electroencephalography were normal. Cerebrospinal fluid analysis was normal. Urine Watson–Schwartz test[4] was positive for porphobilinogen. Electrolyte screening showed mild hyponatremia (serum sodium 126 mmol/L). Serum phosphates and serum potassium were within normal limits. Ultrasonography of abdomen was unremarkable and serum lead levels were within normal limits. Nerve conduction study showed axonal motor neuropathy.

She was treated as a case of AIP with high dose of carbohydrate (300–400gm/day), heme arginate (3 mg/kg/day for 4 days) and gabapentin for seizures control. Precautions were taken not to prescribe any porphyrogenic drugs. At 60th day of her ICU stay ST segment elevation was noticed on the cardiac monitor and 12 lead ECG was ordered which showed [Figure 1] ST segment elevation in lead II. Cardiac enzyme markers were not elevated. Transthoracic echocardiography was done to rule out any cardiac pathology. It showed posterior pericardial calcification with anterior mitral leaflet and papillary muscle calcification [Figures 2 and 3] and relaxation abnormality around mitral valve suggestive of early pericardial constriction. Serum calcium (10.8 mg/dl) and phosphorus levels (5 mg/dl) were

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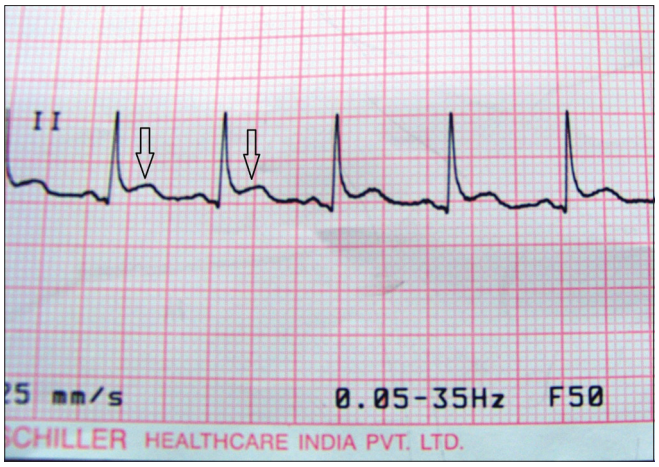

Figure 1: Electrocardiograph of the patient showing ST segment changes in lead II

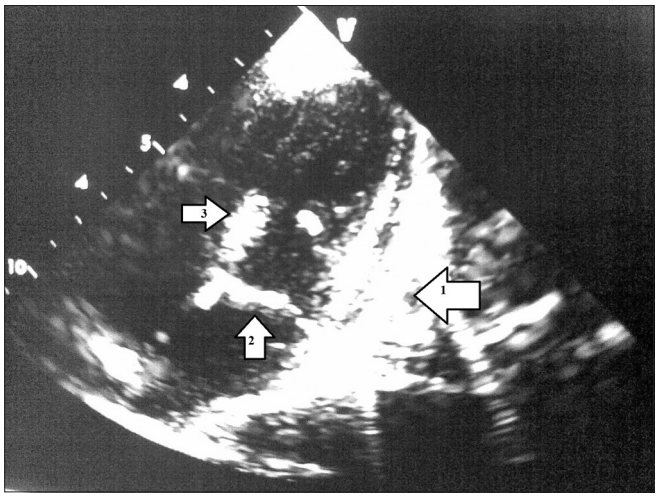

Figure 2: Apical view of heart showing pericardial (1), anterior mitral leaflet (2), papillary muscle, and (3) calcification

within normal limits. Tracheal tube aspirate for acid-fast bacilli was negative for 3 days in continuation. Patient required prolonged ventilator support and was finally discharged with the advise to follow up.

DISCUSSION

AIP is a rare autosomal dominant error of metabolism resulting from a disorder in the hepatic pathway of heme biosynthesis due to decrease of the enzyme uroporphyrinogen I synthetase.[5] Complications of AIP may be neurological such as transient cortical blindness,[6] seizures, quadriplegia, and psychiatric disorders, may be autonomic such as tachycardia, tremor, hypertension, constipation, may be hypothalamic, endocrine and metabolic including hyponatremia, dyslipidaemia, glucose intolerance and hyperinsulinism and disturbances of haemostasis and fibrinolysis.[7] Other long-term complications are chronic arterial hypertension, renal failure[8] and hepatocellular carcinoma.[9] To the best of

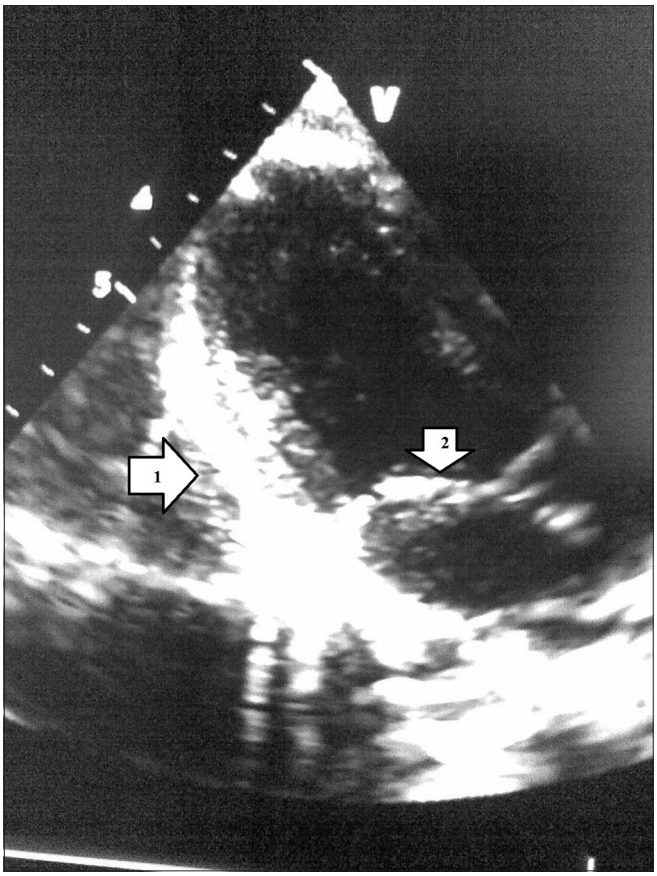

Figure 3: Modified parasternal view of heart showing pericardial (1) anterior mitral leaflet (2) calcification

our knowledge and literature search, we could not find any report of AIP with pericardial or structural cardiac abnormalities.

Calcifications of the pericardium may be present in the absence of constrictive pericarditis,[10] but, 27–50% of calcified pericardium is constricted.[1,3] Exact mechanism of the pericardial calcification is still not clear,[11] but is believed to be due to previous pericardial insult.[1] Pericardial calcification usually occurs over the posterior part, i.e., right atrium and anterior right ventricle, diaphragmatic surface, atrioventricular grooves because fluid preferentially gravitates toward the right side of the heart, where calcium and even bone are slowly deposited in the inspissated fluid.[12] In our case there was posterior pericardial calcification in echocardiography. Echocardiography is the simplest and the most widely used technique for imaging the pericardium.[13] Calcified pericardium will show a brighter echo than normal pericardium.

Our patient is in a very early stage of constrictive pericarditis. As the patient was asymptomatic we had not started any conventional antifailure regime.

This case report shows that cardiac calcification and constrictive pericarditis can occur as a complication

of AIP. At present we cannot comment on the exact mechanism of the pericardial calcification. Deposition of porphyrin precursors in pericardial tissue may be responsible for the changes that result in calcification. Vasospastic mechanisms leading to pericardial injury in such patients cannot be excluded.

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