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Treatment of Thrombotic Thrombocytopenic Purpura with Plasma Exchange

Y. Takenaka, Y. Nishian, K. Hiraoka, O. Inagaki, Y. Takamitsu, M. Ueda*, K. Suehiro*, and E. Kakishita*

*Department of Kidney and Dialysis, and *Second Department of Internal Medicine, Hyogo College of Medicine, Nishinomiya 663, Japan*

Thrombotic thrombocytopenic purpura (TTP) is known to show striking hematologic abnormalities and a fulminant, often fatal, course. However, the treatments with plasma infusion or plasma exchange (PE) have reported to result in remarkable improvement.[1] This paper reports on three patients with TTP who underwent PE with successful results in two cases and no response in one case.

Case 1 was a 40-year-old man admitted to our hospital because of jaundice, convulsion and unconsciousness. A diagnosis of TTP was made from laboratory findings, the hemoglobin concentration 7.5 g/dl, platelet count $1.1 \times 10^4/\mu l$, LDH 9,900 WU, total bilirubin 4.5 mg/dl, and creatinine 2.4 mg/dl. PE was done daily from the admission day to the 6th hospital day. His platelet count was increased to $17.2 \times 10^4/\mu l$ on the 6th day. From the 7th to the 13th day, the patient was treated by PE and fresh-frozen plasma (FFP) infusion alternately every other day. The patient recovered consciousness, and thrombocyte, hemoglobin and LDH returned normal levels on the 15th day. The patient went into remission (Fig. 1).

Case 2 was a 42-year-old man who entered another hospital because of jaundice and purpura. Two days later, he was found to be thrombocytopenic, and to have convulsions with disorientation, then he was transferred to our hospital. From laboratory studies of the hemoglobin concentration (8.5g/dl), platelet count ($1.8 \times 10^4/\mu l$), LDH (3,426 WU), total bilirubin (6.8 mg/dl), creatinine (3.5 mg/dl), and red cell fragmentation, a diagnosis of TTP was made. PE was done daily from the day of admission to the 5th day, however thrombocytopenia was not improved. On the 6th day, the patient was treated by FFP infusion, and from

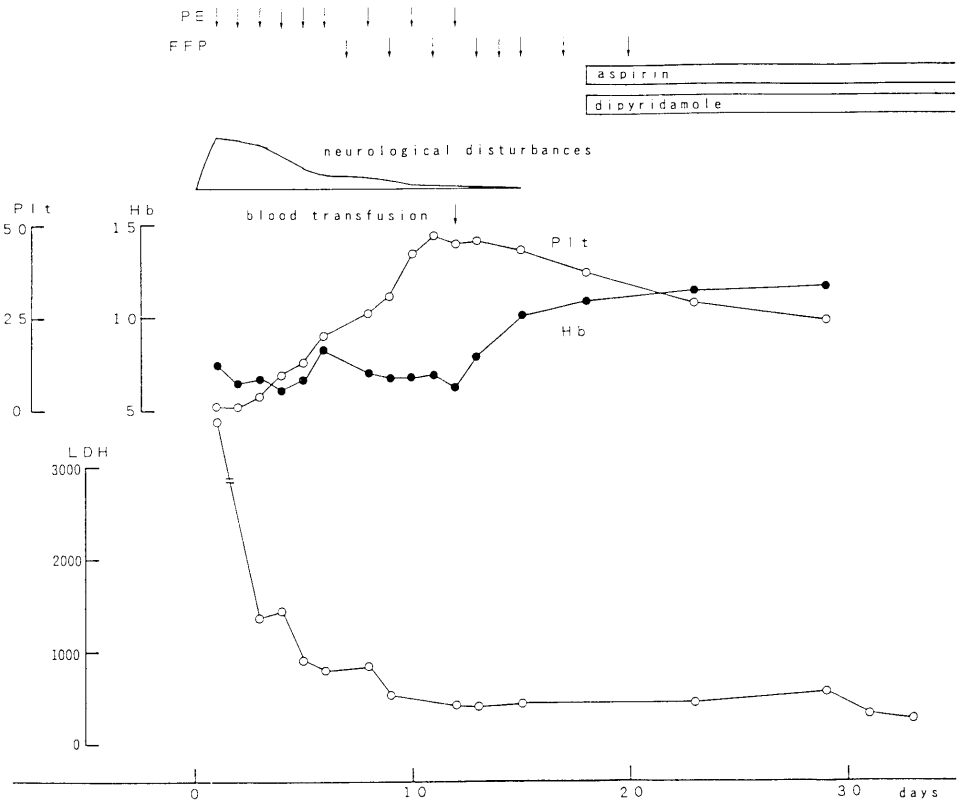

Fig. 1 Clinical course of case 1.

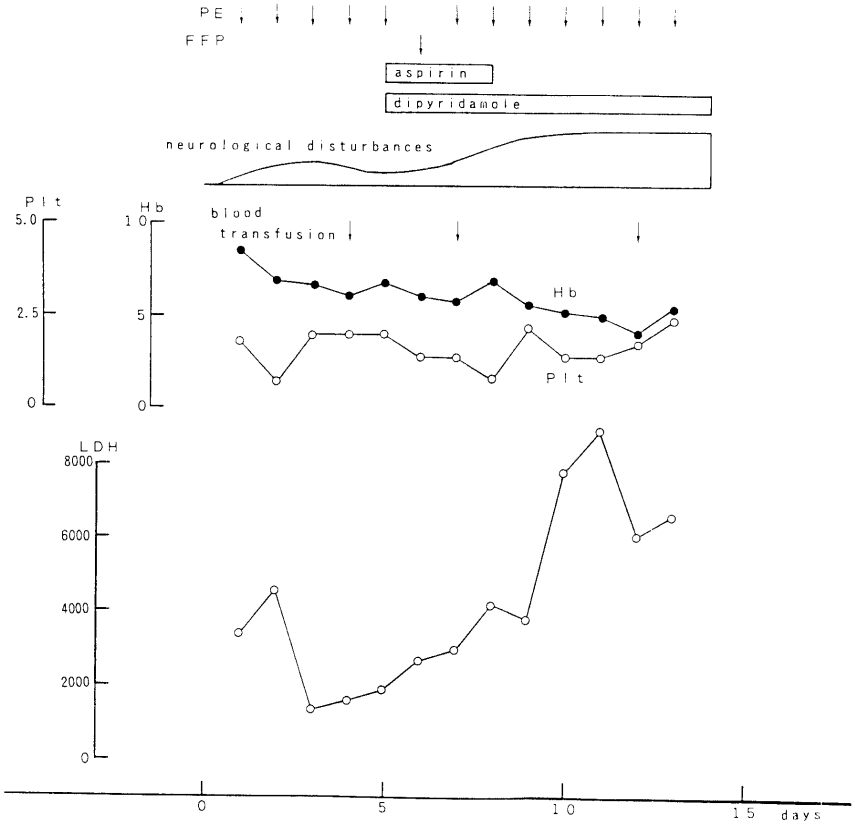

Fig. 2 Clinical course of case 2.

the 7th day to the last day was treated by PE everyday. On the 8th day, the patient became unconscious rapidly, and on the 14th day he died of multiple organ failure despite treatment (Fig. 2).

Case 3, a 14-year-old boy, was admitted to another hospital because of numbness in the right extremities. A diagnosis of TTP was made from thrombocytopenia, hemolytic anemia, red cell fragmentation, and neurological disturbances. The patient entered our hospital because the treatment with pulse therapy and FFP was unsuccessful in inducing remission. The patient had a clear consciousness with laboratory data of the hemoglobin concentration 7.0 g/dl, platelet count $2.8 \times 10^4/\mu l$, LDH 1,637 WU, total bilirubin 3.7 mg/dl and creatinine 1.1 mg/dl. Although PE was performed twelve times for 14 days from the day of admission, thrombocytopenia was not improved. Therefore PE was discontinued and administration of prednisolone 50 mg/day was started, but recurrence of hematuria, convulsion, and elevation of LDH was observed on the 20th day, so that PE was resumed everyday for a total of 7 times. PE therapy led to moderate improvement of thrombocytopenia and neurological disturbances. The patient went into remission with a total of 30 times PE.

There were no apparent differences detected in case 2 compared with the others in terms of the laboratory data and clinical symptoms on admission. It appeared that an immunological mechanism participated in the pathogenesis of TTP in case 3 because antinuclear antibody and PAIgG were positive.

It is generally believed that thrombosis is caused by the presence of the factor which induce platelet agglutination and the absence of factors which inhibit platelet agglutination in plasma. It therefore seems that PE is suited for TTP treatment in supplement and elimination of pathogenesis factor.[2)]

References

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