CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 14 (2015) 69–71

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

A rare cause of claudication treated with IVC reconstruction: A case report

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ARTICLE INFO

Article history:
Received 21 April 2015
Received in revised form 6 July 2015
Accepted 18 July 2015
Available online 26 July 2015

Keywords:
IVC-Inferior Vena Cava
PTFE-Polytetrafluoroehtylene
DVT-Deep Vein Thrombosis

ABSTRACT

INTRODUCTION: Inferior vena cava (IVC) agenesis is one of rare entities of IVC anomalies which presents in young patients with unprovoked deep venous thrombosis (DVT) or unexplained bilateral lower venous insufficiency. We are presenting a case of IVC agenesis which was treated with IVC reconstruction.
CASE: We describe a case of 28 years old male with painful bilateral lower extremity varicose veins and a history of right lower extremity DVT and was on anticoagulation with warfarin. He was found to have extensive bilateral greater saphenous veins (GSVs) and right femoral vein reflux with patent bilateral deep veins. He was treated with bilateral GSV ablation and microphlebectomies. Six weeks later he presented with acute bilateral iliofemoral DVTs treated with tissue plasminogen activator thrombolysis tPA via bilateral popliteal vein access which helped relieve his leg swelling but he continued to have debilitating venous claudication. A computed tomography (CT scan) demonstrated resolution of DVT but revealed IVC agenesis. He underwent IVC reconstruction with prosthetic graft which helped complete resolution of his chronically debilitating bilateral lower extremity claudication.
CONCLUSION: In young patients with severe manifestations of lower extremity venous hypertension i.e. edema, varicosity and DVT, central venous anomaly should be considered. Severely symptomatic cases of IVC agenesis can be treated with IVC reconstruction.

1. Introduction

Congenital anomalies of the inferior vena cava (IVC) are infrequent and the incidence is reported to be 0.7–8.7% in general population. Congenital absence of IVC can either be isolated or associated with other anomalies [1]. In most cases of isolated asymptomatic congenital absence of IVC lifelong anticoagulation is indicated [2]. IVC agenesis is one of the rare IVC anomalies that can present as lower extremity deep venous thrombosis (DVT) typically in younger patients and characteristically unprovoked in nature [3]. We describe a unique case of IVC agenesis that was diagnosed with de-novo bilateral superficial venous insufficiency and treated with superficial venous ablation complicated with bilateral iliofemoral DVT. Subsequent workup revealed IVC agenesis.

2. Case

We are presenting the case of a 28 years old male who presented with painful bilateral lower extremity varicose veins. He had previous history of right lower extremity DVT 2 years before presentation and had taken anticoagulation for 2 months. Noninvasive venous insufficiency study revealed extensive bilateral greater saphenous reflux along with unilateral right sided femoral vein incompetence. The deep veins were all patent. Patient was treated with stripping of right GSV and laser ablation of left GSV with bilateral microphlebectomies. Six weeks later he presented back with acute bilateral lower extremity swelling. Duplex exam revealed bilateral iliofemoral DVTs. This was treated with tissue plasminogen activator thrombolysis via bilateral popliteal vein access. This helped relieve his leg swelling but he continued to have severe venous claudication. A computed tomography (CT) scan demonstrated IVC agenesis [Fig. 1]. Definitive treatment then involved prosthetic reconstruction of the IVC with 14 mm ringed polytetrafluoroethylene graft from right common iliac vein to juxta-renal IVC. Intraoperative findings were consistent with dilated venous collaterals in the abdominal wall. Patient was discharged home on sixth post-operative day on oral anticoagulation.

Over a follow-up of 24 months, patient has had no recurrent symptoms. Interval imaging with CT scan at 3 months and venous duplex at 12 and 18 months revealed widely patent graft [Fig. 2]. Oral anticoagulation was stopped 6-months postoperatively and patient is maintained on low-dose aspirin therapy.

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http://dx.doi.org/10.1016/j.ijscr.2015.07.016

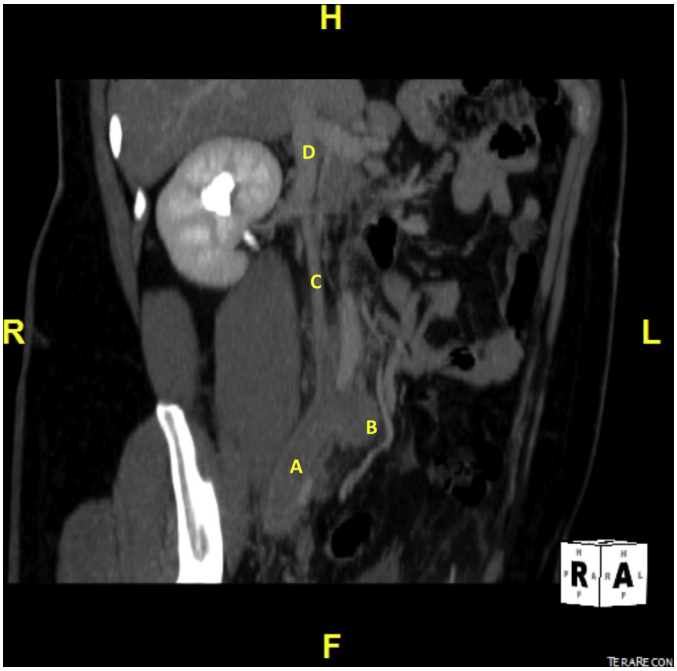

Fig. 1. CT of abdomen and pelvis, coronal section Right Anterolateral, RA projection showing: A, Right dilated common iliac vein. B, Left dilated common iliac vein. C, Atretic inferior vena cava, IVC. D, Proximal normal inferior vena cava, IVC.

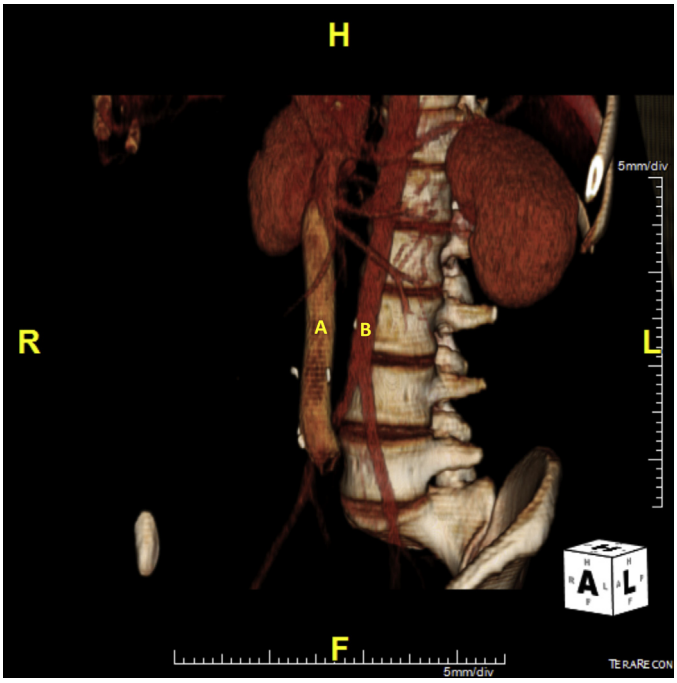

Fig. 2. CT of abdomen and pelvis 3D reconstruction image with Antero-Lateral projection. A, Polytetrafluoroethylene, PTFE graft from Infra-renal IVC to Right common iliac vein. B, Aorta.

3. Discussion

IVC anomalies result from the defects in vascular embryogenesis and agenesis of the IVC can cause of persistently increased venous pressure and predisposes to DVT [4]. The two most common presentations associated with IVC agenesis are thrombosis of deep veins in 76.2% of cases and edema in 23.8% of cases [5]. The first ever case of IVC reconstruction using polytetrafluoroethylene PTFE graft for symptomatic congenital absence of IVC was described in 2008 [1]. Early surgical treatment for IVC agenesis followed by need for oral anticoagulation afterwards has been suggested in the literature as it has been shown potentially reverse the debilitating venous insufficiency associated with it [6,7].

Our patient presented with symptomatic varicosities misleading into a diagnosis of de-novo early superficial venous insufficiency. The treatment of superficial insufficiency however was complicated by severe iliofemoral venous thrombosis likely from compromise of the collateral pathways. The underlying pathology was eventually uncovered once the thrombolysis of iliofemoral system was successful.

This case underlines that symptomatic venous insufficiency in a young patient warrants a search for a central venous cause. The definitive treatment of IVC agenesis is with venous reconstruction with good results.

5. Conclusion

In young patients with severe manifestations of lower extremity venous hypertension i.e., edema, varicosity and DVT, central venous anomaly should be considered. IVC reconstruction is successful in severely symptomatic cases of IVC agenesis.

Conflict of interest

None.

Sources of funding

University of New Mexico, Department of Surgery, Division of Vascular Surgery.

Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Consent

We obtained an informed consent from the patient after explaining the need for reporting the case and its potential contribution to scientific literature keeping all the patient information confidential and not revealing patient identity in the article. We have the consent form which is available on request.

Author contribution

Barkat Ali — performed literature review and write the paper.
Mark Langsfeld — helped conceptualizing and editing the paper.
Mohammad A. Rana — helped with literature review and editing the paper.
John Marek — identified the case and performed writing and editing of the paper.

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