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Rare and Complex Type Of MS, Tumefactive Multiple Sclerosis

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ABSTRACT

Multiple Sclerosis is a disease with remitting and relapsing course and the normal classification is more based on the clinical manifestation and MRI scan findings. There are rare types of multiple sclerosis where demyelination can be monophasic or polyphasic. We present a patient with Tumefactive MS (TMS) with a typical relapsing and remitting course initially but later exhibited secondary progression.

TMS presents like a space occupying lesion or tumour so it is important to make the correct diagnosis, sometimes requiring a biopsy for confirmation.

Treatment choice is tough but needs to be commenced early for better control and outcome. This patient deferred disease modifying agents for a long time but when she finally accepted treatment, it was too late as she developed secondary progression with a relentless course.

Keywords: MS- Multiple Sclerosis; TMS- Tumefactive MS

INTRODUCTION

Multiple sclerosis (MS), an inflammatory condition causes demyelination and axonal loss in the central nervous system (brain and spinal cord). The primary brain insult ("sclerosis") and its clinical sequelae are "disseminated in space", affecting different anatomical sites, and "disseminated in time", appearing episodically over time [1-3].

The sub-division of MS as relapsing and remitting type (RRMS), secondary progressive MS (SPMS) and primary progressive MS (PPMS) is based more on clinical characteristics and not specific biologic pathophysiology. Nonetheless, they provide an organized framework for diagnosis and long-term management.

There are a few rare types of MS described [4,5] , such as the

- Marburg type- This type has a fulminant monophasic course in most cases with poor response to treatment.
- Balo type- By definition shows a peculiar pattern of pathology in cerebral hemispheric white matter consisting of a concentric, mosaic, or floral configuration of alternating bands of white matter whose basis is relatively preserved myelination alternating with regions of demyelination

- Schilder's disease- is an acute MS that occurs in childhood and the clinical course is diverse, but widespread white matter involvement
- Tumefactive MS (TMS) - Rarely the plaque size can be ≥ 2 cm with mass effect, edema, or ring enhancement on magnetic resonance, features pointing to a tumour like space occupying lesion.
- This case report is about a lady who presented with TMS, initially thought to be an astrocytoma but history revealed a relapsing and remitting course and biopsy was not required.

CASE PRESENTAION

A 48 year old female with hypertension, presented with 1 week duration of headache, disorientation and speech impairment. On examination, she had right sided weakness and mild aphasia. MRI brain with contrast showed large, well defined T2 heterointense and T1 hypointense lesion in left frontal lobe with significant oedema, mass effect and midline shift of 13mm to the right. Patchy white matter hyperintensities were noted in right fronto-parietal and occipital lobes and small T2 hyperintensity in right thalamus. Focal gliosis were seen in right cerebellum without significant contrast enhancement. MRS showed elevated Choline and lipid lactates and reduction in NAA and Cr. The differentials included Glioma/ Astrocytoma.

The neurosurgeon was considering surgery however history revealed 3 variable clinical symptoms and MRI findings in the past. In 2014 she had presented with headache with MRI showing multiple hyperintense lesions in the right cerebral white matter, largest confluent lesion in the right parieto – occipital and peri – ventricular region with few tiny hyperintense foci in the left frontal white matter. She improved with IV steroids.

2nd episode was in 2015 with right hemispherical lesion and tiny hyperintensities in the left frontal region. 3rd episode was in January 2018 with giddiness, imbalance and vomiting and MRI brain showing new right cerebellar mass lesion and similar appearance of bilateral fronto parietal white matter hyperintensitie, Figure 1. In all these episodes, she was successfully treated with steroids.

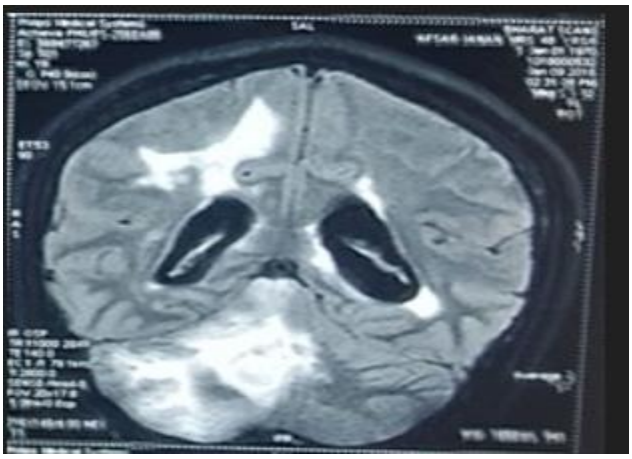

Figure 1: 2018 January MRI Brain- Lesion involving right cerebellar hemisphere displacing the vermis towards left side.

4th episode was in July 2018, when she presented to us for the first time with new left frontal large lesion, Figure 2. Visual evoked potential showed delayed P100 latencies. CSF was negative for Oligoclonal bands.

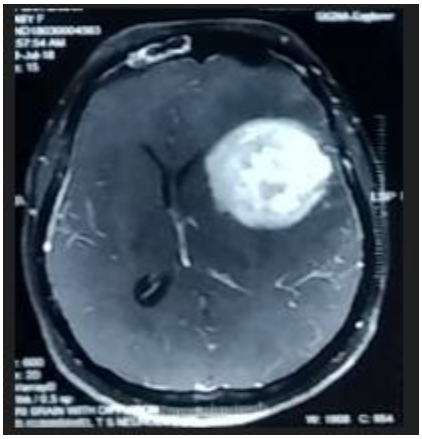

Figure 2: July 2018 Large well defined large lesion in left frontal lobe with significant edema, mass effect and midline shift of 13mm to the right.

Her series of clinical presentation and radiological findings were more suggestive of relapsing and remitting type of Tumefactive demyelination /Multiple sclerosis but she was never commenced on any disease modifying treatment despite multiple clinical presentations [6-8]. On this occasion, we immediately started treatment with IV steroids with initially good response, Figure 3. Her aphasia and motor deficit improved [9,10]. We counselled the patient and family about disease modifying agents for MS and considered Natulizimab although cost was a limiting factor [11].

Within a week, before starting the disease modifying agent, she presented with generalized seizures. MRI brain showed a relapse with increase in size of the left frontal lesion, Figure 4. There was no desirable improvement with repeat IV steroids, other treatment options with IVIG, Rituximab and Plasmapheresis 12 were considered, and Rituximab 13 was chosen and given but patient continued to deteriorate with increased odemea and Intra-cranial pressure. Family did not want decompression craniotomy. She continued to deteriorate in spite of our efforts and died.

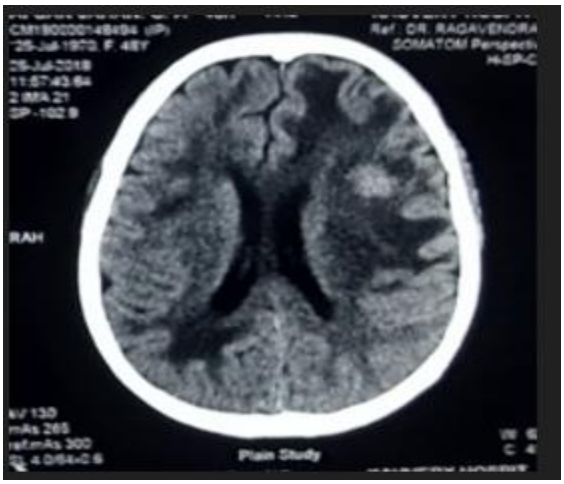

Figure 3: Follow up CT brain after IV steroids- July 2018 with reduction in size of the lesion

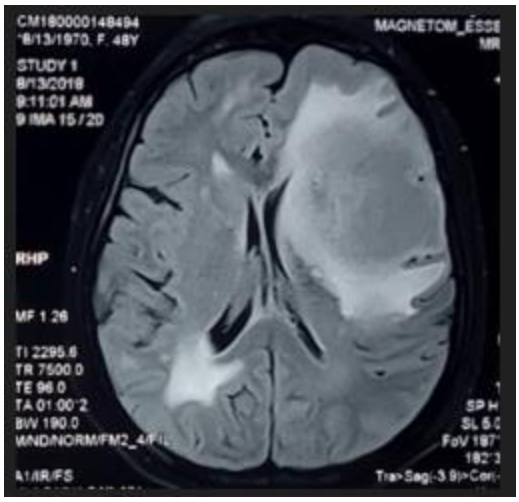

Figure 4: August 2018 MRI Brain with contrast showing increase in size of lesion with associated mass effect.

DISCUSSION

Tumefactive MS (TMS) is very rare with an incidence of 1-2 cases per 1000 MS patients and or 3 cases per million per year in the general population. Although imaging helps, many a times, TMS could be mis-diagnosed as tumour (Astrocytoma) and a biopsy is required for definitive diagnosis especially in the first presentation. CSF analysis reveals positive oligoclonal bands in only 30% of the patients.

Like in MS, females are more affected and majority of the patients are in the range of 20-40 years of age. Like MS, TMS shows evidence for a remitting and relapsing course as seen in our patient.

The condition needs early diagnosis and commencement of disease modifying therapy. Failure of which could lead to secondary progression with a more aggressive course, resistant to treatment as seen in the case we have described [7-9].

Natulizimab has been shown to be effective in many patients. Fingolimoid, a sphingosine 1-phosphate receptor modulator used as oral MS drug can also be used but there are also case reports where Fingolimoid , used as DMT for MS could lead to possible conversion of non-TMS into TMS. Thus this drug is to be considered with caution for long-term treatment [11,14].

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