SCLERODERMA AND PREGNANCY

Anaesthetic Considerations

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Diffuse scleroderma (progressive systemic sclerosis) is a rheumatic disease characterized by inflammation, vascular sclerosis and fibrosis of the skin and viscera. The aetiology is uncertain and there is no effective form of therapy. The prognosis is poor and related to the extent of visceral, rather than cutaneous, involvement (Siegel, 1977). This disorder occasionally occurs in women of child-bearing age, who appear to maintain a normal fertility index (DeCarle, 1964). Consequently, diffuse scleroderma complicating pregnancy may occur.

The pregnant patient with simple acrosclerosis fares better than her counterpart with multisystem involvement. However, pregnancy has an unpredictable effect on the course of the disease (Slate and Graham, 1968). Indeed, morbidity and mortality are substantial among those mothers who develop concomitant hypertension (Fear, 1968; Karlen and Cook, 1974; Smith and Pinals, 1982). Therefore, the anaesthetist may have an integral role in the care of these high-risk parturients, especially if surgical delivery is planned. The recent report of Thompson and Conklin (1983) addresses the management of labour with regional anaesthesia. We offer the following case report to emphasize the considerations necessary for such patients if general anaesthesia is required.

CASE REPORT

The patient was a 24-year-old gravida 3, para 1, spontaneous abortion 1 with no living children. Diffuse scleroderma had been diagnosed by skin biopsy 18 months before admission. She had presented at the labour suite with premature rupture of membranes on the morning of admission. The patient had received no ante-natal care. However, from the history it appeared that the course of her pregnancy had been uncomplicated. The estimated gestational age of her fetus was 34 weeks. She was in active labour and was prepared for Caesarean section under general anaesthesia. Her previous child had also been delivered by Caesarean section.

Her history was unremarkable and she complained only of skin tightness in her limbs. Physical examination revealed normal vital signs and taut, shiny skin on the face, trunk and extremities. She could open her mouth about 4 cm and had full range of neck movements. A rapid sequence oral intubation was thought possible. Examination of the heart and lungs was normal. Serum electrolyte concentrations, urine analysis, blood count, prothrombin and partial thromboplastin times were within their normal ranges. A chest x-ray had been interpreted as normal.

Before the induction of general anaesthesia, the patient received 30 ml of sodium citrate 0.3 mol litre\(^{-1}\) by mouth and glycopyrrolate 0.4 mg via an existing cannula in a peripheral vein. She was positioned with left uterine displacement on the operating table. Following the inhalation of 100% oxygen for 5 min, a moderately difficult rapid sequence induction with endotracheal intubation was performed utilizing cricoid pressure, thiopentone 250 mg i.v. and suxamethonium 100 mg i.v. After the delivery of a vigorous male
infant, anaesthesia was maintained with fentanyl, diazepam, and 50% nitrous oxide and 0.25% isoflurane in oxygen. At the end of surgery, the trachea was extubated when the patient was responsive and her protective airway reflexes had returned. She was taken to the recovery room and subsequently discharged in a stable condition.

Several hours later the patient was noted to be tachypnoeic. Scattered fine crepitations were present on auscultation of her lungs. A chest x-ray revealed bilateral perihilar infiltrates. Blood-gas determination showed $P_aO_2$ 7.3 kPa and a haemoglobin saturation of 86.2%. Volume overload was suspected and fluid was restricted, diuretics given and oxygen administered by face mask. Since these measures did not alleviate her respiratory distress, the patient was transferred to the surgical intensive care unit.

Upon arrival, the patient was sedated and a nasotracheal tube passed. The lungs were ventilated mechanically. Haemodynamic variables were obtained following placement of a pulmonary artery thermodilution catheter. These revealed a pulmonary artery pressure of 48/28 mm Hg, a pulmonary artery occlusion pressure of 22 mm Hg and a cardiac output of 7.5 litre min$^{-1}$. Her initial pulmonary and systemic vascular resistances were 138 dyne s$^{-1}$ cm$^{-5}$ and 1066 dyne s$^{-1}$ cm$^{-5}$, respectively. It was also noted at this time that her urine output was diminishing progressively.

The patient’s subsequent hospital course was complicated by worsening respiratory insufficiency, pulmonary hypertension refractory to vasodilator therapy, sepsis, thrombocytopenia and renal failure. Despite parenteral hyperalimentation, ventilatory support, daily haemodialysis and attempted reversal of her bleeding diathesis, the patient died 1 month post-partum. The cause of death was a massive upper gastrointestinal haemorrhage. Her infant was noted to have multiple congenital anomalies and died at 3 days of age from the complications of necrotizing enterocolitis. The family denied permission for autopsy in both instances.

**DISCUSSION**

The unfortunate outcome of this patient prompted an investigation of the implications which diffuse scleroderma holds for the obstetric anaesthetist. Literature describing the effect of scleroderma on the antepartum course of pregnancy and the management of labour with regional anaesthesia has been cited above. The body of knowledge specifically concerning the complications in these patients following Caesarean section is limited to case reports of pulmonary oedema (Hoffman and Diamond, 1967) and renal failure attributed to malignant hypertension (Ehrenfeld et al., 1977). In neither report was the type of anaesthesia mentioned. Therefore, the method of anaesthesia cannot be related to outcome and it is impossible to recommend a preferred anaesthetic technique for operative delivery.

In this patient a general anaesthetic, rather than a regional technique, was chosen for two reasons. First, scar formation is presumed to be abnormal in scleroderma. It was thought that general anaesthesia would offer greater haemodynamic control should massive haemorrhage occur in the event of rupture of the previous uterine incision. Second, reports exist which attest to the unpredictable spread and prolonged duration of local anaesthetics in such patients (Eisele and Reitan, 1971; Thompson and Conklin, 1983). With this in mind, there seemed to be a very real possibility of sudden respiratory insufficiency with loss of protective airway reflexes should total spinal or total extradural anaesthesia occur inadvertently. This consideration also argued for general anaesthesia to secure the airway initially under controlled conditions.

Although these are the considerations which related directly to the selection of anaesthetic technique, the underlying disease process affects organ systems other than those involved in reproduction. Thus, it would appear pertinent to review those abnormalities which may further influence the choice or conduct of anaesthesia in such patients.

Intravenous access is obviously impeded by dermal thickening. Raynaud’s phenomenon may produce arterial vasospasm and inhibit peripheral perfusion. Musculoskeletal abnormalities such as flexion contractures of the extremities, as well as pre-existing avascular necrosis of the femoral head, have been described (Wilde, Mankin and Rodnan, 1970). Moreover, tightening of facial skin and mandibular resorption may irreversibly narrow the oral aperture (Ryatt, Hopper and Cotterill, 1982). Keratoconjunctivitis sicca exists in some patients and may predispose to corneal abrasions (Osial et al., 1983). Gastrointestinal involvement may produce incompetence of the lower oesophageal sphincter, as well as malabsorption of fat-soluble vitamins from the small bowel.
The kidneys, heart and lungs are major target organs in diffuse scleroderma. Unsuspected renal involvement is common (D'Angelo et al., 1969) and in the pregnant patient may produce the abrupt onset of overt kidney failure (Fear, 1968; Karlen and Cook, 1974). Myocardial dysfunction may occur as a result of sclerosis of the smaller coronary arteries and the conducting system (D'Angelo et al., 1969; James, 1974). Abnormalities of cardiac perfusion may be present, and appear to be attributable to a disturbance of the myocardial microcirculation (Follansbee et al., 1984). Finally, pulmonary fibrosis and arterial sclerosis are expected in far-advanced scleroderma. However, it is not widely appreciated that significant pulmonary hypertension may be present in an asymptomatic patient with a normal chest x-ray and only mild abnormalities of pulmonary function (Sackner et al., 1964; Ungerer et al., 1983).

With the above considerations in mind, we offer the following suggestions for management of such a patient under general anaesthesia. Positioning is best accomplished with gentle guidance, and the co-operation of the awake patient. A centrally-placed catheter may provide the most reliable i.v. access. Maintenance of a warm operating room environment and avoidance of peripheral i.a. cannulation may prevent the vasospasm of Raynaud's phenomenon. Care should be taken that the eyes remain protected at all times. If it appears that a rapid sequence induction will be technically difficult, an awake intubation, perhaps under endoscopic direction, may be advisable to guard against aspiration. Finally, it may be desirable to insert a pulmonary artery catheter before the induction of anaesthesia, especially if impaired renal function, pulmonary hypertension or cardiac dysfunction is suspected. With invasive monitoring, the haemodynamic effects of both parturient volume shifts and anaesthetic agents, including nitrous oxide (Hilgenberg, McCammon and Stoelting, 1980; Schulte-Säse, Hess and Tarnow, 1982), may be rapidly assessed. Moreover, the efficacy of any proposed therapy with pulmonary vasodilators, such as calcium antagonists (Rozkovec et al., 1983) or nitroglycerine (Pearl et al., 1983), may be readily determined.

To summarize, diffuse scleroderma is a disease of multiple organ system derangements, each of which has a specific implication for the conduct of general anaesthesia. The pregnant patient with scleroderma presents a special challenge in anaesthetic management because of her potential for precipitous cardiopulmonary and renal insufficiency. Moreover, should general anaesthesia be selected, an appreciation of the anaesthetic precautions necessary for the care of these patients may prevent serious perioperative complications.

REFERENCES


