Case Report

Anesthetic management of a scleroderma patient undergoing tricuspid valve replacement under cardiopulmonary bypass

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ABSTRACT

Scleroderma is a multisystem disease that starts with the inflammation and atrophy of small blood vessels and leads to fibrosis of the skin and visceral organs. The pathological effects of the disease on various organ systems significantly impact aspects of anesthetic management. In this article, we describe the general anesthetic management of a scleroderma patient undergoing tricuspid valve replacement under cardiopulmonary bypass. We also review for the anesthesiologist perioperative considerations particular to this disease, especially the selection of anesthetic type, method of tracheal intubation, and establishment of venous access. The management of pulmonary hypertension and other anesthesia-related complications during the perioperative period are also discussed.

KEY WORDS: Scleroderma, Anesthesia, Tricuspid valve replacement, Cardiopulmonary bypass.

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INTRODUCTION

Scleroderma is a systemic autoimmune disease that affects various organs and leads to diffuse fibrosis of skin and visceral organs. The pathophysiology of scleroderma involves overproduction and accumulation of collagen, proliferation of fibroblasts, and activation of the immune system.[1,2] Due to multisystem involvement, patients with scleroderma present various challenges for anesthetic management. There are many reports in the literature about perioperative management for scleroderma patient.[2-5] However, anesthetic management for scleroderma patient undergoing cardiac surgery under cardiopulmonary bypass (CPB) is rarely reported in the literature. Here, we report a patient with scleroderma who underwent tricuspid valve replacement under CPB, and we discuss anesthetic concerns that should be considered for perioperative management.

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CASE REPORT

A 25-year-old male (height 176 cm, weight 57 kg) with tricuspid valve regurgitation complicated by pulmonary hypertension was referred to our hospital for tricuspid valve replacement. He had been diagnosed with scleroderma about two years previously. On physical examination, the patient presented with thick and hard skin involving the head, face, neck, chest, upper back, and bilateral forearms. The skin of his hands and feet was normal. Head and neck mobility were seriously limited. Airway examination revealed a limited mouth opening, Mallampati class 4, 3.5-cm thyromental distance, and a 2.5-cm distance between the upper and lower central incisors. Echocardiographic examination revealed right atrial and ventricular enlargement, severe tricuspid regurgitation, pulmonary hypertension, and decrease in right ventricular contractility.

This patient was transferred to a prewarmed operating room and standard monitoring (electrocardiogram, pulse oximeter) was attached. Before induction of anesthesia, omeprazole 40 mg IV was administered, the right radial artery was cannulated under local anesthesia for direct arterial pressure monitoring. Anesthesia was induced with IV 0.2 µg/kg sulfentanil, 0.05 mg/kg midazolam, and 0.2 mg/kg etomidate. The patient, maintaining spontaneous respiration, was then preoxygenated using a face mask with 100% oxygen for five minutes. Nasotracheal fiberoptic intubation was achieved using a flexible, wire-reinforced silicone tracheal tube (7.0 mm internal diameter) and an Olympus fiberscope through the left nostril after nasal ephedrine. To reduce the protective reflex of the respiratory tract, we used 2% lidocaine spray for local anesthesia. The correct position of the tube was confirmed with the fiberscope. The patient was then mechanically ventilated with 2% sevoflurane in 100% oxygen. Ventilation was controlled and the ventilator was set to give a tidal volume of 7 ml/kg at a 16 cycles/minute respiratory rate and a positive end-expiratory pressure (PEEP) of 5 cm H_2O. A 7F, three-lumen catheter was inserted into the left femoral vein for continuous measurement of central venous pressure, and nasopharyngeal and rectal temperatures were also monitored. A loading dose of 0.2 µg/kg sufentanil was administered before sternotomy. Anesthesia was then maintained with sevoflurane and sulfentanil, and repeated doses of vecuronium as needed.

The patient underwent tricuspid valve replacement under CPB. During CPB, The mean nasopharyngeal and rectal temperatures were 32.4 °C (range 30.6-33.2 °C) and 31.9 °C (range 29.4-33.0 °C), respectively. Mean arterial pressure was 68.3 mmHg (range 57-82 mmHg). Rewarming was started and continuous IV infusion of alprostadil (0.02 µg/kg/min), nitroglycerin (0.5-1 µg/kg/min), and dopamine (0.5-3 µg/kg/min) were used for maintenance of hemodynamic stability. Mechanical ventilation was resumed with 100% oxygen at a fresh gas flow of 1 L/min and 5 cm H_2O of PEEP. The patient was weaned from CPB without any difficulty. After weaning and disconnecting the CPB, all IV fluids were prewarmed prior to infusion.

At the end of surgery, the patient was transferred to the intensive care unit (ICU) for close observation and care. The patient regained complete consciousness and enough muscle strength for breathing after 13 hours in the ICU. With the anesthesia care team in attendance, tracheal extubation was performed without difficulty. The patient maintained spontaneous breathing and with good arterial blood gases. Perfusion of the fingers was normal, with a capillary refill time of two seconds and warm temperature. The patient was discharged from the ICU on the third day after surgery. The postoperative period was uneventful.

DISCUSSION

Careful preoperative evaluation of the scleroderma patient is necessary because of the diverse systemic effects of the disease. Involved systems may include the skin, temporomandibular joint, pulmonary, cardiac, renal, and gastrointestinal. Although there are no contraindications to the selection of anesthesia type in scleroderma patients, specific manifestation and pathophysiology of the disease certainly influence the choice of anesthetic procedures during the perioperative period.

Some authors have reported that regional anesthesia was successfully performed in scleroderma patients.[3-5] In many cases, this can provide excellent anesthesia and analgesia, reduce postoperative pulmonary complications, and improve postoperative outcome. However, in the present case, epidural anesthesia would have been difficult due to thickened skin and the affected subcutaneous tissue of the back and neck; the patient's breathing pattern was predominantly abdominal due to the stiff and inelastic skin of the chest and back, it may be difficult to manage respiration under thoracic epidural anesthesia during the intra-operative period. Furthermore, a high level of blockade, which can cause loss of airway, had to be avoided in the present patient. And due to an increased potential risk of epidural hematoma formation, use of epidural techniques in patients undergoing cardiac surgery under CPB remains extremely controversial.[6] Awareness of these considerations, in the present case general anesthesia with tracheal intubation was selected after discussion with the patient & cardiovascular surgeon.

Fibrosis of the skin and subcutaneous tissues may lead to microstomia, which complicates tracheal intubation in scleroderma patients. Our patient presented with thickened inelastic skin of the face, neck, and oral aperture. It appeared that securing the airway by direct laryngoscope would be difficult, due to limited mandibular mobility. Some authors suggest that an awake tracheotomy with local anesthesia could be performed under these conditions.[2] However, a tracheotomy was also ill-advised due to sclerotic involvement of the neck region in this patient. Under these circumstances, an awake nasotracheal intubation while maintaining spontaneous breathing should be considered for securing the airway.

If postoperative mechanical ventilation is required, as in our patient, the nasal tracheal tube is more comfortable than an oral tracheal tube. However, an awake tracheal intubation may induce serious cardiovascular responses. To prevent or attenuate these responses, after adequate sedation and airway topical anesthesia nasotracheal intubation without muscle relaxation was performed via a flexible fiberoptic bronchoscope. At the same time, due to nasal and oral telangiectasias it is necessary to carefully manipulate during intubation to prevent potentially massive bleeding. Meanwhile, there was no profuse bleeding in our patient when heparin was administered during the surgery.

Another consideration particular to scleroderma is that esophageal dysmotility and lower esophageal sphincter incompetence due to progressive esophageal fibrosis may induce gastro-esophageal reflux, increasing the potential for pulmonary aspiration. Before induction of anesthesia, in the present patient IV administration of the proton pump inhibitor omeprazole had the dual benefit of increasing the pH of gastric residue and decreasing the volume of gastric fluid. This may reduce the risk of aspiration pneumonitis.

The placement of a central venous catheter is an essential technique for cardiac surgery, and the internal jugular vein is preferable. However, an inadvertent arterial puncture may lead to cervical hematoma, which could potentially expand and obstruct the airway. The taut skin and neck immobility of the present patient would make it difficult to apply external compression to prevent hemorrhagic complication. To avoid the above potential risks of internal jugular vein cannulation, in the present case femoral central venous catheterization was performed.

Pulmonary involvement often occurs in scleroderma patients and is characterized by interstitial fibrosis, pulmonary hypertension, and an impaired diffusing capacity. Pulmonary hypertension frequently develops even in the absence of clinical signs and symptoms, and is associated with poor prognosis in patients with scleroderma.[7] During anesthetic induction, patients with pulmonary hypertension are prone to develop systemic hypotension and cardiovascular collapse. Etomidate maintains systemic hemodynamic stability without affecting pulmonary vascular resistance (PVR), thus, it is an ideal agent for induction of general anesthesia in patients with limited hemodynamic reserve. Pulmonary hypertension can also be treated with alprostadil. In addition, moderate tidal volumes and low PEEP (4-8 cm H_2O) not only minimize atelectasis and optimize lung recruitment, but also significantly reduce PVR.

The temperature of the patient under CPB can play an important role in cardioprotection. In order to attenuate hypothermia-induced visceral vasospasm, some authors have used normothermic CPB during cardiac surgery in patients with scleroderma.[8] However, in view of a right heart dysfunction, we used mild hypothermia for cardioprotection during CPB in our patient. In order to maintain normal body temperature, we maintained the temperature of the operating room at over 23 ^{o}C, and used prewarmed IV fluids to minimize peripheral vasoconstriction after CPB.

CONCLUSION

This report presents the successful use of general anesthesia with tracheal intubation in a scleroderma patient undergoing tricuspid valve replacement under CPB. We believe that careful preoperative assessment is required to formulate an appropriate anesthetic plan. Although the etiology of scleroderma is still unknown, the selection of anesthetic procedures and agents must be carefully considered to prevent exacerbation of the patient's condition during the perioperative period.

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***Authors contribution*:**

He-Jiang Zhong, Yong Zheng, Bin Zhao and Tian-De Yang provided perioperative care to the patient. He-Jiang Zhong was involved in acquisition of data and wrote the original draft of the manuscript, he was involved in interpretation of data and prepared the final version to be published. Yong Zheng and Bin Zhao were involved in acquisition of data and approved the final version to be published. Tian-De Yang was involved in the critical revision of the intellectual content of the manuscript, and approved the final version to be published.