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Anesthetic Management of a Pregnant Patient with Scleroderma

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The pregnant patient with scleroderma presents multiple obstetric and anesthetic problems. There have been no reports addressing the anesthetic management for such patients. In this article, we describe the management of labor and delivery for a parturient with scleroderma and explore potential problems that should be anticipated by the anesthesiologist.

REPORT OF A CASE

A 25-year-old woman, 39 weeks gestation, gravida 2, para 0, spontaneous abortion 1, was admitted for management of labor and delivery. Progressive systemic sclerosis (scleroderma) was diagnosed 4 years earlier. Since that time, the patient's disease had evolved into the CREST syndrome (calcinoses, Raynaud's phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasias). Pulmonary fibrosis was documented on chest roentgenogram. Severe skin and joint contractures confined her to a wheelchair. Renal function was deteriorating (creatinine clearance 13 ml/min). Esophageal dysfunction prevented the patient from controlling her swallowing mechanisms, and a Hickman catheter was inserted in the left subclavian vein for parenteral hyperalimentation. Therapeutic trials of steroids and plasmaphoresis failed. Chlorambucil therapy was initiated but discontinued when an intrauterine pregnancy was discovered at 10 weeks gestation.

Pregnancy was complicated by a urinary tract infection, gastroesophageal reflux, and anemia, resulting in a short hospitalization for parenteral iron therapy. Attempts to evaluate pulmonary hypertension by cardiac catheterization were unsuccessful.

Physical examination revealed a thin gravid woman weighing 42 kg, with diffuse contractures of skin and joints, multiple telangiectasias, and calcinoses. The mouth was markedly contracted, opening only 2.25 cm. The mandibular prominence was recessed 6 cm beyond the maxillary prominence. No rales were audible. A prominent second heart sound was present. The peripheral pulses and spinous processes were difficult to palpate through the sclerotic skin.

The indwelling Hickman catheter was used for venous access. Blood pressure was monitored with an ultrasonic device (Infrasonde®). The labor room was warmed, and skin temperature was monitored closely. An epidural catheter was inserted at the L3-4 interspace, and when the cervix was 5 cm dilated, 4 ml of 2% chloroprocaine were injected after a 1.5-ml test dose. Analgesia was obtained from T10 to L4. Thirty minutes later, an additional dose of 8 ml 3% chloroprocaine was injected, resulting in anesthesia from T6 to S5. Shortly thereafter, a female infant was born with Apgar scores of 9 and 9 at 1 and 5 min. Anesthesia persisted for 5 hours and 55 min after the final dose of chloroprocaine.

Her cardiovascular status remained stable throughout labor and delivery. Her remaining hospital stay was unremarkable, and she was discharged in good condition on the third postpartum day. The infant, however, died at 3 days of age secondary to multiple congenital cardiovascular anomalies.

DISCUSSION

This patient manifests features typical of the CREST syndrome variant of progressive systemic sclerosis (PSS). PSS is a chronic progressive multisystem disorder characterized by varying degrees of vascular change, inflammation, and fibrosis of the skin and internal organs.[1] Some authors subdivide scleroderma into three forms: acrosclerosis, localized scleroderma (morphea), or diffuse scleroderma. Others prefer the concept of a spectrum of disease, the manifestations of which may vary depending on the degree of progression.[2] These manifestations may include cutaneous changes on the face, scalp, trunk, and extremities; they may progress to include pulmonary fibrosis, pulmonary hypertension, renal artery sclerosis, myocardial sclerosis, esophageal sclerosis, and any or all features of the CREST syndrome.

Pregnancy accelerates the progression of scleroderma in approximately half of the patients.[3] The incidence of spontaneous abortion, stillbirth, premature labor, and perinatal mortality is high.[4] Our patient had a history of spontaneous abortion, and this infant died in the perinatal period.

Some pathophysiological aspects of scleroderma pose problems for anesthetic management, regardless of the type of anesthesia and whether obstetric delivery is by the vaginal route or cesarean section. Lack of venous access is common in the patient who has scleroderma. Sclerotic skin and vasoconstriction frequently prevent access through superficial veins. In our case, the indwelling Hickman catheter was used. However, venous cutdown or central venous catheterization may be necessary.

Vasoconstriction also may interfere with blood pressure monitoring by the usual methods. Blood flow to the periphery (where blood pressure is most frequently measured) is diminished in scleroderma.[5] Korotkoff sounds may be difficult to auscultate adequately and may not accurately reflect central pressures. In our patient, an ultrasonic blood pressure device solved this problem. However, pressures may require monitoring via arterial catheterization. Catheterization of the smaller

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peripheral arteries has been associated with spasm and subsequent necrosis; therefore, these vessels should be avoided.[6]

The anesthetic risk of patients with PSS may be increased because of pathophysiology of various organ systems. Pulmonary hypertension, which is more common in patients with the CREST syndrome than with other PSS variants, is a major cause of death.[7-9] Renal involvement, generally a result of arteriolar intimal proliferation with vascular obstruction, decreases renal perfusion and eventually results in systemic hypertension.[9] Subsequent episodes of left-sided heart failure are not infrequently the cause of death in these patients. The usefulness of vasodilating agents in these situations is unpredictable.[7] Finally, sclerodermatous changes of the gastrointestinal tract, resulting in malabsorption of vitamin K[8], may lead indirectly to a blood clotting abnormality.

The problems above mentioned are common to any patient with scleroderma, irrespective of anesthetic choice or type of obstetric delivery. If a vaginal delivery is planned, the patient may be managed with regional anesthesia, although this may be difficult technically because of the skin, ligament, and joint changes of the back and spine. However, in addition to pain relief, regional anesthesia provides peripheral vasodilatation and increased skin perfusion of the lower extremities and is thus helpful in preventing Raynaud's phenomenon. Other measures to prevent Raynaud's phenomenon, including warming of the delivery room, use of warm intravenous solutions, and warm compresses for the patient's extremities, should be used. If regional anesthesia is used for labor and delivery, smaller doses of dilute anesthetics are recommended, because sclerodermatous patients may exhibit prolonged sensory and motor blockade.[10] This was quite evident in our case.

Cesarean section may be chosen for a patient with PSS. In this situation, the anesthesiologist must choose between regional and general anesthesia. General anesthesia, however, which poses potential risks for even the healthy parturient (*e.g.*, pulmonary aspiration), is complicated further by the presence of scleroderma. Sclerodermatous skin contractures can limit opening of the mouth severely, making conventional intubation difficult or impossible.[11,12] Oral or nasal telangiectasias, which are among the features of the CREST syndrome, may bleed profusely if traumatized during placement of the endotracheal tube.[1,13] The risk of aspiration of gastric contents may be compounded by the presence of gastroesophageal sphincter incompetence and esophageal dysmotility.[1,14] Additionally, patients with scleroderma frequently have constricted blood vessels and abrupt vasodilation after induction of general anesthesia, which may provoke a sudden hypotension.[6]

Sclerodermatous involvement of the lungs poses further risks for the use of general anesthesia. Pulmonary fibrosis causes decreased inspiratory capacity, decreased compliance, decreased diffusion capacity, and increased residual volume.[7] Although dermal sclerosis does not decrease chest wall compliance, as was once believed, lung compliance is diminished by pulmonary fibrosis, and increased pressures may be required for adequate ventilation. Hypoxemia resulting from decreased diffusion capacity is not unusual in these patients, even at rest[7]; thus, increased inspired oxygen concentration may be required for adequate fetal and maternal oxygenation during anesthesia. Also, in the postoperative period, less than full ventilatory effort in the sclerodermatous patient may augment the baseline hypoxemia profoundly. Similarly, the sclerodermatous patient may not tolerate the ventilatory depression caused by narcotics. Thus, assisted postoperative ventilation may be required.

Endotracheal intubation, as mentioned above, may present the greatest problem of all. An awake oral endotracheal intubation will preserve voluntary respiratory effort and provide a margin of safety should poor mouth-opening prevent prompt intubation of the trachea. Awake intubation also will preserve the gag reflex and allow the patient to protect their airway if regurgitation should occur. Use of a fiberoptic bronchoscope may facilitate endotracheal intubation through a small oral opening and permit visualization of telangiectasias, which then can be avoided.[1] A tracheostomy, however, may be needed, should intubation efforts fail, or if time is insufficient to permit adequate preparation for intubation (*i.e.*, urgent cesarean section for which emergency tracheostomy may be faster). Although some may recommend anesthesia via a mask in the head-up position to reduce the risk of aspiration of gastric contents, we feel an airway secured with an endotracheal or tracheostomy tube is more appropriate for obstetric anesthesia. Additionally, anatomic deformities in patients with PSS (as in our case) may make mask anesthesia difficult. Further considerations to prevent aspiration pneumonitis are the use of oral antacids, glycopyrrolate or cimetidine (to reduce gastric acidity), and metoclopramide (to promote gastric emptying and increase gastroesophageal sphincter tone). Finally, although use of regional anesthesia would obviate the difficulties and risks associated with endotracheal intubation and general anesthesia, this may not be the safest approach for the patient with scleroderma. Should a complication of regional anesthesia arise that requires endotracheal intubation for treatment (*e.g.*, total spinal or total epidural), the patient might be at higher risk than if endotracheal intubation was performed as an elective procedure.

In summary, labor and delivery of an infant of a pregnant patient with PSS were successfully managed under epidural anesthesia. Careful preoperative assessment and a thorough understanding of the pathophysiologic interactions of scleroderma, pregnancy, and anesthesia are essential in formulating an anesthetic plan that will provide for all contingencies.

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Prolonged Neuromuscular Blockade Following Succinylcholine in a Patient Homozygous for the Silent Gene

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Prolonged apnea following administration of succinylcholine (SCh) to patients with abnormal cholinesterase (ChE) genes is well documented. However, no investigator has examined an increased sensitivity to SCh with documentation by peripheral nerve stimulation with the extremely rare genotype $E_1^sE_1^s$.[1] In the present study, we stimulated the ulnar nerve and examined the duration and the type of SCh-induced neuromuscular blockade in a patient diagnosed having the $E_1^sE_1^s$ genotype.

Report of a Case

A 44-year-old woman, weighing 60 kg, who was otherwise healthy, was admitted for removal of a rectal cancer. She had undergone cesarean section under spinal anesthesia at age 27 years without difficulty; there was no further surgical or anesthetic history.

In the preoperative examination, EKG, chest roentogenogram, lung function, and all laboratory values, except ChE activity, were within normal limits. Serum ChE activity, measured by the modified butyrylthiocholine method, was zero units (normal ranges 80–140 units). This method used butyrylcholine as substrate and dithiobisnitrobenzoic acid for coloration of thiocholine, a hydrolysis product of the substrate.[2] Coefficient of variation was 4%. Our study plan was approved by the Human Study Committee at the hospital. Premedication consisted of hydoxyzine 50 mg and atropine 0.5 mg im. Anesthesia was induced with droperidol 5 mg, fentanyl 0.05 mg and thiamylal 150 mg iv, and tracheal intubation was facilitated with SCh 1 mg/kg iv. Anesthesia was maintained with fentanyl and nitrous oxide. After the administration of SCh, supramaximal single twitch (0.4 Hz), tetanic (TS, 40 Hz for 5 sec), and train-of-four (TOF) stimulation were administered at 5-min intervals to the ulnar nerve at the wrist by means of steel needle (Ø 0.15 mm) electrodes. The resultant force of adduction of the thumb was measured using a force-displacement transducer and was recorded. The patient remained apneic, and the evoked responses could not be obtained until 45 min after SCh. The fade in the TOF and tetanic stimulation following SCh indicated significant phase II block. The TOF ratios obtained 50 and 105 min after SCh were 0.2 and 0.52, respectively (fig. 1). Spontaneous respiration reappeared about 150 min after SCh; thereafter, muscle paralysis was obtained by administration of pancuronium. After the end of surgery (390 min after SCh), residual paralysis was reversed effectively with neostigmine 1.0 mg and atropine 0.5 mg iv. After the reversal, the TOF ratio increased from 0.48–0.91 (fig. 1), and response to tetanic stimulation was sustained well.

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