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GLIOBLASTOMA MASQUERADING AS A TUMEFACTIVE DEMYELINATING LESION IN A PATIENT WITH MULTIPLE SCLEROSIS: A CASE REPORT

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ABSTRACT

Introduction: Increased incidence of gliomas has been reported in patients with Multiple Sclerosis. Development of new neurological deficits in a patient of Multiple Sclerosis maybe due to relapse of the disease or development of a brain tumor. Distinction between the two is important as a high-grade glioma may mimic a tumefactive demyelinating lesion. Early and correct diagnosis is essential as they have different treatment strategies. **Case report:** A 30 years old female who was diagnosed to be a case of Multiple Sclerosis at 28years of age and was on oral steroids for 2 years presented to the Neurology OPD with development of new symptoms. She showed good response to oral steroids and was clinically stable until recently when she developed gradually progressive neurological deficits. An MRI brain done subsequently elsewhere gave the diagnosis of tumefactive demyelinating lesion. Based on the symptoms and MRI diagnosis, she was suspected of having a relapse of Multiple Sclerosis, so she was started on IV steroid pulse therapy. However, the symptoms of the patient continued to deteriorate despite IV steroids. An MRI examination of brain was then done in our Institute, based on which a provisional diagnosis of a high-grade glioma was made. The diagnosis was subsequently confirmed by histopathological examination to be a Glioblastoma. **Diagnosis:** Based on past and present radiological and histopathological findings, a final diagnosis of Glioblastoma with preceding Multiple Sclerosis was made. The patient was planned for tumor debulking surgery, however, she expired 1 day prior to the surgery.

KEYWORDS: Multiple Sclerosis, High-grade glioma, Glioblastoma, Tumefactive demyelinating lesion, Neurological deficit.

INTRODUCTION

Multiple Sclerosis is a chronic demyelinating disorder of the central nervous system which is characterised by dissemination of lesions in time and space. It is common in young and middle-aged females.

The different variants of the disease are—
1. Classic Multiple Sclerosis
2. Tumefactive Multiple Sclerosis
3. Marburg type (acute malignant)
4. Schilder type (diffuse cerebral sclerosis)
5. Balo concentric sclerosis

Symptoms include visual disturbances due to optic neuritis, gait ataxia due to cerebellar involvement, sensory loss, paresthesia, upper motor neuron signs, urinary incontinence and cognitive decline.[1,2]

McDonald criteria for diagnosis of Multiple Sclerosis—
1. Typical history
2. Oligoclonal bands in CSF
3. Immunoglobulin G in serum
4. Abnormal visual evoked potential
5. MR imaging
6. Lack of alternative diagnosis

Glioblastomas are high-grade astrocytomas with a predilection for spreading along condensed white matter tracts such as corpus callosum and corticospinal tracts to involve the contralateral hemisphere.[3] **Symptoms** include focal neurological deficit, seizures and headache.

Various studies have reported that patients with Multiple Sclerosis have increased incidence of brain cancers, particularly gliomas and lymphomas. In some patients, the demyelinating lesions precede the gliomas, whereas

in others, both the demyelinating lesions and gliomas occur concurrently.[4]

CASE REPORT

A 30 years old female, housewife by profession, presented to the Neurology OPD at the institute.

Symptoms and investigations done 2 years ago—
Chief complaints of the patient: The patient presented with gradually progressive painful vision loss, sensory loss and paraesthesia of bilateral upper and lower limbs (left>right) and mild cognitive decline for 1 month.

Radiological Evaluation:
MRI of brain done 2 years ago:

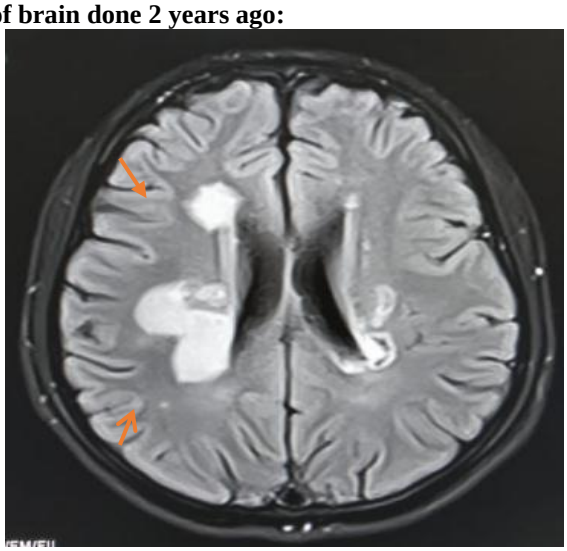

Figure 1

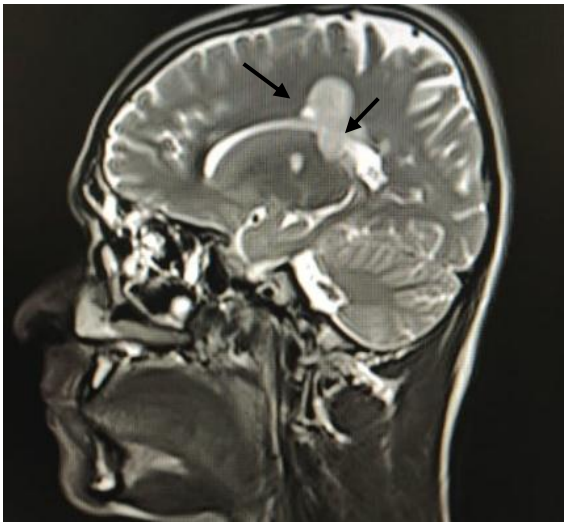

Figure 2

Figure 1: Axial FLAIR image shows multiple (atleast 6) hyperintensities (orange arrows) in bilateral corona radiata (right>left) and periventricular white mater adjacent to body of bilateral lateral ventricles (right>left). Figure 2: The sagittal T2WI shows multiple (atleast 3) triangular hyperintensities (black arrows) in the calloseptal region arranged perpendicular to the body of the right lateral ventricle, these are known as Dawson's fingers. A T2 hyperintense lesion is also noted in the right thalamus.

Past history: There is no past history of head trauma, infection, other autoimmune diseases or radiation exposure.

Family history: There is no significant family history.

On examination:
Table 1.

	Right eye	Left eye
Visual acuity	6/12	6/12

CSF oligoclonal bands and lymphocytes: Positive

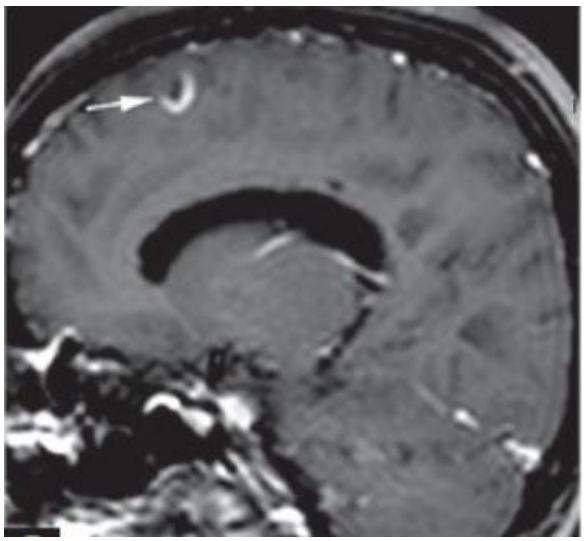

Figure 3: Sagittal post-gadolinium image shows an enhancing juxtacortical lesion (white arrow) in left frontal lobe showing open ring enhancement towards the cortex. However, the periventricular lesions do not show any enhancement. This is consistent with lesions disseminated in time and space.

Radiological diagnosis: A radiological diagnosis of demyelinating disease, possibly Multiple Sclerosis was made based on the history, typical location of the lesions and their dissemination in time and space.

On histopathological examination, a diagnosis of Multiple Sclerosis was made.

Final diagnosis: Based on radiological findings, histopathological examination and presence of CSF oligoclonal bands and lymphocytes, a final diagnosis of **Multiple Sclerosis** was made.

Treatment: She was administered oral steroids with good clinical response to the therapy.

Present symptoms and investigations—
Chief complaints of the patient: The patient presented with gradually progressive right sided hemiparesis and headache for the last 2 months with on and off altered sensorium. She also had 3 episodes of seizure in the last 2 months.

On examination:
Table 2

	Right upper and lower limbs	**Left upper and lower limbs**
Power	3/5	5/5
Reflexes	Exaggerated	Normal
Sensation	Normal	Normal
Babinski reflex	Extensor	Flexor

Table 3

	Right eye	**Left eye**
Visual acuity	6/9	6/9

Pulse:76 beats/min and BP :118/84 mm Hg.

An MRI brain was advised which was done elsewhere and which gave a provisional diagnosis of tumefactive demyelinating lesion. Based on the previous history of Multiple Sclerosis and the MRI brain diagnosis, the patient was suspected of having a relapse of the disease.

So, she was started on IV steroid pulse therapy, however, there was no improvement in her symptoms. She was advised another MRI brain which was done at our institute.

Radiological Evaluation done in our Institute:
Present MRI of brain:

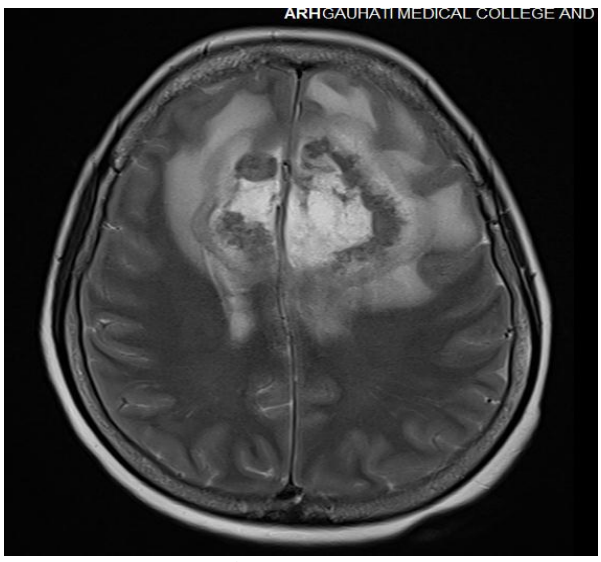

Figure 4

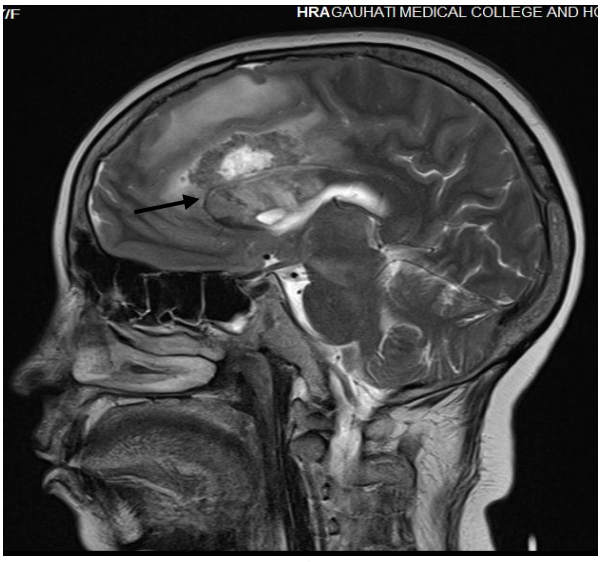

Figure 5

Figure 4: Axial T2WI shows a fairly well-defined lesion showing heterogeneous signal intensity with central necrosis involving the para-sagittal regions of bilateral frontal lobes with extensive peri-lesional edema. Figure 5: Sagittal T2WI shows involvement of the genu and body of corpus callosum (black arrow). There is complete resolution of the previously seen demyelinating lesions.

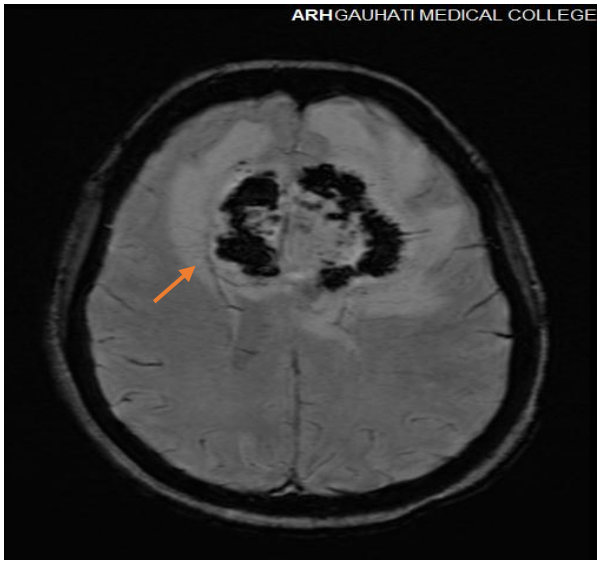

Figure 6

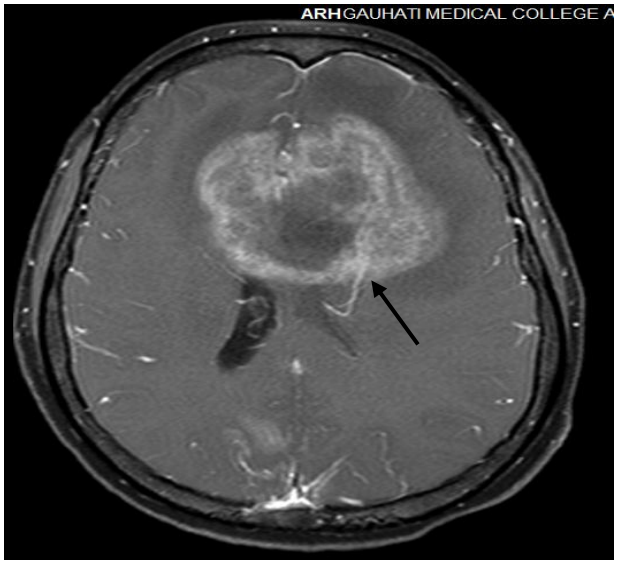

Figure 7

Figure 6: Axial SWI shows susceptibility artifacts (orange arrow) in periphery of lesion consistent with hemorrhage. Figure 7: The lesion shows irregular peripheral hetergeneous enhancement (black arrow) after gadolium administration with central area of non-enhancement suggestive of necrosis.

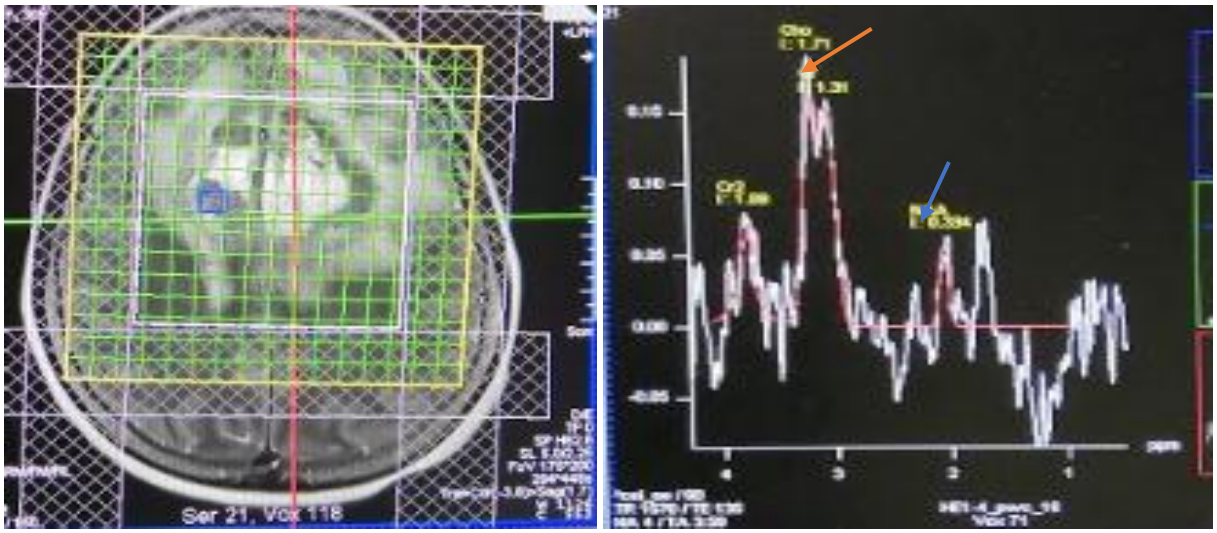

Figure 8

Figure 8: On MRS the lesion shows increased choline peak (orange arrow) and decreased NAA (blue arrow). Increased lipid/lactate peaks were noted in the central necrotic area (not shown). However, there is no Glutamate/glutamine peak at 2.4ppm, thereby excluding tumefactive demyelinating lesion.

Radiological diagnosis: A radiological diagnosis of a high-grade glioma, possibly Glioblastoma, was made on the basis of the afore-mentioned findings.

On histopathological examination, a diagnosis of Glioblastoma was made.

Final diagnosis: Based on the past and present clinical, radiological and histopathological data, a final diagnosis of Glioblastoma with preceding Multiple Sclerosis was made.

Treatment: Tumor debulking followed by adjuvant radiotherapy was planned for the patient. Unfortunately, the patient expired 1 day prior to the surgery.

DISCUSSION

Increased incidence of brain tumors, particularly gliomas, has been reported in patients with Multiple Sclerosis leading to overlapping neurological manifestations and diagnostic difficulty.

Prior to the advent of CT and MRI, Multiple Sclerosis was usually diagnosed clinically. Autopsy of these patients showed that in a few percentages of patients the primary lesion was a primary CNS neoplasm. With the advent of new neuroimaging techniques like MRI, early and accurate diagnosis of demyelinating diseases and brain tumors have been possible. The most common histological sites reported were oligodendroglioma and Glioblastoma.

Development of a new lesion in a patient with Multiple Sclerosis needs to be determined as either demyelinating or neoplastic in character. The gliomas may develop concurrently with the demyelinating lesions or may develop from a pre-existing demyelinating lesion.[4]

Multiple Sclerosis patients have an increased risk for brain tumor due to the following factors[5]—

1. Transformation of Multiple Sclerosis lesions into a tumor maybe due to increased proliferation ratio caused by remyelinating processes.
2. Neoplastic transformation of reactive astrocytes may also be stimulated by an unknown hereditary or acquired factor in some MS patients.
3. Both demyelination and tumor pathogenesis may involve epigenetic mechanisms like DNA methylation and histone protein modifications.
4. Human Polyomavirus/John Cunningham virus (JCV) of Papova virus family is involved in pathogenesis of both Multiple Sclerosis and Glioblastoma.

Glioblastomas and tumefactive demyelinating lesions may have similar radiological appearances with early gliomas sometimes presenting with open ring enhancement, which can be misinterpreted as demyelinating lesion. The first possible hypothesis in Multiple Sclerosis patients is that a tumefactive demyelinating lesion has developed in the course of a consecutive relapse. Though this is the more likely situation, gliomas must be considered in all cases of mass lesions appearing in patients with Multiple Sclerosis. However, the appearance of atypical neurological symptoms like seizures in patients with Multiple Sclerosis should raise the suspicion of other pathologies. Although brain tumors are usually located at frontal and temporal lobes, this localization criterion is not useful for diagnosis. The two conditions can be distinguished on the basis of MR spectroscopy and perfusion study. Both show increased choline and lipid/lactate peaks and reduced NAA but only demyelinating lesions show glutamate/glutamine peak. On perfusion study, Glioblastomas show greater rCBV as compared to demyelinating lesions.

CONCLUSION

A Glioblastoma may masquerade as tumefactive demyelinating lesion in a patient of Multiple Sclerosis. The subsequent development of Glioblastoma could be due to transformation from a pre-existing demyelinating plaque or de-novo tumorigenesis. Both the lesions have overlapping clinical and radiological features, hence, early and correct diagnosis is essential as they have different treatment strategies.

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