MR imaging of demyelinating pseudotumor of the central nervous system

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Abstract
Eight cases of CNS demyelinating pseudotumor were reported. The pathologic features and MR diagnostic value and limitation in this disease entity were discussed. MRI in 7 pathologically proved and 1 clinically proved cases of demyelinating pseudotumor were performed and retrospectively analyzed. The enhancement pattern of vertical distribution to the lateral ventricle was demonstrated, and dorsal white matter enhancement of the cervical cord was revealed. It is difficult to make the diagnosis of demyelinating pseudotumor based on the clinical information and imaging findings. Though analysis of the clinical history and careful observation of MRI would be helpful in diagnosing the disease.

Introduction
The typical MR findings of CNS demyelination disease are multiple abnormal signal intensities in the white matter. Demyelination lesion presenting as space occupying mass is relatively rare, and its pathological finding is similar to, but not exactly the same as that of multiple sclerosis (MS) and acute disseminated encephalomyelitis (ADEM). Therefore, this kind of demyelination is a different disease entity called demyelinating pseudotumor (1,2). The demyelinating pseudotumor is often misdiagnosed as tumors clinically and radiologically (1-5), or even pathologically (5).

Materials and Methods
Seven pathologically proved and one clinically proved CNS demyelinating pseudotumors were retrospectively analyzed. There were 4 males and 4 females. The age ranged from 9 to 53 years. Acute or subacute clinical onset occurred in 5 cases (one week to two months), and chronic process occurred in 3 cases (twelve months to eighteen months). Two patients had the history of vaccination 2-3 weeks before the onset. A 1.5 T (GE Signa) MR scanners were employed using SE T1WI (TR/TE=600/15ms) and FSE T2WI (TR/TE=3000/100ms). T1WI were scanned after intravenous injection of Gd-DTPA (0.1mmol/kg) in 7 cases. Follow-up MRI was performed within the time period of 6 months to 5 years after the operation (7 cases) and steroid therapy (1 case).

Results
Gliomas were misdiagnosed and postoperative pathologic results turned out to be demyelinating pseudotumor in 7 cases. Demyelinating pseudotumor was diagnosed in one case and the cerebral lesion decreased dramatically after steroid therapy.

MR findings: The lesion occurred in cerebral hemisphere in 3 cases and all the lesions mainly involved the white matter. Left basal ganglion, left thalamus, and midbrain were involved in 1 case, hypothalamus in 1 case, right medulla oblongata in 1 case, cervical spinal cord in 1 case, and thoracic spinal cord in 1 case. The lesions presented as isolated mass in all 8 cases. Mass effect was revealed in 6 cases and perifocal edema in 5 cases. On T1WI, the lesions showed homogenous low signal in 5 cases, inhomogenous low signal in 2 cases (Fig.1), and mixed high and low signal in 1 case. On T2WI, the lesions presented as homogenous high signal in 5 cases and inhomogenous high signal in 3 cases. Marked enhancement was seen in all 7 case, and lower signal areas within the lesions were demonstrated in 4 cases (Fig.2). In one case, the long T1 and long T2 lesion showed marked enhancement (Fig.3) and the enhancement pattern of vertical distribution to the lateral ventricle was demonstrated (Fig.4), and the lesion dramatically decreased in size on follow-up MRI (Fig.5,6). Predominant dorsal white matter enhancement of the cervical spinal cord was revealed in another case (Fig.7,8).

Pathologic findings in 4 patients with acute and subacute onset revealed perivascular cuffing of lymphocytes and infiltration of mononuclear leukocytes and macrophages, accompanied by many gemistocytes. The findings in 3 cases with chronic process still showed perivascular lymphocytic infiltration and decreased number of macrophages with marked foamy cytoplasmic changes (myelin degraded into neutral lipid) accompanied by fibrillary astrocytic proliferation. Demyelination existed with intact axons.

Discussion
CNS demyelinating pseudotumor has been classified as an independent disease entity pathologically (1,2). Because of the misleading preoperative clinical and imaging diagnosis, the demyelinating pseudotumor is easily misdiagnosed as gemistocytic astrocytoma in the acute stage and as fibrillary astrocytoma in the chronic stage. Glioma was misdiagnosed in 7 of 8 cases in our group. The misdiagnosis rate is high in the literature (2-5), with single lesions often being mistaken for astrocytomas and multiple lesions as metastasis or lymphomas preoperatively. Some clues to the correct diagnosis were suggested: (1) acute clinical onset, (2) medical history of demyelinating disease, (3) history of vaccination or virus infection before the onset, (4) marked enhanced cerebral mass in children and teenagers, (5)cerebral lesions show vertical sign and intramedullary lesions mainly involve the white matter without secondary syringomyelia. Once the demyelinating pseudotumor is suspected, clinical test steroid therapy should be suggested, thus avoiding the devastating injury caused by surgery or radiation therapy.

References