WORLD JOURNAL OF PHARMACEUTICAL RESEARCH
SJIF Impact Factor 7.523
Volume 6, Issue 17, 774-778. Case Report ISSN 2277– 7105

SCLERODERMA IN PREGNANCY AND ANAESTHETIC IMPLICATIONS: A CASE REPORT

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Article Received on
25 October 2017,
Revised on 15 Nov. 2017,
Accepted on 05 Dec. 2017
DOI: 10.20959/wjpr201717-10379

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ABSTRACT

Systemic sclerosis is a multisystem disorder of unknown etiology influenced by a variety of genetic and environmental factors. The connective tissue disorder involves skin, airway, cardiopulmonary, gastrointestinal, musculoskeletal system and renal system. The various anaesthetic implications of the disease process should be well understood in order to provide safe anaesthesia. Here, we report successful management of primigravida with localized scleroderma posted for elective caesarean section under spinal anaesthesia.

KEYWORDS: Systemic sclerosis (SSc), raynaud's phenomenon, neuraxial anaesthesia, diffuse systemic sclerosis (dcSSc).

INTRODUCTION

Scleroderma is a progressive connective tissue disorder characterized by changes in the microvasculature that lead to organ fibrosis. The disease affects the skin, blood vessels and internal organs. Raynaud's phenomenon is present in 85% of patients with systemic sclerosis (SSc) and is often the presenting symptom. Patients with localized scleroderma have skin sclerosis that is normally limited to the hands, but will occasionally affect the face and neck. The patients may exhibit features of CREST syndrome (Calcinosis, Raynaud's phenomenon, esophageal dysmotility, sclerodactyly and telengectesia). Organ dysfunction may manifest as gastro esophageal reflux, hypertension, myocarditis, arrhythmia, renal insufficiency, dyspnea on exertion with interstitial changes on chest x-ray, pulmonary hypertension all of which can present multiple anesthetic challenges to anesthetists. Potential risks and benefits should be

anticipated carefully and the technique of anaesthesia should be individualized accordingly in order to minimize the risk.

CASE REPORT

A 28 year old female, primigravida was admitted for elective LSCS in view of transverse lie. She was a known case of systemic sclerosis, diagnosed 5 years back when she developed increased sensitivity of digits to cold. The diagnosis was made on the basis of symptoms and presence of antinuclear antibodies. She was on some ayurvedic treatment and no documents were available regarding treatment history. As per the history given by the patient and thorough physical examination, it was confirmed that the disease was localized to skin only. Her vitals were within normal limits. On auscultation bilateral air entry was equal with no added sounds. Patient had increased sensitivity of digits to cold. Skin was thickened and tightened on forearm and hands. Her fingers were shortened and sausage like with slight flexion contracture of fingers. Airway examination revealed microstomia with mouth opening of 2.5 cm and with Mallampati grade III. Neck flexion and extension were normal. All biochemical markers, chest X-ray and ECG were normal.

The patient was shifted to pre-warmed operation theatre. In the operating room, all standard monitors were attached. Her heart rate was 72 beats/minute and blood pressure was 132/84 mmHg. Oxygen saturation was 96% on room air. An 18 G cannula was secured in right External jugular vein because cannulation was difficult in peripheries due to flexion contracture. Ranitidine 50 mg iv and ondensetron 4 mg iv given. We planned spinal anaesthesia for caeserian section. However, Difficult airway cart and an ENT surgeon for tracheostomy was kept ready. Subarachnoid block was given at L3L4 level in left lateral position with 25 guage quincke's needle. Patient was immediately made supine with left lateral tilt. Sensory block was achieved upto T4 level, tested by pin prick test, with complete motor block of lower limbs assessed by modified bromage scale. One episode of hypotension was noted which was effectively managed with intravenous fluids and 6 mg of ephedrine. Surgery lasted for 50 minutes with blood loss of approximately 800 ml. A total of 1500 ml of crystalloids was given. Intraoperative and postoperative course was uneventful. Full sensation was achieved 3 hrs after institution of spinal anaesthesia. Preoperatively and intraoperatively all fluids were warmed, patient was covered with warming blanket and all limbs were wrapped in cottons for warmth and protection. The pulse oximeter probe was alternated between digits to prevent ischaemic damage to the already poorly perfused digits.

Postoperatively, the patient was shifted to the recovery room and after 24 hours of vigilant monitoring shifted to ward.

DISCUSSION

Scleroderma is characterized by inflammation, vascular sclerosis, and fibrosis of the skin and viscera. Microvascular changes produce tissue fibrosis and organ sclerosis. Injury to vascular endothelial cells results in vascular obliteration and leakage of serum proteins in the interstitial space.[1] The spectrum of disease varies from mild raynaud's phenomenon to diffuse skin involvement.[2] In our case, diffuse involvement of skin was present along with raynaud's phenomenon with no evidence of other system involvement. Etiology involves genetic and environmental factors, high incidence of anticentromere antibodies in the limited cutaneous form of the disease and an association with the major histocompatibility complex.[3] Age group 20-40 yrs and women are most often affected. The incidence of spontaneous abortion, premature labour and perinatal mortality is high. Pregnancy accelerates the disease process in approximately 50% of affected parturients.[4] Those with multiorgan involvement[5] and dcSSc are worst affected.[6]

Pathophysiological changes and multisystem involvement poses multiple anaesthetic implications. Venous access may be difficult, as in our case, because of thickened skin and chronic vasoconstriction. Indirect blood pressure measurement may be impaired due to flexion contracture and vasoconstriction, In addition to this, direct arterial cannulation and blood-pressure measurement may be associated with vasospasm and distal necrosis.[7]

There is no specific pre defined anaesthetic technique of choice. Anaesthetic technique should be Based on degree of organ dysfunction and type of surgery. General anaesthesia is associated with potential risk of difficult airway, increased risk of aspiration due to impaired esophageal motility. Sellick's maneuver may be ineffective due to esophageal fibrosis.[8] Nasal intubation is also associated with risk of bleeding due to presence of telengectisia. Tracheostomy may be required which itself may be difficult due to contracture. Regional anaesthesia is preferred over general anaesthesia, wherever possible. Controlled vasodilatation produced by regional anesthetic techniques can also improve tissue perfusion, promote wound healing, and prevent Raynaud's[9,10, 11] phenomenon. Though it should be used cautiously due to prolonged and unpredictable effects of local anaesthetics in systemic sclerosis as reported by Eisele and Reitan[12] who suggested that smaller doses of local anaesthetic are required, as systemic sclerosis patients exhibit prolonged sensory and motor

blockade. Bailey AR et al, Erk G et al, Sulemanji DS et al, Garcia-Sanchez MJ et al, Erol DD and Picozzi P et al reported use of neuraxial anaesthesia in patients with SSc with no prolongation of block. Agents used were bupivacaine, ropivacaine and prilocaine.[13-18]

General care of the patient includes a warm operation theatre, careful positioning, padding to avoid pressure necrosis and wrapping of limbs for warmth to avoid vasoconstriction. Patient's temperature should be monitored and maintained to reduce the risk of vascular crisis. Pulse oximeter probe should be alternated between digits during surgery to prevent precipitation of ischemic changes.[8]

Epidural anaesthesia and combined spinal epidural can also be used successfully as done by Erk G et al, Sulemanji DS et al, Erol DD and Picozzi P et al.

In our case, we planned spinal anaesthesia for the patient. Similarly, A.R Bailey et al reported Spinal anaesthesia for Caesarean section in a patient with systemic sclerosis.[13] Gunther et al also reported successful use of spinal anaesthesia in a patient with scleroderma in pregnancy.[19]

CONCLUSION

Scleroderma, a connective tissue disorder has many anesthetic implications due to its multisystem involvement. So a thorough knowledge of the pathophysiology of the disease, degree of organ involvement and detailed history and evaluation of the patient must be done in order to minimize anaesthetic risk. To avoid the challenges involved in general anaesthesia, regional anaesthesia should be preferred, wherever possible.

ACKNOWLEDGEMENT: Nil.

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