Tumefactive Demyelination

MY Nadkar*, RA Deore**, Raminder Singh***

Abstract
We present a case of 22 year old female who had pulmonary tuberculosis followed by tuberculous meningitis and tuberculomas in past. This time she presented to us with right hemiparesis and altered sensorium. Diagnosis of tumefactive demyelination was made on the basis of typical MRI findings. Patient showed good response to steroids. ©

INTRODUCTION

Multiple sclerosis is the most common demyelinating disease of the central nervous system affecting young adults. The diagnosis is often a clinical one, with evidence of white matter lesions disseminated in time and space.

Tumefactive demyelination is a variant of multiple sclerosis in which plaque size is ≥ 2 cms. On imaging studies it may be confused with intracranial tumours. These lesions have been described to be generally well-demarcated, hypodense on CT, and of high T2 signal and relatively low signal on T1 signal on MRI, various chemical spikes on MR-spectroscopy are nowadays used for diagnosis. The relative lack of mass effect or vasogenic oedema given the size of the lesions is also often a clue to the diagnosis.

CASE REPORT

A 22 years old female patient resident of Sultanpur, U.P, had pulmonary tuberculosis in 1994 at the age of 8 yrs treated with antitubercular therapy for 6 months (details of therapy not available). In 2001 she had tuberculous meningitis with multiple tuberculomas in left frontotemporoparietal and right parietal region (Fig. 1a, 1b). Patient was treated with 5-drug antitubercular therapy (SHRZE), steroids and valproate. Patient was apparently asymptomatic after treatment and continued only on valproate.

Five months back she presented to local physician with generalized tonic-clonic seizures, headache and vomiting followed by right hemiparesis. There was no associated altered sensorium or bowel-bladder incontinence. Patient was started on antitubercular therapy empirically by a local physician at Sultanpur.

In view of worsening symptoms patient was admitted...
at a tertiary care center in Lucknow. CT brain done on 13th February, 2007 showed multiple intracranial ring-enhancing SOLs with perilesional edema, labeled as ?Tuberculomas ?Toxoplasma. MRI brain done same time showed multiple enhancing lesions in bilateral cerebral hemisphere and right cerebellum (Fig. 2).

Since patient was not responding to antitubercular therapy, toxoplasmosis was considered. Serum Toxo IgM was borderline positive (titers not available) but CSF-Toxo IgM and IgG were negative and CSF TB-PCR was also negative. Her HIV ELISA was nonreactive. Patient was started on sulphasadiazine + pyrimethamine empirically with steroids. Patient responded with decreased headache and increased power and discharged on antitoxoplasma therapy and antitubercular therapy was stopped.

Patient was referred for brain biopsy to Neurosurgery OPD at our center but since serum Toxo IgM antibody was earlier positive antitoxoplasma treatment was continued and brain biopsy was deferred and patient was referred to us. Patient was then admitted in our ward for the first time in September 2007 with complaints of increased weakness on right side of body with slurred speech, deviation of angle of mouth and fluctuating level of consciousness.

CSF study repeated to rule out tuberculous meningitis. CSF showed sugar-50 mg%, protein-154 mg%, PMN-4/cmm and lymphocyte-1/cmm. MRI brain (Fig. 3a, b) done showed multiple ring enhancing lesions with significant perilesional oedema, reported as tumefactive demyelination. Brain biopsy was done to confirm diagnosis but was inconclusive.

Patient was given inj. methylprednisolone (1 gm/day for 3 days) followed by oral prednisolone (30 mg/day). Patient improved symptomatically in the form of increased power and improved speech and facial weakness. Patient was discharged on oral prednisolone (10 mg/day). Patient has not followed further as she went to native place after discharge.

**DISCUSSION**

Tumefactive demyelination is a demyelinating condition considered as intermediate between multiple sclerosis and acute demyelinating encephalomyelitis. It is defined as demyelinating plaques > 2 cm in diameter. These lesions are seen in women with an average age of 37 years. Tumefactive demyelination does not usually originate as a postinfectious or post-vaccination response. Symptoms in this condition are atypical for multiple sclerosis and as seen in our patient are due to focal mass lesion leading to focal neurologic deficit, seizures, and aphasia. The most important differential diagnosis is tumour due to its MRI appearance. Gliomas are reported to develop in patients with multiple sclerosis.

MRI showing well-demarcated white-matter lesions, particularly periventricular or infratentorial, with high on T2 signal and relatively low T1 signal is characteristic as seen in our patient. Contrast enhancement, mostly of the rim-enhancement type, is more common in the tumefactive variant than in the standard multiple sclerosis plaque MR spectroscopy shows reduced NAA without corresponding elevation of choline peak relative to creatine peak. Diffusion imaging reveals mildly increased apparent diffusion coefficients within tumefactive demyelinating lesions. This proves to be a useful tool in differentiating ring-enhancing tumefactive demyelinating lesions from cerebral abscesses.

CSF oligoclonal bands can be seen in 65% cases; we could do this as patient could not afford.

Histology of tumefactive demyelinating lesions shows florid cytologically atypical astrogliosis in which mitotic figures can be identified. Biopsy of the gliotic margin may therefore be misinterpreted as the edge of a glioma, In our patient the biopsy was inconclusive and our diagnosis is based on typical MRI findings.

Although the exact pathogenesis is not clearly understood, most patients respond favorably to corticosteroid therapy and do not progress to multiple sclerosis. Most of cases are steroid responsive. Steroid-unresponsive cases treated with plasmapheresis (total 7 exchanges over 14d at 54 ml/kg). We gave inj. methylprednisolone to the patient followed by oral prednisolone. Patient showed good improvement in the focal deficit at the time of discharge.

**Acknowledgement**

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**REFERENCES**

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ERRATUM

Case Report on Carbimazole Induced ANCA Positive Vasculitis in JAPI Vol. 56 October 2008, pg 801-803, Kindly read the authors names as S Pandey, RS Kushwaha, P Mehndiratta, MM Mehndiratta and not as S Pandey, RS Kushwaha, P Mehndiratta. The error is regretted.

Hon. Editor