Rare disease

Paget–Schroetter syndrome: the importance of urgent referral and appropriate management

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Summary

The authors present the case of a young healthy male who was admitted with a case of acute primary subclavian vein thrombosis, otherwise known as Paget–Schroetter syndrome. The patient was successfully treated by catheter directed thrombolysis using a Trellis device and thoracic outlet decompression following prompt referral to the vascular team. The case highlights importance of early referral in the prevention of the long-term sequelae which can occur as a result of this rare condition.

BACKGROUND

Paget–Schroetter syndrome (PSS) is an acute, primary thrombosis of the upper extremities specifically involving the axillary and subclavian veins.^{1 2} Thrombus of the upper extremities accounts for 1–4% of all cases of deep vein thrombosis (DVT) and primary axillary thrombosis accounts for only 10–20% of this already small proportion of cases, making Paget–Schroetter syndrome a rare and unusual condition.^{3–5} If treated conservatively with anticoagulation alone, PSS is a potential cause of significant long-term morbidity and as such should be referred early to vascular surgeons for aggressive treatment.⁶

CASE PRESENTATION

A previously fit and well 18-year-old male student presented to the accident and emergency department with a swollen, painful and discoloured left arm; his symptoms had commenced 3 days prior to his presentation but had subsequently worsened. He had no risk factors for DVT, performed no recent strenuous activity or sustained any trauma to his arm. Clinical examination revealed swelling extending from the shoulder to his hand with prominent dilated superficial veins.

INVESTIGATIONS

An ultrasound scan of his left arm revealed thrombus in the left subclavian vein. He was commenced on an intravenous heparin infusion and referred to the vascular surgeons. A contrast CT scan showed thrombus in the left subclavian and axillary veins, with extension into the left brachiocephalic vein. Multiplanar reconstruction of the CT scan showed a significant narrowing of the left costoclavicular junction. Based on the clinical history and imaging, a diagnosis of Paget–Schroetter syndrome was made.

TREATMENT

The patient underwent a venogram (figure 1) and catheter directed thrombolysis using a Trellis device. The Trellis device allowed the thrombus to be isolated in a thrombolytic zone between a proximal and distal balloon on the

catheter; 10 mg of recombinant tissue plasminogen activator was infused initially over a 10 min period followed by aspiration of clot and repeated for a further 10 min period.

A postprocedure venogram revealed an irregular vein wall overlying the 1st rib and the presence of a short, tight stenosis at the costoclavicular junction. An 8 mm conquest venoplasty was performed to the area of stenosis which established good venous flow.

Subsequently, the patient underwent a 1st rib resection via a trans-axillary approach to resolve the venous impingement caused by the narrowed costoclavicular junction.

OUTCOME AND FOLLOW-UP

The procedure was uneventful and the patient was discharged 7 days later on a 6 month course of warfarin anticoagulation therapy.

DISCUSSION

PSS is regarded as an ‘effort thrombosis’. The rationale behind this nomenclature lies in the underlying pathogenesis: a reduced costoclavicular space due to anatomical abnormalities at the thoracic outlet causes the subclavian vein to be compressed; this results in endothelial distortion of the subclavian vein. Each time ‘effort’ is exerted on the affected side, it causes a resultant cycle of further endothelial trauma, leading to hyperplasia and fibrosis which subsequently causes venous stasis, thrombus formation and further crowding in the costoclavicular space.^{7 8} When the subclavian vein is totally occluded, the forming thrombus can extend into the axillary vein.

Classically, PSS affects previously fit and well young males, in their dominant hand, it is typically preceded by prolonged, strenuous exertion of the upper limb. Despite being rare, it is not surprising that this syndrome is regarded as one of the most common vascular conditions arising in healthy athletes.⁹ However, given the underlying anatomical abnormalities innocuous day-to-day activities can also precipitate thrombosis.^{10 11}

It is important that these rare cases are recognised and referred to the vascular surgeons in the earliest instance.

Figure 1 Venogram showing thrombus in the left subclavian vein.

The optimal treatment window is approximately 2 weeks, beyond this period progressive deterioration can occur and cause significant severe sequelae and permanent disability.⁷

Pulmonary embolism, recurrent thrombosis and post-thrombotic syndrome are among the complications. Post-thrombotic syndrome is perhaps the most debilitating – it manifests as chronic arm swelling, heaviness, pain and early exercise fatigue.^{3 4 7 10} Given the young patient demographics, one can see that there are potentially serious implications for function, future employment and general quality of life.

Long-term data has illustrated an unacceptably high proportion of residual symptoms, complications and long-term physical impairment. Review of case series data suggests that as many as 75% of patients developed long-term consequences, making it evident that conservative management with limb elevation and anticoagulation alone is unsatisfactory.⁴

A consensus exists to employ a multimodal approach to treating PSS; it encompasses thrombolysis in combination with thoracic outlet decompression. A study by Molina *et al* of 126 patients presenting with PSS over a 23-year period has suggested that this combination of treatment for an acute presentation can result in 100% vein patency with no sequelae.⁷ Thrombolysis promptly restores venous patency and thus minimises ongoing endothelial insult.¹²

In our case, we opted to use a method of catheter directed thrombolysis using a Trellis device. The Trellis device allowed us to isolate the thrombus and minimise the quantity of thrombolytic agent used to achieve successful dissolution of the clot.¹³ Catheter directed thrombolysis has been shown to achieve high rates of clot resolution, but

also has the advantage of reducing the risk of incurring the haemorrhagic complications of systemic thrombolysis.¹⁴

The current literature does not offer a consensus on the appropriate time for thoracic outlet decompression surgery to be performed, as such it remains that both immediate and delayed surgery (6–12 weeks post-thrombolysis) are acceptable approaches.⁴ In our case, a first rib resection was performed within 48 h of thrombolysis. Our rationale for this approach was to minimise the insult to the revascularised vessels and break the pathological cycle which caused the initial thrombus formation.

Learning points

- ▶ This case has highlighted that DVT of the upper extremities are both rare and important. The conservative management with anticoagulation that is typically employed with DVT is not appropriate in cases of Paget–Schroetter syndrome. As such, it is important to identify these cases early and refer promptly to vascular surgeons for active management.
- ▶ Thrombolysis is a key component of the treatment, it results in early venous recanalisation and thus prevention of chronic side effects including post-thrombotic syndrome.
- ▶ Catheter directed thrombolysis has been illustrated to be an invaluable method of thrombolysis achieving good results and simultaneously minimising systemic side effects.

Competing interests None.

Patient consent Obtained.

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Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Fenton D, Constantinou J, Srilekha A, Richards T, Harris P. Paget-Schroetter syndrome: the importance of urgent referral and appropriate management. *BMJ Case Reports* 2011;10.1136/bcr.04.2011.4115, date of publication

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