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A Rare Case of Thrombotic Thrombocytopenic Purpura with Normal ADAMTS13 Activity Accompanied by Multiple Brain Infarction and Left Ventricle Thrombus

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Case Study: A 66-year-old male was admitted to emergency department with symptoms of dyspnea and decreased mental status. The laboratory study showed elevation of creatinine level combined with severe thrombocytopenia and positive finding for microangiopathic hemolytic anemia (MAHA). The brain imaging study revealed multiple embolic brain infarctions with focal hemorrhagic transformation. To further evaluate the cause of brain infarction, the transesophageal echocardiography was done, which showed thrombus in his left ventricle. With a suspected diagnosis of thrombotic thrombocytopenic purpura (TTP), the patient was immediately treated with plasma exchange therapy along with steroid pulse therapy.

This case has its significance in two aspects. First, despite appropriate clinical manifestation, the ADAMTS13 (A Disintegrin And Metalloprotease with a ThromboSpondin Type 1 motif, member 13) activity of the patient was 51%, which was within normal range. Additional ADAMTS13 gene sequencing to examine any gene mutation which could be related to congenital TTP had no positive finding. Since there was no complaint of gastrointestinal symptoms, typical hemolytic uremic syndrome (HUS) was less likely, nor was atypical HUS as the additional gene study did not show significant gene variations related to primary atypical HUS. Second, although there have been many cases of thrombotic microangiopathies (TMAs) where patients presented with coagulopathy in small vessels due to endothelial damage, in our case, the patient showed left ventricle thrombus, which is uncommon as compared to other previous reports.

After plasma exchange, the patient recovered his renal function, platelet count and his initially altered mental status improved gradually. Moreover, the left ventricle thrombus and bilateral pleural effusions resolved only with plasma exchange without use of either anti-platelet agents or anti-coagulation therapy.

Figure 1. Tubular-shaped space-occupying thrombus in the left ventricle mid portion

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