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**Case Report**

# May-Thurner syndrome causing venous thromboembolism in a young female with Sturge weber syndrome and connective tissue disease

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## ABSTRACT

May-Thurner syndrome (MTS) is a rare condition characterized by the compression of the left common iliac vein by the overlying right common iliac artery. It is a unique causal reason for development of deep venous thrombosis (DVT) and pulmonary embolism (PE). Young females developing DVT should be examined and investigated with details as triggers can include trauma, pregnancy, autoimmunity. In this case report we report a case of a young female who had Sturge Weber Syndrome, and how a left hip pain was diagnosed as DVT which led to diagnosis of an underlying MTS and autoimmune trigger for her hypercoagulable state.

**Keywords:** MTS, DVT, Connective tissue disease, Sturge Weber syndrome

## INTRODUCTION

Venous thromboembolism (VTE) includes vascular entities of deep vein thrombosis (DVT) and pulmonary embolism (PE). Rheumatologic conditions like connective tissue disease (CTD) are often inflammatory by nature, which can cause vessel disruption and development of thromboembolism. Moreover, MTS due to compressive stasis also has a role in blockage of blood flow and thrombus formation. This case reports discusses the condition of a 22 year old Indian female, who was on antiepileptics and had presented with pyrexia, seizure-like activity and left hip pain. She was found to have MTS with an overlapping CTD presentation. This case report helps us to know the strong implications of MTS and autoimmunity in initiation of a thrombotic cascade of events leading to DVT and PE.

## CASE REPORT

A 22 year old female known case of Sturg Weber syndrome on antiepileptics carbamazepine and valproate, bilateral avascular necrosis of hip with a past history of decompression surgery came with complaints of fever for 20 days, documented up to 102F, associated with chills and rigor, relieved with medication. Patient also had 1 episode of abnormal movements involving right arm and leg lasting for 30 seconds with no loss of consciousness and spontaneous recovery 10 days back. She also complained of left hip pain, aggravated on walking, relieved by taking analgesic. On examination she was conscious, oriented to time, place and person. She was normotensive, tachycardic with a pulse rate of 104/min, febrile with a temperature of 102 F and a respiratory rate of 14/min. She had pallor on examination, with bilateral normal vesicular breath sounds and no abnormalities on examination of cardiovascular and per abdomen. She had no focal neural deficits but had pain on walking and making her lift and rotate her left hip joint.

On investigating she was found to have anemia (Hb-10.8 g/dl), TLC-5.87 thousand/mm$^3$, platelets-255 thousand/mm$^3$; total bilirubin-0.32 mg/dl, direct bilirubin-0.17 mg/dl, SGOT-95 IU/l. SGPT-l69 IU/l, ALP-76 IU/l GGT-202 IU/l. Blood urea nitrogen-3.65 mg/dl,

creatinine-0.58 mg/dl, sodium-132 meq/l, potassium-4.35 meq/l. Infective and inflammatory workup was sent. ESR-115 mm. CRP-97. Serum procalcitonin-0.07 ng/ml. Blood culture and sensitivity (C/S) was sterile. Malaria peripheral smear was negative. Dengue NS1 and IgM were negative. Urine routine microscopy revealed 10-15 WBC, bacteria present. Urine C/S were sterile. Ultrasound abdomen was within normal limits. Prothrombin time-12.4 seconds. INR-1.08. Activated partial thromboplastin time-31.7 seconds. MRI Brain was done to rule out any active foci of seizure which was suggestive of Sturge Weber syndrome with evidence of involvement of left parietal lobe with associated atrophy (Figure 1). An EEG was done which revealed abnormal mild encephalopathy and intermittent dysrhythmic focus over right hemisphere. Valproic acid levels were 24 ug/ml. Carbamazipine level were 4.71 ug/ml.

D dimer was 3.01ug/ml. 2 D ECHO revealed septal motion jerky, no regional wall motion abnormality, trace tricuspid regurgitation, pulmonary artery systolic pressure-36 mmHg, ejection fraction of 60%. In view of persistent pain of left hip radiating to ankle, a venous duplex study of lower limb revealed ilio femoral popliteal and calf vein acute DVT in left lower limb. CT venography was done which was suggestive of distension with non-opacification of the veins of the left lower limb involving the popliteal-superficial femoral vein- deep femoral vein-common femoral vein- external iliac vein and internal iliac vein-proximal common iliac vein with significant attenuated distal left common iliac vein traversing deep to the right common iliac artery-may represent MTS Thrombi was also visualized in the left lower lobe pulmonary artery and paucity in the right lower lobe pulmonary arteries (Figure 2). To ascertain the cause of hypercoagulability extensive investigations were done. Prothrombin mutation was negative. Protein S, protein C were normal, MTHFR gene polymorphism was heterozygous positive, factor V leiden mutation was negative. Antithrombin function assay was normal. Serum homocysteine levels 7.86 umol/l (normal). Direct and indirect Coombs test were negative. Hepatitis B surface antigen, HIV, hepatitis C were negative. C3 levels and C4 levels were 2226 mg/l and 485 mg/l respectively. Autoimmune markers were sent which revealed. ANA IF was positive, speckled pattern, 1:100 tire, + intensity. ANA profile revealed SSA antibody strong positive, RO 52 recombinant Antibody positive. Antiphospholipid antibody syndrome workup was negative.

Hydroxychloroquine was added and mechanical thrombectomy, catheter directed thrombolysis with Intra vena cava (IVC) filter insertion in infra renal IVC was done. She was continued on heparin infusion and monitored with the INR. She was then shifted to Dalteparin 2500 subcutaneously twice daily for the seven days. Patient improved was discharged on oral anticoagulant rivaroxaban and currently is doing well on follow up.

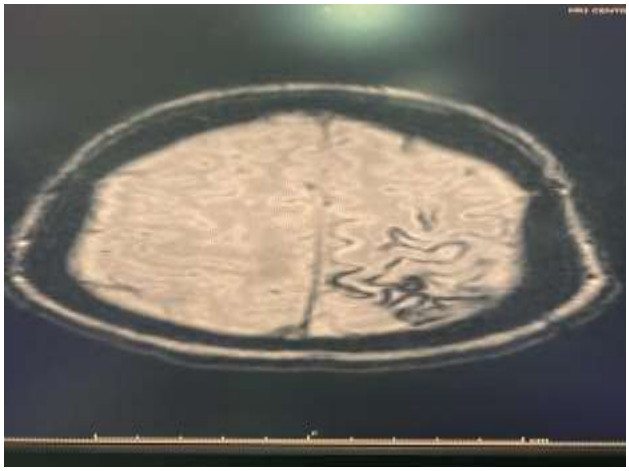

**Figure 1: T2W MRI image of the brain shows well defined thin hypointense signal is seen along the cortical margin of the cerebral parenchyma in the left parietal lobe suggestive of angiomatosis.**

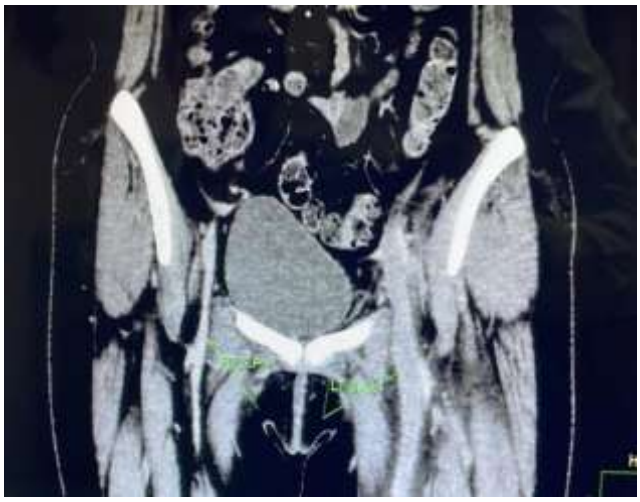

**Figure 2: CT venography image of distension with non-opacification of the veins of the left lower limb with significant attenuated distal left common iliac vein traversing deep to the right common iliac artery representing May-Thurner syndrome.**

## DISCUSSION

MTS leads to venous blockage and pressurization of arterial and venous systems against bones. It presents with a myriad of presentations, frequently found to develop narrowing of the left common iliac vein sandwiching between right common iliac artery and lumbar spine. Other presentations include blockage of the inferior vena cava (IVC) flows by the right common iliac artery.[1-3] Pathogenesis of MTS leading to development of DVT is due to a constant throbbing compression of the common iliac vasculature on the left common iliac vein that leads to stasis of blood, thereby forming thrombus, bands, anciently known as "spurs".[4] These thromboembolic spurs develop due to persistent compression of the veins which causes collection of

collagen and elastin in walls of capillaries causing impeding blood flows and platelet aggregating thrombus. Mechanisms have been attributed to enhanced risk of hypercoagulability like congenital preponderance, mural thrombi, compression of arteries leading to fibrosis.[5,6]

Studies have pointed out to the fact that structural variance of compression of left common iliac vein by the right common artery holds a prevalence of 22 to 32% in autopsied patients.[7] MTS with blockage of inferior vena cava by the right common iliac artery has been seen to be seen rarely.[3]

In a retrospective study done including 79 patients with MTS and secondary DVT, only 1.2 percent incidence was seen of a double lower extremity DVT, whereas majority of the patients had left-sided MTS. In this study, also a total of 79% individuals of non-MTS group and 51% of the MTS group had PE. [8]A case-control study that patients having one-sided DVT, and those with same-sided common iliac vein lumen diameter of less than 4 mm have lower risk for formation of PE.[9] Catheter based contrast enhanced venography has been considered as an investigation of choice for MTS. However intravenous venous ultrasound is regarded as the gold standard for confirming MTS.[10] A similar case of was seen in a 27-year-old individual of Sturge weber syndrome, had presented with a recurrent, nonhealing left lower extremity venous ulcer, was found to have an aplastic left iliac vein, with concomitant MTS. Although there has been a paucity of literature, Sturge weber syndrome can also be a predisposing factor for development of vascular malformations and DVT.[11] Majority of patients with MTS have DVT (77%), whereas without DVT (23%).[12]

In our case although autoimmunity was also present, or some underlying unmasked connective tissue disorder which might have also added to hypercoagulability. Connective tissue disorders have been implicated to cause thromboembolism owing to its inflammatory nature which enhances tumor necrosis factor α (TNF-α) and decreasing protein C.[13] In a metanalysis it was seen that there was a 7.29% cumulative incidence of VTE in lupus, whereas 2.18% in patients with Sjogren's syndrome.[14]

Treatment of thromboembolic cascade is always anticoagulant therapy. Placement of an IVC filter can prevent formation of PE, but it is invasive and expensive. The anticoagulants must be continued for a duration of 6-12 months.[15] Timely mechanical thrombectomy and stenting can help in prolonging life and reducing mortality.[16] Hydroxychloroquine has shown good results in decreasing VTEs in patients with CTD.[17]

## CONCLUSION

MTS is still a very rare entity included as an etiology of DVT along with PE in young women. A prompt management with mechanical thrombolysis and stenting can be safeguarding for a patient's life. Occurrence of DVT also warrants investigating the patient thoroughly for development of other causal agents like autoimmunity and hypercoagulable states.

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