Recurrent lower limb venous thrombosis associated with a congenitally absent infrarenal inferior vena cava

Case report

A 17-year-old man presented with marked lower limb swelling, found to be secondary to spontaneous thrombosis of the left common iliac vein, right external iliac vein and infrarenal inferior vena cava (IVC). Consequently he was anticoagulated with warfarin for 6 months and fitted with Grade 2 compression stockings. Subsequently, he re-attended 7 years later with another spontaneous right sided above knee deep vein thrombosis. Extended duration anticoagulation with warfarin was commenced.

Thrombophilia screening showed only that he was heterozygous for the factor V Leiden mutation. Interestingly however, An Magnetic Resonance (MR) venogram demonstrated congenitally abnormal venous vasculature, with a complete absence of his infrarenal IVC (Figure 1a) and his suprarenal IVC reconstituted from the suprarenal veins. In addition, extensive collateralization was seen within the abdomen, abdominal wall and lower extremities (Figure 1b) and multiple small non-occlusive thrombi were noted in the lower limbs. Furthermore, lower limb lymphatic drainage was also absent, contributing to his post thrombotic syndrome.

Congenital abnormalities of the venous vasculature should be considered in younger patients with apparently spontaneous venous thrombosis. Whilst contrast venography remains the gold standard, MR venography offers a useful, less invasive alternative to imaging the venous system. Despite the absence of the infrarenal IVC in this patient, it is likely that significant alternative routes of embolization still persist through his enlarged collateral vessels. Anomalous venous drainage may also serve to increase the risk for developing post thrombotic syndrome. This should be discussed with patients, together with the importance of extended use of compression stockings.

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