Brachiocephalic Vein Stenosis in Systemic Sclerosis

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Systemic sclerosis (SSc) is a rare disease characterized by widespread collagen deposition resulting in fibrosis.¹ In the literature, central retinal vein occlusion (CRVO)²⁻³ and subclavian vein thrombosis⁴ were reported as venous involvement in SSc.

A 65-year-old female presented with complaints of the prominence of vascular structures gradually in the left upper limb. Her medical history revealed that the patient was diagnosed with SSc for 9 months. She had Raynaud's phenomenon, sclerodactyly, proximal sclerosis, and taking methotrexate (10 mg a week), methylprednisolone (12 mg a day), and nifedipine (30 mg/day). On physical examination, she had telangiectasia on the face and vascular structures were prominent in the left upper limb (Figure 1A). Complete blood count, biochemical parameters, and complement were within normal ranges. Anti-topoisomerase-l was positive and rheumatoid factor, anti-cyclic citrullinated peptide, anticardiolipin immunoglobulin M/G direct Coombs test, and double-stranded DNA antibody were negative.

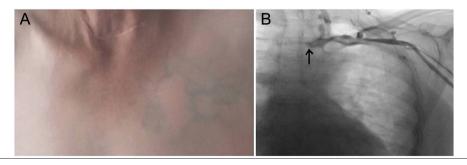
Computerized aortography and upper extremity venography showed stenosis as a thrombus of the left brachiocephalic vein and prominent of the collateral vascular structures (Figure 1B). No hematological malignancy, solid malignancy,

heart failure, kidney, or hepatitis disease was detected in the patient. Antiphospholipid antibodies and thrombophilia panel were negative. The thrombosis was thought to be due to old age and corticosteroid use.⁴ Resting, the elevation of the limb, and warm compresses were recommended as conservative treatments after the detection of brachiocephalic vein stenosis. Written informed consent was obtained from the patient.

Malik et al² reported a case of a 30-year-old male with SSc who developed a sudden decrease of vision in the right eye. He was diagnosed with CRVO. The patient received 3 consecutive intravitreal bevacizumab injections for macular edema. After injections, the best-corrected visual acuity improved from 20/80 to 20/25. Karadžić et al³ reported a 42-year-old SSc patient with acute deterioration of vision in the left eye. This patient was also diagnosed with CRVO. Berriche et al⁵ reported a 56-year-old SSc patient with bilateral subclavian vein thrombosis. Endothelial cell dysfunction which affects the microvasculature system has been mentioned to be the cause of CRVO.² This patient was treated with anticoagulant therapy.

In this article, we report a case of SSc presenting with brachiocephalic vein stenosis. To our knowledge, this is the first SSc

Figure 1. (A) Vascular structures prominence in the left upper limb; (B) upper extremity venography of the left upper limb: stenosis of the left brachiocephalic vein and prominence of the collateral vascular structures.



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patient presenting with brachiocephalic vein stenosis as a rare manifestation. Also, the localization is different from the patients presented in the literature. When the prominence of vascular structures develops in patients with SSc, brachiocephalic vein stenosis should be kept in mind.

Informed Consent: Written informed consent was obtained from the patient.

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