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Bilateral uniportal video-assisted thoracoscopic sympathectomy for managing secondary Raynaud's in CREST syndrome: A case report

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ABSTRACT

INTRODUCTION: Secondary Raynaud's is a manifestation that can present in CREST syndrome as a variant of five different diseases: Calcinosis, Raynaud's phenomenon, Esophageal dysmotility, Sclerodactyly, and Telangiectasia. Secondary Raynaud's presents as a result of an imbalance between vasoconstriction and vasodilation potentially leading to tissue ischemia. The mainstay treatment is medical while surgery treatment preserved as a last resort.

PRESENTATION OF CASE: A 28-year-old female presented with secondary Raynaud's and was subsequently diagnosed with CREST syndrome. The patient failed to respond to medical treatment, and gangrene of the right fourth distal phalanx developed. Stellate ganglion block was successfully used as a bridge to surgery. Uniportal video-assisted thoracoscopic surgery (VATS) sympathectomy was performed via a 2-cm incision, the sympathetic ganglia were identified and transected by cauterization at the level of the 3rd, 4th, and 5th intercostal spaces extending for a distance of 5 cm to ensure that the nerve of Kuntz was transected. The postoperative outcome was satisfactory, and the condition of the patient improved in a few months.

DISCUSSION: Different management modalities have been used to relieve the symptoms of secondary Raynaud's. The treatment ranges from lifestyle modification, medical treatment, and lastly surgical intervention. Sympathectomy has been suggested for the management of refractive secondary Raynaud's owing to its considerable clinical response.

CONCLUSION: The use of uniportal VATS sympathectomy results in favorable cosmetic and clinical outcomes including reduced length of hospital stay and postoperative pain.

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1. Introduction

Raynaud syndrome is a condition affecting the arteries through episodes of vasospasm that reduces the blood supply to the distal parts of the extremities which may result in ulceration and gangrene [1]. It is subdivided into two types: idiopathic, previously known as Raynaud's disease or now primary Raynaud's syndrome and secondary Raynaud's syndrome which is often referred to as Raynaud's phenomenon [2]. Secondary Raynaud's is a manifestation of other underlying pathologies or stressors, resulting from an imbalance between vasoconstriction and vasodilation due to

dysregulation of neuronal control and circulating mediators [3]. Despite recent progress in medical management, surgical intervention is often required for the treatment of secondary Raynaud's if medical therapy fails. Herein, we report the case of a young female with Calcinosis, Raynaud's phenomenon, Esophageal dysmotility, Sclerodactyly and Telangiectasia (CREST syndrome [4]) who was treated with bilateral uniportal video-assisted thoracoscopic surgery (VATS) sympathectomy after conservative treatment and medical management failed. The current case report follows the SCARE guidelines [5].

2. Presentation of case

A 28-year-old female presented to the rheumatology clinic of our institute with severe secondary Raynaud's and digital ulcers of the right third and fourth fingers (Fig. 1). She had a history of dysphasia and bilateral arthralgia of the wrist and proximal

Abbreviations: CREST, Calcinosis, Raynaud's phenomenon, Esophageal dysmotility, Sclerodactyly and Telangiectasia; PIP, proximal interphalangeal; VATS, video-assisted thoracoscopic surgery.

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Fig. 1. (Preoperative finding).

A preoperative photograph demonstrating signs of secondary Raynaud's prior to bilateral uniportal video-assisted thoracoscopic surgery sympathectomy.

interphalangeal (PIP) joints. There was no history of respiratory, cardiovascular, or neurological symptoms. Examination revealed digital ulcers with gangrene in the fourth finger of the right hand and sclerosis in both hands. There was no significant past medical or surgical history; further, the patient was not a smoker. The patient's mother was diagnosed with systemic lupus erythematosus, whereas the remaining of the first-degree relatives were medically free.

Laboratory tests revealed microcytic hypochromic anemia with a hemoglobin level of 9.4 g/dL and normal white blood and platelet counts. The anti-centromere antibody level was 1:1280 IU/mL and results for anti-DNA, anti-cardiolipin, anti-Scl-70, anti-SSA, anti-SSB, anti-RNP, and anti-beta-2 glycoprotein-1 antibodies were negative. Upper gastrointestinal endoscopy showed esophagitis with narrowing of the gastroesophageal junction. A high-resolution computed tomography chest scan revealed no abnormal findings.

Treatment with bosentan 125 mg q12 h, prednisone 5 mg, nifedipine 30 mg, calcium 600 mg, vitamin D 2000 IU, omeprazole 20 mg was initiated, after which the patient showed some improvement, but then gangrene developed in the right fourth distal phalanx and amputation was offered. The patient refused amputation and the dried gangrene fell off after some time. She was then offered sympathectomy and was evaluated by an anesthesiologist for participation in a trial of stellate ganglion block as a diagnostic procedure using local anesthetic (5 mL of 0.25% bupivacaine mixed with 1:200,000 epinephrine) injected into the right-side fingers only to prevent bilateral phrenic nerve or recurrent laryngeal nerve paralysis. In the recovery room, the patient developed temporary right-sided Horner's syndrome, which was expected, and the right-hand temperature increased from 25 °C before the procedure to 34 °C after. This represents a 2nd-degree change, which is considered significant, and the color of the fingers changed from bluish to pinkish. All of these changes indicate that a successful block was achieved and lasted for 3–4 h after the procedure which was predictive of intervention adherence and tolerability and successful sympathectomy procedure.

After confirmation of secondary Raynaud's, bilateral uniportal VATS sympathectomy was performed by a thoracic surgeon as follows: double-lumen left intubation was performed with size (36 Fr) and the position was confirmed by fiberoptic bronchoscopy. The procedure is usually performed on the right first, for which the patient is positioned in the left lateral decubitus position, right side up. A 2-cm incision is made through the 5th intercostal space, near the midaxillary line, and through subcutaneous and inter-

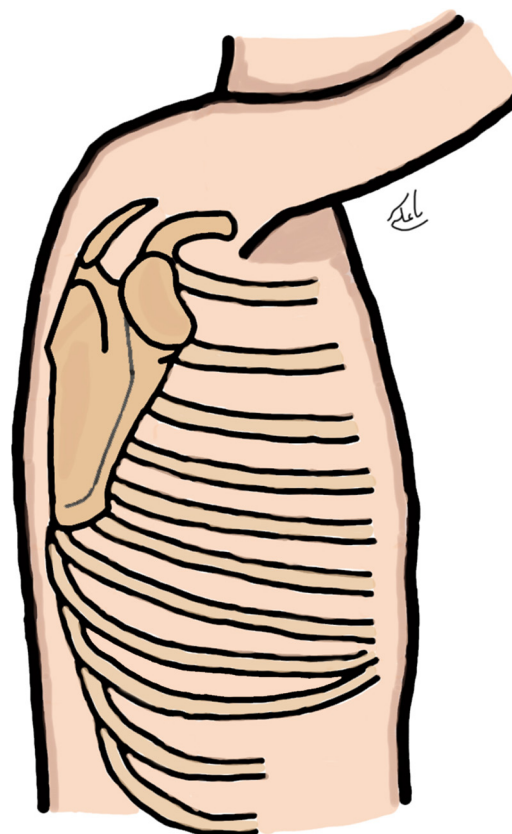


Fig. 2. (Trocar site insertion).

A figure demonstrating the trocar site insertion at the 5th intercostal space near to the midaxillary line (the red line).

costal muscle to open the pleura (Fig. 2). The sympathetic chain was identified below the level of the sympathetic ganglion at the level of the 3rd, 4th and 5th intercostal spaces, and the chain was transected by cauterization extending laterally for a distance of 5 cm to ensure the nerve of Kuntz was transected. Hemostasis was maintained and the lung was inflated by the anesthetist using the Valsalva maneuver at closure of the incision. The patient was repositioned in the right lateral decubitus position to address repeat the procedure on the left side. The postoperative course went smoothly,



Fig. 3. (Postoperative result).

A postoperative photograph shows dramatic improvement in the signs of secondary Raynaud's.

the patient was fully recovered. Incentive spirometry was used, and early mobilization was initiated, with the patient being discharged early. Symptoms disappeared during follow-up and continued to improve satisfactorily for a few months (Fig. 3).

3. Discussion

Raynaud's was first described by Maurice Raynaud in 1862 as "local asphyxia of the extremities", and is considered as the earliest manifestation of CREST syndrome [3,4]. Raynaud's syndrome is subdivided into primary and secondary types [2]; the former occurs without known underlying pathology and is characterized by spasm of the digital arteries, lack of peripheral vascular disease, no evidence of tissue necrosis, normal digital capillary refill, negative antinuclear disease, and lack of complications [3]. However, secondary Raynaud's is associated with underlying connective tissue disease or chemical or physical stress and is characterized by more serious symptoms including the development of digital necrosis and ulceration leading to tissue injury [3]. The prevalence of CREST syndrome is low, and the condition tends to affect women in their 60s and 70s [4].

There have been a number of different management strategies proposed for secondary Raynaud's. Lifestyle modification plays an essential role in decreasing exacerbation of the condition through the avoidance of chemical or physical stressors and treating the underlying connective tissue disease [3]. Medical treatment also makes a major contribution to regulation of the vasomotion through the use of calcium channel blockers, alpha-adrenergic antagonists, angiotensin receptor antagonists, angiotensin-converting enzyme inhibitors, glyceryl trinitrate, selective serotonin reuptake inhibitors, prostacyclin, phosphodiesterase inhibitors, endothelin receptor antagonists, statins, Rho-kinase inhibitors and, lastly surgical intervention [1].

Stellate ganglion block is a minimally invasive procedure performed under ultrasound or fluorescence guidance that is used to treat many medical conditions such as vasomotor symptoms, scleroderma, hyperhidrosis, refractory ventricular arrhythmias, and complex regional pain syndrome [6]. The procedure does carry the risk of complications such as vascular or neuronal injury, thyroid or esophageal injury, pneumothorax, transient Horner's syndrome, infection, and complications associated with intravascular injection [6].

In cases where medical treatment is ineffective, surgical intervention may be considered as the last resort treatment. The use of VATS in sympathectomy offers an approach for the treatment of secondary Raynaud's; uniportal VATS often allows successful, min-

imally intervention surgery, which requires only a small incision (3–4 cm) with no requirement for muscle distribution or rib spreading [7]; in the present case, the port was introduced through a 2-cm incision. The potential benefits of uniportal versus triportal VATS are the reduction of postoperative pain, quicker recovery, reduced length of hospital stay, and favorable cosmetic outcome [7]. Herein, another case report that demonstrated the same effect and result as observed in our case was observed in the trial of uniportal VATS sympathectomy after successful ganglionic block resulted in a positive response with minimal postoperative complication and better cosmetic appearance [8]. A systemic review of sympathectomy for the treatment of digital ischemia showed an initial improvement of digital ischemia in 89% of patients, with improved long-term outcomes [9]. Additionally, 81% of the patients experienced improved healing of ulcers [9]. Moreover, in another study conducted on 30 patients with a median follow-up duration of 40 months, a direct postoperative successful result was observed in 83% of the study participants, with 60% experiencing relapsing symptoms after a while [10]. Furthermore, in a study done on nine patients who underwent endoscopic thoracic sympathectomy for Raynaud's disease, the result of relapse rate was high because 66% of the study participants experienced recurrence of symptoms in 6 months, and the others experienced relapse at 1 year except one [11].

4. Conclusions

Surgical intervention should be considered a last resort for the treatment of secondary Raynaud's. Uniportal VATS sympathectomy offers an effective and safe treatment with favorable cosmetic outcomes, reduced hospitalization, and minimal postoperative pain with patient satisfaction.

Declaration of competing interest

The authors report no declarations of interest.

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Ethical approval

Ethical approval was not required, and patient-identifying knowledge was not presented in the report.

Consent

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Author contribution

- Study concept or design: Yasser Aljehani, Atteia Alhourri.
- Participation in the pre/post-operative management of the patients: Yasser Aljehani, Atteia Alhourri, and Rahma ShahBahai.
- Literature review: Alaa Turkistani and Abdullah AlQatari.
- Drafting of the paper: Alaa Turkistani and Abdullah AlQatari.
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References

- [1] P. Sinnathurai, L. Schrieber, Treatment of Raynaud phenomenon in systemic sclerosis, *Intern. Med. J.* 43 (May (5)) (2013) 476–483.
- [2] S.T. Valdovinos, G.J. Landry, Raynaud syndrome, *Tech. Vasc. Interv. Radiol.* 17 (December (4)) (2014) 241–246.
- [3] M. Baumhäkel, Recent achievements in the management of Raynaud's phenomenon, *Vasc. Health Risk Manag.* 6 (March (1)) (2010) 207.
- [4] A.T. Masi, Preliminary criteria for the classification of systemic sclerosis (scleroderma), *Arthritis Rheum.* 23 (May (5)) (1980) 581–590.
- [5] R.A. Agha, M.R. Borrelli, R. Farwana, K. Koshy, A.J. Fowler, D.P. Orgill, et al., The SCARE 2018 statement: updating consensus surgical case report (SCARE) guidelines, *Int. J. Surg.* 60 (December) (2018) 132–136.
- [6] E. Piraccini, S. Munakomi, K.-V. Chang, *Stellate Ganglion Blocks* [Internet], StatPearls. StatPearls Publishing, 2020.
- [7] D. Nachira, E. Meacci, M. Ismail, D. Gonzalez-Rivas, S. Margaritora, Why to change from multiportal to uniportal VATS? *Video-Assisted Thorac Surg.* 3 (April) (2018), 14–14.
- [8] Y. Aljehani, Bilateral uniportal video-assisted thoracoscopic surgery sympathectomy for managing Raynauds disease, *Int. J. Adv Res.* 7 (July (7)) (2019) 788–789.
- [9] H.M.E. Coveliers, F. Hoexum, J.H. Nederhoed, W. Wisselink, J.A. Rauwerda, Thoracic sympathectomy for digital ischemia: a summary of evidence, *J. Vasc. Surg.* 54 (July (1)) (2011) 273–277.
- [10] T.H. Thune, L. Ladegaard, P.B. Licht, Thoracoscopic sympathectomy for raynaud's phenomenon—a long term follow-up study, *Eur. J. Vasc. Endovasc. Surg.* 32 (August (2)) (2006) 198–202.
- [11] S. Karapolat, A. Turkyilmaz, C. Tekinbas, Effects of endoscopic thoracic sympathectomy on Raynaud's disease, *J. Laparoendosc. Adv. Surg. Tech.* 28 (June (6)) (2018) 726–729.

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