CASE REPORT

May-Thurner Syndrome: A Forgotten Cause of Venous Thromboembolism

Archana Baburao[1], Amandeep Singh[2], Ajay Babu[3], Ankit Pandey[4]

Abstract

The annual incidence rates of venous thromboembolism are approximately 1 per 1,000 persons per year in adult population. Deep vein thrombosis (DVT) most frequently occurs in the setting of underlying illness, and anatomical abnormalities are rarely considered as an etiology for it. A well-described anatomical cause for DVT is "May-Thurner syndrome" (MTS), which occurs as a result of compression of the left common iliac vein by the overlying right common iliac artery. This syndrome most often affects young to middle-aged women. Pulmonary embolism (PE) occurs very rarely in these patients. Anticoagulation therapy alone is not enough in these patients. We report a case of 27-year-old male who had both left DVT and PE caused by MTS and was treated with endovascular management along with long-term anticoagulation.

Keywords: Deep vein thrombosis, May-Thurner syndrome, Pulmonary embolism.

Indian Journal of Critical Care Medicine (2020): 10.5005/jp-journals-10071-23331

Introduction

Venous thromboembolism (VTE) is a major medical problem which includes deep vein thrombosis (DVT) and pulmonary embolism (PE). Pulmonary embolism has been reported in 40–50% of patients with DVT. Deep vein thrombosis most frequently occurs in the setting of underlying illness. Anatomical abnormalities are rarely considered as an etiology for DVT. A well-described anatomical cause for DVT is "May-Thurner syndrome" (MTS). The true incidence of MTS is unknown, and it ranges from 22 to 32% according to the autopsy studies.[1] Despite high prevalence of MTS, the clinical prevalence of MTS-related DVT accounts for only 2–3% of all lower extremity DVTs.[2] Females account for 72% of MTS cases in the age group of 20–40 years.[3] We present a case report of a 27-years-old male who was diagnosed with DVT and PE, ultimately found to have MTS.

Case Description

A 27-year-old male presented to emergency department with 1-week history of cough, left leg swelling, low-grade fever, and streaky hemoptysis. He also had shortness of breath and acute worsening of left leg swelling which has been painful for 2 days. His history was unremarkable and was ambulating well at home. He had no relevant family history and no addiction to tobacco, alcohol, and illicit drugs. On examination he was conscious and oriented, febrile, had tachycardia, and was in minimal respiratory distress with tender erythematous swelling from left ankle to thigh. Systemic examination did not reveal any significant findings.

Complete hemogram revealed leukocytosis. Renal and liver functions were within normal limits. Chest X-ray showed bilateral lower zone haziness. Arterial blood gas analysis showed mild hypoxia. Electrocardiogram showed sinus tachycardia and echocardiography was normal. However, his D-dimer was elevated 2610 ng/mL. Left lower limb venous Doppler showed extensive thrombosis of entire left common femoral, superficial femoral, popliteal vein, posterior tibial vein with superficial extension into proximal great saphenous vein. Therapeutic anticoagulation with intravenous heparin infusion was initiated along with other supportive treatments. In view of underlying DVT and elevated D-dimer levels, CT pulmonary angiogram (CTPA) was done, which showed thrombus in right and left main pulmonary arteries (Fig. 1) with extension into inferior and segmental branches. USG abdomen showed right ectopic kidney and workup for connective tissue disease and hypercoagulability status were negative. Hence, MR venography of lower limb was planned but could not be done as the patient had claustrophobia. CT venogram of abdomen and

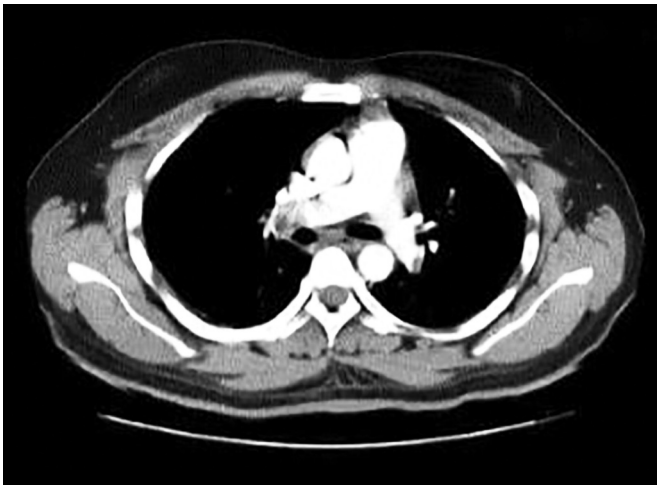

Fig. 1: Thrombus in the right and left main pulmonary arteries

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How to cite this article: Baburao A, Singh A, Babu A, Pandey A. May-Thurner Syndrome: A Forgotten Cause of Venous Thromboembolism. Indian J Crit Care Med 2020;24(1):66–68.

Source of support: Nil

Conflict of interest: None

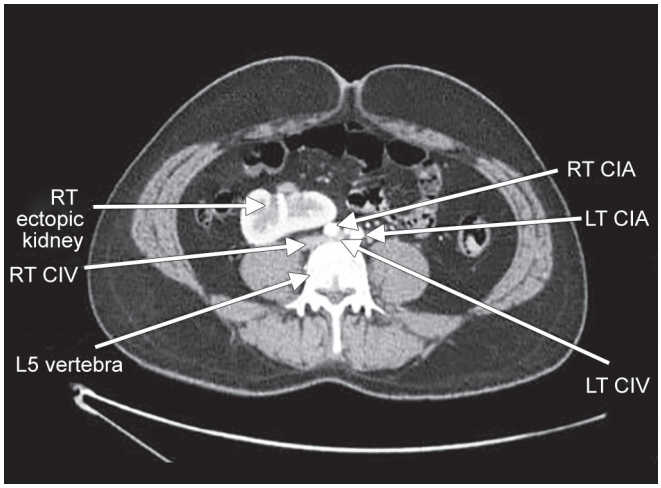


Fig. 2: Transverse section showing compressed left common iliac vein by the right common iliac artery against the fifth lumbar vertebra. RT CIV, right common iliac vein; RT CIA, right common iliac artery, LT CIA, left common iliac artery; LT CIV, left common iliac vein

lower limb was done which showed gross compression of distal left common iliac vein by right common iliac artery (Fig. 2) for about 1.5 cm in length beyond which thrombosis of left common iliac, external iliac, and femoral veins was noted (Fig. 3), and these features were consistent with MTS.

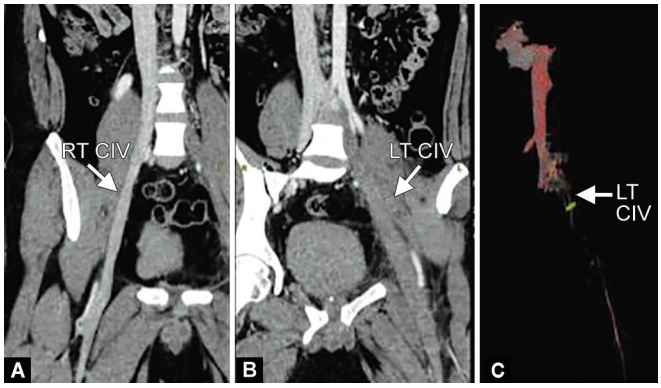
Patient underwent embolectomy followed by stenting of the left common iliac vein and placement of inferior vena cava (IVC) filter. Long-term anticoagulation with rivaroxaban was initiated. He was stable at the time of discharge and remained asymptomatic during follow-up after a month.

Discussion

May-Thurner syndrome is an uncommon anatomical variant characterized by external compression of the left common iliac vein by the right common iliac artery against the fifth lumbar vertebra and known as "iliac vein compression syndrome" or "Cockett syndrome". This entity was first described by McMurrich, in 1908,[4] defined by May and Thurner in 1957,[3] and described clinically by Cockett and Thomas in 1965.[5]

May and Thurner postulated that the chronic pulsations of the overriding right iliac artery led to elastin, collagen deposition, and intimal fibrosis leading to venous spur formation and thrombosis. They noted the absence of these lesions in fetal autopsies, thus supporting their belief that the lesion is acquired rather than congenital,[3] but it is not known why the normal anatomic relationship between the left common iliac vein and right common iliac artery is disrupted and begins to interfere with venous flow.

Our young male patient presented with VTE secondary to a left iliac vein thrombosis, and radiological findings were consistent with the diagnosis of classical MTS. But the case got complicated by the discovery of pelvic kidney. However, on radiological review, the impression was that MTS was independent of pelvic kidney. CT pulmonary angiogram was suggestive of bilateral massive PE, which was in contrast to the reported finding that compression and narrowing of the left iliac vein could protect patients with MTS from PE, or at least from massive embolism.[6] Also, the incidence of PE in patients with MTS is low, and there are few case reports of MTS which presented with acute PE.[7] To the best of our knowledge, there are only four case reports from India, and this is the first report in a male patient presenting with MTS-related VTE.



Figs 3A to C: (A) Coronal section showing patent right common iliac vein; (B) coronal section showing thrombosed left common iliac vein; (C) 3D reconstruction image showing thrombosed left common iliac vein. RT CIV, right common iliac vein; LT CIV, left common iliac vein

The low clinical incidence of MTS-related DVT (2–3%) may be due to missed diagnosis of MTS as the diagnostic workup of DVT is often halted once the underlying common risk factors for DVT are confirmed, such as prolonged bed rest, postsurgery, malignancy, or oral contraceptive pill (OCP). This is supported by a study which showed 55.9% predominance for left-sided DVT.[8] Furthermore, DVT is more common in women, who often have a history of OCP, recent pregnancy and 72% of women diagnosed with MTS are relatively young.[2] Another reason for low incidence of MTS-related DVT is that the diagnosis of DVT is often with ultrasonography, and the anatomical defect associated with MTS occurs high in the pelvis, an area that is not easily visualized by ultrasound.[9]

The gold standard technique for diagnosing MTS is the femoral stick venogram. A femoral stick venogram, although invasive, offers many advantages as it allows other procedures such as thrombolysis, balloon angioplasty, stenting, placing an IVC filter to be done at the same time. Other imaging modalities include CT venography, MR venography, and intravenous ultrasound. Anticoagulation therapy alone is not enough in treating MTS as it does not treat the underlying mechanical compression. Management of MTS has evolved over the past few decades favoring endovascular management as the primary treatment as it treats both mechanical compression with stent placement and thrombus burden with chemical dissolution.[10]

Conclusion

It is important to consider MTS in differential diagnosis when presented with unilateral DVT, especially in a younger age population. If diagnosis is missed, the recurrence of thrombosis, PE, and post thrombotic syndrome will lead to significant morbidity and mortality. May-Thurner syndrome can be well-managed with early recognition and aggressive management. The key to successful treatment is to fix the anatomical lesion along with removal of the clot and use of anticoagulation.

Author Contributions

Archana B, Amandeep Singh, Ajay Babu, and Ankit Pandey contributed to the design of the study. Archana B, Amandeep Singh, and Ajay Babu contributed to definition of intellectual content, literature search, and manuscript preparation. Archana B and Ankit Pandey contributed to clinical studies. Archana B, Ajay Babu, and Ankit Pandey contributed to data acquisition. Archana B and Amandeep Singh contributed to manuscript editing and manuscript review. Archana B was the guarantor of the study.

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