



**ORIGINAL RESEARCH PAPER**

**Anaesthesiology**

**A CASE OF SYSTEMIC SCLEROSIS PLANNED FOR FOOT AMPUTATION UNDER NEURAXIAL ANAESTHESIA : DEALING WITH DECEPTION**

**KEY WORDS:**

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**ABSTRACT**

**Background** Systemic sclerosis is an array of changes resulting from antibody mediated immune dysfunction characteristically observed in integumentary system leading to progressive end organ dysfunction and decreased survival. The microvascular damage leads to vasculopathy and progressive fibrosis of the internal organs. It poses a hurdle for the anesthesiologist due to its rare occurrence, limited evidence-based guidelines and a multi-system involvement. **Case description:** Our patient, a 77 yr-old female patient, who is a known case of systemic sclerosis was admitted with a history of pain in the bilateral lower limbs owing to distal gangrene with critical limb ischemia and is scheduled for amputation. Patient is associated with other comorbidities and is non ambulatory. Patient is on long term medication for the same and has various systemic findings involving the respiratory, cardiovascular and renal system. **Discussion:** Systemic sclerosis is a complex disease with different levels of involvement of different systems. Patient is often exposed to various drugs interactions and review of the current status of the disease becomes a necessity for intervention and to avoid catastrophic results. **Conclusion:** The optimal perioperative follow up and risk stratification can minimise if not annihilate the complications of the disease due to operative stress which is directed to salvage the patient and it should expand beyond the defined challenges of anesthetic technique to multisystem understanding.

**INTRODUCTION**

Systemic sclerosis is a complex clinically heterogenous disease with protean manifestations, a chronically progressive course ,with virtually every organ affected. Patients having lcssc (limited cutaneous SSC with skin involvement, Raynaud's phenomenon, digital ischemic ulcers, calcinosis cutis, oesophageal dysmotility, sclerodactyly, telangiectasia known as CREST syndrome with late lung and renal involvement. Epidural anesthesia which allows gradual incremental titration of block height can be used successfully in such patients without precipitating ischemic attacks.

**CASE DESCRIPTION**

A 77 year old female patient with 59 kg weight and 165 cms in height was scheduled for a Lisfranc amputation of foot owing to gangrene following long standing ischemic ulceration with critical limb ischemia. Patient is known case of SSC on tablet Nifedipine, prednisolone 4mg for 10 years, tab Aspirin 75 mg for 5 years, tab pentoxifylline, tab telmisartan 60 and pre-op injectable antibiotic for the resolution of super infection of the area. Patient has restricted mouth opening, restriction of neck mobility, indicating an anticipated difficult airway, local non pitting edema in ankle area and telangiectasia in the ankle area along with knee joint effusion causing disability. Patient has limited herself to bed and wheelchair with moderate restriction in PFT, bilateral knee joint effusion and power of lower limbs 2/5.

Pre op : CBC, LFT, RFT, Thyroid profile test, urine for protein, RBS, HBA1c including coagulation profile, CXR, a PFT, an ECHO was done. The reports were in acceptable range The Patient had one episode of ICU admission 2 years back with cardiogenic shock. Rheumatologic, dermatologic, cardiothoracic vascular surgery consultation were taken. Doppler Bilateral lower limb showed knee joint effusion, subcutaneous edema and critical limb ischemia.

Immunologic tests showed; ANA positive with titer 1:5130, Anti CCP POSITIVE, Anti-Scleroderma 18.6 units (0-24.9).

Patient was continued for tab aspirin as per ASRA guidelines, loading dose of hydrocortisone 100 mg bolus perioperative dose and anti aspiration prophylaxis given preoperatively injection ranitidine and injection metoclopramide. Warm

intravenous fluid with adequate volume was loaded, eye cover with adequate padding given.



**Fig.1**



**Fig.2**



**Fig.3**

Patient was continued for tab aspirin as per ASRA guidelines, loading dose of hydrocortisone 100 mg bolus perioperative dose and anti aspiration prophylaxis given preoperatively injection ranitidine and injection metoclopramide. Warm intravenous fluid with adequate volume was loaded, eye cover with adequate padding given.

Intraoperative: Vitals were recorded, blood pressure-156/90 and pulse rate 113/min were in normal range although the monitoring with the pulse oximetry was difficult we put two probes in the digits frequently changed, difficult cannulation was encountered, the body temperature probe and

precautions to avoid hypothermia were taken OT temperaturat at 25 degrees, patient had 96% saturation with oxygen support via nasal prongs.

Patient was given epidural anesthesia in L3-L4 space using 18 G Touhys needle and LOR technique and 0.5% Ropivacaine 15 ml was given. Block height was checked for T10 sensory level. Duration of the surgery was 40 mins. No vasopressors were required. The regression of the block height was checked for by pin prick technique and Bromage scale. Duration of sensory block was approximately 3 hours.

**PATIENT OUTCOME :** Patient had excellent sensory analgesia post operatively by VAS score 2/10 and we had achieved a pain free period of 190 minutes. Epidural fentanyl of 25 micgms given post op and patient was able to mobilise within a day. Also the relative vasodilation without major systemic hypotension was our aim to be fulfilled.

**DISCUSSION**

The anesthetic challenges can be traced from the classification of systemic sclerosis done with immunologic markers- limited and diffuse ssc. The organ specific manifestations in the types are given in the table below.

Organ	Manifestations	Anesthetic challenges
SKIN	Thickening and diffuse non pitting edema, ulceration, contractures, telangiectasias	Difficult cannulation and intubation
MUSCULOSKEL ETAL SYSTEM	Limited mobility and altered fascial planes, connective tissue fibrosis, proximal muscle weakness, symmetrical polyarthritis, and joint crepitation, limited TMJ mobility	Difficult for blocks especially PNS guided
EYES	Keratoconjunctivitis sica	Eye padding, covering
CVS	Cardiac dysrthmias, congestive cardiac failure, pulm htn, Raynauds phenomenon, pericarditis	Cardiac arrest, ICU care , pacing
LUNG (second most common manifestation)	Pulmonary vascular disease, interstitial lung disease	Hypoxia, acidosis, high airway pressure, decreased compliance
HEMATOLOGIC	Aplatic anemia	bleeding , transfusion
GASTROINTEST INAL	Dysphagia, decreased lower esophageal sphincter tone	Aspiration, difficult bag ventilation,
RENAL	Renal crisis	Restriction of fluid, nephrotoxic drugs
CNS	Trigeminal neuralgia,	Mask holding difficult, PNS regional anesthesia challenges

The primary goals of using epidural anesthesia was to prevent the hemodynamic fluctuations, cardiorespiratory and renal compromise, positioning of the patient with minimal challenges and adequate post-operative pain control in the geriatric case. In our patient, because of the restricted oral

aperture and decreased neck mobility and lung involvement regional anesthesia could be a better choice.

Controlled vasodilatation produced by regional anesthetic techniques can also improve tissue perfusion, promote wound healing, and prevent Raynaud's phenomenon. Regional anesthesia may be limited, however, by technical difficulties, prolonged sensory blockade, unpredictable spread of local anesthetics, and associated with marked cardiovascular compromise.<sup>1</sup>

The technical challenges encountered in performing peripheral regional anesthesia in SSc patients such as positioning difficulties, altered fascial planes, and altered peripheral sensation, may increase the risk of intraneural injection when using landmarks, fascial clicks, and patient reported paresthesia for location. In addition, the wide range of neurological abnormalities seen in SSc can cause abnormal nerve conduction, marking location of peripheral nerves using nerve stimulators difficult.<sup>1</sup> To avoid complications combined spinal epidural technique may be used which allows dense and low block with intrathecal injection and anaesthesia can be gradually completed using epidural block.<sup>2</sup>

A case of scleroderma with diffuse manifestation was planned for caesarean section under general anesthesia for achieving greater hemodynamic stability and adequate tissue perfusion. However immediate post operative complications were noted. The patient's postoperative course was complicated by pulmonary oedema and pulmonary hypertension, sepsis, thrombocytopenia and renal failure.<sup>3</sup>

Perioperative implications of medications used in the treatment of systemic sclerosis has been complied by Carr ZJ et al. Substantial progress has been made in the treatment of SSc using disease-modifying agents.<sup>4</sup>

SSc patients have an increased risk of unprovoked venous thromboembolic disease (VTE), likely related to the underlying inflammatory state and its pro-thrombotic nature . The highest risk was found in patients within 12 months of SSc diagnosis and is strongly associated with female sex, atrial fibrillation, and heart failure. Trauma and glucocorticoid use were independently associated with DVT, although surgery alone was not, which may be related to the common use of prophylactic anticoagulation in the perioperative period.<sup>5</sup>

**CONCLUSION**

In a case where pain due to Raynaud's phenomenon is a chief manifestation we have decided to utilise the spine for post-operative analgesia. Differential sensorimotor blockade via Ropivacaine has acted in our favor of us as noted in this case. Limited studies are available for epidural anaesthesia in SSc. .With the advent of USG guided blocks and estimation of local anesthetic spread this rare connective tissue disorder will be easier to manage. Proper pre operative assessment can help in planning such cases.

**CONSENT:** due consent was taken for the procedure .

**Conflicts of interests :** none

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