



Upper Extremity Deep Vein Thrombosis (Paget –Von Schroetter Syndrome) - A Rare Case Report

Authors

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Abstract

Primary "spontaneous" upper extremity deep vein thrombosis is uncommon and is defined as thrombosis of the deep veins draining the upper extremity due to anatomic abnormalities of the thoracic outlet causing axillosubclavian compression and next thrombosis. The syndrome is accurately termed venous thoracic outlet syndrome but is likewise called Paget-Schroetter syndrome, and rather as "effort" thrombosis.¹ An aggressive treatment technique that consists of anticoagulation, catheter-directed thrombolysis or thoracic outlet decompression is done towards relieving acute signs and symptoms and minimizing complications, like recurrent thromboembolism and post-thrombotic syndrome. Here we report a 45 old female, a coolie by profession, came with complaints of left arm swelling and pain for one month duration. On evaluation, patient found to have substantial thrombosis of subclavian, axillary and basilic veins, without any other cause and diagnosed to be primary spontaneous in nature. Patient was then managed with anticoagulants and discharged with the advice of regular follow up.

Introduction

Approximately 1-4% of all instances of deep-vein thrombosis involves upper extremities, ensuing in an annual prevalence of 1-2 instances consistent with 1,00,000 people.^{2,3} Upper extremity Deep vein thrombosis (UEDVT) commonly refers to thrombosis of the axillary and/or subclavian veins.⁴ It is classified as primary or secondary on the basis of pathogenesis. Primary upper extremity deep vein thrombosis is defined as thrombosis of the deep veins draining the upper extremity due to an underlying anatomic anomaly on the thoracic outlet inflicting compression or repetitive damage to the underlying axillosubclavian vein.⁵⁻⁸ Primary upper extremity deep vein thrombosis is a

manifestation of venous thoracic outlet syndrome. Thrombosis of the veins draining the upper extremity was initially postulated to be the cause of acute arm pain and swelling by Paget,⁹ and later Von Schroetter related the medical syndrome particularly to the axillary and subclavian veins.¹⁰ This clinical entity became known as Paget-Schroetter syndrome.¹¹ In the mid-twentieth century, the term was changed into "effort" thrombosis,¹² because of the fact that the syndrome often took place in physically active people after an unusual strenuous activity of the arm and shoulder.¹³⁻¹⁵ The term "spontaneous" upper extremity venous thrombosis has also been used, highlighting the frequently dramatic

presentation in an otherwise healthful, younger man or woman. For the purpose of our discussion, we refer the syndrome as primary upper extremity deep vein thrombosis to differentiate it from secondary causes, which are associated with inciting elements which includes indwelling catheters or prothrombotic states. Anatomic abnormalities of the thoracic outlet that bring about compression of the vein may be congenital or acquired. Congenital anomalies include cervical ribs, supernumerary muscles, ordinary tendon insertions, or unusual muscular or tendinous bands.^{16,17} Acquired abnormalities consist of bony overgrowth because of bony fracture (eg, clavicle, first rib)¹⁸⁻²¹ or hypertrophy of anterior scalene muscle or subclavius muscle tissues, regularly associated with repetitive lifting.

Anatomic abnormalities narrows the scalene triangle, or more commonly the costoclavicular space, predisposing the vein to compression between the first rib and muscle or tendon or between anomalous tendon insertions. Under a few circumstances, it appears that an anatomic abnormality is not essential to produce damage to the vein.²² Extremes in range of movement of the upper extremity can lead to movement of the clavicle relative to the first rib sufficient to cause venous compression. Repetitive overhead arm movements or hyperabduction and external rotation of the shoulder are most usually implicated.²³⁻²⁵ Repetitive injury causes perivenous fibrosis, which eventually leads to thrombosis.

Compression of the venous structures that traverse the thoracic outlet occurs in two distinct areas: the scalene triangle and the costoclavicular area.

Scalene triangle – The anterior border of the scalene triangle is formed by anterior scalene muscle which originates from the transverse processes of the 3rd through 6th cervical vertebrae (C3-C6) and inserts on the internal borders and superior surfaces of the first rib. The posterior wall of the scalene triangle is formed by the middle scalene muscle, which arises from the transverse procedures of the second through 7th cervical vertebrae (C2-C7) and inserts widely onto

the posterior border of the first rib. The superior border of the first rib forms the floor of the scalene triangle. The trunks of the brachial plexus and the subclavian artery pass between the anterior and middle scalene muscle tissues, whilst the subclavian vein goes anteromedial to the scalene triangle.

Costoclavicular space – The costoclavicular space incorporates the area between the primary rib and the clavicle. The brachial plexus, subclavian artery, and subclavian vein pass via this space. The subclavian vein is most probably to be compressed at this site, ultimately leads to thrombosis. It is essential to apprehend that the patient frequently presents with acute onset of signs and symptoms associated with the thrombosis, however the underlying trouble may be a persistent repetitive injury that had narrowed the vein.

The occurrence of UEDVT is less than that of the lower extremity DVT probably because:

- Fewer, smaller valves are present within the veins of the lower extremity,²⁶
- Bedridden patients usually have much less cessation of arm actions compared to leg actions,
- Less hydrostatic pressure in arms,
- Increased fibrinolytic activity that has been seen inside the endothelium of the upper arm as compared to the lower arm.^{27,28}

Absence of a similar soleal network of veins might also make contribution to the lesser occurrence of UEDVT. The prevalence of thrombosis within the upper extremity is commonly found within the subclavian vein (18–67%), then axillary (5–25%) and the brachial vein(4–11%)²⁹⁻³¹ with marked predilection for the left side, probably due to anatomical reasons as explained elegantly by Stephen et al., in 1979.³¹ The main complication remains mortality, recurrent thromboembolism and the worrisome post-thrombotic syndrome. Complications of deep-vein thrombosis, which are less common in the upper extremities than lower extremities, includes pulmonary embolism (6% for upper

extremities vs. 15 to 32% for lower extremities),³² recurrence after 1 year (2 to 5% for upper extremities vs. 10% for lower extremities),³² and the post-thrombotic syndrome (5% for upper extremities vs 56% for lower extremities).³² Thrombosis of the axillary, subclavian veins (in comparison with thrombosis at other places) and residual thrombosis at 6 months are associated with an increased risk of post-thrombotic syndrome.³²

Management of UEDVT

The targets of treatment are to relieve the signs of deep-vein thrombosis of an upper extremity and to prevent thrombus progression and its complications like early recurrence, pulmonary embolism, and the post-thrombotic syndrome.³² Strategies consist of anticoagulation therapy, thrombolysis, mechanical catheter interventions, and surgical approaches. Anticoagulation Therapy with low-molecular-weight heparin was associated with a lower rate of recurrent venous thromboembolism at 6 months, death and massive bleeding. Initial anticoagulation treatment for that reason usually includes low-molecular-weight heparin; The use of vitamin K antagonists for 3 to 6 months, is typically encouraged for all types of anticoagulation treatment in patients with deep-vein thrombosis of an upper extremity, except in patients with cancer, for whom low-molecular-weight heparin is preferred. Mechanical Catheter-directed thrombolysis like aspiration, fragmentation, thrombectomy, balloon angioplasty, or stenting need to be considered in patients with recent onset of symptoms with massive swelling and functional impairment of the arm and who are at low risk for bleeding after anticoagulant therapy or thrombolysis. In case of anatomic abnormality, surgical decompression of the venous thoracic outlet is done by means of resection of the primary rib with the aid of the transaxillary route or the use of supraclavicular or infraclavicular incisions. Other methods like resection of the costoclavicular ligament, anterior scalenectomy, and venolysis can also be done.

Postoperative complications include hemopneumothorax, injury to long thoracic nerve or the phrenic nerve, wound hematoma requiring reoperation, and recurrent subclavian thrombosis.³² General protocol includes a multidisciplinary technique to the treatment of patients with deep-vein thrombosis of an upper extremity, wherein patients without a or mild symptoms are dealt with anticoagulation alone and patients with massive swelling or functional impairment of the arm are commonly treated with both catheter-directed thrombolysis and anticoagulation therapy. Patients are reassessed between 1 and 3 months, if symptoms of post thrombotic syndromes or evidence of residual vein stenosis confirmed via positional phlebography are present then surgical decompression of the thoracic outlet with optional percutaneous balloon angioplasty is usually done. Consideration of SVC filter placement have to be generally considered in patients with a contraindication to anticoagulation therapy and to those with thrombus progression or symptomatic pulmonary embolism despite good enough remedy with anticoagulants. Guidelines from Professional Societies Consensus and from the American College of Chest Physicians offer detailed recommendations for the management of deep-vein thrombosis of upper extremity.³²

Case Report

45 year old female came here with complaints of dysphagia, easy fatiguability, pain, swelling over the left arm for one month. The signs have been insidious in onset and gradually progressive for one month. She is a coolie by occupation. On examination, pallor was present, local examination showed swelling over left arm till elbow joint. It was not warmth, mild tenderness was present. No history of any surgeries, OCP consumption, no previous records of hospitalization, no history of recurrent abortions or any large family history. Investigations confirmed Hb- 7.4 gm/dl, peripheral smear shows dimorphic anemia, UGI scopy showed pale mucosa with pan gastritis, Doppler left arm

confirmed the presence of thrombosis in left subclavian, axillary, basilic vein with echogenic luminal contents, non compressible. As the cause for DVT was not known and to rule out hypercoagulable states, BT, CT, PT/INR, antithrombin III, protein C, protein S activity were analysed and all had been found to be within normal limits. As diverse studies revealed, occult lung carcinoma can also present with higher extremity DVT, chest Xray, HRCT thorax was done showing no abnormality except bilateral axillary subcentimetric lymphadenopathy and bilateral cervical ribs without significant transverse procedure. Abdominal organ screening was done seeking out any occult cancer, which turned out negative. ECG was found to be within normal limits. Henceforth, cause of upper extremity DVT was considered to be primary spontaneous in nature probably Paget von schrotter syndrome after ruling out all different possible causes.

Conclusion

In patients with acute discomfort or swelling of the arm, with coexisting risk factors like vigorous arm exercise, an implanted central venous catheter or pacemaker, or a past history of deep-vein thrombosis or cancer, the diagnosis of deep-vein thrombosis of an upper extremity is to be considered. Anticoagulation therapy should be immediately initiated; Catheter-directed thrombolysis have to be taken into consideration in cases of clinically massive deep-vein thrombosis. After initial anticoagulation, oral anticoagulation treatment is suggested for the subsequent 3 to 6 months.

Here, we managed this patient with intravenous unfractionated heparin for the first five days followed by oral anticoagulation. The patient became symptomatically better on the time of discharge. The patient was also advised for regular follow up to monitor the outcome.

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