Thrombotic Thrombocytopenic Purpura: MR Demonstration of Reversible Brain Abnormalities

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Summary: We report a case of thrombotic thrombocytopenic purpura evaluated by MR. Multiple hyperintense foci on the T2-weighted images, observed principally in the brain stem and in the region of the basal nuclei, and neurologic signs disappeared after 15 days of therapy.

Index terms: Thrombosis, arterial; Thrombosis, venous; Nervous system, diseases; Brain, magnetic resonance

Thrombotic thrombocytopenic purpura (TTP), or Moschcowitz disease, is an extremely rare disease of unknown cause. It is characterized by thrombocytopenia, microangiopathic hemolytic anemia, and fluctuating neurologic manifestations, often bizarre. Fever and renal involvement often have been observed (1). Formation of platelet-fibrin microthrombi in the microvascular system is held responsible for the clinical symptoms. Pathologic alterations of the brain parenchyma are mainly manifested by small multiple infarcts (2). In this paper we present a case of postpartum TTP evaluated by magnetic resonance (MR) on the 8th and 23rd day after onset of symptoms.

Case Report

A 28-year-old woman was hospitalized with symptoms of postpartum metrorrhagia. In addition, signs of anemia, hyperpyrexia, low platelet count, and confusion were followed by aphasia, a right VI cranial nerve deficit, and a paresis of the right arm. Diagnosis of TTP was made on the basis of laboratory tests (1215 U/L lactate dehydrogenase, 1.90 mg/100 mL indirect bilirubin, 7g/100 mL schistocytic anemia with Hb, <10.1 mg/100 mL haptoglobin, and 7000 PTL/mm^3 thrombocytopenia). Specific treatment was initiated with repeated plasmapheresis, corticosteroids, immunoglobulins, and antithrombin III.

Unenhanced MR performed immediately before treatment revealed multiple hyperintense foci on the T2-weighted images, principally in the brain stem and in the region of the basal nuclei (Fig 1), areas that were normal on the T1-weighted images. Subsequently, the clinical state of the patient worsened because of high fever and renal involvement. After the 10th day of treatment there were clinical and laboratory signs of regression, and neurologic manifestations of the disease disappeared. A follow-up MR study, performed after 15 days of therapy, demonstrated the absence of abnormal areas observed previously in the T2-weighted images (Fig 2). After 6 months of constant laboratory and clinical follow-up, the patient continued to be free of disease and symptoms.

Discussion

TTP is an extremely rare disease, characterized in the past by a high mortality rate. In fact, therapeutic advances in the last 2 decades have altered the course of the disease, modifying the 85% to 90% mortality of the past to 15% to 20% (1). Neurologic disorders represent the principle manifestations of the disease and are apparent only in the acute phase of TTP, with seizures, aphasia, paresis, and an altered state of consciousness, frequently leading to coma (1). The cause of the microthrombotic phenomenon remains unclear, but it is believed to be associated with a disorder of the endothelial cells (1, 3), which normally play an important role in the regulation of the hemostasis mechanisms attributed to the action of serum factors (4). TTP causes vascular thrombosis resulting in parenchymal lesions. The most frequent alterations of the brain parenchyma are multiple minute infarcts, which involve the cerebral gray matter, brain stem, and cerebellar cortex; lesions seldom involve the white matter. Rarely were petechial hemorrhages observed (2). The alterations observed in our study concur with the neuropathologic findings described above; specifically, involvement of the brain stem and the basal nuclei gray matter (Fig 1) was observed, which may

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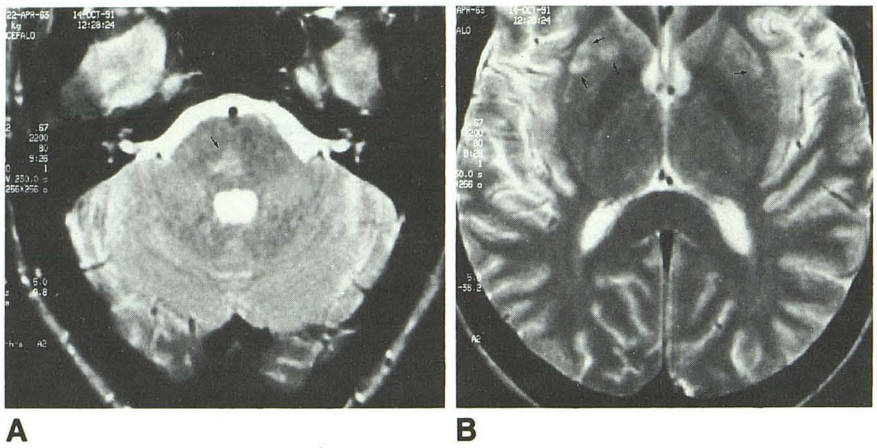

Fig. 1. Spin-echo T2-weighted axial sections (2200/80/1 [repetition time/echo time/excitations]) of the brain stem.

A, Abnormal intensity signal (*arrow*) is noted in the pons.

B, Microfoci of high intensity (*arrows*) in the basal ganglia (putamen).

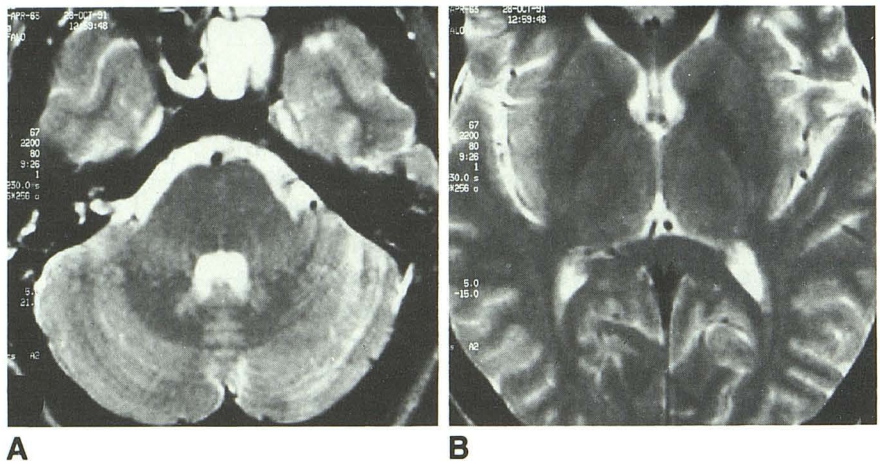

Fig. 2. Spin-echo T2-weighted axial follow-up (2200/80/1) after therapy: resolution of the abnormal signal intensities.

represent multifocal gray-matter edema (5, 6) rather than microinfarctions. The complete disappearance of the high-signal areas on the T2-weighted images after 15 days of therapy (Fig 2) indicates resolution of edema. In conclusion, if the diagnosis of TTP is established immediately and therapy is administered rapidly, the brain lesions identified with MR may regress with time.

References

1. George JN, Aster RH. Porpora trombotica trombocitopenica. In: Williams WJ, Beutler E, Erslev AJ, Licktman MA, eds. *Ematologia.* New York: McGraw Hill, 1991;1399–1403
2. Yamada M, Yoshida Y, Ikuta F, Honma Y. An autopsy case of thrombotic thrombocytopenic purpura. Topography of the vascular lesion in the central nervous system. *No To Shinkei* 1989;41:391–396
3. Burns ER, Zucker-Franklin D. Pathologic effects of plasma from patients with thrombotic thrombocytopenic purpura on platelets and cultured vascular endothelial cells. *Blood* 1982;60:1030–1037
4. Andresen SW and Bukowski RM. Thrombotic thrombocytopenic purpura. In: Rakel RE, ed. *Conn's current therapy.* Philadelphia: Saunders, 1991;344–345
5. Schwaighofer BW, Hesselink JR, Healy ME. Case report. MR demonstration of reversible brain abnormalities in eclampsia. *J Comput Tomogr* 1989;13:310–312
6. Coughlin WF, McMurdo SK, Reeves T. MR imaging of postpartum cortical blindness. *J Comput Tomogr* 1989;13:572–576