Systemic Lupus Erythematosus with Thrombotic Thrombocytopenic Purpura

Sir,

A young male presented with history of fever, bleeding from multiple sites, acrocyanosis, hemolytic anemia with schistocytes, indirect hyperbilirubinemia, central nervous system and renal involvement with SLE features. Patient was diagnosed as SLE with TTP. The case with relevant literature is reviewed.

24 year old male, preserited with history of cough with expectoration and fever since 2-3 months, and hemoptysis, hematemesis and hematochaezia since 2-3 days. Patient had no other significant history. On examination, patient had pallor, icterus, purpuric lesions over face, hands and legs along with ecchymosis over palms and soles. There was ulceration over the hard palate. Patient had alopecia, malar rash and acrocyanosis of limbs. Systemic examination revealed mild hepatomegaly. Patient was drowsy with no focal neurological deficit and both the plantars were extensors. On investigation, patient had hemoglobin - 7.8 gm%; reticulocyte count - 5%; TLC - 5,200/cumm; Polymorphs - 50, Lymphocytes - 30, monocytes - 2, myelocytes - 18 and schistocytes were present on peripheral smear. Platelet count was - 6,000/cumm. T. bilirubin - 4 mg% with D. bilirubin - 1.8 mg%. Urine routine showed albuminuria (2+); PT and APTT were normal. HIV and HBsAg were negative. ANA was positive. Anti dsDNA was negative. Direct and indirect Coomb’s tests were negative. Bone marrow aspiration was normal. C3-35 mg/dl and C4 < 10 mg/dl. Patient could not afford FDP, anti-platelet antibody, aCL antibody and LA, hence were not done.

Thus this patient had hemolytic anaemia with thrombocytopenia with thrombotic event with fever, central nervous system involvement and probably renal involvement. Also patient fulfilled the criteria for SLE-fever, alopecia, oral ulcers, hemolytic anaemia, ANA positivity and low C3 and C4. Hence a diagnosis of SLE with TTP was made. Patient was treated with pulse methylprednisolone, FPPs and platelets. Patient deteriorated in the ward and had a sudden death, the cause of which was not known. A postmortem was performed. The brain showed petechiae and hemorrhages along with one in-situ thrombus in the vessel with minimal surrounding necrosis. Lungs showed subpleural and intra-alveolar haemorrhages with no thrombi. The kidneys showed minimal mesangial proliferation with mild increase in matrix with cloudy tubules suggestive of Type IIa lupus nephritis. The colon showed presence of a clot.

A final diagnosis of SLE with TTP was given based upon the above mentioned findings.

TTP is a diffuse disorder of microcirculation characterized by fever, renal dysfunction, transient and fluctuating neurologic manifestation, microangiopathic hemolytic anaemia (MAHA) and thrombocytopenic purpura. It is a rare disorder, usually seen in women aged 10-40 years following a prodromal illness HSM is seen in 20% of patients. Peripheral smear shows red cell fragments, schistocytes, and helmet cells suggestive of hemolysis. Other features suggestive of MAHA are reticulocytosis, indirect hyperbilirubinina, hemoglobinuria, nucleated red cells and increased LDH. Coagulation parameters are normal except increased FDP. Histopathology shows hyaline thrombi and microthrombi in arterioles and capillaries. Mortality is 50-80%. Various treatment modalities used are steroids, antiplatelet drugs, plasma infusion splenectomy and plasmapheresis.

The association of TTP and SLE is highly debated. It has been recognized in the medical literature since 1939.1 Jonsson OG and Fink CW2 described a 10 year old who presented with TTP and shortly thereafter developed SLE. This is the first report of SLE presenting TTP. Forty cases of TTP in association with SLE are reported in world literature.3 Review of the literature revealed that TTP might occur in the setting of either active or inactive SLE. TTP may be caused by many factors, one of which is SLE. It can present as a terminal event in SLE or SLE has also been shown to initially present several years after TTP. Pediatric TTP with high-grade proteinuria is a predictor of SLE.

Etiology of TTP is elusive. It is unknown if the endothelial damage represents the first lesion or if the platelet hyperaggregability precedes the vascular injury. Certain autoimmune mecanisms, platelet abnormalities, fibrinolytic disorders may be shared with SLE and provide the basis for their association.3 Also a case of primary APLA with TTP has been described in pregnancy. Relationship between these two is controversial. Montecucco et al 4 measured APLA in nine patients with active TTP, eight had primary TTP and one had secondary to SLE. No patient showed circulating LA or false positive test for syphilis.

Autoantibodies to CD36, a platelet glycoprotein have been found in patients with TTP and in those with lupus like anticoagulant with thrombotic complications.5 Koyama T et al described immunological changes in cases of TTP.6 One case that was diagnosed TTP complicated with SLE showed some serological abnormalities of LE test, anti-sterptokinase anti-DNA antibodies, and platelet bound IgG and serum level of IgG by renal biopsy with little changes in complement levels. Platelet associated IgG (PA IgG) was positive in four of six cases and this seemed to be the only abnormality associated with remission and relapse of TTP.

Survival in SLE associated TTP correlated with the use of plasma therapy (infusion/plasmapharesis) rather than with the activity of the underlying autoimmune disease. Patients treated with plasmapharesis/infusion had a lower mortality rate at 25% as compared with 57% mortality in those who were not treated with plasmapharesis. Addition of cyclophosphamide to treatment with plasmapheresis and steroids was beneficial. Management requires timely diagnosis and aggressive treatment by therapeutic plasma exchange.

SS Suralkar*, LS Bichile**, AS Sonawale*, AG Rajyadhyaksha***, Guruprasad#

*Lecturer; **Professor and Head; ***Associate Professor;
REFERENCES


