Short Report

Deep Vein Thrombosis in Azygos Continuation

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Introduction

Agenesis of the inferior vena cava is rare. The blood flow to the right heart is via collaterals via the paravertebral plexus and the azygos and hemiazygos veins. This is called azygos continuation (AC) or azygos continuation syndrome. Within these collaterals venectasis may imitate lymphoma and cause difficulties in differential diagnosis. AC is generally asymptomatic; however, within the last five years two patients with deep venous thrombosis (DVT) together with azygos continuation were treated in our hospital.

Case Reports

Patient A

A 24-year-old male presented with a phlebographically verified isolated DVT in the left common iliac vein following immobilisation due to pain in the left iliosacral joint. There was no previous medical history and no vascular catheterisation at any time. At computer tomography (CT) scan of the abdomen, performed to exclude venous compression proximal to the thrombosis, multiple retroperitoneal masses were seen and an abdominal lymphoma was suspected (Fig. 1). To exclude a lymphoma and a subsequent iliac compression with DVT a laparotomy with biopsy was performed. There was no obvious vascular pathology seen. Histology of the lymph nodes was benign. The subsequent magnetic resonance scan (MR) showed complete absence of the inferior vena cava. The blood from iliac veins returned via the paravertebral plexus and the azygos and hemiazygos veins to the superior vena cava and right heart. The renal veins drained into the paravertebral plexus (Fig. 2) and the liver veins drained into the right atrium. The spleen was normal. The patient was treated with heparin and then orally anticoagulated for six months. The follow-up for almost five years has been uneventful and the left iliac vein recanalised.

Patient B

A 27-year-old female smoker taking ovulation inhibitors presented with a phlebographically verified isolated DVT of the left common iliac vein. There was no previous medical history and no vascular...
catheterisation at any time. The abdominal CT, performed to exclude venous compression proximal to the thrombosis, suggested absence of the vena cava and retroperitoneal masses which were suspected to be venectases. The MR confirmed absence of the suprarenal inferior vena cava. The backflow of the venous blood was via the paravertebral plexus and the azygos and hemiazygos veins. To exclude thrombosis central to the iliac vein, bilateral ascending transfemoral phlebography was performed. Phlebography showed the thrombosed left iliac vein, a normal infra renal vena cava, an absent suprarenal inferior vena cava, a large paravertebral venous plexus and the azygos/hemiazygos system to the right heart (Fig. 3). The liver veins drained normally into the existing last centimetres of the inferior vena cava to the right heart. With the ascending phlebography and repeat MR, a lymphoma could be excluded without performing a laparotomy. Haematological tests revealed an anti phospholipid antibody deficiency. Due to this second factor the patient was started on long term oral anticoagulation. There have been no more symptoms for two and a half years.

Discussion

Azygos continuation is rare.\(^1\)\(^-\)\(^5\) It is described mainly in case reports or in the context of congenital heart disease (with an incidence of 0.5%) and the polysplenial syndrome.\(^4\) The enlarged paravertebral plexus, the azygos–hemiazygos system and the multiple venectases can mimic an abdominal lymphoma. To diagnose azygos continuation and exclude a lymphoma, an abdominal CT, MRI and phlebography may be necessary. A surgical procedure for abdominal lymph node biopsy may thus be avoided.

In both cases a DVT was the main symptom. To our knowledge there are 19 cases of DVT in AC described, including our two cases. In almost all of these patients there is one or more risk factor for DVT described, such as immobilisation after trauma or postoperatively, a haematological disorder, forced diuresis or the intake of ovulation inhibitors. Considering that almost all these patients with DVT in AC had a risk factor for a DVT and only nineteen cases are described it seems likely that AC is a functional equivalent to a normal venous anatomy. In a few cases of DVT\(^1\)\(^-\)\(^3\) in AC a venous thrombectomy or a lysis was performed or an AV fistula created. A successful treatment for a leg ulcer in AC is described: a 13-mm prosthetic bypass had been inserted from the external iliac vein to the intrathoracic azygos.\(^4\) Venous decompressing procedures for enlarged spinal veins causing neurological symptoms are described as well.\(^5\)

To summarise AC may mimic abdominal lymphoma. CT and MR or phlebography can exclude lymphoma and avoid laparotomy. In general symptoms and complications in AC are rare. Treatment of complications is recommended as well as a careful prophylaxis for DVT, especially when a risk factor is present.
References


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