A Case of Relapsing-Remitting Tumefactive Demyelination

Sir,

Tumefactive demyelination (TD) is considered as an unusual presentation of multiple sclerosis (MS) with a prevalence of 1–2/1000 cases of MS.[1] The lesions of TD are usually large (>2 cm) and present as a diagnostic dilemma in patients without a previous diagnosis of MS. According to the current evidence, approximately two-thirds of patients with a TD lesion at initial presentation develop a relapsing-remitting course typical of MS.[2] Patients of TD also mimic space-occupying lesions of the brain and should be considered as a differential diagnosis when the disease course is very short.

A minority of patients with TD relapse with only tumefactive lesions without other lesions characteristic of MS.[3] Possibly, such patients are a distinct subset of central nervous system demyelinating diseases. We report the clinical course of a patient with recurrent TD.

We evaluated a 14-year-old girl with recurrent episodes of weakness over past 4 years. At 10 years of age, the girl developed headache, vomiting, and acute-onset right-sided hemiparesis. There was no history of antecedent vaccination or febrile episode. Computed tomography scan of the brain revealed a large hypodense lesion in the left parieto-occipital lobe with perilesional edema. Initial magnetic resonance imaging (MRI) of the brain was suggestive of a space-occupying lesion with heterogeneous contrast enhancement and significant edema [Figure 1a and b]. The patient was started on intravenous steroids for cerebral edema, which resulted in rapid resolution of clinical features as well as radiological improvement. The patient

was subsequently diagnosed as a case of TD and biopsy was postponed. Follow-up neuroimaging revealed gliosis of the lesion.

Three years after the first episode, the patient had another episode of headache associated with left-sided weakness, for which she was referred to our center. Repeat imaging was done, which revealed a confluent area of white matter involvement in the right parieto-occipital region [Figure 1c]. MRI of the spine did not reveal any demyelinating lesion. Cerebrospinal fluid study revealed slight increase in protein without any oligoclonal bands. Evoked potentials were within normal limits. Inflammatory markers, antinuclear antibody, and vasculitic markers were all negative. Antibody against myelin oligodendrocyte glycoprotein (Anti MOG-Ab) was negative. Anti-aquaporin 4 antibody was also negative. Imaging of the thorax, abdomen, and pelvis did not reveal any space-occupying lesion. The patient responded to intravenous corticosteroid pulse and subsequently oral steroid. She then stopped oral steroid without medical consultation and after 1 year presented with right-sided facial deviation and dysarthria. MRI of the brain was done after the relapse which showed a new T2 hyperintense lesion in the right frontal region with surrounding edema but without significant contrast enhancement [Figure 1d]. The patient was restarted on corticosteroid which lead to improvement of her symptoms. To prevent relapses, azathioprine was added to her treatment regime and she is being followed up till date without further relapses.

Recurrent TD is a rare clinical scenario which may not be associated with MS. While significant proportion of patients were shown to convert to MS according to McDonald's criteria in a mean time of 4.4 months,[4] our patient did not have any lesion typical of MS during follow-up of 4 years. We propose that such cases belong to a separate subset of demyelinating diseases of the central nervous system and require further clinical evidence on natural history and optimum treatment.

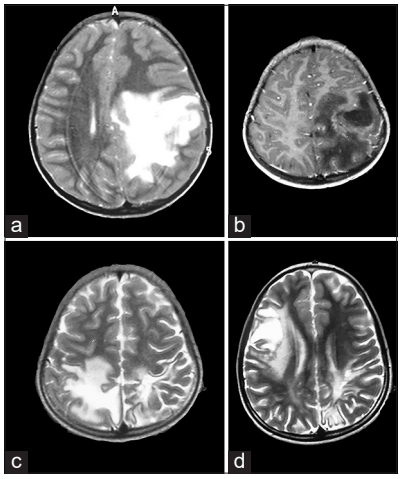

Figure 1: (a) T2-weighted axial magnetic resonance imaging of the brain shows large hyperintense lesion in the white matter of the left parieto-occipital lobe during the first attack of tumefactive demyelination. (b) Postgadolinium T1-weighted axial magnetic resonance imaging section showing peripheral incomplete enhancement. (c) T2-weighted axial magnetic resonance imaging of the brain during the second attack shows the appearance of a new T2 hyperintense lesion in the right parieto-occipital lobe; there is evidence of gliosis in the left occipital lobe. (d) T2-weighted axial magnetic resonance imaging of the brain during the third attack shows a lesion in the right frontal lobe, with resolution of the right occipital lesion (A in the figure indicates anterior)

Treatment of TD requires special considerations. In the acute stage, the patients respond to pulse corticosteroid and in some cases require plasma exchange.[2] TD has been reported in patients being treated with fingolimod[5] or natalizumab.[2] Therefore, these agents should not be used in treating patients with TD. Patients with isolated recurrent TD has been shown to be well responsive to corticosteroids without disease-modifying therapy. Due to frequent relapses in our case, we had used azathioprine in addition to corticosteroid, which showed good clinical response. Our patient had a high frequency of relapses and yet developed minimal neurological disability in the long term. In a clinical series of TD, mean time to relapse was approximately 16 months and majority of patients showed good clinical recovery, similar to our patient.[6]

The patient showed response to intravenous methylprednisolone pulse during relapses. However, there is a lack of robust clinical evidence on long-term treatment of such patients. Azathioprine, mycophenolate mofetil, cyclophosphamide, and methotrexate have been used effectively in relapsing-remitting TD.[6]

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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