ANSWER TO PHOTO QUIZ (PAGE 420)

A RARE CAUSE OF ABDOMINAL MASS AND DEEP VENOUS THROMBOSIS

DIAGNOSIS

The computed tomography scan and MRA showed absence of inferior vena cava. Left renal hypoplasia was also noted and a renal perfusion study demonstrated decreased blood flow to this kidney. The right common iliac drained into the azygous vein and the left common iliac drained into hemizygous vein. The deep venous thrombosis (DVT) was initially treated with heparin and then bridged to warfarin, but he only attended follow-up in the clinic intermittently and had a poor medication compliance. After 4 years, a repeat ultrasound showed a subacute left femoral thrombus with persistence of the right iliac thrombus, so he was advised to continue warfarin therapy. Because follow-up and medication compliance proved challenging for this patient, he was offered treatment with a direct oral anticoagulant, rivaroxaban, when he was 25. For the past 3 years, he has been stable with no clinical evidence of a new DVT.

DISCUSSION

Inferior vena cava agenesis (IVCA) is a rare abnormality[1] that predisposes patients to DVT. In the absence of any associated symptoms or visceral defects, patients are generally undiagnosed unless the condition is discovered as an incidental finding. When symptomatic, patients typically present with lower extremity swelling and pain, and are subsequently diagnosed with unprovoked DVT.

The increased risk of DVT in patients with IVCA is most likely multifactorial. In these cases, the veins of the lower limbs drain into the azygous and hemizygous veins which have smaller lumens. Thus, there is congestion and stasis with consequently an increased risk of DVT. In addition to this anatomical predisposition, patients with IVCA may have an increased incidence of other prothrombotic conditions such as protein C deficiency, factor V Leiden mutation, and hyperhomocystenaemia.[2] The development of new thromboses, as in our patient, emphasises the importance of meticulous follow-up.

Renal aplasia is commonly reported in IVCA with right renal aplasia being more common as the right metanephros drains directly into the IVC[3] while the left metanephros drains into the left gonadal vein which drains into the lumbar perforators. There are some reports of left renal agenesis with IVCA[4] but these are uncommon and the cause is less clear.

Ultrasound is the preferred imaging modality to diagnose DVT but CT or MRI is ideal to identify IVC abnormalities. The dilated collaterals in these patients pose diagnostic challenges as, when in the abdomen or thorax, they can even mimic malignant masses.[2] Appropriate imaging of these patients is vital to formulate treatment plans.

There are no recommended guidelines for the treatment of IVCA and previously reported therapies have ranged from chronic anticoagulation to surgical reconstruction of the IVC. Anticoagulation with warfarin is the most commonly used treatment modality. Given the continued predisposition to thrombus, these patients require lifelong anticoagulation.[5]

In conclusion, a high index of suspicion for IVCA is advised especially for the younger patient who presents with an unprovoked DVT. Patients require lifelong anticoagulation due their predisposition to thrombosis, and as such, require close follow-up in the outpatient setting.

REFERENCES

1. Ruggeri M, Tosetto A, Castaman G, Rodeghiero F. Congenital absence of the inferior vena cava: a rare risk factor for idiopathic deep-vein thrombosis. Lancet. 2001;10;357:441.
2. Gayer G, Luboshitz J, Hertz M, et al. Congenital anomalies of the inferior vena cava revealed on CT in patients with deep vein thrombosis. Am J Roentgenol. 2003;180:729-32.
3. Gayer G, Zissin R, Strauss S, et al: IVC abnormalities and right renal aplasia detected on CT: a possible link? Abdom Imaging. 2003;28:395-9.
4. Iqbal J, Nagaraju E. Congenital absence of inferior vena cava and thrombosis: a case report. J Med Case Reports. 2008;2:46.
5. Sagban TA, Grotemeyer D, Balzer KM, et al. Surgical treatment for agenesis of the vena cava: a single-centre experience in 15 cases. Eur J Vasc Endovasc Surg. 2010;40:241-5.