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Case Report

Anaesthesia Section

Unravelling Anaesthetic Challenges in Patient with Diffuse Systemic Sclerosis: A Case Report

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ABSTRACT

Systemic sclerosis or scleroderma is an uncommon autoimmune condition with a global incidence of 8 to 56 new cases per million per year, which commences from skin and progresses to affect multiple systems in the body. It is marked by abnormalities in blood vessels, sclerosis of connective tissues and atrophy of skin and internal organs. The systemic sclerosis treatment depends on the disease manifestation and are usually treated with vasodilators (for vasculopathy improvement and prevention of Raynaud's phenomenon), prostacyclin analogues (for the prevention of Raynaud's phenomenon refractory to oral vasodilators), immunosuppressants like methotrexate, cyclophosphamide, mycophenolate mofetil, low dose corticosteroids (for the treatment of skin hardening, interstitial lung disease and inflammatory arthritis) and antifibrotics like endothelin receptor antagonists (for the treatment of pulmonary artery hypertension). Avascular necrosis in systemic sclerosis can arise as a result of the macrovascular and microvascular effects of vasculitis and corticosteroid therapy. The multisystem involvement of systemic sclerosis can impact every aspect of anaesthetic care especially airway management. During perioperative management, numerous systemic manifestations like pulmonary artery hypertension, interstitial lung disease and cardiac arrhythmia should be considered. The regional anaesthesia serves as a safe alternative to general anaesthesia and useful adjunct in the management of postoperative pain, but can be technically challenging. This case report described a 61-year-old female patient who had a history of systemic sclerosis for past 25 years and was scheduled for a total hip replacement due to avascular necrosis and secondary osteoarthritis of the left hip. The patient was having difficult cannulation, interstitial lung disease and anticipated difficult airway because of microstomia, limited mouth opening and limited flexion and extension at atlantooccipital joint. But the patient was successfully managed with combined spinal epidural anaesthesia, which provided effective pain control and minimised the perioperative risk associated with general anaesthesia.

Keywords: Collagen disorder, Microstomia, Raynaud, Regional anaesthesia

CASE REPORT

A 61-year-old female with a known case of systemic sclerosis since last 25 years presented with interstitial lung disease, sclerodactyly, Raynaud's disease with autoamputation of distal phalanges [Table/Fig-1a], avascular necrosis and secondary osteoarthritis of left hip. She was scheduled for total hip replacement. The patient was receiving tab prednisolone 5 mg once a day, tab mycophenolate mofetil 1 g twice a day, tab tadalafil 20 mg once a day, tab hydroxychloroquine 100 mg once a day and tab pregabalin 75 mg once a day.

General physical examination revealed pallor, tightly adherent facial skin, sclerodactyly with amputation of distal phalanges and ulceration of overlying skin. No peripheral venous sites were accessible. Heart rate was 86 bpm, blood pressure of 130/80 mmHg and oxygen saturation of 98% on room air. On airway examination there was restriction of mouth opening of 2 cm and Mallampati grading of 3. There was limited flexion and extension of atlantooccipital joint. On systemic examination, air entry was decreased on bases on bilateral sides. Heart sounds were normal.

Laboratory investigations revealed normal Kidney Function Test (KFT), Liver Function Test (LFT), serum electrolytes and coagulation profile. Complete Blood Count (CBC) revealed decreased haemoglobin of 9.8 g/dL. Electrocardiography (ECG) revealed left axis deviation with left bundle branch block [Table/Fig-1b]. On Two-dimensional Echocardiography (2D Echo) there was normal chamber dimension with ejection fraction of 56%, grade 1 diastolic dysfunction and mild aortic regurgitation. On chest X-ray there were bilateral opacities more prominent in lower lobe of lungs [Table/Fig-1c]. Arterial Blood Gas (ABG) analysis was unremarkable. Spine X-ray anteroposterior and lateral view was advised to look for intervertebral spaces [Table/Fig-1d].

[Table/Fig-1]: (a) Hands showing shiny skin and autoamputation of digits; (b) Electrocardiogram (ECG) showing left axis deviation and left bundle branch block; (c) Chest X-ray of the patient showing opacification in all lung fields, more prominent in lower lobes of lung; (d) Anteroposterior and lateral view of the spine with visible intervertebral space in lower lumbar region and crowding of space in lower thoracic and upper lumbar region; (e) Paediatric SpO_2 sensor probe on ear lobule for continuous saturation monitoring.

There was straightening of spine in cervical and thoracic region with visible invertebral spaces in lower lumbar region [Table/Fig-1d].

The patient was classified as American Society of Anesthesiologists (ASA) grade 3 physical status and written informed consent was obtained. Preoperatively, the patient was given tab pantoprazole 40 mg and tab anxit 0.25 mg the night before and 2 hrs before surgery. Since no peripheral veins were accessible, so under all aseptic precautions ultrasound guided right internal jugular vein cannulation was done. A combined spinal epidural anaesthesia was planned. However, all the preparation for general anaesthesia was kept ready including a difficult airway cart for anticipated difficult airway. An 18 gauge Tuohy needle was inserted at L3-L4 intervertebral space in the midline and the identification of epidural space was done using loss of resistance technique. A 27 gauge Sprotte® spinal needle was inserted through a Tuohy needle into subarachnoid space where free flow of Cerebrospinal Fluid (CSF) was observed followed by injection of 14 mg of 0.5% heavy bupivacaine. An epidural catheter was threaded into the epidural space and secured over the patient's back. The procedure was reported as atraumatic with no bloody tap or blood in the catheter. The upper limit of sensory blockade was T6. Intraoperatively, basic monitoring was done. There was difficulty in getting the saturation, so paediatric SpO_2 sensor probe was applied on the ear lobule [Table/Fig-1e]. Surgical procedure was completed in three hours. During this period, the patient remained haemodynamically stable with no episode of hypotension. The blood loss was 500 mL and one unit packed cell volume was transfused. Postoperatively, monitoring of vitals and epidural catheter care was advised and epidural infusion of 0.125% bupivacaine and 2 mcg/mL fentanyl was started at 5 mL/hr. The epidural catheter was removed three days after surgery. The patient was discharged after five days in stable condition.

DISCUSSION

Scleroderma, a connective tissue disease is associated with excess production and deposition of collagen types I and III, fibronectins and glycosaminoglycans within connective tissues, which can involve the skin, airway, musculoskeletal, gastrointestinal, cardiopulmonary, and renal systems [1]. It can present multiple challenges to anaesthetists ranging from difficult intravenous access, difficult mask ventilation and intubation, difficult regional anaesthesia, risk of aspiration, cardiac arrhythmias to decreased pulmonary compliance requiring high airway pressure to facilitate ventilation [2].

In systemic sclerosis, characteristic changes in blood vessels are produced by three separate processes [3]. Firstly, autoantibodies are produced, and there is activation of cell-mediated autoimmunity by abnormalities in the innate and adaptive immune system. Secondly, defective endothelial cells and fibroproliferative vasculopathy of small vessels develops, leading to vasoconstriction and obliteration of macro- and microvasculature [4]. Thirdly, because of the fibrogenic characteristics of endothelin and abnormal fibroblasts, qualitatively normal collagen is excessively produced. The occurrence of avascular necrosis in systemic sclerosis can be attributed to both macrovascular and microvascular effects of vasculitis, and corticosteroid treatment represents an additional risk factor for its development [5].

Dermal fibrosis manifests in a sequential manner, commencing with the hands and extending to the face, arms, and upper chest, leading to a waxy, non pitting oedema accompanied by flexion contractures, ischaemic ulceration, and resorption of distal phalanges. In around 70% of patients, there may be pinching of the face with atrophy of the nasal alae and restriction of mouth opening due to shrinkage of soft tissues and temporomandibular joint fibrosis. The thickened and inelastic skin may lead to limitations in neck extension [2,6,7].

The digestive system is most frequently affected and accounts for more than 90% of patients. The Gastrointestinal (GI) abnormalities include Gastro-esophageal Reflux Disease (GERD), dysmotility, small bowel bacterial overgrowth and pseudo-obstruction. In the present case, there was no history of GERD, but the authors prophylactically gave antacid and antiemetic prior to surgery [8]. Almutairi R and Alkhudair D, reported a life-threatening complication of systemic sclerosis secondary to aspiration because of oesophageal dilatation and reflux, which contributed to the development of Acute Respiratory Distress Syndrome (ARDS) in an already damaged lung due to interstitial lung disease [9].

Central venous access is often required when peripheral venous access proves challenging because of skin thickening, flexion contractures, and vasoconstriction [2]. Here, also there was difficult cannulation, so to avoid multiple puncture, decision for central venous access was taken earlier. There may also be difficulty in oxygen saturation monitoring on fingers due to autoamputation of digits, so paediatric SpO_2 sensor probe was applied to the ear lobule for uninterrupted oxygen saturation monitoring [10].

Interstitial lung disease may develop as pulmonary infiltration leads to fibrosis of the interstitium and peribronchial areas and proliferation of the bronchial epithelium. Pulmonary function tests show decreased compliance, reduction in vital capacity, and an alteration of diffusion capacity associated with hypoxaemia [11]. Pulmonary vascular disease, principally pulmonary arterial hypertension, may develop in 10-40% of patients with systemic sclerosis, leading to cor pulmonale and subsequently right-sided heart failure [2,12]. So, the patient was advised 2D-Echo in the pre anaesthetic check-up but there was no pulmonary artery hypertension in the patient.

Wherever feasible, regional anaesthesia is the preferred method for systemic sclerosis patients to mitigate the risk of aspiration and intubation failure. As per the available literature, surgeries of the lower limb have been done successfully under regional anaesthesia in patients with systemic sclerosis [13-15]. Awake fibreoptic intubation is preferred in cases where general anaesthesia is the only choice, especially when mouth opening is limited. It also preserves spontaneous respiration, which is helpful if difficulty arises, and the cough reflex if regurgitation occurs. Rapid sequence induction should be avoided, in spite of the high risk of aspiration, and the risk of difficult intubation. Due to the oesophageal fibrosis, Sellick manoeuvre may not be very useful in these patients. It may be necessary to consider awake tracheostomy with local anaesthesia in particularly difficult situations [2]. There are various case reports of difficult airway in these patients. Ye F et al., described the use of Shikani optical stylet for intubation in a patient with history of localised scleroderma and anticipated difficult airway, posted for the surgery of acetabular fracture [16]. Kanter G and Barash PG, presented a case of a patient with difficult airway with previously unrecognised scleroderma which required retrograde wire intubation [6].

In cases of regional anaesthesia, caution should be taken to prevent potential complications such as total spinal anaesthesia or insufficient anaesthesia necessitating tracheal intubation [7]. When the available literature was reviewed, it was found that smaller dose of local anaesthesia is required as systemic sclerosis results in the prolongation of sensory and motor blockade [17]. It may be due to fibrosis of muscles, connective tissue, and inelasticity of the fascial planes surrounding peripheral nerves, which may lead to a rise in compartment pressure on injection of local anaesthetic agent [18]. However, in this case, a routine dose of local anaesthetic (hyperbaric bupivacaine 0.5%, 14 mg) was used without any complications. Due to vasoconstriction commonly seen in systemic sclerosis, patients often exhibit reduced intravascular volume, potentially resulting in refractory hypotension during and after anaesthesia induction. These individuals are often unresponsive to vasopressors, which can also exacerbate vasoconstriction in the upper limbs [17]. Therefore, adequate preloading is crucial, although caution must be exercised to prevent rebound pulmonary oedema upon resolution of neuraxial block and the return of intravascular tone. To mitigate these risks, the combined spinal epidural technique offers a viable approach,

allowing for a dense and controlled block via intrathecal injection with the option to gradually prolong the duration of anaesthesia using an epidural block.

During the intraoperative period, it is crucial to closely monitor and regulate the patient's body temperature through standard methods to decrease the chances of vascular crisis. In individuals with systemic sclerosis, reduced sweating requires cautious measures to prevent overheating, as it can manifest as malignant hyperthermia. Proper positioning, especially considering flexion contractures, should be gently guided with the patient's cooperation while paying special attention to protecting pressure points with additional padding [2,6].

CONCLUSION(S)

In summary, the present case illustrated the successful use of combined spinal epidural anaesthesia in a systemic sclerosis patient undergoing total hip replacement surgery, taking into consideration all known risk factors and possible complications in such cases. A meticulous preoperative evaluation to assess the systemic involvement and a thorough clinical examination are essential to devise an appropriate anaesthesia strategy for these patients.

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