A curious case of the vanishing bone disease: Anesthesia management

Vanishing bone disease, also known as Gorham-Stout (GS) syndrome, wiggly bone syndrome or phantom bone disease is a rare idiopathic musculoskeletal disorder, of yet unknown etiology, characterized by the clinical and radiological disappearance of the bone caused by proliferation of non-neoplastic vascular and lymphatic tissue. It was initially described by Jackson in a case of massive osteolysis of the humerus in a 12-year-old boy in the year 1838 and was later classified by Gorham and Stout.[1] The syndrome affects one or multiple bones including the spine, upper and lower extremities, skull and pelvis.

We recently managed a 25-year-old male patient, a known case of GS syndrome, with history of C7-T1 fixation who presented with implant failure due to cervical vertebrae resorption. The patient had complaints of bilateral lower limb weakness with swelling of the left lower limb. Ultrasonography of the left lower limb revealed acute–subacute complete lumen occluding thrombus in left external and common iliac vein brought upon by prolonged immobilization. Preoperatively, inferior vena cava filter was inserted. CT chest showed left-sided chylothorax which was drained 12 hours prior to the surgery. Manual in-line stabilization (MILS) was done and patient was intubated using C-MAC D blade. Positioning of the patient intraoperatively was done with due precautions. Intraoperative course was uneventful.

The anesthetic considerations were difficult airway, cervical instability requiring MILS, careful positioning, pulmonary complications due to chylothorax, risk of thromboembolism due to pre-existing deep vein thrombosis, the prolonged surgical duration, risk of hypothermia and excessive blood loss. Bone loss in GS syndrome may cause joint instability and problems during airway management and positioning for surgery. The affection of the spine leads to neurological decline, which is the main cause of patient immobilization and the subsequent comorbidities.

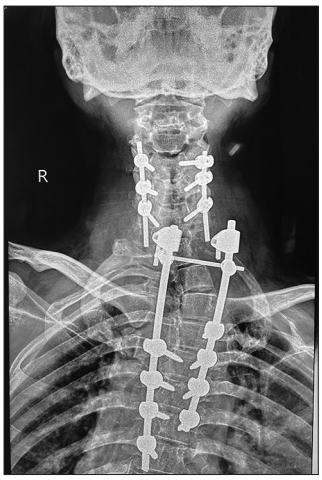

Figure 1: Pre operative X ray showing anterolisthesis of C7 over T1

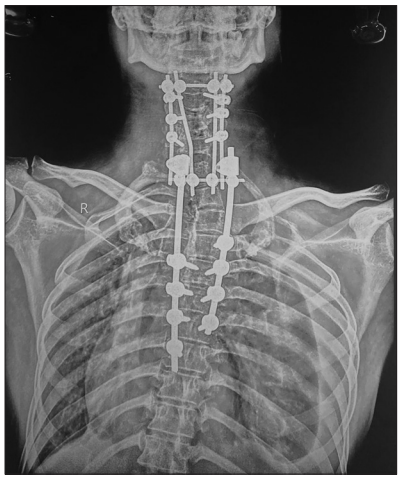

Figure 2: Post operative X ray

Among patients with primary osteolysis like GS syndrome, difficult intubation was encountered in 64% and postoperative ventilation was required in 27%. Therefore, preoperative assessment of a patient with primary osteolysis should include an appropriate strategy for managing a potentially difficult airway.[2] Awake fiber optic laryngoscopy or video laryngoscope such as C MAC should be used to avoid airway disasters.

Chylothorax may occur due to the extension of the lymphangiectasia of affected thoracic skeleton into pleural cavity or by the invasion of thoracic duct.[3] Pleural effusion and chylothorax (reported to be a complication of GS syndrome in up to 17% of cases) can dramatically influence respiratory function.[4] Complete respiratory assessment should be done preoperatively including chest X-ray, HRCT, arterial blood gases, spirometry and oxygen saturation.

Drainage of effusions results in increased oxygen delivery and oxygen consumption coinciding with a decrease in pulmonary capillary wedge pressure. The pulmonary arteriovenous shunt decreased, implying an increase in functional residual capacity and improved oxygenation.[5] Thus, we preferred to perform thoracocentesis and drain an adequate amount of chylothorax. With possibility of hypoproteinemia due to excessive formation of chylothorax, highly protein-bound drugs should be cautiously used. Even though difficult airway is anticipated in patients of GS syndrome, succinylcholine should be avoided as uncontrolled fasciculations may cause fracture of osteoporotic bones. Neuromuscular monitoring helped in judicious use of muscle relaxant.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Dhwani S. Walavalkar, Deepa Kane, Priti Devalkar, Ramesh Chandan, Nimisha Thakur

Department of Anaesthesiology and Critical Care, Seth GSMC and KEM Hospital, Mumbai, Maharashtra, India

Address for correspondence: Dr. Dhwani S. Walavalkar, B 605, Manish Mahal, Off Veera Desai Road, Andheri West, Mumbai - 400 053, Maharashtra, India.
E-mail: dhwani.walavalkar@yahoo.com

References

1. Jackson J. A boneless arm. Boston Med Surg J 1838;18:398-9.
2. Herd RS, Sprung J, Weingarten TN. Primary osteolysis syndromes: Beware of difficult airway. Pediatric Anesthesia, 25, 727-37
3. Patel DV. Gorham's disease or massive osteolysis. Clin Med Res 2005;3:65-74.
4. Tie ML, Poland GA, Rosenow EC 3rd. Chylothorax in Gorham's syndrome. A common complication of a rare disease. Chest 1994;105:208-13.
5. Ahmed SH, Ouzounian SP, Dirusso S, Sullivan T, Savino J, Del Guercio L. Hemodynamic and pulmonary changes after drainage of significant pleural effusions in critically ill, mechanically ventilated surgical patients. J Trauma 2004;57:1184-8.

Access this article online	
Quick Response Code:	Website: https://journals.lww.com/joacp
	DOI: 10.4103/joacp.joacp_195_22

How to cite this article: Walavalkar DS, Kane D, Devalkar P, Chandan R, Thakur N. A curious case of the vanishing bone disease: Anesthesia management. J Anaesthesiol Clin Pharmacol 2024;40:171-2.

Submitted: 25-May-2022
Accepted: 26-Jun-2022 **Published:** 08-Feb-2024