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Thrombotic Thrombocytopenic Purpura: MR Findings

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Summary: Neurologic manifestations occur in over 90% of patients with thrombotic thrombocytopenic purpura. Neuropathologically, thrombi produce occlusion of terminal arterioles and capillaries resulting in diffuse small infarcts. In the great majority of surviving patients, brain CT does not disclose any abnormalities. The authors report a case of thrombotic thrombocytopenic purpura in which brain MR examination showed multiple punctate lesions in the white matter.

Index terms: Thrombotic thrombocytopenic purpura (TTP); Brain neoplasms, magnetic resonance

Thrombotic thrombocytopenic purpura (TTP) is a life-threatening disease of unknown cause, characterized by hemolytic anemia, thrombocytopenia, fever, renal involvement, and neurologic manifestations. The clinical manifestations of TTP are the consequence of widespread hyaline thrombosis and occlusion of capillaries and arterioles without surrounding inflammatory reaction. Neuroradiologic investigations are rarely performed and computed tomography (CT) of the brain generally does not disclose any specific abnormalities. We report a case of TTP in which magnetic resonance (MR) showed multiple punctate lesions in the white matter.

Case Report

A 25-year-old man was admitted in December 1988 with microangiopathic hemolytic anemia, severe thrombocytopenia with normal coagulation tests, hematuria, rectal bleeding, petechia on his ankles, mild renal impairment, and fluctuating neurologic signs including obtundation and generalized seizures. Neurologic examination did not show any focal deficit. TTP was diagnosed and the patient was treated for 2 weeks with plasma exchanges without effect. Intravenous prostacyclin (epoprostenol, Flolan, Wellcome Laboratories, London, England), a potent inhibitor of platelet aggregation that has been reported to be decreased in patients with TTP, was instituted from 4 ng/kg/minute to 9 ng/kg/minute for 6 days and the patient's clinical and

hematologic status normalized (1). No infectious disease, neoplastic disorders, or connective tissue disorder was found and a human immunodeficiency virus-antibody test was negative. During the next 2 years the patient had three occasions of biologic relapses that were treated by plasma infusions. After the last remission, the patient had four episodes of unexplained transient loss of consciousness. Neurologic examination and standard electroencephalograms (EEG) on each occasion were normal. CT scans were obtained on two occasions and these were also normal. On January 1991, a witnessed episode of transient loss of consciousness established the diagnosis of generalized seizure, and EEG with megimide stimulation showed generalized spikes. Brain CT scan without and with contrast enhancement remained normal and MR showed multiple hyperintense punctate lesions in the white matter of the two hemispheres on T2-weighted images (Figs. 1 and 2).

Discussion

Neurologic manifestations occur in over 90% of patients with TTP; the illness initially presents with neurologic signs or symptoms in 60% (2). In order of frequency, neurologic manifestations include confusion, headache, altered mental state, paresis, aphasia, coma, seizures, and visual problems. A transient and fluctuating nature of neurologic signs is characteristic for TTP and is best explained by brief episodes of focal ischemia caused by microthrombi (3). Neuropathologically, thrombi produce occlusion of terminal arterioles and capillaries resulting in diffuse small infarcts and petechial hemorrhages that are generally confined to the gray matter. Extensive infarct is unusual (4). In the great majority of surviving patients, brain CT does not disclose any abnormalities; only a few cases of lacunar and small cortical infarcts have been reported in patients with permanent focal deficits such as hemiparesis (5). In our patient, brain CT scan was normal on three occasions. Numerous hyperintense punc-

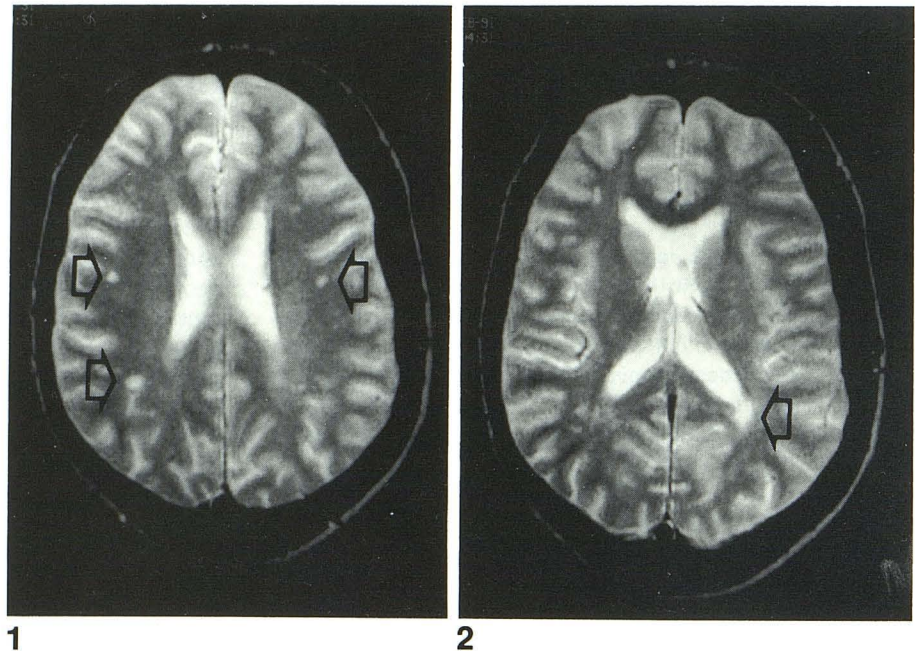
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Fig. 1. T2-weighted MR image: multiple hyperintense small areas in the white matter.

Fig. 2. T2-weighted MR image: multiple hyperintense small areas in the white matter.



tate lesions compatible with ischemic lesions were shown on T2-weighted images while T1-weighted images were considered normal. These punctate lesions were seen exclusively in the white matter, as opposed to the accepted opinion that lesions are predominantly confined to the gray matter. However, occlusions of the vessels of the white matter have been described, although most often in association with lesions in the gray matter. In a study of six patients with TTP in which examination of the central nervous system was performed, typical vascular lesions were found in the vessels of the white matter in three patients (4). In these cases mainly the subcortical white matter adjacent to the marked cortical involvement was affected, as is observed in this case report. Two additional cases have been recently reported. In one case, hyperintense T2 signals were found in the periventricular white matter; in the other, a left parietal infarct in the white matter was observed on T2-weighted images (6, 7). There is no particular reason that lesions were seen exclusively in the white matter in our case. In our

patient, MR images disclose no obvious cortical infarcts that would best explain the persistence of seizures. Here we would like to focus attention on brain MR, which may document cerebral abnormalities in patients with TTP. It should be useful during the acute phase or after remission.

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