Rare disease

Duplicated inferior vena cava with associated ilio-femoral deep vein thrombosis

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Summary

We present the case of a fit and well 20-year-old gentleman who presented to our emergency department with unilateral lower limb pain and swelling. Subsequent imaging revealed a left ilio-femoral deep vein thrombosis, with associated duplication of his inferior vena cava. He was treated conservatively with a heparin infusion, warfarin and compression therapy prior to being discharged following a short inpatient stay.

BACKGROUND

The Department of Health estimates that 25 000 patients die annually in English hospitals as a result of a venous thromboembolism. Congenital venous anomalies have been identified as a risk factor in the development of deep vein thrombosis (DVT) in the young though only very few cases have been reported.¹ By publishing such a rare case, it is hoped that guidelines will eventually be designed to optimise both its management in the acute setting and in the longer term.

CASE PRESENTATION

A 20-year-old gentleman presented to A+E with a 2-h history of left lower limb cramps with associated swelling, which had started suddenly while walking. He was known to have a dysplastic right kidney and was otherwise fit and well, taking no medication. Five weeks previously he had flown back to the UK from Egypt following a holiday but there were no other known risk factors to promote a DVT, including no family history.

On examination, he was comfortable at rest having received morphine for severe limb pain. His observations were normal and he complained of no other symptoms apart from those in his left leg. This limb appeared significantly swollen and mottled along its length with a blue discoloration of the toes, though the skin temperature was palpably normal. Sensation was intact but diminished and motor function was preserved. Pulses in the left lower limb were impalpable (most likely due to the degree of swelling) compared with being easily palpable on the contralateral limb. A provisional diagnosis of a DVT was made and the patient was commenced on a heparin infusion.

INVESTIGATIONS

All haematological (Hb 14.2 g/dl, INR 1.0) and biochemical tests were normal on admission. ECG showed sinus rhythm and biphasic flow was elicited in both lower limbs with a bedside doppler. A venous duplex confirmed a left femoral DVT extending into the external iliac vein (long

saphenous vein was patent). A CT was organised to determine the extent of the thrombosis.

This showed duplication of the inferior vena cava (IVC) with (figures 1–4):

- ▶ A thin right IVC ascending to the level of the right renal vein. The right internal and external iliac veins were distended.
- ▶ A patent, distended left IVC draining into the left renal vein. The internal and external iliac veins were of normal calibre, with the known ilio-femoral thrombosis extending to the level of the confluence.
- ▶ Collateral veins crossing from above the right iliac confluence, behind the aorta and into the left IVC (figure 5).
- ▶ A known hypoplastic right kidney, with compensatory hypertrophy on the left.
- ▶ A hypoplastic aorta and iliac arteries.

Thrombophilia screen, including lupus anticoagulant, antithrombin III, protein S, protein C and factor V Leiden were negative.

TREATMENT

In addition to the prescription of compression stockings, the patient was commenced on warfarin. The heparin infusion was ceased once the INR had reached a therapeutic level (2–3) for 2 consecutive days prior to discharge. His anticoagulation therapy is initially planned to continue for a period of 6 months, following which the authors intend to re-image the venous system.

DISCUSSION

Duplication of the IVC has previously been reported to have an incidence of 0.2–3%,² and the majority of cases are identified incidentally during imaging for various reasons. This duplication is a result of a failure in the embryological formation of the IVC. This complex process involves three pairs of veins and takes place between the sixth and tenth weeks of gestation.³

The author has identified only 13 previous cases of a duplicated IVC with associated DVT in the literature (age

Figure 1 CT slice displaying a duplicated IVC.

Figure 2 Imaging of the large left renal vein crossing the midline, with a small right renal vein and associated hypoplastic kidney.

Figure 3 A thrombus is clearly seen in the left iliac vein.

Figure 4 Pelvic imaging revealing four iliac veins.

range 19–81). In all previous cases, the right IVC is noted to be wider than the left, making this case the first of its kind. Interestingly, a recent American case⁴ describes managing a similar scenario with thrombolysis, thrombectomy and

stenting of a popliteal vein occlusion and both IVCs. Such dramatic variation in management highlights the fact that there are currently no guidelines to advise on the management of DVTs associated with IVC anomalies.

Figure 5 Schematic reconstruction of the altered venous anatomy.

Chee *et al* identified IVC anomalies in 5–6.7% of patients (<40 years) presenting with a DVT; a figure much higher than the expected rate, suggesting an increased risk. Some suggest that any anomaly in the IVC promotes thrombus formation due to venous stasis. However, Anne *et al*⁵ believe that any correlation between a duplicated IVC and DVT is purely incidental.

Learning points

- ▶ Consider venous anomalies in patients with unexplained DVTs, particularly in the young.
- ▶ If the placement of an IVC filter is deemed appropriate, these patients require one in each system.
- ▶ Surgeons and radiologists must be cautious. Several adverse events have been reported in the literature due to a failure to appreciate these anatomical variations.

Competing interests None.

Patient consent Obtained.

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