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Case Report

Thrombocytopenic Thrombotic Pupura Presenting with Neurological Symptoms: A Case Report

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ABSTRACT

Thrombotic thrombocytopenic purpura (TTP) is thrombotic microangiopathy caused by decreased activity of ADAMTS13, a von Willebrand factor-degrading metalloprotease. Here, we present a male patient with neurological symptoms, diagnosed with TTP and successfully treated with plasmapheresis.

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Introduction

Thrombotic thrombocytopenic purpura (TTP) is a thrombotic microangiopathy (TMA) caused by severely reduced activity of the von Willebrand factor-cleaving protease ADAMTS13.[1] Complete pentad includes thrombocytopenia, microangiopathic hemolytic anemia (MAHA), fever, neurological findings and renal failure.[2] However, the complete pentad may not be detected in most patients. In this article, we aimed to present a case of TTP presenting with neurological symptoms.

Case Report

A 53-year-old male patient was admitted to the emergency service with the complaints of slurring of the tongue, headache and weakness. The patient's temperature was 36.2 °C at the time of admission. Cranial computed tomography (CT) or magnetic resonance imaging (MRI) was found to be normal in the patient whose neurological and systemic examination did not reveal any obvious pathology. Complete blood count (CBC) revealed anemia and thrombocytopenia (leukocyte 6,070/mm^3, neutrophil 4,220/mm^3, hemoglobin 8.9

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g/dL, platelet 20,300/mm^3), and features of hemolysis (lactate dehydrogenase [LDH] 821 U/l, total bilirubin 2.9 mg/dL) was detected. In biochemical parameters, creatinine was 1.17 mg/dL and urea was 35 mg/dL. In the peripheral blood smear; schistocytes were seen in 3-4 high power fields and also one orthochromatophilic cell. The platelet count and frequency were consistent with the hemogram. Evaluated with these findings, the patient was admitted to the hematology clinic with a preliminary diagnosis of TTP. The blood sample was stored at -80 °C for the ADAMTS13 test at the patient's admission. Therapeutic plasma exchange (TPE) and 1 mg/kg methylprednisolone were initiated. After 15 sessions of TPE, the platelet count increased to 154,400/mm^3, and LDH levels decreased to 207 U/l. The complaints of slurring of the tongue and headache disappeared. After the patient received 19 sessions of TPE, the platelet count were 183,500/mm^3 and the LDH levels were within the normal range, so it was decided to open the intervals between the TPE sessions. The diagnosis of TTP was confirmed after ADAMTS13 activity was reported as 6.84% (low), ADAMTS13 antigen 0.096 (low), and ADAMTS13 inhibitor 16.07 (high). TPE was performed 2 days a week for 3 weeks. Afterwards, he was discharged with stable vitals and hemodynamics. After discharge, TPE was performed once a week for a total of 5 weeks. Methylprednisolone treatment was tapered and discontinued. The patient is being followed up.

Discussion

Deficiency of the ADAMTS13 is seen in 90-95% of TTP cases. Microangiopathic hemolytic anemia (MAHA) and thrombocytopenia are hallmarks of TTP.[1-3] Neurological symptoms are seen in 39-80% of cases, fever in 27-42%, renal failure in 10-75% of cases, while the complete pentad is seen in only 7%. According to a review of 78 patients with acquired TTP from Oklahoma, published by Page et al.[4] in 2017, 41 (53%) had neurological symptoms, 8 (10%) had fever, 11 (14%) creatinine level ≥2.5 mg. /dL, 4 (5%) had acute kidney injury.[4]

In the first presentation of our patient, neurological findings, microangiopathy and thrombocytopenia were present, but there was no fever and elevated creatinine. In conclusion, considering that complete pentad is seen in only 7% of patients, possibility of TTP should be suspected in any patient presenting with MAHA findings and thrombocytopenia with or without symptoms of organ involvement and without an alternative explanation.

Acknowledgment

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Conflict of interest

The authors declared that there are no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Authors' Contribution

Study Conception: YC, VO; Study Design: YC, BO, VO; Supervision: YC, VO, FO; Materials: YC, BO, VO; Data Collection and/or Processing: YC, BO, FO; Statistical Analysis and/or Data Interpretation: YC, BO, FO; Literature Review: YC, BO, VO; Manuscript Preparation: YC, BO, VO; Critical Review: YC, BO, VO.

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