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A CASE OF REFRACTORY THROMBOTIC THROMBOCYTOPENIC PUPURA

Internal Medicine

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ABSTRACT

Thrombotic thrombocytopenic purpura (TTP) is a rare blood disorder characterized by clotting in small blood vessels (thromboses), resulting in a low platelet count. In its full- blown form, the disease consists the pentad of microangiopathic hemolytic anemia, thrombocytopenic purpura, neurologic abnormalities, fever and kidney disease

KEYWORDS

Thrombotic thrombocytopenic purpura, microangiopathic hemolytic anemia, thrombocytopenic purpura, neurologic abnormalities, fever and kidney disease

INTRODUCTION

Thrombotic thrombocytopenic purpura (TTP) is a rare blood disorder characterized by clotting in small blood vessels (thromboses), resulting in a low platelet count. In its full- blown form, the disease consists of the following pentad [1]:

- Microangiopathic hemolytic anemia
- Thrombocytopenic purpura
- Neurologic abnormalities
- Fever
- Kidney disease

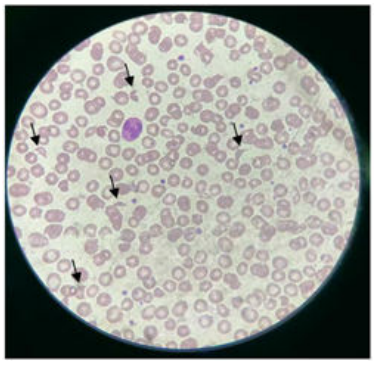
Here is a case report of a patient with TTP with severe deficiency of ADAMTS13 refractory to treatment.

Case

71 year old male with a history of fever since 12 days, cough with minimal expectoration since 10 days, abdominal pain since 10-15 days and 2 episodes of GTCS followed by decreased responsiveness. Intubated electively in view of low GCS.

- Routine evaluation was done and blood investigations revealed low HB, low platelets ,elevated LDH and indirect hyperbilirubinemia. Peripheral blood smear showed anisopoikilocytosis with schistocytes- 7.4% confirming that patient had developed MAHA.
- Possibility of autoimmune hemolysis was ruled out as coombs test (direct and indirect) was negative, hence ruling out possibility of AIHA.
- The 3 causes for MAHA i.e. HUS,TTP & DIC were considered.
- DIC was ruled out as Fibrinogen, APTT, PT, INR levels were within normal range & HUS was ruled out as patient didn't show any signs of renal involvement.

Investigation

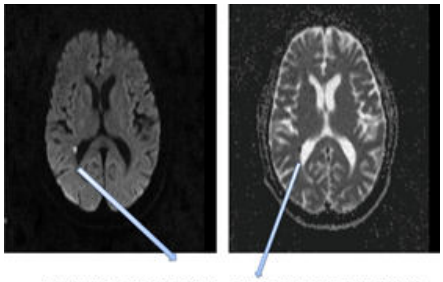


After evaluation a diagnosis of TTP was considered and TMA (Thrombotic microangiopathic) panel done was to evaluate the cause and stratify the severity of TTP.

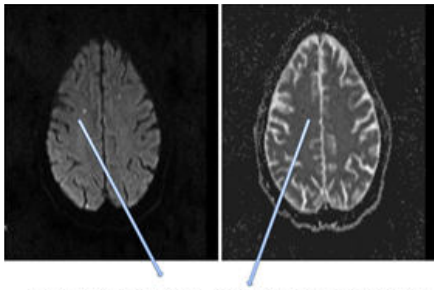
PERIPHERAL SMEAR 25/09/2023
• **RBCs show anisopoikilocytosis.**
• **RBCs are both microcytic hypochromic and macrocytic.**
• **Many polychromatophils are seen.**
• **nRBCs (10/100 WBCs) are seen.**
• **Microspherocytes and fragmented RBCs are seen.**
• **Schistocyte count = 7.4%**
• **There is a decrease in total red cell mass.**
• **WBCs show increased total count with increase in absolute neutrophil count.**
• **Neutrophils appear reactive.**
• **DC = Neutrophils 84%, Lymphocytes 15%, Eosinophils 00%, Monocyte 01%, Basophil 01%.**
• **Platelets are markedly reduced.**

NAME	VALUE	REFERENCE RANGE
ADAM TS 13 FACTOR ACTIVITY LEVELS	< 1 %	40-130%
ADAM TS 13 INHIBITOR LEVELS	80.28 IU/ml	Negative < 12IU/ml Positive> 15 IU/ml
HUMAN COMPLEMENT FACTOR H ASSAY	832.886 ug/ml	60-800 ug/ml

MRI brain with angiography-Acute lacunar infarcts in right frontoparietal lobe, right peritrigonal region, bilateral centrum semiovale, cerebellar vermis and left cerebellar hemisphere.



DWI AND ADC IMAGES - ACUTE INFARCT IN RIGHT PERITRIGONAL REGION

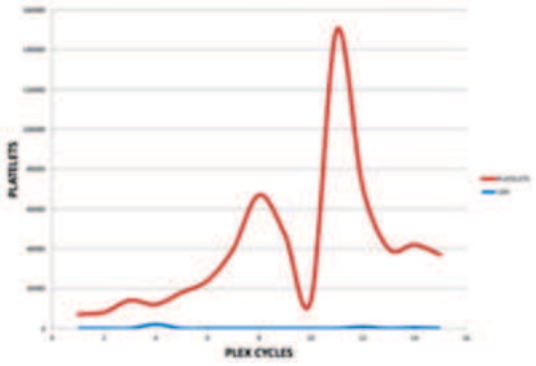


DWI AND ADC IMAGES - ACUTE INFARCT IN BILATERAL FRONTOPARIETAL LOBES

Treatment

- Patient was started on PLEX along with IV methylprednisolone (1.5 gm X 3 days).

- Once daily PLEX was done for 7 days. Over the course of 7 cycles elevation of platelet level from 7,000 (24/09/2023)to 40,000(01/10/2023) was noted.
- Post 7 cycles of once daily PLEX platelet levels were 67,000 and patient was planned for rituximab infusion on 02/10/2023.
- Patient was initiated on single antiplatelets i.e. Tab. Ecosprin 150mg OD as platelets were above 50,000.
- Rituximab infusion a total dose of 600mg was administered and consecutively platelets were monitored. Once weekly for four weeks.
- However on consecutive days drop in Platelets were noted and patient was resistant to treatment with PLEX and rituximab.



CONCLUSIONS

- The diagnosis of TTP should be treated as a medical emergency.
- Due to the high risk of preventable early deaths in TTP, treatment plasma exchange should be initiated as soon as possible.
- Pre- treatment samples should be obtained to measure ADAMTS13 activity levels and to detect anti-ADMATS 13 antibodies.
- Platelet transfusions are contraindicated in TTP unless the patient has life-threatening haemorrhage.
- In cases of severe TTP and patients presenting with neurological symptoms starting of PLEX and Rituximab has better response rates and prognosis.

REFERENCES:

1. Stanley M, Killeen RB, Michalski JM. Thrombotic Thrombocytopenic Purpura. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 [cited 2024 Aug 6]. Available from: http://www.ncbi.nlm.nih.gov/books/NBK430721/