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Agenesis of the Inferior Vena Cava: A Case Report

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Abstract: Congenital anomalies of the Inferior Vena Cava (IVC) are rare and result from aberrant development of the venous system during embryogenesis. The authors describe a case of a 23-year-old man with complete absence of the inferior vena cava. He was admitted to the emergency room with edema and pain of his left lower limb. A doppler ultrasound revealed thrombosis of the left iliac, femoral and popliteal veins. The patient was treated with low molecular weight heparin and discharged with oral acenocumarol therapy. The three main influences in the pathogenesis of thrombosis are: Abnormalities of the vessel wall, blood components and dynamics of flow (Virchow triad, described over 100 years ago). An anomalous inferior vena cava should be suspected in patients 30 years of age or younger who have thrombosis involving the iliac veins.

Key words: Inferior Vena Cava (IVC), agenesis, thrombosis

INTRODUCTION

Venous system development is a complex process, during which several malformations may occur, such as absence or duplication of some sections, malpositions, etc. During embryogenesis, a process involving the development, regression and anastomosis of three sets of paired veins forms the inferior vena cava: the posterior cardinal, subcardinal and supracardinal veins (Obernosterer *et al*., 2002). If the originally paired structures do not unite, anomalies of the inferior vena cava may result.

Estimated prevalence of the anomalies of inferior vena cava in the general population is about 0.07-8.7% (Yigit *et al*., 2006). Agenesis of the IVC has an incidence of 0.0005-1% in the general population (Gil *et al*., 2006). This anomaly was first described by Abernethy in 1793 and the first series was reported by Dwight in 1900 (Gavin *et al*., 2003).

Frequently the diagnosis is merely coincidental, during abdominal surgery or radiological procedures, the patient not having any clinical symptoms; however, anomalies of the IVC may present themselves with deep venous thrombosis of the iliac or femoral veins (Vermeulen and Van Urk, 1996).

Until the advent of angiocardiography all reports were based on autopsy studies, thus, it was not until the early 1950s that clinical reports were seen (Gavin *et al*., 2003).

We present a case of deep venous thrombosis in a young healthy men: the abdominal CT scan showed absence of the IVC.

CASE REPORT

A 23 year old caucasian man was admitted after 3 days of left limb pain and oedema. He was a healthy non-smoker student, without previous trauma or surgery. There was no similar episode in the past, no family history of hypercoagulation and he was taking no medication.

Physical examination revealed oedema of the left limb; peripheral arterial pulses were normal. The routine laboratory investigations showed no abnormalities. Coagulation studies (antithrombin, protein C, protein S, lupus anticoagulant, thrombin time, FV, FVIII: C, FXI and antiphospholipid antibodies) did not show thrombosis predisposing. Eco-doppler ultrasound scan revealed a thrombosis of the left iliac, femoral and popliteal veins.

Chest X-ray was normal. Toraco-abdominal computed tomography scan and subsequently, Angio CT scan showed absence of the entire IVC, enlarged azygous and hemiazygous veins (Fig. 1a and b) hepatic veins entering directly into the right atrium (Fig. 2a, b and c) and dilated paravertebral venous collaterals (Fig. 3a and b). A malformation of the IVC was diagnosed.

The patient was treated with rest in bed, compression bandage, leg elevation and heparin (heparin low molecular

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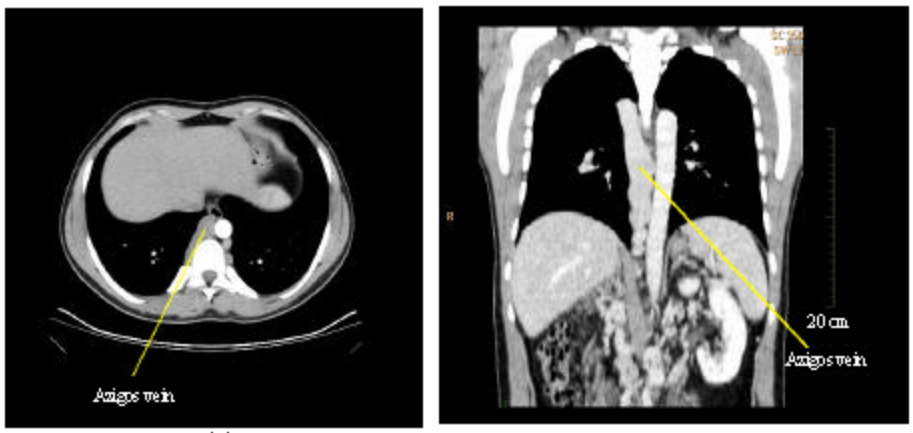


Fig. 1: Azigos vein

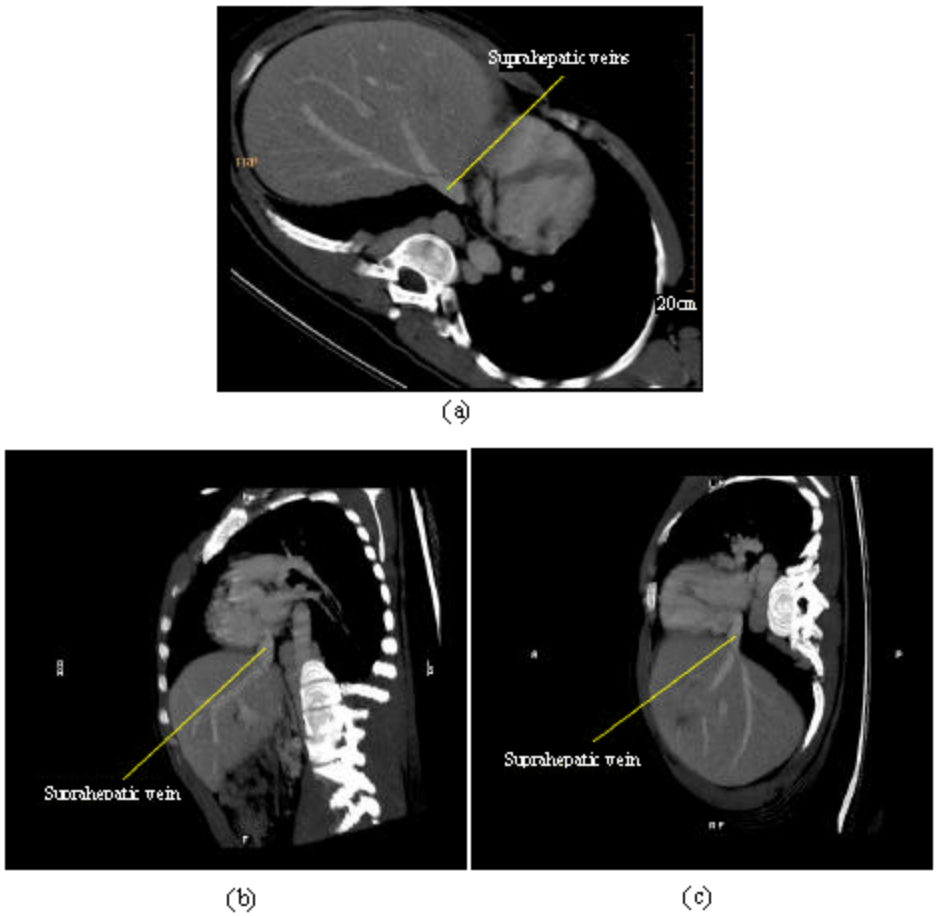
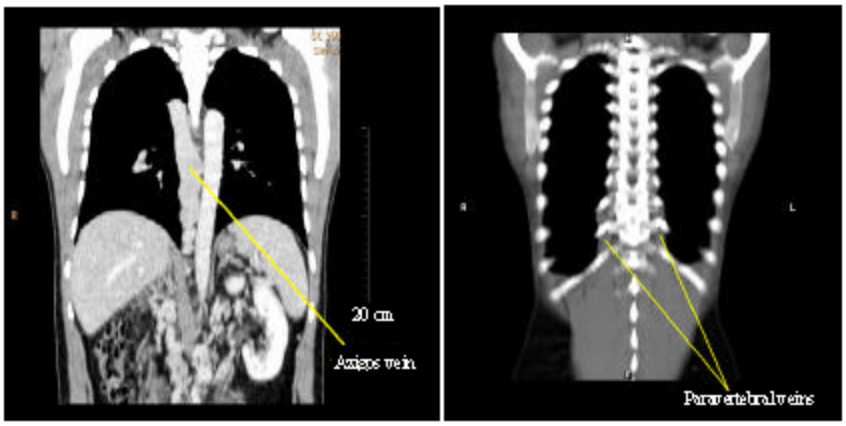


Fig. 2: Suprahepatic veins

weight); the pain and oedema decreased and he was discharged eight days after admission with acenocumarol therapy.

We advised the patient to avoid prolonged immobilization. Oral anticoagulation is been done for 6 months; it is now without symptoms.



(a) (b)

Fig. 3: Azigon and paravertbrel veins

DISCUSSION

The three main influences in the pathogenesis of thrombosis are: Abnormalities of the vessel wall, blood components and dynamics of flow (Virchow triad, described over 100 years ago). In more than 80% of patients presenting with deep venous thrombosis, a risk factor can now be identified. However, the presence of congenital venous malformations has been estimated at 1% of the general population (Sakellaris *et al*., 2005).

The venous system begins its development in the earliest stages of intrauterine life, being even and symmetrical in its origin and asymmetrical in its final anatomical structure.

Normal inferior vena cava consists of 4 components: Post renal segment, renal segment, pré-renal segment and hepatic segment.

Anomalies of the inferior vena cava are caused by aberrant development in the 6-8 weeks of gestation (Obernosterer *et al*., 2002), which coincides with the development of the spleen, liver, heart, lung, pleuroperitoneal membrane and rotation of the gut (Artico *et al*., 2004).

The phenomenon of abnormalities of IVC has been described in a variety of ways such as absence, agenesis, ablated, anomalous and interruption of a particular segment (infrahepatic, prerenal, renal or infrarenal). Double IVC is the most common anomaly involving this vessel (Minniti *et al*., 2002).

The causal reasons for the developmental failure are not clear; hypotheses are either embryonic dysontogenesis that affects separate segments or the entire IVC and intrauterine thrombosis or umbilical vein complication postpartum as causal events (Schneider *et al*., 2002).

Some cases are incidentally found in abdominal surgery or in radiologic workup; in adults, such an anomaly can cause diagnostic problems in the paravertebral area because of their tumor-like appearance.

Patients with IVC agenesis may present with symptoms of lower extremity venous insufficiency or idiopathic deep venous thrombosis; venous drainage of the lower limbs occurs through anastomosed channels, including, the sacral, lumbar and epigastric veins (Sakellaris *et al*., 2005).

If the deep venous collateral system is sufficiently developed and drains the venous blood from the lower extremities to the heart, venous stasis secondary to inadequate blood return through collaterals resulting in subsequent deep venous thrombosis is likely to be prevented (Yigit *et al*., 2006).

Alternative routes of venous return are the vertebrolumbar route, the anterior abdominal wall veins and the transumbilical portocaval route.

Diagnosis of vena cava agenesis is usually formulated indirectly on the basis of symptoms of iliac and femoral vein thrombosis, even if in the majority of cases patients may be asymptomatic.

Diagnostic tests include Doppler ultrasound, CT and MRI. MRI has the ability not only to visualize the venous circulation, but also to visualize other associated soft tissue or congenital anomalies.

Therapy must be focused on the prevention of complications such as thrombosis. It is necessary to anticoagulate for at least 6 months without the occurrence of any other risk factor.

Treatment of DPT is anticoagulation, whereas more aggressive intervention, such as surgical construction, may be indicated for recurrent, complicated and non-responsive cases (Sakellaris *et al*., 2005). Although reconstruction of the venous drainage system has been reported, most patients can be treated conservatively. (Artico *et al*., 2004).

Patients who are known to have anomalies of the inferior vena cava should be advised to avoid additional thrombogenic risk factors, such as unusual physical efforts, prolonged immobilization and oral contraceptive use. An anomalous inferior vena cava should be suspected in patients 30 years of age or younger, who have thrombosis involving the iliac veins, after exclusion of other concurrence of risk factors (like hypercogulable states or underlying malignancy).

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