

# Anaesthetic management in a patient of Systemic sclerosis with CREST syndrome - A Case Report

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**Abstract:** Systemic sclerosis, previously known as scleroderma, is an autoimmune disease characterized by excessive fibrosis that occurs more commonly in females. In some patients, the disease evolves into the CREST syndrome. The prognosis is poor related to the extent of visceral involvement. We report the case of a 38 year old female, known case of systemic sclerosis with Interstitial Lung Disease, Pulmonary Arterial Hypertension, left lung collapse and consolidation and Esophageal dilations, Reynaud's disease, with gangrene left foot for below knee amputation under unilateral low dose spinal anesthesia.

**Keywords:** Systemic sclerosis, CREST Syndrome, unilateral, spinal.

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## Introduction

Systemic sclerosis, previously known as scleroderma, is an autoimmune disease characterized by excessive fibrosis that occurs more commonly in females.<sup>[1]</sup> Raynaud's phenomenon is the most common occurrence besides skin thickening. Localized scleroderma involves first the skin without involvement of other organs. Limited cutaneous systemic sclerosis is "limited" to the skin of the face and upper extremities as the cutaneous manifestation, but with systemic involvement, the GIT (dysphagia, reflux) and Lungs (Interstitial Lung Disease) can be affected. Diffuse cutaneous systemic sclerosis causes generalized skin thickening and multiple end-organ damage.<sup>[2]</sup>

In some patients, the disease evolves into the CREST syndrome. The prognosis is poor related to the extent of visceral involvement. No drugs or treatments have proved safe and effective in altering the underlying disease process in scleroderma.<sup>[3]</sup>

## Case Report

A 38 year old female, known case of systemic sclerosis since last 13 years, with Interstitial Lung Disease (ILD); Pulmonary Arterial Hypertension (PAH) with HRCT documented left lung collapse and consolidation; Esophageal dilations; Raynauds disease, presented with gangrene left foot and was planned for below knee amputation. The patient was receiving cilastazol, pregablin, nifedipine and aspirin from the last 1 year. General physical examination revealed Pallor, Periorbital puffiness, tightly adherent facial skin. Pulse was 110bpm, regular, good volume; BP 100/60mmHg. No peripheral venous sites were accessible. Sclerodactyly with flexion contractures in fingers was present. Gangrene left foot was present.

Airway examination revealed Mouth opening < 2 finger breath and MPS – III.

In the Systemic examination, diffuse bilateral coarse crepts (L>>R) were present. A short systolic murmur could be heard. Laboratory Investigations revealed a normal KFT, LFT, Serum electrolytes and coagulogram. CBC revealed ↓Hb (8.8g/dl). ECG revealed sinus tachycardia with right axis deviation with P-pulmonale; Chest X-ray showed bilateral diffuse infiltrates (L>>R) and obliteration of CP-angles. HRCT Chest showed Left lung collapse and consolidation with Esophageal dilations. ECHO revealed severe Tricuspid Regurgitation with moderate to severe PAH with no systemic congestion, ABG was unremarkable with SpO<sub>2</sub> of 90% (on Room Air). The patient was put on supplemental oxygen through nasal prongs preoperatively.

### **Anesthetic Management:**

Patient was classified as ASA Grade III physical status and a written and informed consent was obtained. Preoperatively, the patient was put on tablet ranitidine 150mg and tablet alprazolam 0.25mg the night before and 2 hours prior to surgery. She also received injection metoclopramide 10mg 1 hour prior to surgery. Preoperative vitals were – HR-115 bpm, BP-115/75 mmHg, SpO<sub>2</sub> 90% (on Room Air). Since no peripheral veins were accessible, Central venous catheterization was done through sub-clavian approach. CVP reading at start was 23 cmH<sub>2</sub>O. A unilateral, low dose spinal anaesthesia was planned. However, all preparations for a general anaesthesia were made including a difficult airway cart in case the spinal anaesthesia failed or in situations like a total spinal anaesthesia etc. Preloading was done with 300ml of Ringer lactate. Patient was put in left lateral decubitus position and a unilateral subarachnoid block was performed with 1.5ml(7.5mg) of 0.5% Hyperbaric Bupivacaine along with 0.5ml(25mg) preservative free Tramadol. Thereafter, patient was kept in that position for 15 minutes. Confirmation of adequate unilateral block was done and then patient was put in supine position. The level of block was upto T<sub>10</sub> dermatome.

Intraoperatively, basic monitoring as well as Central Venous Pressure (CVP) monitoring was done and CVP guided intravenous fluids were administered. O<sub>2</sub> supplementation via nasal prongs was given which was continued in postoperative recovery room.

Surgical procedure was completed in 1 hour and 30 minutes. During this period patient remained hemodynamically stable with no episode of hypotension. However, the patient had persistent tachycardia. SpO<sub>2</sub> remained in the range of 93% to 97% during the duration of surgery CVP remained in around 25 to 28 mmHg. The blood loss was approximately 300 ml and 1 Unit Packed RBC's was transfused.

Post operatively, patient's monitoring of vitals and post spinal anaesthesia care was advised along with continuation of O<sub>2</sub> inhalation via nasal prongs. Patient was discharged after 15 days in a stable condition.

### **Discussion**

The term scleroderma is derived from the Greek word 'sclerosis' (hard or indurated) and derma (skin) and it is used to describe a disease characterized by progressive skin hardening and induration. Hippocrates first described this condition as thickened skin.<sup>[4]</sup> Systemic sclerosis is a complex and heterogenous disease and clinical forms ranging from limited skin involvement (limited cutaneous systemic sclerosis) to forms with diffuse skin sclerosis and severe and often progressive and occasionally a fulminant course (fulminant systemic sclerosis). Limited cutaneous systemic sclerosis involves areas distal to the elbows and knees but may involve the face and neck. CREST syndrome (Calcinosis, Raynaud's phenomenon, Esophageal dysmotility, Sclerodactyly, and Telangiectasis although not needed for the disorder to be called CREST) is an older term used to describe this subset of limited cutaneous systemic sclerosis. Exact etiology of systemic sclerosis is not known. Systemic sclerosis is not inherited, although a genetic predisposition plays an important role in its development. Environmental factors (e.g. triggers or accelerators) may contribute to the development of systemic sclerosis in the proper genetic background.<sup>[5]</sup> Risk of systemic sclerosis is 4-9 times higher in women than in men.<sup>[6]</sup> Peak onset occurs in individuals aged 30-50 years. Numerous cases occur in children, however, as well as in very old individuals.

### **Treatment Approach:**

Current treatment of systemic sclerosis is directed toward managing complications and providing symptomatic relief. In addition, a range of disease modifying treatments have been investigated. Disease modifying treatment aims at inhibiting tissue fibrosis and vascular and immune system alterations which are three crucial components of disease pathogenesis<sup>[7,8,9]</sup> like methotrexate, mycophenolate mofetil.

### **Anaesthetic Implications:**

Literature regarding anaesthetic management reveals that there are no specific contraindications to the use of any type of anaesthesia, although the selection must be guided by identification of organ dysfunction. General anaesthesia may involve difficult tracheal intubation due to fibrosis and tight facial skin which may markedly hinder active and passive motion of the temporo- mandibular joint. Awake, fiberoptic laryngoscopy may be required, tracheostomy may be necessary in severely affected patients. Patients are at substantial risk for aspiration pneumonitis during the induction of anesthesia because of high incidence of esophageal dysmotility and gastroesophageal reflux.

Regional anaesthesia may be administered, although the response to local anaesthetics may be prolonged. The anaesthesiologist is often consulted as to the efficacy of sympathetic blockade for the treatment of vasospasm secondary to Raynaud phenomenon.<sup>[10]</sup>

The type of anaesthesia has no substantial effect on perioperative morbidity and mortality.<sup>[11]</sup> Unpredictable spread and prolonged duration of action of local anaesthetics may occur and regional anaesthesia may be unacceptable for the patients with Systemic sclerosis.<sup>[12]</sup> In difficult to intubate cases despite the presence of high risk of spinal anaesthesia, some authors recommend regional anaesthesia for patients with severe Systemic sclerosis.<sup>[13]</sup> In addition to this, it is widely believed that regional anaesthesia is beneficial for patients with severe pulmonary disease. Harald et al. reported that, even high thoracic regional anaesthesia is well tolerated in patients with severe pulmonary disease, although FEV<sub>1</sub> and vital capacity may be slightly decreased.<sup>[14]</sup>

There are not many case reports of the use of regional anaesthesia in Systemic Sclerosis patients. In a relatively recent publication, a 56 year old female with a 12 year old history of scleroderma was safely administered a combined spinal epidural anaesthesia for total hip replacement.<sup>[15]</sup>

We preferred a unilateral low dose spinal anaesthesia for our patient to avoid hemodynamic instability. There are many benefits of this technique including less hemodynamic changes,<sup>[16]</sup> decreased urinary retention, more satisfied patients and the restriction of selective nerve block to the relevant limb.<sup>[17]</sup> In a study, unilateral spinal anaesthesia was given with hyperbaric bupivacaine 0.5% (1.1–1.8 ml) prior to lower-limb surgery in elderly patients with ASA classification of III or IV (average age, 60). The authors found no significant hemodynamic changes.<sup>[18]</sup> In our case also, the patient remained hemodynamically stable with no episode of hypotension. We avoided establishing invasive arterial access for haemodynamic monitoring because of Raynauds disease and instead relied on measurements of CVP for this purpose.

### Conclusion

Since systemic sclerosis is a multi-systemic disorder, it requires a careful and vigilant preoperative assessment, selection of type of anesthesia by consideration of various organ dysfunctions. Among regional anaesthesia techniques, unilateral, low dose spinal anaesthesia is a safe alternative in patients undergoing lower extremity, short duration procedures.

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