CASE REPORT

Paget-Schrotter syndrome and complications of management

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SUMMARY

An 18-year-old man presented to the emergency department with an acutely swollen right upper limb having spent the previous day canoeing. Venography confirmed right subclavian venous thrombosis at middle one-third of right clavicle with no evidence of cervical rib or other structural abnormalities. Following heparinisation, catheter directed thrombolysis was performed which restored luminal flow. Postprocedure the patient developed spontaneous small-volume haemoperitoneum and acute renal failure. The patient was admitted to the intensive care unit for haemodialysis and supportive management. The patients' renal function improved and he was discharged well following his 28th day of admission. Paget-Schroetter syndrome or effort thrombosis involves subclavian venous thrombosis associated with strenuous activity of upper extremities. In these cases, catheter directed thrombolysis with first rib resection provides greater patency rate than anticoagulation therapy alone. Acute kidney injury following mechanical thrombolysis is rarely reported in the literature.

BACKGROUND

Paget-Schroetter syndrome (PSS) or effort thrombosis involves subclavian venous thrombosis associated with strenuous activity of upper extremities. It is uncommon with an incidence of 1–2/100 000 population. PSS accounts for 10–20% of upper extremity deep venous thrombosis (DVT) and up to 40% of spontaneous axillary-subclavian venous thrombosis (ASVT).

DVT of the upper limb can be treated by a variety of modalities including anticoagulation, systemic thrombolytics and catheter-directed thrombolytics (CDT). Anticoagulation therapy is the treatment of choice for mild, asymptomatic cases of DVT, specifically low-molecular-weight heparin (LMWH). However, anticoagulation therapy alone may not be optimal treatment in patients presenting with larger, symptomatic DVT owing to increased risk of pulmonary embolism (PE). There exists a paucity of best practice treatment guidelines in this setting, and further randomised controlled trials are required to inform guidelines for optimal management.

CASE PRESENTATION

We describe the case of an 18-year-old man who presented to the emergency department (ED) with a 2-day history of right upper limb swelling. This was associated with mild generalised arm discomfort without numbness, paraesthesia or weakness. There was no history of external limb trauma;

however, the patient had spent the preceding day canoeing. He was haemodynamically stable with a heart rate of 78 bpm and a blood pressure of 135/80 mm Hg. On examination he was found to have pitting oedema of the right arm extending from his hand to axilla. There were visibly dilated the superficial veins. Ulnar, radial, brachial and axillary pulses were palpable and capillary refill time was less than 2 s. He had full and pain-free range of motion of his wrist, elbow and shoulder joints with no sensory or neuromuscular deficit.

Biochemical and haematological investigations were carried out. Full blood count revealed haemoglobin, white cell count and platelet count within normal range. Similarly renal profile was normal with creatine of 76 ng/mL. D-dimers were not raised at 0.29 µg/mL (normal range 0.01–0.50 µg/mL). Radiological investigations on presentation to the ED included chest X-ray which showed no cervical rib or bony injury. The primary differential diagnosis was venous thrombosis of the right axillary or subclavian vein. Intravenous heparin infusion was started at 1000 iu/h following a 5000 iu bolus.

Subsequently a right upper extremity venography was performed via the right basilic vein. This study confirmed a right subclavian venous thrombosis at the level of the middle one-third of right clavicle with delayed filling of the superior vena cava and collateral venous formation (figure 1). The patient was scheduled for CDT. An Angiojet catheter was used to pulse spray a combination of tissue plasminogen activator (t-PA) and heparinised saline. Suction thrombectomy was performed using this device in combination with balloon venoplasty, following which there was a successful restoration of

Figure 1 Right upper extremity venography via right basilic vein; right subclavian vein thrombosis with collateral venous formation.

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luminal flow. No residual thrombus was evident on postprocedure check angiography.

One day following CDT, the patient reported of central chest pain, abdominal discomfort, nausea and vomiting. Biochemical and haematological investigations were repeated. A decrease in haemoglobin was noted with haemoglobin of 10.9 g/dL. The patient was noted to be newly thrombocytopenic at this stage with a platelet count of $82 \times 10^9/L$. In addition, a repeat renal profile demonstrated acute kidney injury (AKI) with a creatine of $291 \mu\text{mol/L}$. CT of thorax, abdomen and pelvis were performed. This study demonstrated no obvious intrathoracic pathology or large pulmonary embolus. However, a small volume of free intra-abdominal fluid was visible at the inferior pole of liver indicative of spontaneous haemoperitoneum secondary to thrombolysis.

The patient was admitted to the intensive care unit at this stage for specialist multidisciplinary care and haemodialysis following further deterioration of renal function. The patient's renal function improved and haemodialysis discontinued after 5 days. Oral anticoagulation therapy was initiated with warfarin. The patient was discharged home after 28 days in hospital with resolution of symptoms at the time of discharge. MR venography at 1 month postdischarge revealed a patent axillary, subclavian and brachiocephalic vein (figure 2).

DISCUSSION

Effort thrombosis, also known as PSS, refers to ASVT associated with strenuous and repetitive activity of the upper extremities with an incidence of 1–2/100 000 population.¹ We describe the case of an 18-year-old man with no other predisposing factors who developed acute AVST following a canoeing trip. PSS accounts for 10–20% of upper extremity DVT and up to 40% of spontaneous ASVT.² Patients age at presentation is predominantly in the 30s, with a male-to-female ratio of 2:1. In the majority of cases (80%) the dominant arm is affected.

The pathogenesis of PSS has been considered by Thompson *et al*³ who proposed mechanisms of development following repetitive trauma. They suggested that hypertrophy of the pectoral muscles causes extrinsic compression of the vein. They also reported that repetitive trauma damages the venous wall leading to thickening and fibrosis, and that damage to the intima specifically leads to a rough, thrombogenic surface. Other anatomical abnormalities such as cervical ribs, congenital bands and variation in the insertion point of the costoclavicular ligament are also thought to be risk factors for PSS.⁴

DVT of the upper limb can be treated by a variety of modalities including anticoagulation, systemic thrombolytics and CDT. Anticoagulation therapy is the treatment of choice for mild,

asymptomatic cases of DVT.⁵ Compared with unfractionated heparin, LMWH has a lower risk of recurrent thromboembolism and allows for a shorter inpatient stay.⁶ Oral anticoagulation is also started before discharge from hospital for a period of up to 6 months.⁷

Anticoagulation therapy alone is not felt to provide appropriate therapy for patients presenting with larger, more symptomatic DVT.⁵ It is suggested that in such cases consideration is given to the use of thrombolytics. With anticoagulation alone symptoms of post-thrombotic syndrome (PTS) are reported in 41–91% while 39–68% can suffer from permanent disability.⁸ Acute PE is reported in 6–15% of patients with anticoagulants alone.⁸ Early CDT therapy (within 10–14 days) resolves upper limb DVT and prevents bleeding complications and PTS when compared with anticoagulants alone.⁹ CDT achieves a luminal patency of at least 50% after 6 months follow-up.¹⁰ Systemic thrombolytics such as t-PA, recombinant human-tissue-type plasminogen activator and alteplase cannot be given in instances of recent trauma or surgery, gastrointestinal bleeding and history of a bleeding disorder.¹¹ Systemic thrombolytics also have the disadvantage of being diffusion dependent because the rate of fibrinolysis is influenced by the degree of obstruction of the thrombus.¹²

There is evidence to suggest that elective first rib resection may be beneficial in this patient cohort in reducing recurrence. Urschel and Razzuk¹³ reported a 95% rate of excellent or good outcomes in 199 patients treated with thrombolysis and subsequent first rib resection. Similarly a recent study reported a 100% patency and 0% rate of significant symptoms at 5 years after thrombolysis and first rib resection for acute effort thrombosis.¹⁴ Owing to the rarity of this condition no randomised control trials available to definitively guide treatment.

AKI following mechanical thrombolysis occurs rarely. The AngioJet device destroys thrombi by trapping and hydraulic recirculation. This process results in the lysis of red blood cells.^{15 16} Haemolysis following AngioJet has been reported to cause a transient increase in serum creatine levels.¹⁷ Haemoglobin by-products such as haeme have pro-inflammatory effects, while high amounts of haeme proteins (haemoglobin and myoglobin) can cause oxidative stress leading to nephrotoxicity.¹⁸ The exact mechanism of nephrotoxicity by haemoglobinuria may be unknown but it is thought that haeme proteins precipitate cast formation leading to obstruction of renal tubules resulting in acute tubular necrosis.¹⁹ The subject of this case report experienced dramatic deterioration in his renal function immediately following thrombolysis and a further disimprovement following urgently performed CT studies of thorax, abdomen and pelvis with intravenous contrast.

Figure 2 Follow-up MR venogram 12 weeks postcatheter directed thrombolysis; patent right subclavian vein without stenosis.

Learning points

- ▶ This case demonstrates a classical presentation of Paget-Schroetter Syndrome in an 18-year-old man. His management was complicated by dialysis dependent acute kidney injury from a combination of catheter-directed thrombolysis (CDT) and radiological contrast medium.
- ▶ CDT with first rib resection provides greater patency rate than anticoagulation therapy alone.
- ▶ Current treatment is based on small series and case reports. Randomised controlled trials are needed to provide an evidence base to inform the optimum management of this rare pathology.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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