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A rare case of systemic lupus erythematosis with severe thrombocytopenia leading to Terson syndrome

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Abstract: *A rare case report of SLE with Terson syndrome was made when a young woman of reproductive age presented initially with history of epistaxis , bleeding gums ,2 episodes of blood stained vomitus, melena, menorrhagia, hematuria , generalized rash , headache, and low grade fever since 3 days. She was referred to eye department for defective vision both eyes. UCVA both eyes was 6/36. Anterior segment examination revealed defective abduction in right eye with a fundus picture of preretinal and sub retinal haemorrhages over the posterior pole of both the eyes .Complete blood investigation including ESR, ANA profile, Platelet count and CT brain was done and a diagnosis of SLE with Secondary immune thrombocytopenia leading to Terson syndrome. She was started on I.V. Methyl prednisolone and followed by oral prednisolone, oral acetazolamide .On follow up her vision improved to 6/9.*

Keywords*: systemic lupus erythematosis , Secondary immune Thrombocytopenia, Terson syndrome.*

I. Introduction

Terson syndrome was originally defined by the occurrence of intraocular hemorrhage in association with subarachnoid hemorrhage. Intraocular hemorrhage includes pre retinal, sub retinal, sub hyaloid , or vitreal bleed .The classical presentation being sub hyaloid haemorrhage.

II. Case report

19 year old female admitted with history of epistaxis, bleeding gums, 2 episodes of blood stained vomitus, melena, menorrhagia, hematuria, generalized rash , headache, and low grade fever since 3 days. She gave history of fever with rashes and treated as varicella 2 weeks before.

Her UCVA on admission was 6/36 BE, with restriction of abduction in the right eye.

Fundus examination both eyes showed a clear media, hyperemic optic disc with blurred margins. Cup was filled up and hemorrhages seen over the disc margins. Multiple pre retinal and sub retinal haemorrhages seen over the posterior pole and hemorrhages also noted in the macular region [Fig 1]

On General Examination she was found to have petechial rashes all over the body and soft palate, with bleeding gums and oral mucosal erosions. Investigations revealed an elevated ESR of 105 mm/hour, positive CRP suggesting inflammation and a prolonged Bleeding time of 20 minutes. She had a normal Prothrombin time, INR and Activated partial thromboplastin of 10.9, 0.7, 33.5 respectively. Her platelet count was 7000/c.mm indicating severe thrombocytopenia, ANA profile analysis revealing high positive ANA titres (1:1000 dilution), positive Anti smith of 24 Ru/ml which is a specific marker for SLE, positive Anti RNP of 24Ru/ml, positive Anti SSA-Ro of 180 RU/ml. Rheumatoid factor was negative. A diagnosis of SLE leading on to a secondary Immune thrombocytopenic purpura was made. MRI Brain revealed Bilateral fronto parieto temporal and Right Occipital Sub dural haemorrhage with increased intracranial pressure [Fig 2]. This association of sub dural haemorrhage with pre retinal and sub retinal haemorrhages suggested the possibility of Terson syndrome in this case of SLE. She was treated with Inj. Methyl prednisolone 1 gm I.V. OD for 3 days, oral prednisolone 45 mg od x 10 days, Inj Mannitol 20% 100ml tds infusion run over 20 minutes, oral acetazolamide 250 mg bd, oral Sodium valproate 200 mg tds, I.V. Antibiotics as per Neurologist advice. Blood transfusion in the form of whole blood and platelets were transfused. The UCVA of both eyes improved to 6/9 with improvement in RE abduction.

III. Discussion

Terson syndrome refers to an association of intracerebral haemorrhage with intraocular haemorrhage. Most common cause is a ruptured aneurysm. A primary increase in intracranial pressure is transmitted through optic nerve sheath to optic nerve head, which in turn occludes the retinal venous system, resulting in rupture of the superficial retinal vessels & intraocular bleeding.[3,4]In Terson syndrome blood typically clears completely & usually visual acuity returns to normal. [2] In some cases it may lead to complications such as epi retinal membrane rarely RD which may require surgical intervention.[5]

SLE is an autoimmune disease in which tissue binding auto antibodies & immune complexes are responsible for the damage. 90% of cases are women of child bearing age. Eye involvement in the form of ocular surface epitheliopathy secondary to Keratoconjuctivitis sicca, stromal keratitis (rare), episcleritis,

scleritis, retinal vasculitis can occur in SLE. Retinal vasculitis is the second most common ocular manifestation of SLE next to Keratoconjuctivitis sicca.25 to 50 % of patients with SLE usually have a mild thrombocytopenia with platelet count between 1,00000 to1,50000/micro L. Counts less than 50,000/micro L occurs only in 10% of SLE. [7] Immune mediated platelet destruction is most often the cause in SLE. Thrombocytopenia also serves as a marker of severe systemic disease. [8]

In our case, the platelet count is less than 10 thousand which is a rarity and also indicates the severity of the systemic disease. This severe thrombocytopenia is responsible for causing both intra cerebral and intraocular haemorrhage, favoring the diagnosis of Terson syndrome, which is again a rare association with SLE. An association of SLE with Terson syndrome has not been reported to the best of our knowledge.

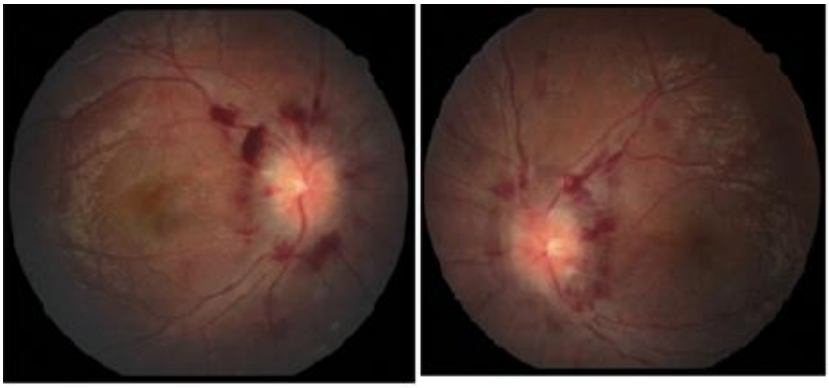

Figure 1: Fundus photo both eyes showing retinal haemorrhages

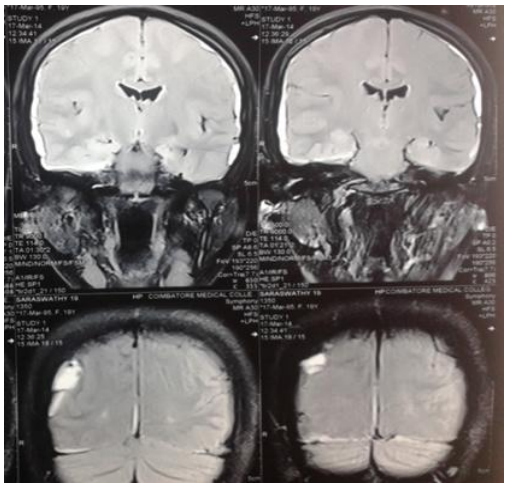

Figure 2: MRI brain coronal section T2W flair showing Right fronto temporo parietal SDH (maximum thickness - 10mm) with midline shift 3mm.

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