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HYPOCALCAEMIC CARDIOMYOPATHY AFTER PARATHYROIDECTOMY IN A PATIENT WITH TURNER SYNDROME: A CASE REPORT

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ABSTRACT Hypocalcaemic cardiomyopathy is a rare postoperative complication after hemi-parathyroidectomy which reverses completely on the restoration of normocalcaemia. Patients of Turner syndrome have subtle cardiovascular anomalies due to the underlying oestrogen deficiency. A 16-year-old female patient with Turner syndrome underwent hemi-parathyroidectomy for parathyroid adenoma. The patient developed signs of heart failure on postoperative day two, which was precipitated by tachycardia due to fever. She developed tachycardia, tachypnoea, hypertension, and pulmonary oedema with an ejection fraction of 20-30% at the time without any underlying cardiac disease. The patient was managed with intravenous diuretics, beta-blockers, and non-invasive ventilation. Other probable causes such as sepsis, pheochromocytoma, thyrotoxicosis, and Takotsubo syndrome were ruled out. A differential diagnosis of hypocalcaemic cardiomyopathy was suspected, and the patient was given intravenous calcium and vitamin-D supplementation. The patient responded promptly to restoration of normocalcaemia, and the cardiomyopathy reversed completely. Echocardiography done on follow-up showed normal ejection fraction.

KEYWORDS : Cardiomyopathy, hypocalcaemia, Turner syndrome, acute heart failure, postoperative.

INTRODUCTION

Hypocalcaemia can lead to severe impairment of left ventricular contractility, which manifests as dilated cardiomyopathy, and is defined as hypocalcaemic cardiomyopathy [1]. Heart failure resulting from this type of cardiomyopathy is refractory to conventional therapy, but responds completely to the restoration of normocalcaemia and normal Vitamin-D levels [2]. Oestrogen deficiency in Turner syndrome may contribute to arterial wall defects and intimal thickening [3].

Case Report

A 16-years old female, weighing 52 kilograms, with left-sided facial swelling, came to our institute for the surgery of a maxillary giant cell tumour. In the pre-anaesthetic check-up (PAC), her serum calcium level was 14.8 mg/dl. As a workup for hypercalcaemia, serum parathormone levels were sent which were high (246 pg/ml). Ultrasound neck was done which showed a left upper parathyroid mass. The patient was then scheduled for the excision of the parathyroid gland before the removal of the primary tumour. The patient gave a history of delayed menarche and features suggestive of delayed puberty. The patient was sent to the endocrinologist for the management of hypercalcaemia and workup for delayed menarche. Karyotyping revealed a single X chromosome consistent with the diagnosis of Turner syndrome. Other than a bicuspid aortic valve, the echocardiography showed no anomaly with normal ejection fraction. The patients had no history suggestive of any cardiovascular event with normal effort tolerance. Ultrasonography abdomen was normal. She received intravenous (IV) zoledronic acid 5mg three days before surgery. On the day of surgery, serum calcium levels were 10.1 mg/dl and excision of parathyroid adenoma was done under general anaesthesia. The peri-operative course was uneventful. At the end of the surgery, the patient was extubated and shifted to the intensive care unit (ICU). The patient remained haemodynamically stable and was shifted to the ward the coming morning. On postoperative day two, the patient developed fever with tachycardia followed by hypertension, tachypnoea, and sudden desaturation. The patient was immediately shifted to ICU. On auscultation, the patient had bilateral crepitations with pulse oximetry showing saturation of 70% on room air. Bedside transthoracic echocardiography displayed an ejection fraction of 20-30% with global hypokinesia (Figure1A-C). Treatment was initiated with IV furosemide (80mg/24 hrs) for pulmonary oedema, IV labetalol (10 mg stat followed by 5mg/hour infusion) for heart rate control and non-invasive ventilation (NIV) was initiated with the patient having high oxygen requirement (80-100%). Electrocardiography revealed sinus tachycardia and generalized T wave inversions with a long QTc interval of 0.54 seconds (Figure2A), and a negative Troponin-T card test. The AP view chest X-ray exhibited signs of pulmonary congestion with hilum enlargement (Figure2B). Serum metanephrine and normetanephrine levels were sent subsequently, which were normal, hence pheochromocytoma was ruled out. Thyroid function test was done to rule out thyrotoxicosis and was normal. Total leukocyte counts were normal with no obvious source of infection, hence ruling out sepsis. The patient required continuous NIV despite all the measures. Serum calcium at that time was 6.8 mg/dl. Differential diagnosis of hypocalcaemic cardiomyopathy was suspected, and the patient was given 10 ml of IV calcium gluconate 10% (90 mg of elemental calcium) every 6 hours and an oral vitamin-D sachet 60,000 IU/week. The patient improved within 24 hours and was kept on intermittent NIV. Daily serial echocardiography showed gradual improvement in ejection fraction and contractility, which normalized by day seven. Subsequently, beta-blockers, diuretics, and ACE inhibitors was stopped after features of acute heart failure had resolved. In the ward, the patient maintained well on room air with normal hemodynamics. She was discharged home on oral tablet calcium (1.5 gram of calcium carbonate) and vitamin-D (125 IU of Vitamin D3 per tablet) given three times a day. The patient was sent for evaluation to a cardiology centre, but no abnormality was detected on echocardiography. The patient was followed up postoperatively for one month and her serum calcium and vitamin-D levels stayed normal. She was asymptomatic and returned to normal activity after discharge from the hospital.

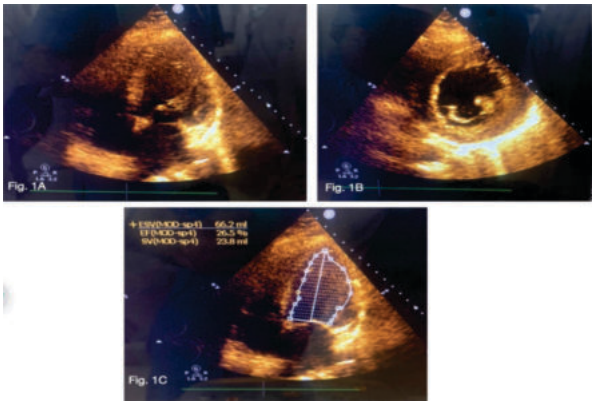
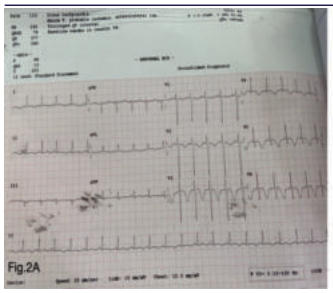


Figure 1: Cardiac ultrasound results. (A) : Apical 4-chamber view showing dilated cardiac chambers. (B): Parasternal short axis view showing poor contractility with generalised hypokinesia. C): Ejection fraction calculation using Simpson's method.



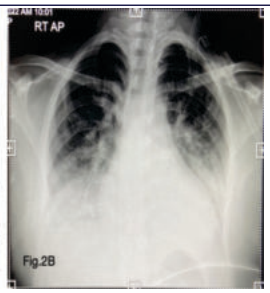


Figure 2A: Electrocardiography showing sinus tachycardia, generalised T-wave inversion and prolonged QTc interval(0.54 seconds).

Figure 2B: Chest X-ray showing pulmonary congestion with hilar enlargement.

DISCUSSION:

Hypocalcaemic cardiomyopathy is a rare complication in a patient after hemi-parathyroidectomy. Parathormone has a vital role in the maintenance of calcium homeostasis and can affect cardiovascular functions. Bashour et al. in 1980 described two patients with hypocalcaemia secondary to hypoparathyroidism showing features of congestive heart failure refractory to standard heart failure treatment but responded completely to calcium therapy [4]. They labelled this condition as "hypocalcaemic cardiomyopathy." Calcium plays an important role in cardiac contractility as it acts as a messenger via changes in intracellular calcium levels [5,6]. Various experimental results indicated that hypocalcaemia reduces myocardial contractility and presents with reduced left ventricular function [7,8], and can result in prolongation of the corrected QT interval, potentially leading to life-threatening arrhythmias [8].

Our patient was incidentally diagnosed to have primary hyperparathyroidism with Turner syndrome and underwent hemi-parathyroidectomy. She developed hypocalcaemia on postoperative day two leading to cardiomyopathy which was decompensated by the episode of tachycardia due to fever. Multiple electrocardiograms and cardiac markers did not show any evidence of acute coronary syndrome or Takotsubo syndrome [9]. The patient responded completely to calcium supplementation. Therefore, we believe that our patient had reversible cardiomyopathy due to hypocalcaemia.

Cardiomyopathy is a rare manifestation of hypocalcaemia. The role of calcium in the cardiovascular system is well known, though the mechanisms underlying cardiomyopathy induced by hypocalcaemia remain unclear. However, cardiomyopathy may be related to genetic susceptibility or other factors that have not yet been determined. Turner syndrome may present as vasculopathy such as aortic root dilatation with higher aortic size index (ASI), coarctation of the aorta, and intimal thickening due to oestrogen deficiency, which was not present in our patient in the initial evaluation [3]. Subtle cardiac and vascular anomalies with increase in ASI may herald the development of life-threatening complications.

We could not find any case report or literature regarding a patient with Turner syndrome presenting with hypocalcaemia induced cardiac failure. Therefore, our case report may serve as a unique entry point for additional research.

We want to emphasize through this case that in a postoperative patient with hemi-parathyroidectomy, we must monitor serum calcium levels routinely. Hypocalcaemia can lead to cardiomyopathy, which may present as acute cardiac failure. Patients with Turner syndrome have subtle cardiovascular anomalies, hence the pre-operative cardiac work-up should be done diligently. Prompt diagnosis and rapid management may help to decrease perioperative morbidity as this condition is completely reversible.

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