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Case report

Perioperative management of a case of Paget's disease with dilated cardiomyopathy: A report

Parnandi Bhaskar Rao [a], **Neha Singh** [a,*], **Sidhartha Mohanty** [b], **Chandra Sekar Pradhan** [b]

[a] *Department of Anaesthesiology, Critical Care and Pain Medicine, AIIMS, Bhubaneswar 751019, Odisha, India*
[b] *Department of Anaesthesiology, Critical Care and Pain Medicine, IMS and SUM Hospital, SOA University, Bhubaneswar 751030, Odisha, India*

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Abstract Paget's disease (PD) of bone is a nonmalignant disease of unknown etiology involving accelerated bone resorption followed by deposition of dense, chaotic, and ineffectively mineralized bone matrix. It is frequently asymptomatic and the patient may present with symptoms depending on the bones involved. We are reporting perioperative management of a case of Paget's disease with dilated cardiomyopathy for emergency orthopaedic surgery.

1. Introduction

Paget's disease is also known as osteitis deformans and it is more prevalent in males than in females [1]. It is rarely diagnosed in people less than 40 years of age [2]. Any bone can be affected but PD occurs most frequently in the spine, skull, pelvis, femur and tibia. The symptoms progress slowly and the disease does not spread to normal bones. Treatment can control Paget's disease and lessen symptoms but there is no cure. PD may lead to high-output congestive failure. Here we are reporting perioperative management of a case of Paget's disease with diabetes with hypertension with dilated cardiomyopathy. Institutional ethical committee clearance and consent from the patient were obtained before reporting the case.

2. Case report

A 74 year old, 148-cm, 48 kg, ASA four lady was presented in the casualty with history of slip and fall one week back. She was diagnosed with left subtrochanteric fracture (Fig. 1). She was a known case of Paget's disease with diabetes mellitus with hypertension for last six years and was on irregular medication which includes Tab digoxin ø OD, Tab Lasix 20 mg BD and Tab amlong 5 mg BD.

General examination revealed pallor and blood pressure was 170/90 mmHg. On auscultation, an ejection systolic murmur was heard in aortic area. Chest X-ray was showing prominent bronchovascular markings and cardiomegaly (Fig. 2). Electrocardiography (ECG) showed sinus tachycardia with ST segment depression and T inversion in lead two, three,

* Corresponding author at: Department of Anaesthesiology, Critical Care and Pain Medicine, AIIMS, Bhubaneswar 751019, Odisha, India.
E-mail address: drneha.com@gmail.com (N. Singh).
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Fig. 1 Left subtrochanteric fracture.

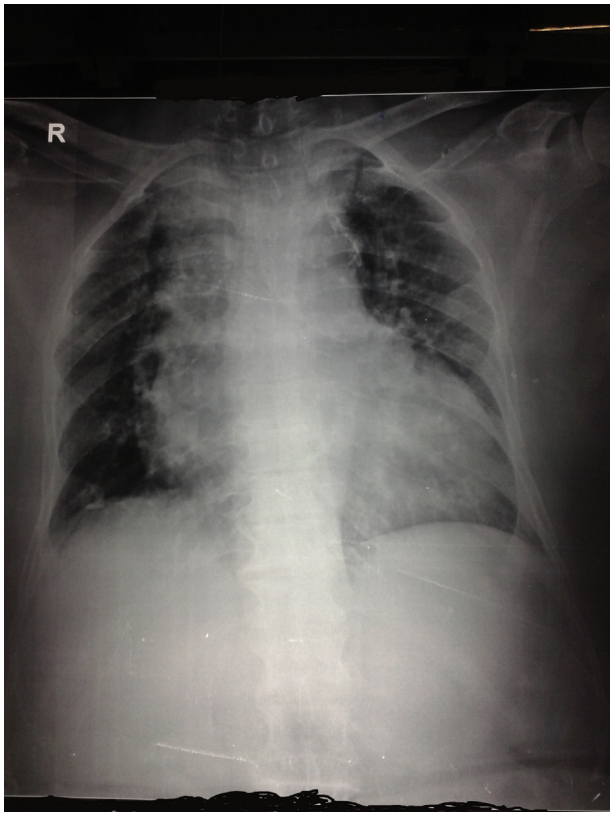

Fig. 2 Chest X-ray showing cardiomegaly.

aV_f, aV_l, V_4, V_5, and V_6 along with left ventricular hypertrophy and left bundle branch block. Two-dimensional echocardiography (2D Echo) revealed severe LV dysfunction with ejection fraction of 32% along with global hypokinesia, predominantly involving anterior wall. Mild AS with a gradient of 15/4, minimal MR and mitral annulus calcification was also present. Her hemoglobin was 9.2 gm/dl, serum urea −71 mg/dl and creatinine – 3.2 mg/dl. Coagulation profile, blood sugar and electrolytes were normal. Mouth opening was adequate but neck extension was restricted with modified mallampati class 3. Patient refused regional anesthesia in view of severe backache so we have decided to go with general anesthesia. She was explained about the risks and informed written consent was taken.

She was premedicated with inj. metoclopramide (10 mg) and inj. ranitidine (50 mg). Pre-induction monitoring included electrocardiogram (ECG) lead II and V, invasive blood pressure (IBP), Heart rate (HR) and Peripheral oxygen saturation (SpO_2). Venous access was established using 18G cannula on the dorsum of non-dominant hand. On table, she received intravenous midazolam (1.5 mg) and fentanyl (200 μg). After checking for adequate ventilation inj. vecuronium bromide (0.1 mg/kg) was given and trachea was intubated in second attempt with 7.0 size tube using True view video laryngoscope. Tube was fixed after checking bilateral air entry and right internal jugular vein was cannulated with 7.5 Fr triple lumen catheter for central venous pressure (CVP) monitoring which varied between 10 and 12 cm of water. She was catheterized and surgery was started. Anesthesia was maintained with Air:O_2: 50:50, sevoflurane 1–2%, vecuronium infusion 1 mcg/kg/min infusion and fentanyl infusion 2 mcg/kg/hr. The intraoperative period was uneventful except for occasional ventricular ectopics for which inj. lignocaine 1 mg/kg was given. She received 200 ml of colloid and 2 units of packed cell. Urine output was 120 ml and blood loss was approximately 500 ml. The duration of surgery was four hrs and neuromuscular blockade was reversed with inj. neostigmine 0.05 mg/kg and inj. glycopyrrolate 0.2 mg/1 mg of neostigmine. The patient was extubated on table and shifted to ICU for monitoring. She was discharged from hospital on sixth postoperative day.

3. Discussion

Paget's disease is a disorder of bone turnover and remodeling of unknown etiology but probably caused by infection with a common and widespread virus superimposed on genetic variation. The excessive breakdown and formation of bone tissue lead to weak bones resulting in pain, bone deformity, fractures, and arthritis in the joints near the affected bones.

Unlike osteoporosis that usually affects all the bones in the body, Paget's disease typically is localized, affecting just one or a few bones. It commonly involves the axial skeleton, but it can affect any area mostly at least two bones [3]. Although there is no cure for Paget's disease, medications such as bisphosphonates and calcitonin can help control the disorder and lessen pain and other symptoms.

Our patient was having PD with dilated cardiomyopathy (DCM) with hypertension and diabetes mellitus. PD itself can lead to aortic stenosis and left ventricular hypertrophy. Goals of perioperative management include prevention of myocardial depression, maintain normovolemia, avoid ventricular after load and sudden hemodynamic changes [4].

We have chosen General anesthesia over regional block in view of patient refusal due to backache and the presence of PD. PD patients may have a characteristic, 'picture frame' appearance, or collapse of vertebral bodies, ossifications and positioning problems. Smaller gauge needles may not bypass ossifications [5]. Anesthetic management needs to be customized for patients according to their hemodynamic status. Drugs such as etomidate and narcotics have minimal depressing effect on cardiac function and are used frequently [6]. Oxygen carrying capacity should be adequate. The main

determinants of oxygen carrying capacity are cardiac output and hemoglobin. Therefore, hemoglobin should be maintained at higher level [7]. Skeletal muscle paralysis is to be provided by nondepolarizing muscle relaxant that lacks significant cardiovascular effects. As it was an anticipated difficult airway, true-view video laryngoscope was used and trachea was intubated in second attempt. Transillumination guided nasal intubation was done in a patient with PD with anticipated difficult airway [8]. Regional anesthesia techniques such as subarachnoid block or epidural may be an alternative to general anesthesia in selected patients with DCM [9].

In conclusion, our experience with this patient suggests that successful outcome in these patients depends on thorough preoperative assessment, optimization of cardiac status, perioperative plan including prompt diagnosis and management of complications along with proper post operative monitoring. However, intensive invasive monitoring can be individualized according to patients.

Conflict of interest

We have no conflict of interest to declare.

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