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THROMBOTIC THROMBOCYTOPENIC PURPURA MIMICKING CEREBRAL VENOUS THROMBOSIS

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ABSTRACT

A 21-year-old female was admitted with GTCS due to thrombosed cerebral cortical vein . Laboratory tests showed hemolytic anaemia and thrombocytopenia. Thrombotic thrombocytopenic purpura (TTP) was diagnosed. Therapeutic plasma exchange (TPE) was performed with complete resolution of symptoms. The gold standard treatment of TTP is TPE, and its delay can be fatal.

Key Words: Thrombotic Thrombocytopenic Purpura (TTP), Ischemia, Haemorrhage, Anaemia.

INTRODUCTION

Case report

A 21 year old female had presented to our emergency department (ED) with fever and headache for last 3 days with 3 intermittent episodes of GTCS, not associated with vomiting. She had family history of consanguineous parentage (2nd Degree). She was blind since birth and had persistent swelling of legs for last 6yrs. Her only sibling (younger brother) is also blind and also has swollen legs.

On examination, blood pressure was 140/80 mm Hg, pulse 94 beats/min regular, respiratory rate 14 breaths/min, and temperature 38.0 °C. She was conscious, fully alert , moving all four limbs, had bilateral phthisis bulbi , No limb or facial weakness was observed and had Bilateral non-pitting leg edema upto knees (figure 1).

CT scan brain showed venous haemorrhage in left occipital cortex (Figure 2) , later on MRI including MR Venogram showed thrombosed cortical vein in left occipital lobe with haemorrhagic infarct (figure 3).

Laboratory tests revealed were Hb-12.4g%, PCV - 36%, TLC – 8600/cumm; N- 60%, L-31%, M-8%, E-1%, Platelet count 111,000/cumm, RBS 111 mg / dl, PT/INR 13.5/1.18. Kidney and liver function test were normal. Patient was started on Inj. Phenytoin, Inj. Heparin (5000 units IV bolus followed by 1000 units per hour), Inj Mannitol and simultaneously started on tablet warfarin. Her headache subsided and no seizure recurred.

While under treatment, 5days later, she developed intense headache with right hemiplegia. Repeat NCCT scan showed increase in size of left occipital bleed extending to parietal lobe with significant mass effect and significant SAH on right fronto-parietal lobe (figure-4).
Many more problems developed, there was drop in Hb from 12.4 →7.5, drop in platelet count 111,000 → 14,000, S. Creatinine raised 0.8 → 5.8 And fall in urine output over the next 24hrs progressing to anuria .There was no fever, hematemesis or malena.

PBF showed - Numerous Schistocytes and low platelet count. Suspecting of intravascular haemolysis, LDH was done which was significantly raised (6983).

The intravascular haemolysis, thrombocytopenia, fluctuant neurological findings and ARF were consistent with a diagnosis of TTP. The patient was treated with hemodialysis, plasmapheresis and regular FFPs. Heparin and Warfarin were stopped. Complete remission was achieved on day 21 after admission, and she was discharged on Aspirin and Phenytoin.

DISCUSSION

TTP is a life threatening disease, characterised by

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thrombocytopenia, microangiopathic hemolytic anaemia, fluctuating neurological signs, renal failure, and fever. The condition is rare with an annual incidence in adults of 3.7 per million. It is seen predominantly in women, usually between 30 and 40 years of age-[1].

The pathogenesis of TTP is attributed to the presence of unusually large von Willebrand factor (vWF) multimers that lead to platelet clumping and subsequent microvascular thrombosis. These vWF multimers are normally cleaved into smaller protein units by a metalloprotease enzyme, the von Willebrand factor cleaving protease (ADAMTS- 13). Impairment of ADAMTS- 13 activity leads to an excessive accumulation of vWF, which then may lead to the onset of TTP. TTP is mostly idiopathic, but may be triggered by clinical situations such as bacterial or viral infections, pregnancy, drugs (for example, clopidogrel, ticlopidine, quinine, ciclosporin), and autoimmune disorders (systemic lupus erythematosus, thyroiditis, and antiphospholipid syndrome) [2, 3].

Identification of the first acute TTP episode is challenging as no specific clinical symptom or biological criterion is available for the diagnosis. Suspicion of TTP relies on the association of several clinical symptoms and laboratory results. The brain is the commonest target for ischaemia, and abnormal neurological findings present in most cases of TTP. Patients usually develop headache, confusion, ataxia, seizures, and mental status and focal abnormalities. Other frequent symptoms are fever, weakness, arthralgia, myalgia, jaundice, nausea, vomiting, diarrhoea and abdominal pain. Skin and mucosal bleeding secondary to the thrombocytopenia is also common [2, 3]. Patients may have renal abnormalities including oligoanuria, acute renal failure, albuminuria, and microscopic haematuria. Consumption thrombocytopenia is present in most cases, with platelet counts often below $20\,000 \times 10^9$. The widespread deposition of fibrin in the vessels leads to mechanical trauma to the passing red blood cells, resulting in fragmented red blood cells on the blood smear. Haemoglobin is usually below 80 g/l (8 g/dl) secondary to haemolytic anaemia. Elevated lactate dehydrogenase and bilirubin levels may represent intravascular haemolysis [1-3].

Currently plasma exchange treatment is indicated for all adult patients with a clinical diagnosis of TTP. Since the availability of plasma exchange treatment, mortality has fallen from 90% to about 10%. As delay in the initiation of plasma exchange correlates with treatment failure, therefore early diagnosis and treatment is essential [4].

Fig 1. Showing Bilateral Pedel edema upto Knee

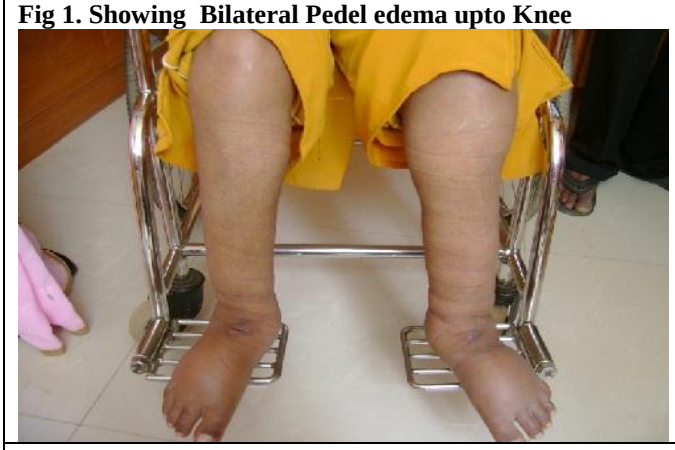


Fig 2. CT scan brain showed venous haemorrhage in left occipital cortex

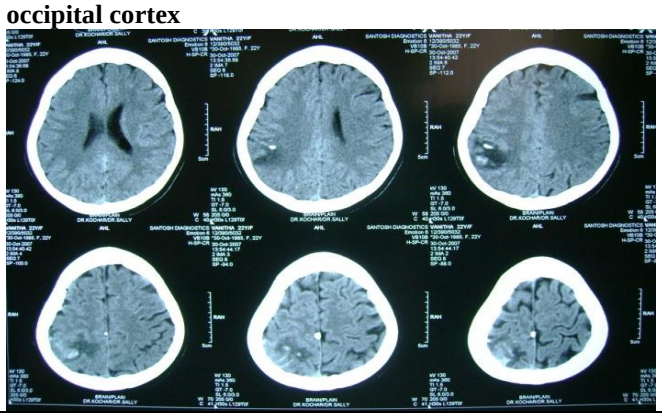


Fig 3. MR Venogram showing thrombosed cortical vein in left occipital lobe with haemorrhagic infarct.

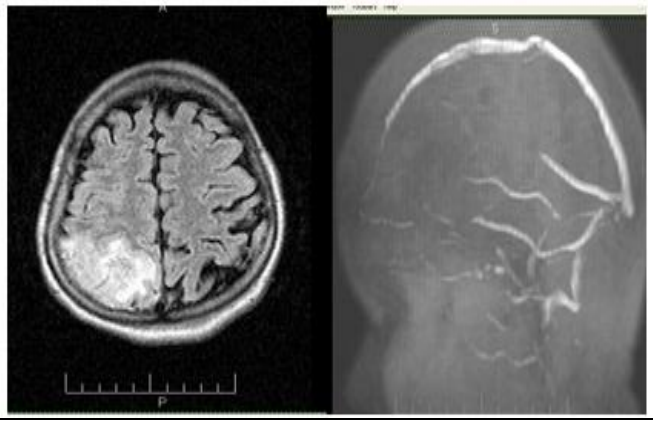
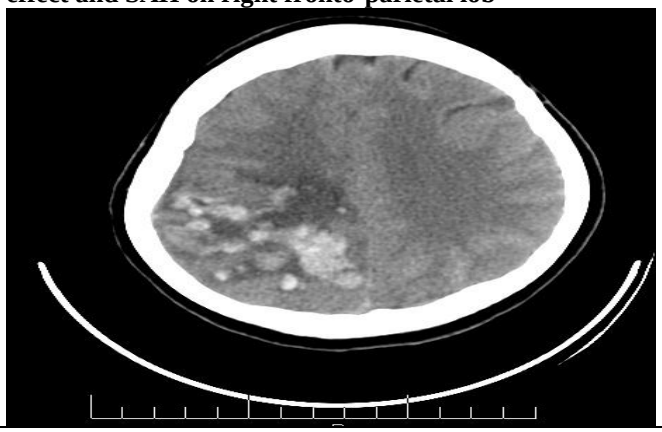


Fig 4. CT head showing left occipital bleed with mass effect and SAH on right fronto-parietal lob



CONCLUSION

Rapid diagnosis and treatment are necessary for decreasing the risk of fatal outcome in patients with TTP. Emergency physicians should be familiar with the clinical presentation and laboratory abnormalities of TTP to be able to make the diagnosis early on. In patients presenting with fluctuant neurological findings, thrombocytopenia, heamolytic anemia, and fever, the diagnosis of TTP should be considered.

STATEMENT OF HUMAN AND ANIMAL RIGHTS

All procedures performed in human participants were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. This article does not contain any studies with animals performed by any of the authors.

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Nil

CONFLICT OF INTEREST

No interest

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