Case Report

Anaesthesia for a Patient with Gorham's Disease A Very Rare Disease to Report

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Abstract

Gorham disease, which is also known as massive osteolysis, vanishing bone disease or lymphangiomatosis, is an extremely rare condition, characterized by proliferation of vascular channels resulting in destruction and resorption of osseous matrix leading to bone loss, which in turn leads to joint instability and problems during airway management and positioning for surgery. Respiratory involvement may further complicate anaesthesia management. We are reporting the anaesthesia care of a 11-year-old boy of known GS with left sided chylothorax who came for amputation of left arm due to closed injury arm resulting from RTA.He was on cyclical Pamidronate therapy and had completed radiotherapy.

Keywords: *Gorham's disease, Vanishing bone disease, Osteolysis, Airway management, Positioning.*

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Introduction

Gorham disease is an extremely rare condition with fewer than 200 cases reported in the medical literature. First noted in 1838, the disease was not described in detail until 1954 by Gorham and Stout. [(1)]It is described as gradual and often complete spontaneous resorption of bone tissue.Characterized by proliferation of vascular channels resulting in destruction and resorption of osseous matrix leading to bone loss, which in turn leads to joint instability and problems during airway management and positioning for surgery. The exact etiology of the disease is unknown and it usually is not recognized until a fracture occurs, with subsequent improper bone healing. The condition is non familial, affect people of all ages and is slightly more prevalent in males than females. [(2)] The osteolytic process can affect any bone although shoulder and pelvis are commonly affected. Spine involvement can lead to acute neurological deficit requiring emergency surgery. These patients require great care during positioning including cervical spine protection. Here, we report the uneventful perioperative care of a 11-year-old boy with GS for amputation of left arm.

Case report

A 11-year-old boy of 38 kg presented with the known case of GS of left arm with left sided pleural effusion who came for amputation of left arm due to closed injury arm resulting from RTA. He was on cyclical Pamidronate therapy and had completed radiotherapy. He had a history of fall on 2012 and got sustained closed injury to left arm. At first treated locally

He was diagnosed as GS by biopsy of right clavicle. Physical examination, baseline investigations and vital signs were within normal limits. Chest X-ray showed ICD (left) in situ and almost complete destruction of left humerus and scapula.X-ray left arm also reveals the same.

Just before shifting to the operating room, his pulse rate was 82 beats / min, blood pressure was 110/50 mmhg and room air saturation was 98%.

The patient was shifted to the OR and standard monitors were attached. Patient were premedicated with midazolam 2mg and glycopyrrolate .2mg intravenously followed by 100% inspired oxygen administration for 5 min. Anaesthesia was induced with fentanyl 40 microgram, ketamine 10 mg and propofol 50mg.After the loss of consciousness, manual in

line stabilization (MILS) of the neck was provided by a second anaesthesiologist and after confirming the adequate mask ventilation, rocuronium 30mg was given intravenously following which trachea was intubated with 6 mm ID cuffed endotracheal tube using fibreoptic bronchoscope, taking a great care of neck extension and mandible. Anaesthesia was maintained with 50 % oxygen in nitrous and end tidal sevoflurane 1-1.5 % using circle system. Intraoperative monitoring included ECG, Noninvasive BP, capnometry, pulse oximetry, temperature, urine output. The surgery lasted for 1.5 hour with around 200 ml of blood loss. The patient received 1000 ml of lactated ringer solution and blood loss was not replaced as it was within allowable blood loss (ABL). Vitals were stable throughout the surgery. At the end of the surgery, the patient was reversed with neostigmine 2mg and atropine .6mg intravenously and the trachea was extubated. Then the patient was shifted to the post anaesthesia care unit (PACU) with oxygen 5L/min using face mask. In the PACU, patient's vitals were normal. Post operatively pain was managed with IV paracetamol 1gm and then 6 hourly, IV Ketorolac 30mg and titrated dose of IM Pethidine 40 mg aliquots to maintain VAS (0 means no pain, 10 worst pain)of < 3. The patient was observed in the post-operative care unit for 2 hours and then shifted to his room without any complications.

Discussion

GS or disappearing bone disease is a mysterious bone disease of unknown etiology which affects patients from 1.5 t0 72 years, although it occurs most commonly in the 2nd and 3rd decades[3] and is slightly more prevalent in males than females. There is no evidence of any endocrine, metabolic, neoplastic, infectious or neurologic disturbances. The pathologic process is the replacement of normal bone by an aggressively expanding but non neoplastic vascular tissue, similar to haemangioma or lymphangioma.[4] The cavernosum type of lymphangioma affects organs in the thorax, abdomen and bones and may affect just one bone or extend into adjacent bones, viscera or soft tissues, resulting in local fibrosis.[5,6,7] Bone loss occurs and progresses in certain areas of the body with no new bone growth. Affected areas may include the hands, arms, shoulders, ribs, spine, pelvis, femur and jaw. In addition, nonchylous pleural effusion or clylothorax which is the presence of lymphatic fluid in the pleural space secondary to leakage from the thoracic duct or one of its main tributaries, may occur in association with this disease.[5]

Clinical manifestations usually are related to the area of involvement. When the lower or upper jaw, tooth sockets or other bones in the face, neck, or head are affected, symptoms may include pain, loose teeth, fractures, facial deformity and recurrent meningitis. Peripheral bone involvement in the young may result in shortening or deformity of the affected bone. Rib or thoracic vertebral bone involvement may manifest as chronic or acute pain in cases of pathological fracture. Spinal involvement may produce vertebral collapse, which could result in neurological disability or lead to chest deformity and respiratory compromise.Non chylous pleural effusions occur as the result of lymphangiomatosis and osteolysis of the thoracic cage. Chylothorax is associated with invasion of the thoracic duct for which the mechanism has not been determined.[5-7]

Diagnosis is based on clinical manifestations and radiological findings, which is confirmed by biopsy of the bone.[8] Various treatment modalities include radiation therapy,[9,10] anti-osteoclastic medication and alpha-2b interferon[11]. Surgical treatment options include resection of the lesion and instrumentation using bone grafts and prostheses.

The formulation of an anesthetic plan for a patient with Gorham Disease, should begin by considering the potential for increased anaesthetic risk caused by co morbid conditions and the surgical procedure to be performed. A weakened cervical spine prone to subluxation could increase the risk of spinal cord injury, particularly during hyperextension for intubation. Preoperative cervical spine x-ray would be needed to detect this problem. Manual in line stabilization of the cervical spine during airway maneuvers could be used prophylactically in patients without radiographic abnormalities, however patients who have abnormal findings of the head, neck, or face require awake intubation via fibreoptic bronchoscopy.[2]

Respiratory system involvement in the form of chylothorax in GS patients is not an infrequent complication and hence, needs special mention.

Patients might present for surgery with this complication or they might develop it post operatively.[3,12,13] Diseases of ribs, scapula, or thoracic vertebra may lead to the development of chylothorax from direct extension of lymphangiectasia into the pleural cavity or via invasion of thoracic duct. These patients should undergo complete respiratory assessment (arterial blood gas analysis, spirometry). If there is restrictive lung disease, ventilation should be undertaken by using low tidal volume and high respiratory rate. In these patients, pressure controlled ventilation seems more appropriate. [(12,13)] Ourpatient did not have any respiratory involvement and complication as well.

GS generally does not affect hepatic, renal, metabolic or muscular function. Therefore, the pharmacokinetics and pharmacodynamics of commonly used anaesthetics remain unaffected. However, chylothoarx induced hypoproteinemia dictates cautious use of high protein bound drugs. [(13)]succinylcholine may produce unpredictable fasciculation and the possibility of fracture in osteoporotic bones is a concern, hence better tobe avoided.

Fortunately, our patient had an uneventful recovery without any positional or neurological sequelae.

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