Congenital vena cava inferior agenesis
Thrombosis of the deep veins of the lower extremities with concomitant iliac, portal and mesenteric vein thrombosis

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Summary
A patient aged 25 years with lower extremity deep vein thrombosis in the course of vena cava inferior agenesis is described. This very rare malformation can be connected with chronic venous insufficiency or thrombotic complications. In such cases other congenital abnormalities should be excluded and suspicion of vena cava inferior agenesis should always be taken into consideration in young patients with idiopathic deep vein thrombosis.

In 2002 a man aged 25 years was admitted to the Department of Surgery (Specialist Hospital in Tychy) because of a right hypogastric region pain. The reported ailments started a few days before. No history of previous surgery, trauma, immobilisation or any data indicating hereditary coagulation disorders was found. According to the physical examination, the presence of a large, irregular and painful tumour in the right hypogastric region was reported. There were no signs of peritonitis and laboratory investigations did not reveal any abnormalities. In the ultrasound examination, a heterogenous tumour containing liquid spaces in the right hypogastric region was found.

In the CT scan of the above-mentioned pathological structure in the right retroperitoneal space, a tumour (size: 7 × 6 cm) was present. The continuity of an infiltration between the ascending colon and tumour was documented and the presence of enlarged lymphnodes in the tumour surrounding was described. Additionally, in the same investigation, iliac vein thrombosis and right kidney urosthisis was found.

The patient was treated conservatively (bed rest, antibiotics, anti-inflammatory drugs, anticoagulants). The further course was complicated by the right leg oedema that occurred 7 days after admission. Because of suspicion of periappendix abscess, the patient was qualified for surgery. Intraoperatively, a tumour consisting of multiple thrombosed vessels in the right retroperitoneal space was found. Due to its usually uncomplicated course, it is accidentally recognized during procedures connected with retroperitoneal space exploration or in radiological investigations performed for other reasons (4). The frequency of VCI malformation ranges from 2 to 5%. Usually the cases of VCI duplication or cases of left inferior caval vein are recognized (3, 13).

According to the small number of reported cases, agenesis of VCI is one of the rarely described congenital pathologies. We describe the case of a young man with lower extremity deep vein thrombosis in the course of VCI agenesis.
hypogastric region was recognized. There were no changes concerning other abdominal cavity organs and the diagnosis of the previously suspected abscess was excluded.

In the angio-CT (performed postoperatively), the absence of the vena cava inferior or below the hepatic vein confluence was recognized (Fig. 1). The lack of the inferior caval vein in this region was compensated by the presence of collateral circulation along the vertebral column and also inside the vertebral canal (anterior and lateral to the spine). The collateral circulation was also present within the anterior wall of the abdominal cavity and in the posterior compartment dilated azygos veins were described.

Additionally, a thrombus inside the mesenteric, hepatic and right iliac and femoral veins was documented. Intraoperatively, in the right hypogastric region enlarged and thrombosed veins were found. In the colour coded doppler examination thrombosis of the portal, mesenteric, femoral and iliac veins was confirmed. The patient was treated with unfractioned heparin for 10 days (continuous infusion according to aPTT elongation) and subsequently with low molecular weight heparin administration. Additionally, antibiotics and non steroid anti-inflammatory drugs were continued. A decrease of the leg circumference and local condition improvement (pain release) were observed. The patient was discharged. In the CT performed 7 weeks later a partial recanalization of the portal and right iliac vein was documented. Because of the very slow progress of the recanalization, after CT examination, the long term anticoagulant treatment with oral anticoagulants was started and is still continued.

Discussion

The reasons for vena cava abnormalities are embryogenesis disturbances, usually between the 6th and 8th week of pregnancy. Embryogenesis of the major vein system is related to the proper development of the conjunctions between 3 pairs of embryonal veins. Various anatomical variants are possible. Agenesis is a very rare vascular malformation and can occur in the form of isolated pathology or can be combined with other disturbances (6, 12, 15) such as:

- congenital heart diseases,
- situs inversus,
- multiple spleen,
- agenesis of the portal vein or
- hypoplasia of the biliary tract.

Few cases of the lack of VCI complicated by deep vein thrombosis are described (11, 15). The blood outflow from the lower extremities leads through the compensatory enlarged azygos, lumbar and para-vertebral veins.

The pathology discussed is usually asymptomatic and the diagnosis is made accidentally during surgery or radiological investigations performed because of other disease. The occurrence of deep vein thrombosis can be related to coagulation disorders and presence of some risk factors. In cases of insufficient collateral circulation development, agenesis of the VCI can be an important deep vein thrombosis risk factor. According to the data reported by Obernoster et al., 16% of the investigated cases with deep vein thrombosis revealed VCI abnormalities, especially among young patients (9, 10). The rate of thrombosis recurrence in this group is significantly higher (9). According to others, deep vein thrombosis of the lower extremities seems to be one of the most common clinical manifestations (14, 17).

In these cases, the presence of iliac vein thrombosis is very often recognized and the complaint usually concerns young patients (<30 years) without any additional risk factors (10, 15). Some very surprising reports are described, suggesting the diagnosis of other pathology such as haematuria, colica renalis or abdominal cavity complaints including acute abdomen ailments (1, 11). Massive dilation of the collateral circulation in the thorax can result in haemaphysis related to bronchial vein rupture (1, 7). On the other hand, widening of the mediastinum can suggest a development of myeloproliferative diseases (16).

In the presented case the manifestation of disease was rather unusual, too. Finally, it should be emphasized that abnormalities of the VCI cannot always be diagnosed by means of ultrasound and CT, which are the methods of choice in the diagnosis of uncomplicated deep vein thrombosis. This may explain the relatively rare diagnosis of this congenital pathology. That is why, in some patients younger than 30 years with idiopathic deep vein thrombosis and with no risk factors, additional investigations should be proposed, e.g. angio-CT or angio-MR.

In cases of deep vein thrombosis related to agenesis, long-term anticoagulant treat-
ment should be prescribed. There are still doubts concerning asymptomatic patients with the above-mentioned malformation and uneventful course (without deep vein thrombosis). In the literature two cases of surgical treatment of this pathology are described. In one case, venostasis was treated by means of bypass from the external iliac to the azygos vein (15). In the other case, venous thrombectomy in a deep vein thrombosis patient with arteriovenous fistula was performed to increase flow through collateral circulation (8).

For our patient, the diagnosis of his pathology was realized after thrombosis had occurred. The advanced thrombosis of the peripheral vessels with their occlusion and the lack of the proper preoperative diagnosis excluded the possibility of the surgical treatment and restoration of the outflow from the lower extremities (e. g. by means of the saphenous vein extra-anatomical bypass from the femoral to the subclavian vein or by the anatomical reconstruction to the patent caval vein above the diaphragm).

Conclusions

Agenesis of the VCI is a very rare malformation. Its presence may be connected with chronic venous insufficiency or thrombotic complications. In such cases other congenital abnormalities should be excluded. Suspicion of VCI agenesis should be taken into consideration in young patients with idiopathic deep vein thrombosis. Because of the high risk of DVT recurrence, symptomatic patients with deep vein thrombosis and VCI agenesis should receive long term anticoagulant treatment.

References


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