

Postoperative thrombotic thrombocytopenic purpura(pTTP) in open heart surgery : A rare case report

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Postoperative thrombotic thrombocytopenic purpura is a rare, life-threatening clinical syndrome characterized by microangiopathic hemolytic anemia, thrombocytopenia, renal failure, neurologic involvement, and fever. pTTP should be differentially diagnosed from other post-op thrombocytopenias such as heparin-induced thrombocytopenia, disseminated intravascular coagulation, infection. In pTTP, there is an autoimmune-mediated mechanism related to antibodies against a disintegrin and metalloproteinase with a thrombospondin type 1 motif, member 13(ADAMTS-13). A 54-year-old female had a descending thoracic aortic aneurysm replacement operation(Fig.1). After surgery, platelet count decreased from 179 to 34K/ μ L. Post-op 7days, BUN/Cr increased to 105/4.4mg/dl, so hemodialysis was started and performed for 4days. Post-op 9days, she presented seizure due to subarachnoid hemorrhage.(Fig.2) Peripheral blood smear showed microcytic anemia with anisopoikilocytosis, leukoerythroblastic reaction, and severe thrombocytopenia. Hb decreased to 6.5g/dl, plasma Hb increased to 20mg/dl, haptoglobin decreased to less than 10 mg/dl, indirect bilirubin increased to 1.04mg/dl, LDH increased to 2306U/L, while coomb's test was negative. PT was 1.06 INR and PTT was 30.1 sec. ADAMTS13 activity was 92%(reference range, 44~121). Platelet concentrates were transfused, but platelet did not increased. Oral high-dose corticosteroid was administered on post-op 10days. 5days after, platelet count increased to 215K/ μ L, BUN/Cr decreased to 59/1.99mg/dl, LDH decreased to 206U/L without plasmapheresis.(Table.1) The patient was discharged 5weeks after surgery and corticosteroid was tapered off. This case was suspected of pTTP because of MAHA, elevated LDH, decreased haptoglobin in spite of normal ADAMTS13 activity. DIC was ruled out because of normal PT and PTT. Plasma exchange is recognized to remove the antibody inhibitor of ADAMTS13 and it is a primary treatment for pTTP. In our case, ADAMTS13 activity was normal and the patient was managed without plasmapheresis, but only with a high-dose corticosteroid. It should be noted that normal or mild reduction of ADAMTS13 activity does not rule out pTTP.

Figure 1

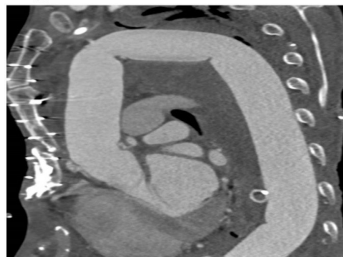


Figure 1. Chest CT angiography showed ascending aorta, partial arch, and descending thoracic aorta replacement .

Figure 2

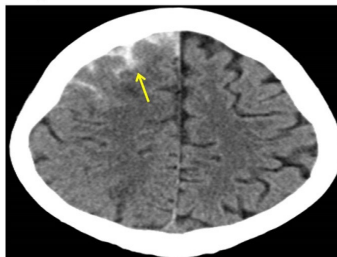


Figure 2. Brain CT showed SAH in right frontal area.

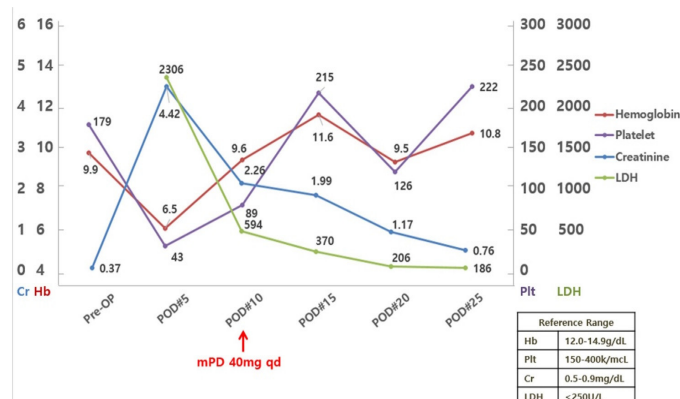


Table 1. Clinical course after oral high-dose corticosteroid therapy