Effort-Induced Venous Thrombosis of the Upper Limbs

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Introduction

Spontaneous thrombosis of the upper extremity was postulated by Sir James Paget in 1875 as acute pain and swelling of the arm. Von Schrötter in 1884 was the first to relate the clinical syndrome to thrombotic occlusion of the axillary and subclavian veins. Spontaneous thrombosis of the upper-extremity veins was subsequently termed Paget-von Schrötter syndrome. The term effort-induced thrombosis was coined in 1960 [1]. We report 3 patients presenting with a diagnosis of effort-induced thrombosis of the deep venous system of the upper limb.

Case Reports

Case 1

A 42-year-old Egyptian manual worker with no known previous medical problems presented with a 2-week history of fever, shortness of breath, and increasing swelling of the neck which occurred after lifting heavy objects as a part of his job. On examination, he

Key Words
Effort-induced venous thrombosis • Thrombosis, upper limbs • Subclavian vein

Abstract

Objective: To report 3 patients who presented with effort-induced thrombosis of the upper limbs. Case Presentations and Intervention: The 1st patient presented with a 2-week history of fever, shortness of breath, and increasing swelling of the neck after strenuous manual work. The 2nd patient presented with a 2-day history of pain, swelling, and discoloration of the right upper limb following a session of intense weight lifting. The 3rd patient presented with a 2-day history of swelling, pain, and discoloration of the left upper limb due to repeated rotary movements of the arm at work. In all 3 cases, the diagnosis of effort-induced thrombosis of subclavian and axillary veins was made. In the 1st case, the lesion was bilateral and complicated by superior vena cava obstruction. In the 2nd and in the 3rd case, right and left subclavian veins were thrombosed, respectively. After consulting the vascular team, anticoagulation therapy was initiated in all 3 cases. The patients described showed gradual recovery with eventual recanalization of the thrombosed veins on follow-up. Conclusions: These cases indicate the need to consider thrombosis of the subclavian vein as a part of the differential diagnosis in a patient with a history of strenuous manual work. Therapeutic options include immediate anticoagulation or thrombolysis, while vascular surgery should remain an option for cases with underlying anatomical anomalies.
was noted to have a low-grade pyrexia with a plethoric complexion and congested neck and upper chest veins (fig. 1). The lungs were clear and fundi normal.

A clinical diagnosis of thoracic outlet obstruction was made. A chest X-ray showed slight widening of the mediastinum and a small left pleural effusion. A chest CT scan showed extensive thrombosis of superior vena cava and right internal jugular and left innominate veins, with no evidence of a mediastinal mass. Urgent upper-limb venography revealed bilateral thrombosis of subclavian and innominate veins. Both axillary veins were patent. All blood investigations, including coagulation profile and subsequent thrombophilia and immunology screens, were within normal ranges. Echocardiography showed no extension of the thrombus into the right atrium, and a CT scan of the abdomen and a gallium-67 citrate scan ruled out occult malignancy or infection.

The diagnosis of effort-induced thrombosis of bilateral subclavian veins complicated by superior vena caval obstruction was confirmed. The vascular team was consulted, and it advised anticoagulation therapy. Thrombolysis was not initiated because of the patient’s late presentation.

The patient was treated with infusion of unfractionated heparin. An initial dose of 80 U/kg followed by continuous infusion of 18 U/kg/h aimed at obtaining 2–2.5 times the baseline activated partial thromboplastin time, as determined by frequent monitoring. After 1 week, the patient was on long-term oral warfarin for 6 months. Follow-up visits were scheduled every 3 months, and at each visit, the patient underwent clinical evaluation. Venography was done at the first visit and ultrasonography of both upper limbs at the subsequent visits. The symptoms improved gradually, the follow-up evaluations showing good recanalization and no evidence of recurrence or postthrombotic venous insufficiency.

Case 2
A 37-year-old healthy Kuwaiti bodybuilder presented with a 2-day history of pain, swelling, and discolouration of the right upper limb following a session of intense weight lifting. Clinical examination revealed a swollen, tender right upper arm with palpable pulses. Ultrasonography showed a thrombus in the right subclavian vein, with patent brachial and axillary veins. A chest CT scan ruled out any compressive lesion, and a thrombophilia screen was normal.

He was subcutaneously treated with low-molecular-weight heparin (nadroparin 85 IU/kg) for 7 days, followed by oral warfarin for 6 months. Follow-up every 3 months with venography and then ultrasonography of the affected limb after 6 months showed full clinical and radiological recovery with a patent vein.

Case 3
A 43-year-old diabetic Jordanian presented with a 2-day history of swelling, pain, and discolouration of the left upper limb. He related the symptoms to the electrical work he was doing which required repeated rotatory movements of the arm. Examination showed a swollen left arm with bluish discolouration which was markedly tender over the biceps tendon. An ultrasound scan ruled out haematoma or evidence of a ruptured biceps tendon. A color duplex scan confirmed the diagnosis of left subclavian and axillary vein thrombosis. Both CT scan of the chest and thrombophilia screen were normal, and he showed a good response to anticoagulation therapy with resolution of the thrombus on follow-up imaging. The same therapeutic protocol and follow-up as for case 2 was used for this patient.

Discussion
Most patients presenting with venous thrombosis of the upper extremity can be divided into two categories: one group includes those with primary disease associated with repetitive active use of the upper limbs or anatomic abnormalities known as Paget-von Schröter syndrome [2]; the other category includes patients with secondary thrombosis due to recognized thrombotic risk factors such as compressive lesions or invasive venous catheter use [3].

Deep vein thrombosis (DVT) of the upper extremities is estimated to be present in 1–4% of all cases [4]. Primary upper-extremity DVT is recognized in 30% of the cases [5].

The patients presented in this report did not have any history of any invasive intervention or thrombotic risk factors and hence were diagnosed as having primary effort thrombosis. Primary effort thrombosis often presents as a dramatic unexpected event in healthy young individuals, usually after strenuous use of arm and shoulder, manifesting with dull, aching pain in shoulder and/or axilla and swelling of arm and hand. Common precipitating factors include repetitive arm use, hyperextension of shoulder, and rotatory movements of the arm.

Fig. 1. Neck swelling and congested neck and upper chest veins in patient 1.
activities involve hyperabduction and external rotation of the arm or backwards and downwards rotation of the shoulder. Causative activities may include playing cricket, tennis, or baseball, weight lifting, or chopping wood. Strenuous movements lead to intimal injury of the axillo-subclavian vein which initiates thrombosis [6].

Previous reports [7] have suggested that there is generally an underlying compressive anomaly of the thoracic outlet. Compression of the vein between the first rib and a hypertrophied scalene or subclavian muscle or between tendons of these muscles, compression between the clavicle and a cervical rib, and partial occlusion of the vein by a congenital web have also been reported [3, 7]. The abnormalities of the thoracic outlet are often bilateral and predispose to eventual thrombosis of the contralateral venous system [7, 8]. Embolic complications occur in about 36% of the patients with Paget-von Schrötter syndrome [9]. However, none of our 3 cases had embolic complications.

Postthrombotic syndrome occurs in almost 25% of the patients within the first 2 years and is related to residual thrombosis and, to a lesser degree, to the extent of the initial thrombosis [10]. The syndrome can result in occupational disability, especially in those individuals whose livelihood depends on the ability to perform vigorous physical activity, while chronic compression can lead to perivenous fibrosis. Superior vena cava syndrome, venous gangrene, loss of venous access, and/or brachial plexopathy are other possible complications [11], as in our 1st case. A recent study [5] showed that the recurrence rate of upper-extremity DVT was low with 1.6%, but that the rate tends to be higher in patients with thrombophilia.

The optimal management of this disease is not standardized and remains controversial. Therapeutic options include anticoagulation, thrombolysis (systemic or local), and surgery. There are currently no randomized controlled trials comparing the use of anticoagulants with thrombolytic or surgical management. There is general acceptance of initial thrombolytic therapy (systemic or catheter directed, if expertise is available) and anticoagulation to rapidly restore patent veins [2, 7, 8].

Thrombolytic therapy is acceptable, if it is started within the first 5–7 days of presentation. On the other hand, there are no agreed criteria on the indication or timing of surgery, with some vascular teams preferring immediate thrombolytic therapy, followed by prompt thoracic outlet decompression, while others adopt a more selective approach, offering surgery only to patients with persistent symptoms following initial thrombolytic and anticoagulant therapy, followed by a period of close observation [7, 12].

Most investigators agree that patients who benefit most from surgery are those with persistent symptoms of thoracic outlet compression and venous obstruction after an initial period of thrombolysis and close observation [13, 14].

In our patients, thrombolysis was not initiated in case 1 due to his late presentation; the other 2 cases showed a favourable response to anticoagulation with remarkable improvement, subjectively and objectively, with venography showing patent veins.

Conclusions

This report indicates that there is a need to consider primary effort-induced venous thrombosis of the upper limb as part of the differential diagnosis in otherwise healthy young patients presenting with thoracic outlet obstruction and having a history of strenuous manual work. The diagnosis should be promptly confirmed and immediate anticoagulation or thrombolysis initiated, while vascular surgery remains an option for those cases with an underlying anatomical abnormality.
References


