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CASE REPORT

Endovascular Y-reconstruction of chronic ilio-cava occlusion

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ABSTRACT

INTRODUCTION: Inferior vena cava (IVC) agenesis is a rare pathology, associated with an increased risk of iliofemoral deep venous thrombosis (DVT), a frequent cause of disabling post-thrombotic syndrome (PTS).

CASE REPORT: Authors present a case of bilateral iliofemoral thrombosis in a patient with IVC agenesis, successfully treated at a European reference center. Patient was submitted to an endovascular Y reconstruction of the IVC and iliac veins.

Keywords: inferior vena cava agenesis; deep vein thrombosis; post-thrombotic syndrome; endovascular reconstruction.

INTRODUCTION

Inferior vena cava (IVC) agenesis is a rare pathology, of controversial etiology, with a prevalence of 0.0005-1%.[1,2] It accounts for up to 5% of unprovoked DVTs in young patients aged 20–40 years.2 In 90% of cases, it involves a suprarenal defect and only 6% of cases involve the renal and infrarenal segments.[1] It is associated with an increased risk of bilateral iliofemoral Deep Venous Thrombosis (DVT), a frequent cause of post-thrombotic syndrome (PTS) and impaired cardiac venous return, with compromised tolerance to cardiopulmonary exercise.[1,3,4,5] A homogeneous profile has been detected in patients with IVC agenesis: young patients, < 40 years of age, male gender, unilateral or bilateral DVT.[1,2]

There are different theories about IVC agenesis etiology. IVC agenesis, also called atresia or aplasia, can be congenital or acquired and is thought to be due to embryonic dysgenesis or thrombosis during the intrauterine or perinatal period.[2,6]

Some authors stated that it is one of the more than 60 different congenital IVC anomalies, described since 1793.[1,2,5,6] The embryological development of the IVC is a complex process consisting of multiple steps, all of which occur in a specific order.1 During embryogenesis (6–8 weeks of gestation), the IVC is formed by the fusion of three sets of paired veins (posterior cardinal, subcardinal, and supracardinal veins).[2] Failure of these paired veins to fuse into a unilateral right-sided venous structure results in anomaly of IVC.[1,2]

Acquired perinatal venous thrombotic events due to central venous catheters or heritable thrombophilic disorders have also been suggested to cause secondary IVC atresia.[6] Data suggests that approximately one-third of patients with IVC atresia had associated hypercoagulability disorders; hence screening is recommended to rule out a hypercoagulability state.[1,2,6] It is unlikely that the presumptive embryologic origins of IVC atresia would also predispose a patient to hereditary thrombophilia.[6] Rather, it is more likely that thrombotic events during the perinatal period or early life, such as those incited by heritable thrombophilia, lead to thrombotic occlusion of the IVC and secondary IVC atresia.[2,6] From this point of view, this condition frequently called "congenital" IVC agenesis can actually be an acquired or secondary process, rather than a primary embryologic dysgenesis.[6]

Patients with IVC agenesis develop a robust collateral deep venous system with drainage through the azygous, hemiazygous, lumbar, para-vertebral and abdominal wall veins.[1,2] The majority of cases remain asymptomatic because of the extent of the collateral venous network in the abdomen and lower extremity.[1] However, the collaterals may be unable to cope with the demands of increasing blood flow, thereby generating venous stasis, ulceration and

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recurrent DVT.[1,2]

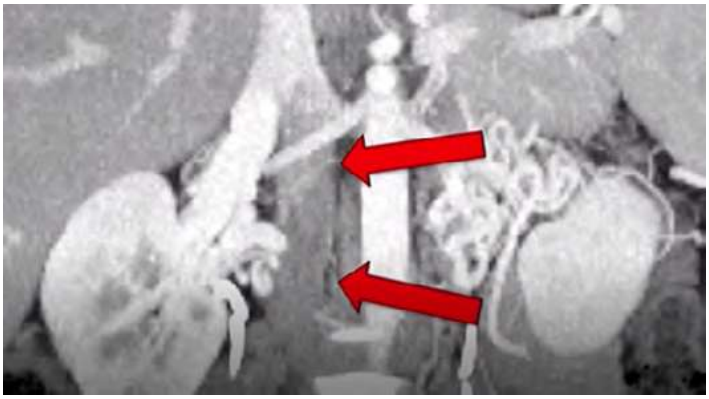
Obstruction of IVC is characterized by a wide clinical spectrum, depending primarily on the degree of venous collateralization.[4] Occlusion of the inferior vena cava (IVC) is often associated with venous claudication, swelling, skin changes, including venous ulcers and reduced exercise capacity.[7] The extent of venous occlusion correlates with the probability of a future post-thrombotic syndrome and the degree of PTS correlates with impairment of the quality of life.[7]

Owing to the young age of the patients and frequent disabling PTS, aggressive DVT treatment should be proposed.[1] We present a case of bilateral iliofemoral thrombosis in a patient with IVC agenesis, treated at a European reference center.

CASE PRESENTATION

Authors report a case of a 24 years old male who went four times to the Emergency Room due to low back pain lasting for 17 days, after returning from a long car trip. He had no edema of the lower limbs. Pain was previously considered to be of muscular origin, but for the fourth time, on 18/9/2018, a CT scan was performed. CT scan revealed complete agenesis of the entire IVC to the suprahepatic veins, with marked bilateral renal and azygos collateral venous circulation and extensive bilateral iliofemoral subacute thrombosis to the confluence of the deep femoral veins (Figure 1). Collaboration of a vascular surgeon was then required and a triplex scan was performed, which confirmed the extension of the thrombosis to the confluence of the deep femoral veins, with permeability of popliteal veins.

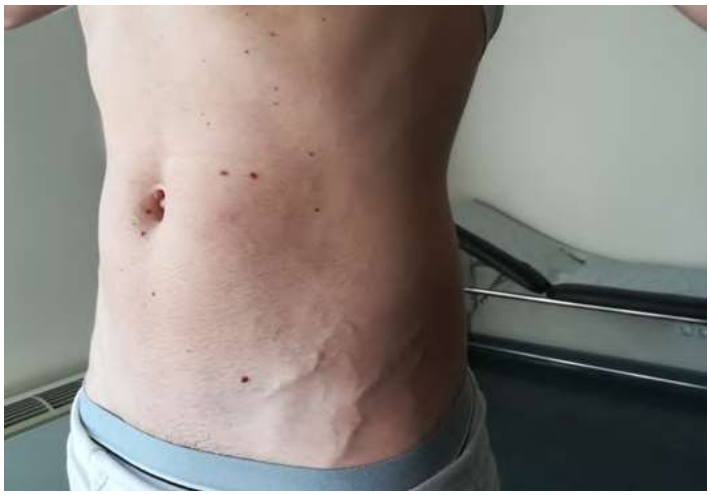
Figure 1. Initial CT scan.



IVC agenesis, marked collateral circulation and venous thrombosis are noted.

The clinical case was discussed within service peers and with other national and foreign institutions, regarding the perspective of intervention, but given the anticipated technical difficulty, conservative treatment with hypocoagulation and elastic stockings was proposed. Thrombophilia screening rulled out a hypercoagulability state. During follow-up, the patient developed venous circulation in the abdominal wall (Figure 2), venous claudication and decreased exercise tolerance – post-thrombotic syndrome with Villalta-Prandoni Score of 6 and spiro-ergometry with a predicted oxygen uptake (VO2) of 49%, which represent a severe cardiopulmonary functional capacity impairment, according to the Ludwigshafen scheme.

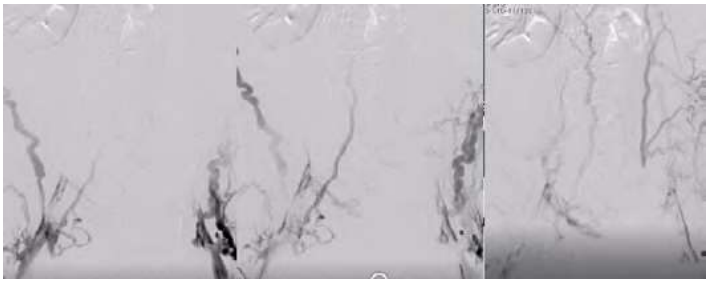
Figure 2. Abdominal wall aspect.



Development of collateral venous circulation is showed.

We reviewed the literature and made contact with an experienced European Hospital in the treatment of this pathology (62 cases treated) and the colleague agreed to observe the patient. Through the National Health Care System patient mobility platform for medical assistance abroad, the patient was referred to the institution. Treatment and trips to the Hospital had no costs to the patient. He was successfully treated, with direct transmission of the procedure in LINC 2019 (22/01/2019). Procedure was performed under general anesthesia and under full anticoagulation with intravenous unfractionated heparin, maintained for the first 24 hours after the intervention. After ultrasound guided bilateral femoral vein access with 10F sheaths, baseline venograms were obtained (Figure 3).

Figure 3. Baseline venograms.



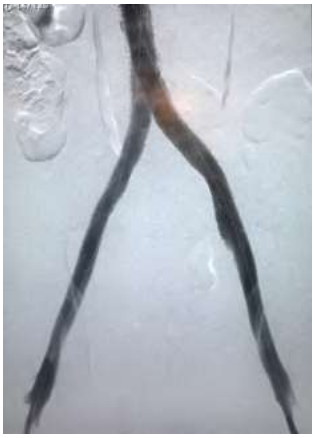
IVC agenesis, marked collateral circulation and extensive iliofemoral thrombosis are evidenced.

Venous occlusions were crossed with a 0.018-inch chronic total occlusion wire (Astato 30TM; Asahi Intecc) and a standard angiographic 4F diagnostic catheter. After predilation of the IVC and of the iliac veins with high-pressure balloon catheters (Atlas GoldTM; Bard), intravascular ultrasound (IVUS; Visions PV 0.035TM; Philips Volcano) was performed to identify the proximal and distal stent landing zones and the entire venous tract of the IVC and the iliac and femoral veins. Patient underwent Y-shaped IVC reconstruction from the suprahepatic veins to the femoral veins, using VenovoTM Bard venous stents (one 20*160 mm stent, two 16*160 mm stents and two 14*140 mm stents). Stent implantation with overlapping stents was performed from proximal to distal. The stent overlapping zones were 2 cm in the IVC and 1.5 cm in the iliofemoral veins. For reconstruction of the iliac bifurcation, kissing stents were deployed with a 2-cm double barrel within the proximal IVC stent. All implanted stents were post-dilated with high-pressure balloon catheters. Excellent imaging and clinical results were achieved (Figure 4 and 5).

Figure 4. Completion venogram.

IVC Y-reconstruction with total permeability of the IVC and iliac veins.

Figure 5. Completion venogram.



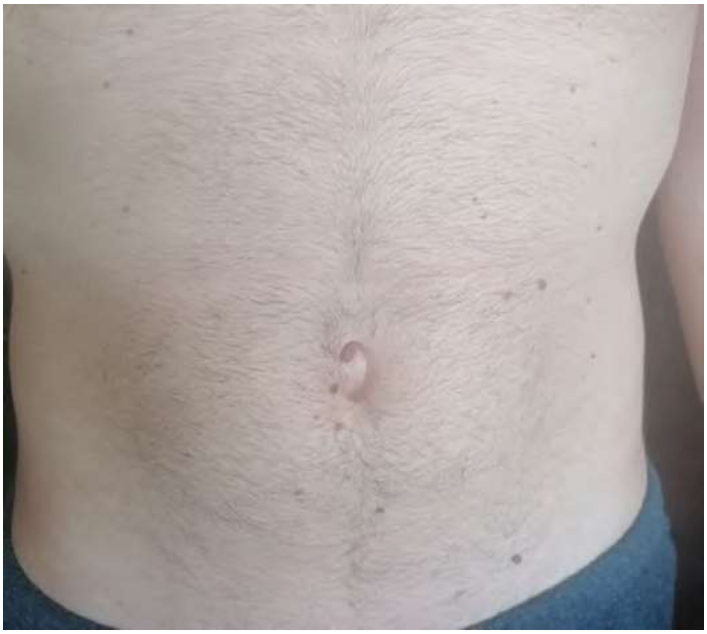
IVC Y-reconstruction with total permeability of iliac and femoral veins upon completion.

After endovascular therapy, oral anticoagulation therapy with apixaban was initiated. Routine follow-up visits were performed at 1, 3, 6 and 12 months, with yearly visits thereafter. At each visit, signs and symptoms of PTS were assessed and Duplex ultrasound was performed. There was remission of the abdominal venous circulation (Figure 6), resolution of venous claudication and increased exercise tolerance (VO2 of 92% - normal according to the Ludwigshafen scheme). After 29 months of follow-up the patient continues under hypocoagulation with apixaban, clinically well and with permeability of the all stents.

DISCUSSION

VCI agenesis is a rare vascular anomaly and often presents with extensive iliofemoral DVT.[1,2,5] Although ultrasound is an excellent tool in identification of DVT, it can often miss the diagnosis of IVC agenesis.[2]

Figure 6. Abdominal wall aspect during follow-up.



Remission of collateral venous circulation is shown.

CT or MRI studies are more effective in identifying IVC anomalies and are recommended if there is history of DVT in younger patients.[2]

Data suggests that chronic IVC obstruction reduces venous return to the heart with impaired cardiac preload.4 This may cause cardiopulmonary exercise intolerance, with limited peak oxygen uptake during cardiopulmonary exercise testing.[4]

The restoration of venous flow in the IVC and iliac veins aims to prevent and treat PTS and restore venous flow to the heart.[7] Cardiopulmonary functional capacity was graded according to the Ludwigshafen scheme [14]: Normal cardiopulmonary capacity was defined as achievement of at least 85% of predicted oxygen uptake (VO2) at peak exercise; in patients with <85% of predicted VO2 uptake, extend of impairment was graded into mild (70–84%), moderate (50–69%) and severe (<50%).[4] Endovascular stent reconstruction of the IVC has emerged as a potentially effective treatment for symptomatic patients with PTS and as a way to improve cardiopulmonary exercise capacity.[4]

By the time of treatment of this patient, the European Hospital to which he was referred to had already performed endovascular reconstruction of the inferior vena cava (IVC) in 62 patients with IVC obstructions, with a technical success of 98% with 2-year primary, primary assisted and secondary patency rates of 57%, 76%, and 87%, respectively.[7] They reported that 43% of patient where free from symptoms or signs of venous hypertension and another 48% showed significant clinical improvement.[7] Reported complications during follow-up were early stent thrombosis, symptomatic stent occlusion and stent stenosis.[7] Authors concluded that technical and clinical outcomes of endovascular reconstruction of the IVC were favorable and IVC endovascular reconstruction can be recommended for symptomatic IVC obstruction.[7]

There are multiple alternative techniques and materials (steel alloy stents, self-expanding arterial stents, dedicated venous stents) that may be used when treating iliocaval

obstructions and primary and secondary patency described in literature varies from 57-87% and 78-98%, respectively.[7-11]

Predictors of loss of stent primary patency are patient and procedural related.[7] Patients with post-thrombotic venographic changes of the femoral veins at baseline or a history of DVT were more likely to lose primary patency compared with patients with normal leg inflow veins and no history of DVT.[7,8]

It´s very important that bilateral caudal extension is performed as necessary to cover the entire obstructive lesions and to ensure an adequate inflow.[8]

In another study from the Zurich Center, authors stated that in patients with chronic IVC obstruction, cardiopulmonary exercise intolerance as a result of impaired cardiac filling is at least partially reversible following endovascular IVC reconstruction.[4] Our patient had exceptionally great clinical enhancement, with resolution of venous claudication and improved cardiopulmonary exercise capacity, documented and quantified by spiro-ergometry, passing from a VO2 of 49% before the intervention (severe cardiopulmonary functional capacity impairment) to 92% (normal) after the endovascular reconstruction.

The rarity of the pathology mirrored in this clinical case and the anticipated technical difficulty of the necessary endovascular procedure sometimes hinds the intervention. However, regarding the facility of scientific discussion without borders and the gratuitousness of our National Health Care System, conservatism cannot be the limiting factor to treat these patients, generally young and with incapacitating PTS.

ETHICAL RESPONSIBILITIES

Protection of humans and animals

The authors declare that the procedures followed were in accordance with the regulations established by the heads of the Clinical Investigation and Ethics Committee and in accordance with those of the World Medical Association and the Declaration of Helsinki.

Confidentiality of Data

The authors claim to have followed the protocols of their work center regarding the maintenance of confidentiality of patient data.

Right to privacy and written consent

The authors declare that no patient data appear in this article.

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Conflicts of interest None
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