A Multidisciplinary Approach in Managing Raynaud’s Disease: A Case Report

ABSTRACT
Raynaud disease is a benign episodic digital vasospasm mainly of upper limbs, resulting in ischaemia of other peripheral tissues. The chief complaints are constant excruciating pain, burning type, aching sensation in the affected extremity, can be unilateral or bilateral, and discolouration of finger tips along with allodynia and hyperalgesia. A multidisciplinary approach is the key behind successful outcome of treatment which includes drugs, exercise, and interventions in form of diagnostic sympathetic blocks followed by radiofrequency sympathetic denervation.

KEYWORDS vascular, sympathectomy, pain.

INTRODUCTION
Pain takes a central position in a varied group of disorders, due to insufficient blood supply to the extremities. It results in ischemia of the peripheral tissues, which causes pain and often functional limitation in the patient. Pain is a signal indicating a serious problem. Two causes for such disorders are critical vascular disease and the Raynaud’s phenomenon. The latter can be of primary and a secondary type. The context and the cause of each of these three groups are different, hence good diagnostics are essential to identify and influence the prognosis.

CASE REPORT
A 28-year-old aesthetic build female presented in pain clinic with chief complaint of severe burning pain associated with numbness, tightness, and aching sensation in both hands since last 2 years. The pain was more severe in right hand involving index, middle, and ring fingers with bluish discolouration of fingers at the tip. Pain was severe at night and precipitated by cold. On exposure to cold, the fingers turned white, then blue and red during the acute attack. She was working in a beedi factory since last 7–8 years and left the job. She was emotionally depressed (felt like committing suicide) as she was unable to perform her daily activities. On examination, allodynia and hyperalgesia was present. The distal fingers were thin with contractures and showed signs of atrophy, ulceration, and discolouration. She also complained of pain and similar symptoms in right foot off and on. Her systemic examination revealed no respiratory or cardiac problem. There was no history of any significant illness or surgery in past. Her radial pulse was felt in right hand but was very feeble. Her blood investigations revealed normal reports except ESR which was 30 mm at the end of 1 h. Colour Doppler study showed dampened flow in radial and ulnar artery on right side. The flow was triphasic up to right dorsal pedis artery (DPA) in right lower limb. Her chest X-ray was normal. She was not on any treatment since the last 5–6 months. Considering it to be a case of Raynaud’s disease, the plan of treatment was made. Physician’s opinion regarding systemic involvement was taken. The patient was reassured that the condition is benign and generally remains stable and will slowly regress. She was advised to avoid exposure to cold and to protect the extremities by wearing warm gloves and clothes. She was also advised to quit the job, as it was the main causative factor for progress of the disease. Since the disease was in severe form, vasodilators (amlodipin 5 mg bd) and steroids (omnacort 5 mg tds) were started. Tramadol with paracetamol was given for pain relief. Since the pain was very severe with inability to perform any activity...
Regional Sympathetic Denervation was planned. A series of 4–5 diagnostic as well as therapeutic Stellate Ganglion Blocks were given under image intensifier at right C6 level with long acting local anaesthetic Bupivacaine 0.25% in dose of 5–8 ml. After these successful blocks, the VAS score came down from 9/10 to 3/10. Since these blocks produced adequate vasodilation and pain relief, radiofrequency (RF) ablation of the sympathetic plexus\textsuperscript{12,13} was done for pain relief. A small volume of local anaesthetic was injected before proceeding RF. The electrode was directed to the most medial part of the transverse process (Figs. 1–3) at 2 Hz, with 0.1–1.5 V, to locate or identify the phrenic nerve laterally and the recurrent laryngeal nerve anteriorly and medially. The patient should be able to say “ee” to preserve the motor function. There should be no motor response on proper localization. Second, seek paraesthesia for localization. For that, stimulate the nerve at 50 Hz with 0.1–1.5 V. Generally, a patient feels paraesthesia at a setting of 0.5 V. The patient should be awake to respond to the stimulation. At this stage, RF lesioning of the stellate ganglion was done by injecting a small volume of local anesthetic before lesioning. RF was applied for 60 seconds at 70°C at three different positions after repeated sensory and motor stimulation before each lesioning.

DISCUSSION

The treatment of the primary form of Raynaud’s phenomenon is usually conservative and not pharmacological. In case of primary Raynaud’s, it is generally sufficient to inform the patient well and advise them to avoid provoking factors by wearing warm clothes, stopping smoking, taking sufficient exercise, and avoiding vasoconstrictive medication. If pharmacological treatment is required, the vasodilators nifedipine (Ca\textsuperscript{2+} - antagonist) and prazosin (alpha 1-blocker) have been studied most, but their effects have been disappointing\textsuperscript{4,5}. The main problems encountered with these drugs are the adverse effects and the loss of efficacy in long-term.

The treatment of secondary Raynaud’s is initially aimed at the underlying disease. Sympathectomy is not often performed in patients with Raynaud’s. However, it can be considered in patients with dystrophic changes leading to ulceration\textsuperscript{13,14}. In their retrospective study (n = 28), Matsumoto et al.\textsuperscript{28} found an initially favourable result in 92.9\% after endoscopic thoracic sympathectomy (ETS); however, recurrent symptoms were subsequently noted in 82.1\%. Despite recurrent symptoms, these patients did not exhibit ulcerations during the study period.
Selective block of the sympathetic trunk was first reported by Sellheim and, shortly thereafter, by Kappis in 1923 and Brumm and Mandl in 1924. After 1930, the technique and indications were established by White and Sweet in the United States, and by Leriche and Fontaine in Europe.

The type of analgesia or sedation needs to be ascertained before performing the procedure. Intravenous fentanyl, midazolam, and/or propofol may be used judiciously for the patient’s comfort.

The etiology of peripheral vascular disease (PVD) is multifactorial and therefore the multidisciplinary approach to pain management is appropriate. The commonly encountered conditions are:

- TAO (thromboangiitis obliterans)
- ASO (arteriosclerosis obliterans)
- Embolism
- Collagen vascular diseases
- Raynaud’s phenomenon
- Raynaud’s disease
- DVT
- Thrombophlebitis
- Varicose veins
- Post thrombotic syndrome

**Mechanism of pain in PVD:**
- Inadequate perfusion of tissues
- Sudden change in dimension of vessels: aneurysm, vessel spasm
- Secondary changes: ulceration, gangrene
- Rupture of vessels
- Impaired venous return

**The clinical features of PVD are:**
- Pain
- Decreased or absent pulse
- Abnormal skin colour
- Diminished/elevated skin temperature
- Swelling/oedema

**Pathophysiology**

Trophic changes of skin and its appendages. The exact pathophysiological mechanism remains as yet largely unclear. However, it has been shown that the physiological vasoconstriction on noradrenaline is enhanced by cold and that there is an increased sensitivity to alpha 2-agonists and serotonin. The vasoconstrictive endothelin-1 would also be involved, and the calcitonin gene related peptide (CGRP) and cyclooxygenase supposedly play a (modulating) role. The primary or idiopathic form (Raynaud’s disease) often presents without an apparent cause and has a favourable course over time. In case of the secondary form (Raynaud’s syndrome), there is often a disorder of the connective tissue, collagen, or a rheumatic disease, often with autoimmune features (scleroderma, Sjogren’s disease, rheumatoid arthritis, systemic lupus erythematosus, polymyositis) or a peripheral vascular disease (thromboangiitis obliterans or Buerger’s disease). In rare cases, it occurs in combination with a malignancy or chemotherapy (cisplatinum, bleomycin, and vincristine).

**Diagnostic investigations:** Apart from routine Doppler flowmetry, Doppler ultrasound, angiography, and digital subtraction angiography (DSA) play an important role in diagnosing the type of PVD. The role of pain physician is in managing the intractable pain when the conservative measures are inadequate in managing the pain.

**Pain:** The pain in TAO or ASO is intermittent and is precipitated by exercise and relieved by rest. Continuous pain is produced by sudden arterial occlusion, ulceration, gangrene, inflammation of arteries, veins or lymphatics, and by venous or lymphatic congestion. The intensity of pain varies according to the etiology, degree, and acuteness of circulatory imbalance. The pain of intermittent claudication is usually sharp and often burning in nature. The pain of sudden arterial occlusion is usually sharp when it involves somatic structures and dull and aching in character when it involves the viscera.

**MANAGEMENT**

Cessation of smoking is important.

**Sympathetic blocks:** With local anaesthetics (lumbar sympathetic block, Stellate Ganglion block) may give relief from vasospasm and pain, but it is of little value in relieving claudication. Before proceeding to chemical sympathectomy with alcohol or phenol or RF sympathectomy, several prognostic blocks are given with 0.25% bupivacaine, about 10 ml in volume to predict the long-term effect of prolonged interruption.

**The successful block means:** reduction in pain intensity, improved colour, raised temperature, plethysmographic response and walking tolerance test is used to establish the degree of increase in peripheral blood flow as a result of sympatholysis.

**Analgesics:** non narcotic analgesics, potent narcotics such as morphine are used to control severe pain.

**Epidural infusions:** LA with opioid infusions of bupivacaine with fentanyl is used to control acute attack of vasospasm and pain, but it is of little value in relieving claudication.

**Surgical sympathectomy** is not preferred over percutaneous chemical or RF sympathectomy as the latter is least invasive, done on outpatient basis and has low morbidity.

**Other measures include:** cessation of smoking, adequate control of DM, and weight reduction in obese patients. Foot care, avoidance of exposure to cold, exercise...
program, adjuvant medications, vasodilators, beta blockers, and haemorrhheologic agent.

**Spinal cord stimulation:** PVD is one of the most common indication for SCS. Pain relief of about 60–90% can be achieved by SCS, limb salvage rate is 60–80% and it also improves healing of ischemic foot ulcers.\(^{15–18}\)

**CONCLUSION**

Chronic pain remains a frequent diagnostic feature of many vascular diseases. The control of pain in these disorders is an integral part of managing these patients not only because it mitigates human suffering, but it also allows aggressive physical and occupational rehabilitation to be carried out. The exact incidence and prevalence of PVD has not been studied, but it remains a significant problem among Indian population. Apart from conservative, surgical procedures interventional procedures have a very important role, wherein they not only serve diagnostic but also as a therapeutic purpose in managing pain.

Raynaud’s disease is a benign, episodic digital vasospasm in response to cold or emotional stimuli. All the criteria for diagnosis of Raynaud’s disease (Allen and Brown Criteria, 1932) were present in this young patient. Hence the conservative, pharmacological, and interventional technique (Stellate ganglion blocks)\(^{19}\) showed drastic improvement in the patient’s condition. Detailed evaluation and multidisciplinary approach is mandatory in successful outcome in such case. Excellent results are obtained with interventional procedures and RF denervation in such case, hence one must consider these interventions before the surgical management.

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**REFERENCES**


