ORIGINAL RESEARCH PAPER

General Medicine

A CASE OF TTP COMPLICATED WITH SEPTICEMIA AND PULMONARY HAEMORHHAGE : IN A TERTIARY CARE HOSPITAL SOUTH GUJARAT.

KEY WORDS: THROMBOTIC THROMBOCYTOPENIC PURPURA , PLASMIC SCORE , NEUROLOGICAL MANIFESTATION

Dr.Sanket Thakkar	3rd Year Resident Doctor
Dr.yash Thakkar	2nd Year Resident Doctor
Dr.uttam Gondaliya	2nd Year Resident Doctor
Dr. Ashvin Vasava*	Associate professor , government medical college , surat*Corresponding Author

INTRODUCTION

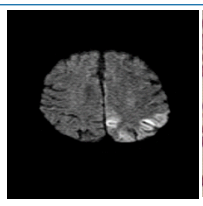
Thrombotic thrombocytopenic purpura (Moschocowitz's syndrome or T.T.P.) is a micro-angiopathy characterised by thrombocytopenic purpura, intravascular hemolysis, fluctuating neurological manifestations and renal failure.[6] it is more common in females and occurs around the age of 40 years. It is a rare disease and is fatal in 50 to 80 per cent of cases. It is thought to be due to increased platelet adhesiveness secondary to reduce prostacyclin production by the vascular endothelium.[4],[6] Very few cases have been reported in the Indian literature.[8],[9] The present paper describes one such rare case admitted under our care at the government medical college,surat.

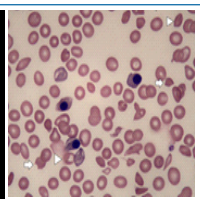
CASE DETAILS :

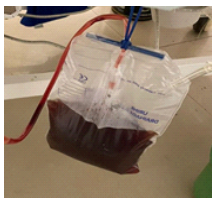
A 39 years old female presented with history of Fever,Nausea, vomiting,loose stool since 1 month.Fever was low grade, intermittent;loose stool were watery,large in quantity,5-6/day.This was followed by Right sided convulsions and post ictal Loss of consciousness.After which she was admitted in local CHC where primary treatment done with 2 unit PRBC transfusion and she regained consciousness & discharged on anti epileptic drugs .After 7 days she again developed Rt sided convulsions followed by loss of consciousness but this time despite of primary treatment she was unconscious and then she was taken to Gmc, surat on arrival her pulse-100/min , respiratory rate-28/min, blood pressure in supine position - 90/50.There were petechial & echhymotic rashes all over the body but more on extremities. she also had puffiness of the face & edema of feet.she was pale .

On respiratory system examination showed that Right lower zone coarse crepitation .on central nervous system examination pt was unconscious ,she responded to painful stimuli by moving all four limb. Hypertonia in Right upper limb and lower limb. Bilateral planter were extensor. Peripheral smear showed severe anemia(Hb-6.1gm/dl) with RBC morphology showing schistocytes10-12/100 Rbcs, thrombocytopenia (platelet count -29000/CMM) Rc-6.5%,indirect bilirubin -3.5mg/dl,s creatinine - 1.38mg/dl,urea-108 mg/dl,LDH-3811 U/L,direct coombs testnegative, ANA -Negative ,FDP-positive, urine colour -red to brown with clot ,urine routine and micro showed albumin +3 and 80-100 Rbcs/hpf. MRI brain showed left occipital and posterior parietal post ictal edema. plasmic score was 7/7 suggestive of severe ADAMTS13 def. Bringing The diagnosis of TTP in favour. At the same time pt's GCS was worsening,so pt was intubated and urgent plasmapheresis and steroids were started. Even after 5 cycle of plasmapheresis there was no significant improvement noted.

Her subsequent hemogram and metabolic panel showed worsening of intravascular hemolysis in form of falling Hb and rising LDh despite of multiple PRBC transfusion.







DISCUSSION AND CONCLUSION :

The diagnosis of TTP is based on the presence of hemolysis, thrombocytopenia, neurological involvement and renal failure.[6] The other common clinical features seen in more than 90% of the cases include fever, anemia, petechiae and erythema, all of which were present in this case. The age and sex of this patient matched with those commonly affected by this disease. The biochemical, hematological and immunological investigations were compatible with the diagnosis of TTP. Worsening of neuroloical and hematological parameter may be due to late presentation leading to late plasmapheresis. The only differential diagnosis to be considered was adult hemolytic uremic syndrome (HUS). Onset with fever and gastroenteritis, no progression into chronic renal failure and elevated fibrin degradation products would favour this diagnosis. However, when HUS occurs in adults, it is in relation to pregnancy or oral contraceptives.[6] The diagnosis of TTP is more likely because of persistence of fever, neurological manifestations dominating the clinical picture and lesser degree of renal involvement.[2] The precipitating or causative factors implicated include pregnancy, surgery, antibiotics, oral contraceptives and abortions. This patient had a preceding febrile illness with gastroenteritis.[3] TTP has a worse prognosis than HUS. Among those who survive the acute episode, chronic renal failure develops within 9 months. This patient needs to be carefully followed up to check for its occurrence. Plasma exchange offers the best mode of therapy for this disease.[5] This probably acts by replacing a prostacyclin-stimulating factor which is missing in patients with TTP. As a result, the increased platelet adhesiveness is reversed.[1],[4] When used with fresh plasma, corticosteroids and anti-platelet agents, patients may show dramatic improvement. Splenectomy is reserved for refractory cases.[7]

early presentation and early diagnosis could have saved the life of patient.

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