

Paget-Schroetter Syndrome

Manoj Meena¹, Sabarigirivasan Harish¹, Jai Prakash Kewlani¹, Neeraj Gupta¹, Vinay Kumar Meena²

¹Department of Respiratory Medicine, Jawaharlal Nehru Medical College, Ajmer, Rajasthan, India

²Department of General Medicine, SMS Medical College, Jaipur, Rajasthan, India

To the Editor: A 40-year-old male working as a manual labor with a history of smoking presented with a swelling in the neck and right upper extremity with the breathlessness since 5 days. The swelling in the neck region was acute in onset, pain associated with the generalized swelling in a right upper extremity. Breathlessness was acute in onset but was not associated with chest pain. There was no history of cough, fever, trauma and family history of other coagulopathies.

On physical examination, there was tender swelling in right supraclavicular region, around 4 cm × 5 cm in size with cord-like structure extends toward the angle of mandible [Figure 1a] and generalized edema of right upper limb, which was nonpitting in nature [Figure 1b]. Movements of the limb were slightly restricted. Measurements of the right upper limb at the time of presentation was arm – 34 cm, forearm – 22 cm and left upper limb was arm – 30 cm, forearm – 18 cm. Radial pulse in the right upper limb, and other peripheral pulses were felt. Blood pressure was normal. The left upper limb was normal. Examination of other system revealed normal. Routine blood investigations, electrocardiogram, echocardiography and chest X-ray were normal. Contrast-enhanced computerized tomography (CECT) of the chest and soft tissue neck showed the thrombus present in the right subclavian and brachiocephalic vein [Figure 1c and 1d]. It was also known as effort induced thrombus. To confirm it, ultrasonography (USG) Doppler was done, which showed the thrombus in right subclavian vein [Figure 1e]. The patient was started on low molecular weight heparin and planned to put on oral anticoagulant (warfarin) for 6 months with PT-INR monitoring. The patient was improved symptomatically on discharge.

Paget-Schroetter syndrome is a rare condition characterized by the presence of axillo-subclavian thrombus vein in an otherwise normal individual.^[1] Primary upper extremity deep vein thrombosis (DVT) comprises of two categories (a) Paget-Schroetter syndrome, and (b) idiopathic. In contrast to the patient with Paget-Schroetter syndrome, the patient with idiopathic upper extremity DVT has no known trigger or obvious underlying disease. Primarily there is no difference between Paget-Schroetter syndrome and DVT except that former is an idiopathic thrombosis of the axillary or subclavian vein in the upper limb, whereas DVT can occur either in both upper or lower

limb. It is a venous syndrome associated with the thoracic outlet compression. The subclavian vein rests in the groove anterior to the scaleus anterior muscle with the subclavian artery and brachial plexus lying posterior to it. The clavicle runs in close opposition directly over the neurovascular structures. As the clavicle swings backward, the vein, artery, and nerves become compressed like a nut cracker between the clavicle and the first rib, thereby giving rise to a collection of symptoms known as the thoracic outlet compression syndrome.^[1] There is usually temporal and causal relationship between the activities involving hyper-abduction of the upper limb and the genesis of thrombosis. Hyper-abduction and retroversion of the arm, especially with sudden jerky movements can cause repetitive trauma to the endothelium of subclavian-vein^[2] or can strain the subclavian/axillary artery and vein causing it to be crushed between the clavicle and the first rib.^[3] This can cause venous intima damage and initiate venous thrombosis. Significant thrombosis may occur with the repeated insults to the vein wall, especially if mechanical compression by adjoining cervical rib, muscles or fibrous muscular bands is also presents.^[4]

Person involved with strenuous activities such as wrestling, weight lifting, and sports man have the higher risk of developing thrombosis due to repetition injury of the subclavian vein from repeated mechanical compression of the vessels between clavicle, the first rib, and costoclavicular joint.^[5]

A detailed history and thorough physical examinations are often sufficient for suggesting the diagnosis. The Paget-Schroetter syndrome is usually present in younger and otherwise healthy patients, involves the dominant hand, males are more often affected. Symptoms include sudden onset of pain, warmth, redness and swelling in the arm. Diagnoses are confirmed by chest and soft tissue neck CECT and USG-Doppler. Finally, differential diagnoses for this condition are a lymphatic

Address for correspondence: Dr. Manoj Meena,
Department of Respiratory Medicine, Jawaharlal Nehru Medical College,
Ajmer, Rajasthan, India
E-Mail: manojchest@gmail.com

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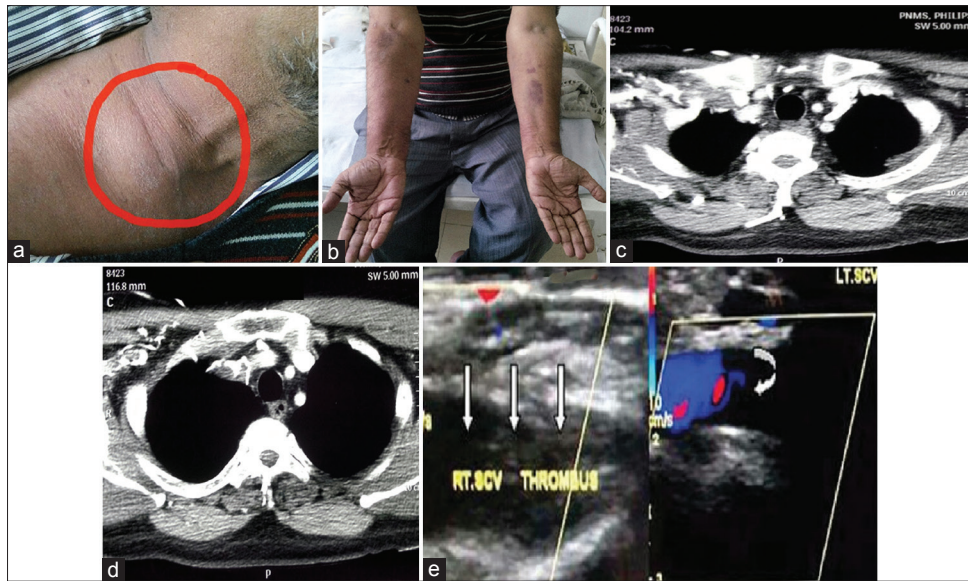


Figure 1: (a) Swelling in right supraclavicular area. (b) Generalized edema of right upper limb. (c) Thrombus in right subclavian vein. (d) Thrombus in right brachiocephalic vein. (e) Ultrasonography Doppler shows thrombus in right subclavian vein (arrows).

obstruction, intramuscular hemorrhage. Anticoagulation therapy is the mainstay of treatment of upper limb DVT. The aim is to prevent the further propagation of thrombosis and maintains the patency of collateral veins. Surgery should be done for the patient with persistent symptoms of thoracic outlet compression syndrome and venous obstruction after an initial period of thrombolysis and click send observation. Therefore, the surgery is not required in all cases of Paget-Schroetter syndrome.^[6]

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Conflicts of interest

There are no conflicts of interest.

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