C A S E R E P O R T

Paget-Schroetter syndrome

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Paget-Schroetter syndrome, or 'effort' thrombosis of the axillary-subclavian vein, is an uncommon deep vein thrombosis usually caused by excessive upper limb activity. It may cause post-thrombotic syndrome, leading to significant disability if not treated appropriately. The optimal management for this syndrome is still controversial because the outcomes of different treatment strategies are based on case studies and series in small numbers. We report a case of Paget-Schroetter syndrome in a young male weight-lifter and discuss treatment strategies suggested by the current literature.

Introduction

Primary axillary-subclavian vein thrombosis was first described by Paget in 1875 and von Schroetter in 1884 and was named the 'Paget-Schroetter syndrome' by Hughes in 1948.[1-3] Its aetiology is complex and multifactorial. It is most often the consequence of chronic compression of the subclavian vein at the level of the thoracic outlet.[4] Optimal management of this syndrome is still controversial because the outcomes of different treatment strategies have been reported in case studies and small series only. We report a case of Paget-Schroetter syndrome in a young male patient who had been weight-lifting regularly for 5 months and discuss treatment strategies in the light of the current literature.

Case report

An 18-year-old male was admitted in January 2003 with a painful swelling of the right arm for more than 4 weeks. He had been exercising regularly, usually weight-lifting, for 5 months, in order to apply for a sports academy. He had been diagnosed as having a soft tissue infection and treated with cefuroxime axetil and an analgesic anti-inflammatory drug (flurbiprofen) 10 days earlier, in another medical centre. On physical examination, oedema and dilated superficial veins were noted in the right upper limb. Brachial, radial, and ulnar pulses were palpable and both Adson's sign and the hyperabduction sign were negative. The arterial blood pressure was 120/70 mm Hg in both upper limbs. No lymphadenopathy was detected. There were no respiratory, cardiovascular, or abdominal abnormalities. Routine laboratory tests—including a coagulation profile, chest X-ray, and echocardiography—were within normal limits. A Doppler ultrasound of the right upper limb showed a thrombosis of the right subclavian vein (Fig 1). Venous angiography of the right upper limb confirmed the diagnosis. Ventilation-perfusion scintigraphy of the lungs revealed normal ventilation with no perfusion defects.

The patient was given urokinase with a loading dose of 250 000 U by bolus then 200 000 U/h was given intravenously for 24 hours. Partial dissolution of the thrombus was noted. He was then given intravenous unfractionated heparin followed by oral warfarin. The patient's symptoms improved and he was discharged on the 10th day of hospitalisation. The international normalised ratio was kept at 2.0-2.5. He was then referred to the thoracic surgery unit for evaluation of the need for decompression of the thoracic outlet. A magnetic resonance imaging study of the upper limb revealed no masses or bone structures causing external venous compression and the decision was made to manage him conservatively. Oral anticoagulants were continued for 12 months. He was followed up every 3 months in the first year and then every 6 months for another 2 years. Doppler ultrasonography was performed at every visit. The patient's right arm oedema improved gradually and he had no complaints after the third month of treatment, with no evidence of post-thrombotic syndrome. A Doppler ultrasound of the right upper limb revealed a patent subclavian vein at the 9-month visit (Fig 2).

Discussion

Leg and pelvic veins are the most common sites for deep vein thrombosis. Paget-Schroetter syndrome, or 'effort' thrombosis of the axillary-subclavian vein is a relatively uncommon condition. It develops more frequently in the dominant arm, usually after sports activities, including long

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Paget-Schroetter綜合徵

Paget-Schroetter綜合徵，又稱腋—鎖骨下靜脈運動血栓，是一種特別的深靜脈血栓，通常由上肢過度活動造成，可引致血栓形成後綜合徵，假如沒有妥善治療，會使患者失去相當的活動能力。至於治療本症最有效的方法目前仍未有定案，因為不同治療方案的結果都是來自個別病例和人數少的病例系列。本文報告一位年輕男性所患的Paget-Schroetter綜合徵，患者經常有舉重活動。本文亦討論目前文獻所建議的治療方案。

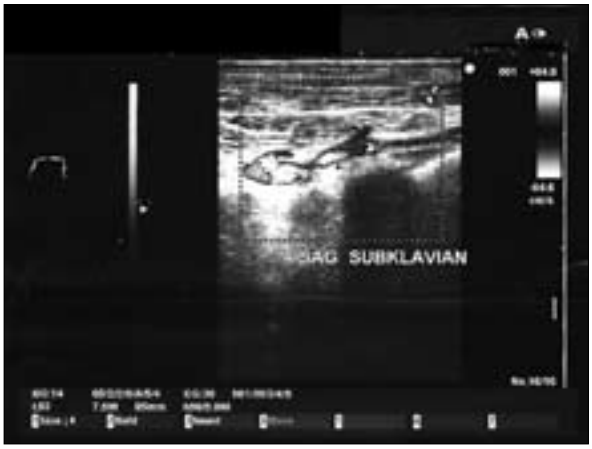

FIG 1. Doppler ultrasonographic image showing thrombus in the right subclavian vein

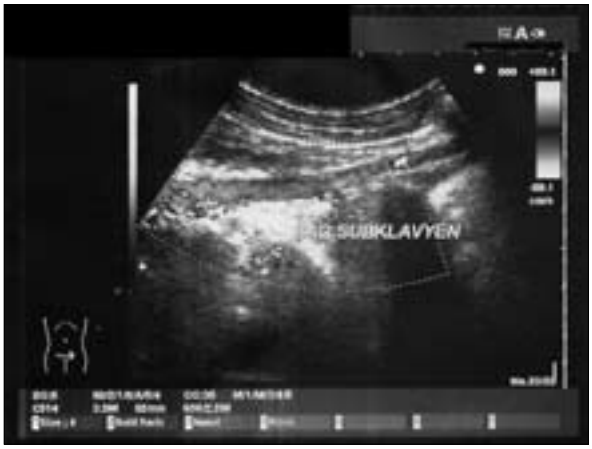

FIG 2. Doppler ultrasound revealing patency of the right subclavian vein in the ninth month of treatment

distance swimming, body building exercises, wrestling, handball, and baseball throwing. Most of the activities are vigorous, strenuous, or last for a long time.[5,6] It is caused by the strain put on the axillary-subclavian vein by retroversion or hyperabduction of the arm. This process causes microtrauma of the venous intima and the damaged endothelium initiates thrombus formation, ultimately blocking the vein.[6] The most common symptoms are painful congestion and oedema of the arm. The clinical presentation can easily be confirmed by Doppler ultrasonography. A contrast venogram is the standard means of determining the anatomy and can be done in preparation for interventions such as catheter-directed thrombolysis and percutaneous transluminal angioplasty (PTA).[7]

Upper limb deep vein thromboses are more frequently associated with occult malignant disorders than those in the lower limb.[8] Screening for a possible underlying malignancy, including a clinical history, physical examination, complete blood count, blood sedimentation rate, basic biochemistry including hepatic and renal function tests, a chest X-ray, and an abdominal ultrasonography, was performed in our patient. There was no evidence of malignancy.

In Paget-Schroetter syndrome, the earlier the diagnosis and treatment, the better the results.[9] The treatment goals are to relieve the acute symptoms of venous occlusion, prevent pulmonary embolism, reduce the likelihood of recurrent thrombosis, and avoid development of the post-thrombotic syndrome.[10] The treatment for this condition includes non-surgical and surgical interventions. Thrombolysis and anticoagulation are the mainstay of treatment. Early diagnosis provides an opportunity for rapid venous recanalisation with an effective thrombolytic therapy. It has been suggested that the optimal period for thrombolytic treatment is within 6 weeks of the thrombosis.[11] The general consensus is that urokinase or recombinant tissue plasminogen activator should be used for thrombolysis.[7] Anticoagulants are used to prevent further deposition of thrombus, allowing an established thrombus to stabilise and to undergo endogenous lysis, reducing the risk of recurrent thrombosis.[12]

Decompression of the thoracic outlet can be performed later, using various techniques. These include first-rib and/or clavicle resection after conservative treatment, especially in cases of proven extrinsic venous compression and persistent or recurrent symptoms. Transaxillary resection of the first rib is the most popular surgical intervention. Urschel and Razzuk[9] suggested that prompt transaxillary first-rib resection and neurovascular decompression can be safely and effectively performed after thrombolytic treatment. If this approach fails to re-establish patency, leaving some residual arm disability, axillary-subclavian vein revascularisation can provide good mid-term results.[4]

Vascular procedures including PTA, 'Roto-rooter' techniques, and venous stenting have also been used to treat this syndrome. Percutaneous transluminal angioplasty after first-rib resection and venous stenting

in patients with short-segment venous strictures that persisted after PTA, produced long-term patency and symptomatic relief in a report by Kreienberg et al.[13] The 'Roto-rooter' technique is a minimally invasive endovascular procedure used for mechanical removal of the thrombus or the plaque from the arterial or venous walls by using a rotational catheter. The occlusion material is shaved away by vibration of the tiny blades at the tip of the catheter and is sucked into the catheter head. Urschel et al[9,11] reported that thrombolysis or 'Roto-rooter' techniques have not been successful after 3 months. They did not recommend the use of stents in the treatment of this condition.

Post-thrombotic syndrome is a common complication of upper limb deep vein thrombosis with a frequency ranging from 7 to 46% (weighted mean, 15%).[14] There is currently no validated, standardised scale for assessing upper limb post-thrombotic syndrome, and little consensus on the optimal management of this condition. Quality of life is impaired in patients with upper limb post-thrombotic syndrome, especially if the deep vein thrombosis was in the dominant arm.

In conclusion, Paget-Schroetter syndrome should be considered a possible cause of painful swelling of the upper limb, particularly in young and active patients with excessive arm activity. It may cause post-thrombotic syndrome resulting in significant disability if not treated appropriately. There is no consensus on the optimal management for this condition. Treatment choices should be based on the cause of the thrombosis and should be individualised. A multimodal approach including thrombolytic treatment, anticoagulation, vascular techniques, and surgical thoracic outlet decompression (in selected patients) can effectively restore venous patency and avoid the development of post-thrombotic syndrome.

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