SUNTEXT REVIEWS

SunText Review of Case Reports & Images
ISSN: 2766-4589

Open Access
Case Report
Volume 4:8

Management of Life Threatening First Acute Attack of Thrombotic Thrombocytopenia Purpura (TTP) with Multiorgan Dysfunctions and Sequelae Using Pulse Steroids/Plasma Exchange and Monoclonal Antibodies (Rituximab)

Abdelrahman A*, Bilali A, Ahmed M, Vadkout M, Syed S, Nair D, Faisal B, Gupta P and Chetty D

Senior Critical Care Specialist, Lifecare hospital, Abudhabi, UAE

***Corresponding author:** Abdelrahman A, Senior Critical Care Specialist, Lifecare hospital, Abudhabi, UAE

Received date: 27 November 2023; **Accepted date:** 03 December 2023; **Published date:** 10 December 2023

Citation: Abdelrahman A, Bilali A, Ahmed M, Vadkout M, Syed S, Nair D, et al. (2023) Management of Life Threatening First Acute Attack of Thrombotic Thrombocytopenia Purpura (TTP) with Multiorgan Dysfunctions and Sequelae Using Pulse Steroids/Plasma Exchange and Monoclonal Antibodies (Rituximab). SunText Rev Case Rep Image 4(8): 114.

DOI: https://doi.org/10.51737/2766-4589.2023.114

Abstract

TTP is a life-threatening disorder in which the blood clots that form in the small blood vessels throughout the body can limit or block blood flow to the major organs. This, in turn, can damage the organs like the brain, kidneys, and heart and prevent them from functioning correctly. Here, we present a case of a 52-year-old he experienced abdominal pain, mild fever and thrombocytopenia. Things took a serious turn when, he developed seizures altered conscious level, hemiplegia and worsening of thrombocytopenia, he was electively intubated and was on mechanical ventilation till stabilization and improving symptoms.

Keywords: Steroids/Plasma Exchange; Monoclonal Antibodies; Rituximab

Introduction

TTP can affect any organ system, but involvement of the peripheral blood, the central nervous system, and the kidneys causes the clinical manifestations. The classic histologic lesion is one of bland thrombi in the microvasculature of affected organs. These thrombi consist predominantly of platelets, with little fibrin and red cells compared with thrombi that occur secondary to intravascular coagulation.

Patients with thrombotic thrombocytopenic purpura (TTP) typically report an acute or subacute onset of the following symptoms related to central nervous system (CNS) dysfunction, anemia, or thrombocytopenia:

- Neurologic manifestations include alteration in mental status, seizures, hemiplegia, paresthesias, visual disturbance, and aphasia
- Fatigue may accompany the anemia
- Severe bleeding from thrombocytopenia is unusual, although petechiae are common

Clinical manifestations may also include the following:

- Fever occurs in approximately 50% of patients
- Patients may notice dark urine from hemoglobinuria.

Case Presentation

A 52-year-old he presented to emergency department with complaint of abdominal pain, mild fever further evaluation he was found to have thrombocytopenia anaemia and renal impairment suddenly patient developed altered conscious level, hemiplegia and seizures, and was intubated electively and connected to mechanical ventilation. In view of laboratory tests which shows sever thrombocytopenia, anaemia and renal impairment. MRI brain which shows cerebral odema and suspicion of PRESS. Thrombotic Thrombocytopenia Purpura (TTP) – a rare disorder of the blood coagulation system that usually affects 4-6 persons per million population- was suspected and The ADAMTS 13 test

confirmed he had TTP, a rare disease that can be fatal if left untreated. We started Pulse steroid /Plasma exchange and rituximab to rapidly control this blood crises Patient remained hemodynamically unstable and critical under mechanical ventilation and deep sedation to avoid worsening of neurological status .After multiple PLEX and rituximab doses platelet count improved and patient successfully weaned of mechanical ventilation. Patient later extubated and becomes fully conscious oriented laboratory results become better [1-9].

Investigations

N-terminal pro B-type natriuretic peptide 168.20 pmol/lit. Troponin T (High Sensitive) 172.60 pgram/mil. Lactate Dehydrogenase 1472 iu/lit. Lipase 181.20 iu/lit. Hb 8.0 gram/deciliter. Ht 21.90%. Platelets 11000. Blood smear, peripheral: Findings are suggestive of hemolytic anemia with severe thrombocytopenia, probably of microangiopathic hemolytic. ADAMTS13 ACTIVITY less than 2.0%, ADAMTS13 ANTIBODY 65 units per milliliter, Creatinine 199 mmol/ml, Haptoglobin report was 0.03 g/lit .Urea Nitrogen 19.80 mmol/lit. USG abdomen possiblity of medical renal disease, bilateral renal cyst. CT abdomen was suggestive of acute pancreatitis.

Management

A 12-member multi-disciplinary team of doctors to analyze his health condition and plan the next steps. The team designed the treatment protocol and started therapeutic plasma exchange (TPE) – a process of replacement of plasma obtained from healthy individuals. The patient received a total of 10 sessions of TPE daily with almost 30 litres of plasma exchanged. He was also treated with steroids (high dose methylprednisolone 1G IV daily for 5 days than 1 mg/kg IV daily) and monoclonal antibodies. Rituximabis typically given in a dosage of 375 mg/m^2 weekly for 4 weeks. Ideally, at least 4 hours should elapse between administration of rituximab and plasma exchange.

Follow up

The patient to be seen every week for 2 weeks, weekly measurement of a complete blood count and lactate dehydrogenase (LDH) are performed and were good. Regular monitoring of ADAMTS13 activity is also recommended during follow up of patients - with TTP in remission. Patient flyed to home country to continue follow up there.

There is no method of preventing TTP or its relapse. According to doctors, when a patient is diagnosed with this disease, it is better to follow a healthy lifestyle by avoiding smoking and ensuring adequate hydration, a low-carbohydrate, low-salt, low-fat diet, and proper sleep.

Discussion

Patient was admitted in the Intensive Care Unit with chief complaint of abdominal pain associated with vomiting and epigastric tenderness. Patient was suspected to have acute pancreatitis. Blood smear were suggestive of hemolytic anemia with sever thrombocytopenia, probably of microangiopathyic hemolytic anaemia. In view of severe thrombocytopenia and active hemolysis, need for Therapeutic Plasma exchange was explained to the patient. Patient developed altered sensorium and convulsions and developped MODs, So poor prognosis.

Patient was connected to mechanical ventilation and deeply sedated. ADAM TS13 and ADAM 13 antibody was sent, which came back positive. Patient was diagnosed to have Thrombotic Thrombocytopenic Purpura as per the finding and guidelines. The International Society on Thrombosis and Haemostasis (ISTH) issued guidelines on the treatment of TTP in 2020. For the treatment of a first acute event, the panel suggests the addition of rituximab to corticosteroids and TPE, So Patient was continued on plasmapheresis therapy, intravenous high dose of steroids, the anti-CD20 monoclonal antibody rituximab, antiepileptics and supportive care. Patient respond later MODs corrected and platelets count improved. Proper weaning and later successful extubation from mechanical ventilation done.

Conclusion

Management of life threatening Thrombotic Thrombocytopenia Purpura (TTP) complicated with multiorgan dysfunction syndrome and sequelae is life threatening condition, this require multidisciplinary team approach to control multiple organ dysfuction/failure. Treatment of the first acute event, using rituximab, corticosteroids and TPE shows efficacy in inducing remission.

References

1. Stanley M, Killeen RB, Michalski JM. Thrombotic Thrombocytopenic Purpura. 2023.
2. Sukumar S, Lämmle B, Cataland SR. Thrombotic Thrombocytopenic Purpura: Pathophysiology, Diagnosis, and Management. J Clin Med. 2021; 10.
3. Cauchois R, Muller R, Lagarde M, Dignat-George F, Tellier E, Kaplanski G. Is Endothelial Activation a Critical Event in Thrombotic Thrombocytopenic Purpura?. J Clin Med. 2023; 12.
4. Mariotte E, Azoulay E, Galicier L, Rondeau E, Zouiti F, Boisseau P, et al. Epidemiology and pathophysiology of adulthood-onset thrombotic microangiopathy with severe ADAMTS13 deficiency (thrombotic thrombocytopenic purpura): a cross-sectional analysis of the French national registry for thrombotic microangiopathy. Lancet Haematol. 2016; 3: e237-245.
5. Scully M, Thomas M, Underwood M, Watson H, Langley K, Camilleri RS, et al. Thrombotic thrombocytopenic purpura and

pregnancy: presentation, management, and subsequent pregnancy outcomes. Blood. 2014; 124: 211-219.

6. Markham-Lee Z, Morgan NV, Emsley J. Inherited ADAMTS13 mutations associated with Thrombotic Thrombocytopenic Purpura: a short review and update. Platelets. 2023; 34: 2138306.
7. Scully M, Cataland SR, Peyvandi F, Coppo P, Knöbl P, Kremer Hovinga JA, et al. Caplacizumab Treatment for Acquired Thrombotic Thrombocytopenic Purpura. N Engl J Med. 2019; 380: 335-346.
8. Schofield J, Hosseinzadeh S, Burton K, Pavord S, Dutt T, Doree C, et al. Drug-induced thrombotic thrombocytopenic purpura: A systematic review and review of European and North American pharmacovigilance data. Br J Haematol. 2023; 201: 766-773.
9. Wu N, Liu J, Yang S, Kellett ET, Cataland SR, Li H, et al. Diagnostic and prognostic values of ADAMTS13 activity measured during daily plasma exchange therapy in patients with acquired thrombotic thrombocytopenic purpura. Transfusion. 2014.