Case Report

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Thrombotic Thrombocytopenic Purpura: Atypical Presentation in Er/An Approach to be Considered by Er Physicians

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Abstract

To Consider the Diagnosis of TTP in cases of Thrombocytopenia Presenting to Er with altered mental status. A 28 year young male known case of seizure Disorder Juvenile Myoclonic Epilepsy Presenting to Er with complaints of sudden onset of altered mental status since 1 day with history of fever for 1 day Documented 3 days back and his labs revealed thrombocytopenia with Aki. Patient was initially managed conservatively with Antiepileptics, IV fluids, Antibiotics and supportive therapy. In course of hospital stay, patient developed Hemolysis with generalised tonic clonic seizures and was later diagnosed as TTP (Sepsis Induced) with Neurological Involvement with AKI with status epilepticus.

A case of thrombocytopenia with altered mental status presenting in Er, A Differential Diagnossis of TTP must be considered in Initial Period.

Keywords: Sepsis Induced Thrombotic Thrombocytopenic Purpura; Emergency Department; Neurological Involvement; Atypical.

INTRODUCTION

Background

Thrombotic Thrombocytopenic Purpura (TTP) is a type of Microangiopathic Hemolytic anemia that classically has been characterized by the pentad of fever, thrombocytopenia, hemolytic anemia, renal dysfunction, and neurologic dysfunction. TTP results from either a congenital or acquired decrease/absence of the von Willebr and factor cleaving protease ADAMTS 13 (a disintegrin and metalloproteinase with a thrombospondin type 1 motif member. Low levels of ADAMTS 13 result in microthrombi formation, which leads to end organ ischemia and damage.[1,2,3] This is due to the inability of the ADAMTS 13 to inactivate the large multimer von Willebrand factor (VWF) that is necessary to prevent spontaneous coagulation. Unchecked, the large multimers have a tremendous avidity to bind platelets and initiate thrombi formation. The central nervous system (CNS) and kidneys are the two most common organ systems affected by TTP. Timely diagnosis is very important because TTP is a medical emergency which, without treatment, has a mortality of about 90%.

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Patient Information

A 28 year old Male known case of Juvenile Myocloic Seizures living in urban area of Delhi, NCR region.

Clinical Finding and Diagnostic Assessment

28 year Old Male Patient Presented to Er with Altered Mental Status since 1 day sudden in onset, with History of Fever for 1 day documented at home 3 days Back-Low Grade not Asssociated with chills with no particular foci. He was a known case of Juvenile Myoclonal Epilepsy on Antiepileptics.

His General Examination was within normal limits with GCS-E4v1m4, Patient was irritable with no signs of Meningeal Irritation. Patient was haemodynamically Stable and Baseline Labs were sent with Toxicology Screening. Toxicology Screening Turned to be negative and routine labs revelaed rise in TLC with differentials of Neutrophilia and Thrombocytopenia with Aki. In course of Stay, Patient developed drop in HB with Hemolysis Picture in Peripheral Smear (Rwticulocyte Count 5%, LDH-591, Haptoglobin Less than 30, Peripheral Smear revealed shistocytes and Severe Thrombocytopenia Platetelt Count-5000. Blood C/S Staph. Hominis Isolated. Labs were sent for Tropical Diseases Causing Thrombocytopenia Such as Dengue, Scrub Typhus and it all turned out to be Negative. Adams T13 Levels were sent which were low and Lab Panels for Hemolysis were sent such as Ana Levels, Complement Levels, Coombs Test which Turned out to be Negative.

Therapeutic Intervention

Patient was Started on Broad Spectrum Antibiotics in view of sepsis, Dual Antiepileptics was started Dose was stepped UP and Platelets and Prbc was transfused in view of Thrombocytopenia and anemia. Plasma Exchange was done for the patient with steroid cover due to Hemolysis.

In the course of Hospital, Patient Condition Improved Gradually after Plasma cxchange.

DISCUSSION

Zhu H, Liu JY.(11) Reported a Case of TTP with Cerebrovascular Disease with Posterior Reversible Encephalopathy Syndrome.

As the patient did not develop Hemolysis in the initial Hospitalisation, the Approach of the Patient was AFI with thrombocytopenia with AKI under evaluation Prompting towards the tropical diseasescausing thrombocytopenia such as Dengye Fever, Scrub Typhus and also his Past History of Juvenile Myoclonal Epilepsy Prompted Towards Status Epilepticus and Supportively Managed Accordingly.

CONCLUSION

The Patients Presenting to Er with Thrombocytopenia with renal and Neurological Involvement must be considered for differential Diagnosis of Thrbotic Thrombocytopenic Purpura. Atypical clinical Presentation of TTP poses a diagnostic challenge especially in Patients with known Neurological Diseases which causes a delay in Prompt Treatment and also the case report Highlights the Above.

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