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Case Report Open Access

Interrupted Inferior Vena Cava and Deep Vein Thrombosis - An Underdiagnosed Pediatric Disease

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Abstract

Interrupted inferior vena cava is a rare, but underdiagnosed congenital anomaly. Commonly it presents with bilateral deep vein thrombosis in teenagers and young adults. Lifelong anticoagulation treatment is mandatory. We describe three cases of teenagers with interrupted inferior vena cava, who presented with collapse, back pain and deep vein thrombosis.

Keywords: Developmental defect; Adolescent medicine; Vessel anomaly

Abbreviations:

DVT: Deep Vein Thrombosis; IIVC: Interrupted Inferior Vena Cava

Introduction

Interrupted inferior vena cava is a rare developmental defect; it is usually accompanied with azygos and hemiazygos continuation and is asymptomatic most of the time. The incidence of IIVC is 1:5000. In the vast majority (90%) of the case, it is an isolates variant not associates with isomerism or fetal anomalies. In all the cases of isolates interrupted inferior vena cava, the outcome is favorable [1]. We describe three cases of teenagers with isolated interrupted vena cava inferior who presented with acute deep vein thrombosis.

Case Presentation

CASE I

A 15 year old previously healthy girl presented with a ten days history of lower back pain spreading to the left leg, accompanied with swelling in the left lower extremity. Three month ago the patient was started on oral contraception.

Colour-Doppler-ultrasound revealed a massive thrombus filling of the lumens of the popliteal, femoralis communal and superficial veins. A CT-scan of the abdomen and pelvis showed a completed thrombosis of the abdominal inferior vena cava, the right ilical communis vein and the Vena iliaca communis externa and interna at the left side, caused by isolated atresia of the inferior Vena cava subhepatal, in a length of 5cm and a markedly dilated azygos and hemiazyogs vein with azygos continuation. Laboratory values for thrombophilic screening including protein C and S, Antithrombin III, Protein C resistance and homocystein levels were all normal. The long term therapy in this case is a lifelong anticoagulation therapy and reduction of all other risk factors for thromboembolism (such as smoking, oral contraception, etc.). Additionally compression therapy was recommended to prevent development of superficial venous collaterals in the lower extremity.

CASE II

A seventeen year old boy was brought to our hospital due to breathing difficulty after collapsing. He was suffering from deep vein thrombosis in both legs diagnosed two month ago. Therefore he was already put on oral anticoagulation. At this time the patient´s international normalized ratio was 1.8.

CT-Scan of the thorax was performed showing no evidence of pulmonary embolism. Colour Doppler ultrasonography revealed a thrombus filling the lumens of the iliaca communis vein, the left iliaca external vein and the proximal femoral vein. It also showed a postthrombotic change in the inferior Vena cava just below the passing of the superior mesenteral arterial vessel with venous collaterals at this level.

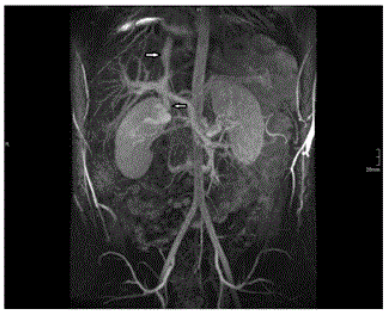

Figure1: MR angiography image showing no detectable flow in the atretic segment of the IVC. Paravertebral collateral vein (arrows)

In the right leg no thrombus was provable. Further on a MR angiography of the abdomen discovered a congenital isolated agenesis of the inferior Vena cava subhepatal with extensive venous collaterals

and a markedly dilated Vena azygos. There was no thrombus in the lumen of the collaterals. Laboratory values of thrombophilic screening were all normal and the patient had no other risk factors. So his therapy with oral anticoagulation was continued (Figure1).

CASE III

A sixteen year old boy presented with a swollen left leg. Interrupted vena cava inferior with azygos continuation was known since July 2010. He was already taking oral anticoagulation but the international normalized ratio at this time was far below his therapeutic range of 2.5-3.5. Colour Doppler ultrasonography of the leg showed a postthrombotic change in the left leg, but a thrombus could not be clearly established. Further on numerous collaterals could be detected in the retroperitoneal and paravertebral area.

Two years ago a CT scan of the abdomen showed an isolated agenesis of the vena cava inferior just below the kidney with azygos continuation (Figure 2).

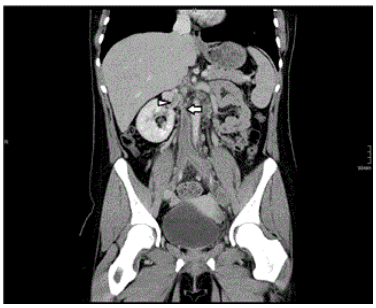

Figure 2: CT scan of the abdomen: Only the intrahepatal part of the inferior vena cava is contrasted, as the interruption is just below the hepatic level. Note numerous collaterals channels replacing the interrupted suprarenal portion of the inferior vena cava. Thrombosed segment of interrupted IVC (arrow). Renal vein thrombosis (arrowhead).

Discussion

Acquired risk factors for DVT include trauma, surgery, prolonged immobilization and pregnancy [2]. Predisposing genetic risk factors have also been established including protein C, protein S and antithrombin deficiencies as well as factor 5 Leyden and hyperhomocysteinemia [2]. In addition, several congenital anomalies of the IVC have also been recognized as risk factors for DVT, especially in young patients [3]. Several anomalies of the IVC and renal vein have been described including left sided IVC, double IVC, interrupted IVC with azygos continuation and absence of the infrarenal or entire IVC, retroaortic left renal vein and circumcaval ureter [4].

A thorough coagulation study in each case reveals no defects that predisposed to thrombophilia and these patients had no other risk factors for DVT such as trauma or immobilization. The authors suggested that an anomaly of the IVC alone may be a predisposing factor for development of thrombogenic events [5]. Increased venous blood pressure in the lower extremities leads to venous stasis and subsequent deep venous thrombosis [3]. Strenuous muscular exercise, physical effort and long periods of inactivity (e.g. long air travel or car rides) may also trigger acute thrombotic events. Those patients with DVT typically present with painfully swollen legs, lumbar or inguinal pain and occasionally hematuria and high grad fever [3,6]. The prevalence of interrupted IVC with azygos continuation is 0,5% [5]. In one study IVC anomalies were detected in 4 (5)% of 75 patients younger than 30years of age who developed lower extremity DVT during the 5 year investigation [5]. The mean age of DVT in the general population is 53 years while it is 27 years in patients with IVC anomalies [3]. Furthermore, the extent of DVT is greater in patients with IVC anomalies than in patients without, it is frequently bilateral and involves the IVC, the common, internal, and external iliac and femoral veins [6,7].

In the acute situation the first line therapy is systemic anticoagulation with unfractionated heparin to prevent pulmonary embolism and the prolongation of the thrombus [6,8]. In addition a compression therapy is obligate to avoid postthrombotic syndroms or chronic venous insufficiency. Later on a lifelong therapy with oral anticoagulation with vitamin K antagonists is mandatory as the risk for rethrombosis or embolism is very high, especially with supplementary risk factors such as smoking, immobilisation, trauma etc [9]. A surgical therapy for the purposes of a vascular reconstruction or bypass arrangement is described only in isolates cases and was applied exclusively with not healing venous ulzerations [10]. In conclusion interrupted inferior vena cava is more common as estimated. If teenagers present with DVT and a thorough coagulation study is normal just think of isolated IIVC as therapeutic regime changes.

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