Case Report

Anaesthetic consideration in patients with Gorham's syndrome: A case report and review of the literature

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ABSTRACT

Gorham's syndrome (GS) is a rare disorder characterized by proliferation of vascular channels resulting in destruction and resorption of osseous matrix leading to bone loss. Bone loss leads to joint instability and problems during airway management and positioning for surgery. Respiratory involvement further complicates anaesthesia management. We report the anaesthetic care of a 21-year-old male patient of known GS for spine decompression and fusion in prone position. Airway management, induction technique, pathophysiology of the disease, drug selection and other concerns of anaesthesia for major spine surgery has been discussed reviewing the sparse literature available.

Key words: Airway management, anaesthesia, Gorham's syndrome, spine surgery

INTRODUCTION

Gorham's syndrome (GS) is characterized by proliferation of vascular channels that results in destruction and resorption of osseous matrix. Since its initial description by Jackson as a case of massive osteolysis of the humerus in a 12-year-old boy in 1838[1] and then by Gorham and Stout in 1955,[2-4] the precise aetiology of the disease is largely unknown. The osteolytic process can affect any bone, although shoulder and pelvis are commonly affected.[5] 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 21-year-old patient with GS, for decompression and fusion of upper thoracic spine.

CASE REPORT

A 21-year-old, 78 kg male patient presented with pain in the upper back. Since the last 1 month, he was having weakness of both lower limbs leading to inability to walk and sit for a long duration. Past history revealed disappearance of right clavicle between 3 and 12 years of age and, subsequently, he was diagnosed as GS by biopsy of right clavicle. Physical examination and vital signs were within normal limits. Neurological examination revealed weakness of both lower limbs. Chest X-ray showed thoracic (D1-2) destruction and kyphosis. Cervical spine X-ray and biochemical investigations were within normal limits. Magnetic resonance imaging of the spine revealed increasing kyphosis at the upper thoracic level centering at D1-2, and cord compression mainly at the D1-2 level. Therefore, he was planned for decompression and fusion of C3 – D5 spine.

Just before shifting to the operating room (OR), his pulse rate (PR) was 78 beats/min, blood pressure (BP) was 126/76 mmHg and room air saturation was 96%. He was premedicated with butorphanol 1 mg, promethazine 12.5 mg and glycopyrrolate 0.2 mg intramuscular 45 min prior to surgery.

The patient was shifted to the OR and standard monitors were attached and midazolam 1.5 mg was given intravenously (IV), followed by 100% inspired oxygen administration for 5 min. Anaesthesia was induced with IV fentanyl 140 mcg and propofol 140 mg. After loss of

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consciousness, manual in-line stabilisation (MILS) of the neck was provided by a second anaesthesiologist, and after confirming adequate mask ventilation, IV rocuronium 50 mg was given following which trachea was intubated with 8.5 mm ID cuffed flexometallic tube. Anaesthesia was maintained with 40% oxygen in nitrous oxide and end-tidal sevoflurane 1–1.5% using circle system. Intraoperative monitoring included ECG, non-invasive BP, capnometry, pulse oximetry, temperature, urine output, train of four using TOF Watch and gas analyser. Subsequently, the patient was positioned prone for the surgery over the bolsters, taking great care to avoid eye and bony compression. Surgery lasted for 2.5 h, with around 800 mL of blood loss. Posterior decompression and fusion of C3 to D5 and interbody cage insertion at D1-2 was done. The patient received 2 L of lactated ringer solution and blood loss was replaced with tetrastarch (130/0.42) 500 mL. Vitals were stable throughout the surgery. At the end of the surgery, the patient was reversed with neostigmine 3 mg and glycopyrrolate 0.5 mg IV, and the trachea was extubated. Then, the patient was shifted to the post-anaesthesia care unit (PACU) with oxygen 5 L/min using face mask. In the PACU, the patient's vitals were normal. Pain management was done with IV Paracetamol 1 g and then 6th hourly, over 15 min, IV ketorolac 30 mg and titrated dose of IV morphine using 2-mg aliquots to maintain visual analogue score (0 means no pain, 10-worst pain) of ≤3 using syringe pump. The patient was observed overnight in the PACU and shifted to his room the next day without any complications.

DISCUSSION

GS or disappearing bone disease, or massive osteolysis, is a mysterious bone disease of unknown aetiology, which affects patients from 1.5 to 72 years, although it occurs most commonly in the 2nd and 3rd decades.[6] It occurs with equal sex distribution. 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.[7]

Clinical manifestation varies from pain and swelling of the affected extremity, to limitation of motion and weakness of the involved limb. Many a times, they present with pathological fracture and respiratory and neurological complications. Diagnosis is based on clinical and radiological findings, which is confirmed by biopsy of the bone.

Various treatment modalities include radiation therapy,[5,8] anti-osteoclastic medication and alpha-2b interferon.[9] Surgical treatment options include resection of the lesion and instrumentation using bone grafts and prostheses.

Anaesthesia for major spine surgery, such as spinal stabilization, present a number of challenges. Prolonged anaesthesia, significant blood loss, hypothermia, positioning, concerns of air embolism, spinal cord monitoring and respiratory complications make anaesthetic management very challenging. A proper plan should be made and discussed with surgeons pre-operatively regarding anticipated duration of surgery, blood conservation strategies (use of antifibrinolytics, hypotensive anaesthesia, cell saver, acute normovolaemic haemodilution), neurologic monitoring to prevent spinal cord damage, wake-up test, post-operative ventilation and pain management. Most of the commonly used anaesthetics affect the interpretation of neuromonitoring. Hence, management should be tailored according to the method of neuromonitoring.[10] Although we had motor-evoked potential monitoring in our hospital, we did not use them, as our patient had lower limb power of 1/5 pre-operatively.

Review of the anaesthesia literature showed only few case reports till date.[2,6,11,12] Apart from the above-mentioned concerns for major spine surgery, additional problems in our patient were cervical spine protection during induction and positioning. MILS of the cervical spine during airway manoeuvres should always be used in patients without radiological abnormalities. However, in patients with cervical cord compression, awake fibreoptic intubation should be considered.[2,6] Anaesthesia induction (intravenous or inhalational) might need to be modified depending on the presence or not of lesions involving maxillary or mandibular bone in which inhalational induction, especially in children, and mask ventilation might not be possible. Great care should be taken while positioning prone for surgery, especially pressure over eyes, bony points and abdomen should be avoided.

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.[2,6,11] Diseases of ribs, scapula or thoracic vertebrae 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.[2,11] Post-operative ventilatory problem leading to re-intubation and prolonged ventilation has been described in the literature. Therefore, extubation has to be planned carefully, with due preparation for prolonged Intensive Care Unit management. Fortunately, our patient did not have any respiratory involvement and complication as well.

In a very recent case report, a nulliparous lady with GS was managed successfully under spinal anaesthesia for urgent caesarean section.[12] The same patient had a fused cervical spine due to surgery of cervical spine thus posing a serious challenge for airway management. Hence, neuraxial anaesthesia can be safely employed in infra-umbilical surgeries provided there is an absence of lumbar spinal osteolytic lesions.

GS generally does not affect hepatic, renal, metabolic or muscular function. Therefore, the pharmacokinetics and pharmacodynamics of commonly used anaesthetics remain unaffected. However, chylothorax-induced hypoproteinaemia dictates cautious use of high protein-bound drugs.[11] Succinylcholine may produce unpredictable fasciculation and the possibility of fracture in osteoporotic bones is a concern; hence, better to be avoided.

Fortunately, our patient had an uneventful recovery, without any further neurological sequelae. Apart from regular concerns for major spine surgery, special attention should be given to airway management and positioning, and anticipation of post-operative pulmonary complication will go a long a way in safe anaesthesia for this rare disorder.

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