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Conclusion

We described, in one case, pre-cardioversion TEE findings of a thin, linear, mobile, and nonobstructive membrane within the cavity of the LAA. The clinical implications and origins of these kinds of membranes are not clear; however, they may represent an anatomic variant. The echocardiographer should pay attention to the LAA during examination.

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Double etiology of recurrent thrombophlebitis: Behçet's disease and inferior vena cava agenesis

Tekrarlayan tromboflebit çift etiyolojisi: Behçet hastalığı ve vena kava inferiyor agenizisi

Sonia Hammami, Amel Barhoumi, Salem Bouomrani, Olfa Harzallah, Silvia Mahjoub

Department of Internal Medicine, University Hospital of Monastir, Monastir, Tunisia

Introduction

Behçet's disease (BD), a systemic vasculitis with unknown origin, mostly involves vascular lesion (1). Thrombosis was a commonly feature of the disease which it may affect large vessels, such as vena cava (2). There are different considerations about the pathogenesis of the vascular complications and the tendency for thrombosis in BD. However, less knowledge considering vascular malformations was clarified. Absence of the inferior vena cava (IVC) which is an uncommon vascular anomaly is exceptionally associated to BD. In this field, we report a case.

Case report

A 36-year-old man was admitted because of the fifth episode of left leg thrombophlebitis. He had suffered from recurrent erythema nodosum, oral and scrotal ulcers for five years. Since one year, when he developed a bilateral pan uveitis, he had diagnosed as Behçet's disease and treated by colchicine, platelet suppressive agent and over dose of corticosteroid. He fulfilled all criteria of International Study Group of Behçet's disease (3) and he had positive HLA B 51. Despite regular intake of his treatment, he had noticed engorgement of his left leg and he was hospitalised for further exploration. On admission, the classical signs of poor venous drainage were present. There were several oral and scrotal aphtous showing a BD flare.

Ultrasonography examination showed acute deep venous thrombosis extending from the popliteal to the distal external iliac vein. Thrombophilia testing was in normal value; it included antithrombin III, protein C, protein S, homocysteine, activated protein C resistance test, and presence of antiphospholipid antibodies. Importance of collateral superficial vein of chest, abdomen and lower limb led to practice thoraco-abdominal angiographic tomography to search vena cava obstruction. It showed absence of retro-hepatic portion of inferior vena cava (Fig. 1) and developed collateral veins. No visceral malformations were detected. Heart sonographic exploration was normal. Treatment was consisting in low molecular weight heparin for a week associated with adjusting dose of acenocoumarol. No place for surgical treatment

Address for Correspondence/Yazışma Adresi: Sonia Hammami, MD, University Hospital of Monastir, Department of Internal Medicine, Monastir, Tunisia
Phone: 0021698913188 Fax: 0021673460737 E-mail: sonia.hammami@fmm.rnu.tn

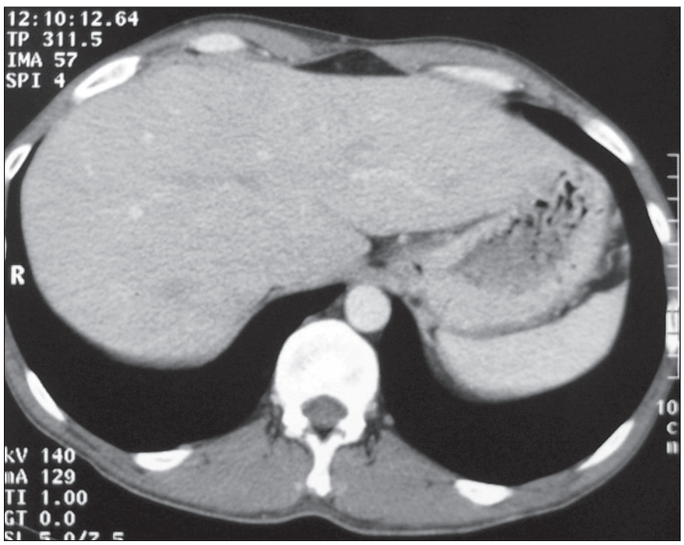

Figure 1. Contrast-enhanced axial computerised tomography scan obtained at the level of hepatic dome shows absence of the retrohepatic segment of the inferior vena cava

since compensated blood outflow via a system of developed collaterals. The patient was in excellent clinical condition as his BD had been in remission.

Discussion

Deep venous thrombosis (DVT) is an illness of clinical interest, due to the associated morbidity and mortality consequences. The aetiology in young patients was frequently associated with congenital or acquired coagulation abnormalities, immunologic diseases, and vasculitis (4). However, recent radiological advances have identified vena cava malformations as a new etiologic factor to be considered. This congenital malformation can give rise to anatomic malformations that impede vein drainage and favour the development of thrombosis (5). Recurrent low extremities thrombosis presented the revelation's mode of IVC agenesis in our patient. Behcet's disease itself may explain thrombophlebitis in this case, but an additional risk factor, like venous aberrancy, could be a reason of thrombosis recurrence. Our case is the second in the world literature which it describe an eventual association between BD and IVC agenesis (6).

Although, our patient had no any congenital visceral malformation, heart diseases should be searched since they had been described in 0.6 - 2% of IVC aplasia cases (7). According to publications, this pathology was first diagnosed at the end of the 18th century during autopsy (8). Due to recently developed examination methods such as ultrasonography and helicoidal computerised tomography the detection frequency of IVC anomalies has improved, and these anomalies have been found in healthy individuals (9).

The treatment of congenital IVC anomalies is currently conservative for a majority of patients (5, 6, 9). Anticoagulant therapy conducted during the acute DVT stage or its relapse and later vasotonic medications together with elastic leg compression demonstrate a slight improvement in hemodynamics. In cases of venous occlusion complicating IVC, the prognosis is not always poor because of the development of collateral veins and recanalisation (5, 7). If the patient has severe collateral insufficiency, surgical intervention may be considered. Considering an angio-Behçet syndrome in our case, colchicine, prednisone and heparin have been prescribed and still used by a patient. The role of cytotoxic agents, such as cyclosporine-A, azothioprine and cyclophosphamide, in the treatment of vascular lesions in BD is discussed (10).

Conclusion

Thrombophlebitis deserves an exhaustive exploration in young patients. Our observation illustrates the possibility of association of two causes to phlebitis: a congenital (IVC agenesis) and an acquired (Behcet's disease) factors.

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