A Case of Gorhams Syndrome: An anesthetic challenge

Gorham's syndrome or vanishing bone disease is a rare disorder with massive osteolysis and idiopathic fractures, which may result in neurological and respiratory complications. For anesthetist, Gorham's syndrome presents various challenges such as long duration, blood loss, air embolism, positioning, and respiratory complications. We present a case of an 18-year-old male, who was diagnosed with the disease 2 years back, following a fall. He developed numbness and tingling sensation over chest wall with motor weakness in both lower limbs,with a gradual change in shape of the chest with increasing scoliosis of spine over the next few months. He was operated for decompression of dorsal vertebrae and grafted with mother's fibula. Postoperatively, excisional specimen was sent to histopathology confirming the diagnosis of Gorham's syndrome.

The patient suffered a fall again and came with power grade 4/5 in lower limbs, which progressed to 1/5 within 1 week of admission. Airway examination revealed a normal mouth opening and neck movements. On palpation, there was a retraction of right upper hemithorax and the upper ribs could not be palpated, and scoliosis toward right side was noted. Chest X-ray revealed right 4-8th rib's destruction on anterior, posterior aspect [Figures 1]. Magnetic resonance imaging spine: Severe scoliosis of the dorsal spine with convexity to the right. D2-8 vertebra appears osteoporotic with bony erosions. Pulmonary function tests show restrictive lung disease with total lung capacity 38% of normal forced vital capacity, and forced expiratory volume in 1 s 38% and 41% of normal. Arterial blood gas showed normal values.

Inside theatre, anesthesia was induced with midazolam 1.5 mg, fentanyl 150 mcg and propofol 120 mg, and rocuronium 50 mg. He was intubated with 8.5 mm endotracheal tube with manual inline stabilization. The right internal jugular vein was cannulated. The patient was given lateral position for the surgery. Maintenance was with O_2:N_2O 50:50 as per institutional protocol, isoflurane 1 minimum alveolar concentration, dexmedetomidine infusion at 0.7 mic/kg/h and buprenorphine 120 mcg. Volume control mode was used with a tidal volume of 450 ml, respiratory rate of 16/min, and PEEP of 5 cm. Maximum peak airway pressures intraoperatively were 17 cm of H_2O. There was no intraoperative episode of hypotension, desaturation, or fall in end-tidal CO_2.

Intraoperative blood loss was 1000 ml, which was replaced with five pints of ringer lactate and 500 ml of colloid. No blood products were used as preoperative, hemoglobin was 15.4 and maximum allowable blood loss was calculated to be 2343 ml. Duration of surgery was 4 h. The patient was extubated uneventfully.

Gorham's syndrome is a disease of unknown etiology, most commonly in the second and third decades with equal sex distribution. The pathologic process shows replacement of normal bone by an aggressively expanding but non neoplastic vascular tissue similar to hemangioma or

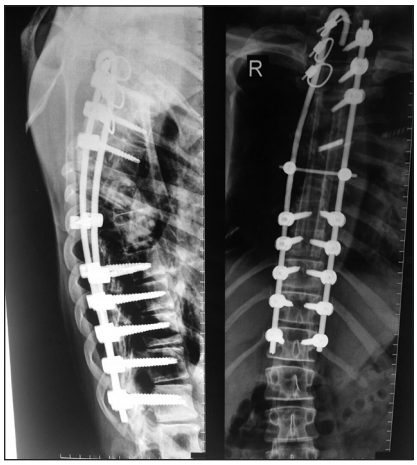

Figure 1: The chest X-ray posteroanterior view and lateral view shows right 4, 5, 6, 7, 8th rib's destruction on anterior and posterior aspect with blunting of costophrenic angle on the right side. Thoracolumbar scoliosis with convexity toward right. Lung parenchyma appears normal. Graft used in previous surgery along with instrumentation done is visible in the images

lymphangioma.[1] Patients present with pain in the involved region with pathological fractures. Diagnosis is by clinical examination, X-ray and histopathological findings. The treatment includes radiation therapy, bisphosphonates, and alpha-2b interferon.[2] Definitive treatment is by surgery.

For anesthetist, Gorham's syndrome presents various challenges. Complications specific to surgery such as long duration, blood loss, air embolism, and positioning and those related to the disease such as respiratory complications, chylothorax, and effusions. If there is restrictive lung disease mainly due to the disease process and accompanying scoliosis, ventilation using low tidal volume and high respiratory rate, pressure-controlled ventilation seems more appropriate. Postoperative ventilatory problem leading to re-intubation and prolonged ventilation may be seen. Extubation has to be planned carefully with preparation for prolonged Intensive Care Unit management.[3]

Thus to conclude, Gorham's disease is a challenge for both the surgeon and anesthetist, and a team effort is needed to manage these cases successfully.

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Supriya Lynette Dsouza, Adarsh Kulkarni, Hashim Sageer, Naveen Pajai, Nirav Kotak

Department of Anaesthesiology, Seth G.S Medical College and King Edward Memorial Hospital, Mumbai, Maharashtra, India

Address for correspondence: Dr. Supriya Lynette Dsouza, Department of Anaesthesiology, Seth G.S Medical College and King Edward Memorial Hospital, Mumbai, Maharashtra, India.
E-mail: supriyalynettedsouza@yahoo.com

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