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Case Report

THROMBOTIC THROMBOCYTOPENIC PURPURA RELATED ACUTE MYOCARDIAL INFARCTION

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Abstract:

Introduction: Thrombotic thrombocytopenic purpura (TTP) is a rare life-threatening thrombotic microangiopathy. The central nervous system, kidneys, and myocard can be affected by TTP.

Case Presentation: A 47-year-old female patient with no chronic disease came to the emergency department presented with chest pain, blur in seeing. Electrocardiogram was performed and there was biphasic T wave pattern. Laboratory tests showed an elevation in white blood cell ($14.10^3/mm^3$), decrease in thrombocyte ($3.000/mm^3$). Blood urea nitrogen (BUN) was 129.3 mg/dl, serum creatinine was 1.87 mg/dl, lactate dehydrogenase (LDH) was 1581 U/l. Cardiac markers were elevated. These tests results revealed non ST-elevation myocardial infarction and acute renal injury. Antithrombotic agents were not applied because of thrombocytopenia. ADAMTS 13 activity was found lower than 0, 2 % and ADAMTS 13 antibodies were positive. Plasmapheresis was started to perform, immediately and repeated for ten days. After the treatment, hematologic parameters and LDH values came to normal ranges.

Conclusion: Thrombotic Thrombocytopenic Purpura (TTP) is a type of thrombotic microangiopathic anemia. If it is not diagnosed early, the mortality rate can be about 90%. Plasmapheresis should begin immediately.

Key Words: Thrombotic Thrombocytopenic Purpura, myocardial infarction, acute renal injury

Introduction

Thrombotic Thrombocytopenic Purpura (TTP) is a type of thrombotic microangiopathic anemia which is manifested with thrombotic microangiopathy, neurological symptoms, fever and renal failure. It is a rare and life-threatening disease. The incidence of TTP ranges from 3 to 11 cases per million per year (1). If it is not diagnosed early, the mortality rate can be about 90%. TTP is classified as acquired which is caused by

anti-ADAMTS13 autoantibodies (disintegrin-like and metalloprotease with thrombospondin type 1 motif 13) and congenital which is caused by a reduced activity of ADAMTS13. The reduced of ADAMTS13 activity results in microthrombosis which cause end-organ ischemia. The central nervous system, kidneys, and myocard can be affected by TTP (2). We presented a 47 years old female patient who has myocardial infarct and cerebrovascular accident secondary to TTP.

Case

A 47-year-old female patient with no chronic disease admitted to the emergency department with chest pain, blur in seeing. Physical examination was performed. Cardiovascular and respiratory system examination was normal. Electrocardiogram was performed and there was biphasic T wave pattern. Laboratory tests showed an elevation in white blood cell ($14.10^3/mm^3$), decrease in thrombocyte ($3.000/mm^3$) and decrease in hemoglobin (11.1 g/dl). Blood urea nitrogen (BUN) was 129.3 mg/dl, serum creatinine was 1.87 mg/dl, lactate dehydrogenase (LDH) was 1581 U/l, c-reactive protein was 51.6 mg/dl, troponin I was 1.97 ng/dl, creatinine kinase (CK) was 221 U/L and CK-MB was 18.1 ng/ml. Prothrombin time and INR values were normal. These tests results revealed non ST-elevation myocardial infarction and acute renal injury. Cranial magnetic resonance was performed and displayed multifocal acute ischemic focuses in cerebral-cerebellar hemispheres. The patient was hospitalized and taken under intensive care unit. Antithrombotic agents were not applied because of thrombocytopenia. Renal functions and electrolytes were followed during hospitalization and replaced by parentral medication. A peripheral blood smear was made due to anemia and displayed schistocytes. It was considered TTP according to these findings. Then, ADAMTS 13 activity was found lower than 0.2 % and ADAMTS 13 antibodies were positive. Serologic tests for acute infection by human deficiency virus, hepatitis A, B, C were negative. Antinuclear antibody (ANA), anti-dsDNA, antineutrophil cytoplasmic antibodies (ANCA), anti-glomerular basement membrane, anti phospholipid antibody, anticardiolipin antibody were negative. Plasmapheresis was started to perform, immediately and repeated for ten days. After the treatment, hematologic parameters and LDH values came to normal ranges. Anti-Islamic therapy was given.

Discussion

Thrombotic thrombocytopenic purpura (TTP) is a rare life-threatening thrombotic microangiopathy. It is characterized by a microangiopathic hemolytic anemia, thrombocytopenia, neurologic abnormalities, and fever and organ ischemia damage due to microthrombosis. TTP is classified as acquired and congenital (3). Inherited form is caused by genetic mutations in the ADAMTS13 gene. Therefore, reduced activity of the von Willebrand factor-cleaving protease (ADAMTS13) reduces. Antibody against ADAMTS13 caused acquired TTP. ADAMTS13 cleaves von Willebrand factor (vWF) multimeric strings. Whether acquired or inherited, reductant of the ADAMTS13 enzyme activity causes that large vWF strings remain uncleaved after endothelial cell secretion and they bind to platelets, so microthrombosis occur. Our patient presented with myocardial infarct and cerebrovascular accident due to microthrombosis. Laboratory tests and peripheral blood smear are important for confirming the diagnosis. We can see schistocytes and thrombocytopenia at peripheral blood smear. Lactate dehydrogenase level increases. ADAMTS13 enzyme activity level and antibodies against ADAMTS13 level are used. If ADAMTS 13

activity is less than 10%, it is important for diagnosis (4). ADAM TS 13 activity of our patient is found lower than 0.2 % and antibody against ADAMTS13 is higher than 90%.

The first type of treatment is plasma exchange therapy. The goal of plasmapheresis is to remove the antibodies against ADAMTS13 and replace ADAMTS13 enzymes. We performed plasmapheresis until the thrombocytopenia, LDH returned to normal values. Immunosuppressive therapy is important, too. Glucocorticoids are thought to reduce production of the ADAMTS13 inhibitor. It is suggested glucocorticoids are thought to reduce production of the ADAMTS13 inhibitor (autoantibody) by mechanisms similar to those in other autoimmune diseases. 1 gram glucocorticoid is suggested intravenously per day for three days. After three days oral prednisone should begin 1 mg/kg per day. As a result, TTP is a rare life-threatening disease. It should be diagnosed early and it should begin emergent therapeutic plasma exchange for survival.

References:

1. Terrell DR, Williams LA, Vesely SK, Lammle B, Hovinga JA, George JN. The incidence of the thrombotic thrombocytopenic purpura-hemolytic uremic syndrome: all patients, idiopathic patients, and patients with severe ADAMTS-13 deficiency. J Thromb Haemost 2005; 3: 1432-6.
2. Sadler JE, Moake JL, Miyata T, et al. Recent advances in thrombotic thrombocytopenic purpura. Hematology Am Soc Hematol Educ Program 2004;407-423.
3. Scully M, Cataland S, Coppo P, et al. Consensus on the standardization of terminology in thrombotic thrombocytopenic purpura and related thrombotic microangiopathies. J Thromb Haemost. 2017;15(2):312-322.
4. Barrows BD, Teruya J. Use of the ADAMTS13 activity assay improved the accuracy and efficiency of the diagnosis and treatment of suspected acquired thrombotic thrombocytopenic purpura. Arch Pathol Lab Med. 2014;138(4):546-549.