CASE REPORT

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Primary Hypoparathyroidism: An Uncommon Presentation with Reversible Cardiomyopathy

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Abstract

Primary hypoparathyroidism is an endocrine disorder with numerous causes leading to a low level of Parathormone (PTH) and consequent hypocalcaemia, leading to metabolic disturbances in the body. The authors here describe a case of 53-years-old female who presented with complaints suggestive of hypoparathyroidism. On further evaluation, she was found to have left ventricular dysfunction with valvular abnormality. Her blood investigations showed low calcium, low iPTH and high phosphorus levels s/o Primary hypoparathyroidism. Reversible cardiomyopathy could very well be the first presentation of an endocrinological disorder. The present case highlights that correction of hypocalcaemia is one of the potentially reversible causes of left ventricular dysfunction hence, diagnosing and treating this condition early is very vital.

Key words: Reversible cardiomyopathy, hypoparathyroidism, hypocalcaemia.

Introduction

Parathyroid Hormone (PTH) is involved in the regulation of calcium and phosphorus levels in the body with its neuro-hormonal effects on kidney, bone and gastrointestinal tracts. The most common cause of hypoparathyroidism is post-surgical iatrogenic removal of parathyroid gland (in around 80% of the cases), while less than 20% caseshave autoimmune aetiology[1]. Due to any cause, if the PTH levels decrease, hypocalcaemia develops, leading to various clinical manifestations. The laboratory hallmark of hypoparathyroidism is hypocalcaemia and hyper-phosphataemia.

Hypoparathyroidism presenting with severe hypocalcaemia leading to cardiomyopathy with valvular dysfunction is potentially reversible when hypocalcaemia is corrected after early diagnosis. In sporadic cases, it is reported that long standing hypocalcaemia can lead to irreversible structural damage to the myocardium[2-4]. Various cardiac manifestations that can develop as a consequence of hypocalcaemia are arrythmias (long QT syndrome), and diminished myocardial performance (hypocalcaemic cardiomyopathy).

Case report

A 53-year-old middle-aged female, presented in our emergency department with the complaints of acute onset carpopedal spasm, multiple joint pains, bilateral hand tremors with numbness and paraesthesias of hands and feet. Her spouse gave a history of irritability of behaviour in recent times; however, she denied any other neurological symptoms, or history of fatigue, exertional dyspnoea, chest pain, palpitations, fever, neck pain, lumps, skin changes, hair fall, joint pains, oral ulcers or photosensitivity. She gave no past history of any neck surgery, trauma, or any other chronic disease, nor there was any positive family history suggesting any genetic causes or polyendocrinopathy. There were no symptoms during her childhood or adulthood, until the day she presented to our hospital.

On examination, her vitals were normal (BP - 140/90 mm hg, HR - 78/min, RR - 16/min, afebrile). On neurological examination, there were resting fine bilateral hand tremors, but there was no rigidity, Gait was normal. Both Chvostek's and Trousseau's signs were positive. There were no dysmorphic features, muco-cutaneous manifestations (including evidence of candidiasis/vitiligo) or dental malalignment. Cardiovascular examination revealed no cardiomegaly but had auscultatory evidence of mitral regurgitation (i.e., Pan-systolic murmur at mitral area). Abdomen and respiratory exam were unremarkable. Ocular examination revealed no cataracts or any other abnormal ocular findings.

Patient's blood biochemistry showed initial serum calcium level of 3.9 mg/dl (normal range - 8.4-10.2 mg/dl); phosphorus 7.7 mg/dl (normal range - 2.5 to 4.5 mg/dl); serum PTH level of < 4 pg/ml (normal range 9.2 - 44.60 pg/ml); serum albumin - 4.4 g/dl (normal range - 3.5 - 5 g/dl); serum magnesium 2 mg/dl (1.6 - 2.3 mg/dl); 24-hour urine

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calcium level was normal 200 mg/24 hrs (abnormal > 250 mg/24 hrs), and serum vit D level, liver function and kidney function tests were normal.

ECG showed prolonged corrected QT interval (554 ms) and 2D Echo revealed LV dysfunction with EF - 40% and global LV hypokinesis with severe mitral regurgitation (Fig. 1).

Ultrasonography of neck did not reveal any lump or other abnormality and USG abdomen was normal. CT head showed multiple focal symmetrical areas of calcification in and along the gyri in bilateral frontoparietal region, caudate nucleus, basal ganglion, bilateral medial temporal lobes and bilateral cerebral hemisphere, suggesting long standing hypoparathyroidism.

Serological tests (rheumatoid factor, anti-CCP antibodies, hepatitis B, C and HIV) were negative. Hormonal tests (thyroid profile, serum cortisol, serum ACTH) and serum iron profile were normal.

She was diagnosed with primary idiopathic hypoparathyroidism with extensive CNS calcifications and asymptomatic LV dysfunction (40%) with severe MR. She was started on intravenous calcium initially followed by oral calcium later on after resolution of acute tetanic spasms, and with vit D supplements, ACE-inhibitors and thiazide diuretics, to which she responded well. Patient was further followed-up in OPD after two months, echocardiography revealed increase in EF to 50%.

Discussion

The manifestations of resultant hypocalcaemia, hyperphosphataemia from hypoparathyroidism are both acute and chronic. Acute hypocalcaemia can present with tetany due to neuromuscular irritability, which may be mild like perioral numbness, paraesthesiac of hands and feet, muscle cramps, or severe in the form of carpopedal spasm, focal or generalised seizures, and laryngospasm. Some patients may present with non-specific symptoms of fatigue, irritability, anxiety and depression as well. Some features which are limited to chronic hypoparathyroidism are extrapyramidal disorders due to intracranial calcifications which are detected by CT scan when routine skull radiography fails to identify any calcifications[5]. In particular, the patient may present with movement disorders or parkinsonism due to basal ganglion calcifications[6]. Other manifestations are ocular (subcapsular cataracts, keratoconjunctivitis)[7], ectodermal manifestations (dry coarse skin, patchy alopecia, brittle nails, etc.), dental abnormalities (dental hypoplasia, failure of tooth eruptions, carious teeth, etc.), skeletal defects (osteosclerosis, cortical thickening, cranio-facial thickening) and occasionally cardiac dysfunction.

Any young or middle-age patient presenting with heart failure should be evaluated for reversible causes of cardiac dysfunction. Impact of hypoparathyroidism on cardiac function has been less commonly mentioned in the literature which is mainly due to resultant hypocalcaemia. The presentation of primary hypo-parathyroidism is variable but all patients should be screened for cardiomyopathy, whether they are symptomatic or asymptomatic. The main cause of cardiac abnormality is the hypocalcaemia, with its metabolic effects on cardiac muscle contraction and relaxation[8]. Cardiac contraction occurs when there is a influx of extracellular calcium, leading to generation of myocardial action potential. Hypocalcaemia leads to initiation of the action potential to a lower membrane electro-potential thus increasing excitability[9].

The various cardiac manifestations of hypocalcaemia are heart failure (may be asymptomatic in early stages) with or without reduction in ejection fraction; valvular dysfunction; and cardiac arrythmias like long QT syndrome[4]. Patients with hypocalcaemia can sometimes present with hypotension particularly if hypocalcaemia rapidly develops leading to depressed myocardial performance. Hypocalcaemia causes prolongation of QT interval as it lengthens phase 2 of the action potential leading to depressed myocardial functioning and dysrhythmias like *torsades de pointes*. There are few case reports mentioning association of hypocalcaemia due to hypoparathyroidism leading to significant cardiovascular manifestations. Saini N *et al* observed that hypocalcaemia is a cause of dilated cardiomyopathy when the patient presents with heart failure due to left ventricular systolic dysfunction in association with seizures and other neurological manifestations in hypoparathyroidism. Hypocalcaemic cardiomyopathy is reversible, with early intervention (as in the present case) and can drastically reduce morbidity and mortality[10]. Development of irreversible cardiac manifestations in long standing hypocalcaemia has been reported infrequently. Jariwala, in their case report, mentioned that hypocalcaemia along with hypomagnesaemia and low PTH levels can leads to dilated cardiomyopathy with its neuro-hormonal effects on cardiac contractility[11]. Although it is known, in children, that uncorrected hypocalcaemia can leads to cardiac dysfunction but in adults, primary idiopathic hypoparathyroidism as a cause of cardiomyopathy is reported uncommonly.

The present case highlights that, despite classical clinical manifestations of chronic hypoparathyroidism, LV dysfunction was overlooked. As in the present case, progress of LV dysfunction can be slowed with correction of hypocalcaemia. It should be stressed in the management of primary hypoparathyroidism that cardiovascular

manifestations like LV dysfunction or any valvular abnormality should be actively sought, despite the patient being asymptomatic, as it is reversible with timely treatment.

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