

Original research article

## Utility of MRI in Clinico Radiological Evaluation of White Matter Diseases

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### Abstract

**Background and Objectives:** The purpose of this study is to evaluate the role of MRI as an investigative modality in white matter diseases and to document clinic-radiological value in early diagnosis and management. This was done by studying the MR findings of different white matter diseases at our hospital with clinical presentation correlation along with most prevalent diseases and their location identification.

**Materials and Methods:** Forty patients with a strong clinical suspicion of demyelinating disorder were evaluated by MRI using 1.5 Tesla (GE Optima MR360) scanners in a prospective study period of 1 year after ethical approval at GIMS, Patna, Dept. of Radiology. Patients were referred from neuromedicine department.

**Results:** PRES (MS) was the most common white matter disease encountered in our one year study after ethical approval. Out of 40 cases, 16(40%) cases presented with altered sensorium, seizure and visual disturbances among which most of cases were of nephrotic syndrome and few hypertensive post delivery female patients. Among 16 cases of PRES, all 16(100%) cases showed posterior parieto-occipital periventricular and deep white matter involvement on MRI as T2/FLAIR axial images hyperintensity and T1 hypointensity.

**Conclusion:** MRI is the modality of choice for early lesion detection, disease progression and in follow up cases in both subclinical and clinical settings due to its multiplanar images and better resolution. Therefore, early management for better patient cure is possible.

**Keywords:** Hyperintensity and T1 hypointensity, ADEM, MRI

### Introduction

White matter diseases are the pathological condition which affect specifically white matter<sup>1,2,3</sup> of the brain by altering normal myelination process and result disease progression. They comprise a vast heterogeneous group and have a variety of appearances and clinical presentations. These are classified into demyelinating, dysmyelinating and hypomyelinating disorder. Introduction of MRI and broad clinical evaluation definitely helped in diagnosis of more cases of demyelinating diseases so that they can be managed earlier. MRI has a vital role in early diagnosis and the best imaging modalities than ct due to its high sensitivity and its multiplanar imaging. We can do simultaneous imaging of spinal cord and orbits .CT is less

sensitive to detect early demyelinating lesion. In this article we have studied patients with suspicion of demyelinating disease showing different specific clinical findings and simultaneously studied the role of MRI in early diagnosis, follow-up and therefore early management of these conditions.

### **Aim & Objectives**

The aim and objective was to evaluate the role of MRI as an investigative modality in white matter diseases and to document clinic-radiological value in early diagnosis and management.

### **Materials and Methods**

Forty patients with a strong clinical suspicion of demyelinating disorder were evaluated by MRI using 1.5 Tesla (GE Optima MR360) scanners in a prospective study period of 1 year after ethical approval at IGIMS, Patna, Dept. of Radiology. Patients were referred from our neuromedicine department. Images according to MR protocol; were obtained in sagittal, axial and coronal planes using SE T1, PD and T2, T1C and FLAIR sequences<sup>11</sup> with Slice thickness of 5mm, and FOV of 24 x 24 and 256 x 256.

### **Inclusion criteria**

- Strong clinical suspicion of white matter disorder presenting visual, sensory and motor symptoms.
- All age group including male and female child and adult.
- Neurological symptom with Fluid overload, Underlying connective tissue disorder, Alcohol & drug overload, viral infection.

### **Exclusion criteria**

- Absence of clinical suspicion regarding white matter disorder.

### **Results**

PRES (fig.3) (MS) was the most common white matter disease encountered in our one year study after ethical approval. Out of 40 cases, 16 (40%) cases presented with altered sensorium, seizure and visual disturbances among which most of cases were of nephrotic syndrome and few hypertensive post delivery female patients. Among 16 cases of PRES, all 16 (100%) cases showed posterior parieto-occipital periventricular and deep white matter involvement on MRI as T2/FLAIR axial images hyperintensity and T1 hypointensity. No cases showed diffusion restriction. No post contrast enhancement seen. Two cases (12.5%) showed hemorrhagic changes on gradient sequences. Out of 16 cases 10 (62.54%) showed B/L temporal involvement. 4 cases (25%) showed asymmetrical frontal subcortex involvement. MS and ADEM<sup>4</sup> were second most common condition encountered after PRES. 5 cases of MS with prevalence of 12.5% seen who were presented with limb sensory loss or paraesthesias<sup>10</sup>, upper motor neurone signs, urinary incontinence. One case presented with visual loss and one case was follow up case of MS.

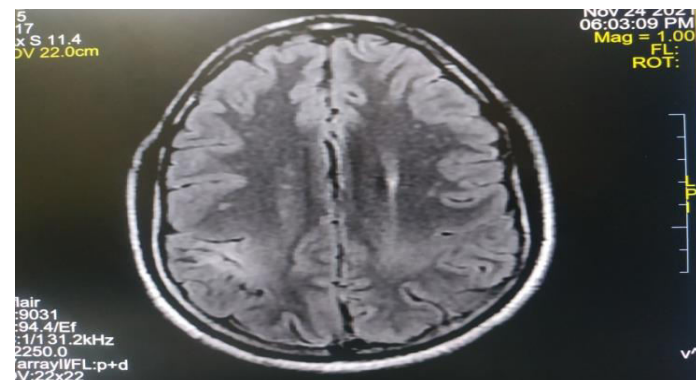
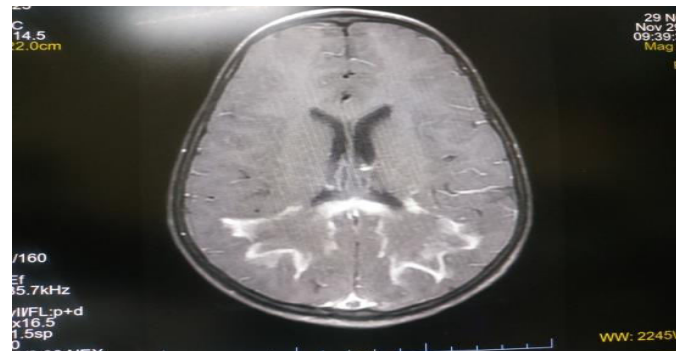
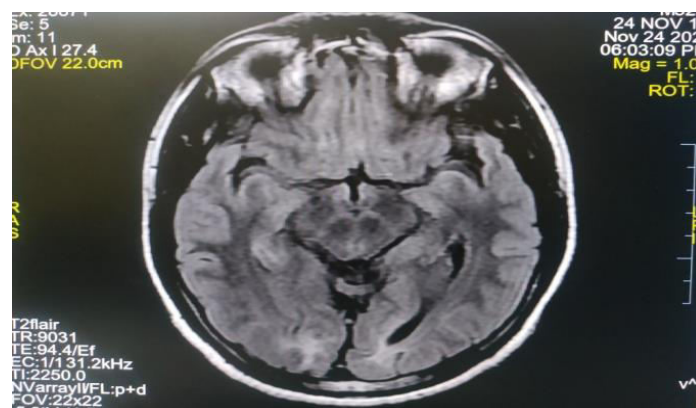
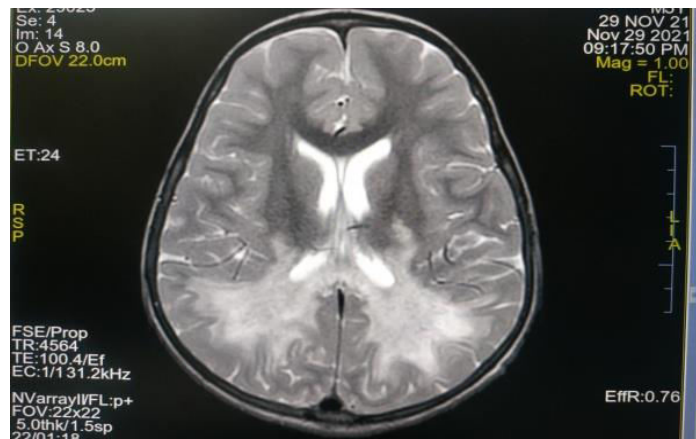
5 out of 5 MS (fig.4) cases (100%) cases showed T2/FLAIR periventricular with periventricular Dawson finger hyperintensities as typical feature with T2 hypointense appearance corresponds to T2 black hole sign. All showed faint peripheral and few nodular post contrast enhancement. Few lesion showed diffusion restriction with ADC dark corresponded to active plaque. MRS in some lesion showed decreased NAA peak within larger lesion. 3 of 5 cases (60%) showed deep white matter involvement. 2 cases (40%) showed juxtacortical involvement. 1 out of 5 cases (20%) showed spinal cord and b/l optic nerve involvement. One

Follow up cases showed few DIS & DIT lesion on T2 images. Five patients of acute disseminated encephalomyelitis (ADEM) were evaluated by MRI, all having a history of fever prior to the onset of clinical symptoms of hemiparesis and altered sensorium. The age group varied through a wide range of 10 to 50 years. Majority of the lesions in our study were located in the cerebral white matter (100%) with asymmetric and patchy involvement followed by basal ganglia involvement (80%). Brainstem involvement were noted in 2 patients (40%) with one case showed periventricular involvement. On MR, T2 high signal, with surrounding oedema noted in subcortical locations with few cases showed peripheral enhancement and most were non enhancing. All cases responded well with corticosteroid therapy at neuromedicine department and follow up scan showed complete resolution of lesion.

Four patients of osmotic myelinolysis (10%) presented with quadriparesis, pseudobulbar palsy and few were referred cases of hyponatremia having h/o rapid electrolyte correction from outside remote areas, one was alcoholic and one transplant case. Out of four patients 2(50%) showed extra pontine involvement in addition to 2(50%) showed characteristic involvement of central Pons. All lesion showed patchy diffusion restriction with T2/FLAIR hyperintensity. 3 out of 40 (7.5%) cases of SSPE presented with myoclonus, spasticity & neuropsychological deterioration having incomplete vaccination history. Most of them were child and one was young adult. EEG finding was periodic slow wave complex in those cases. MRI was done in which asymmetric T2/FLAIR temporo-parietal white matter hyperintensity were seen in all 3 cases (100%) along with b/l symmetrical basal ganglia involving putamen were noted in 2 cases (66.67%). Possible diagnosis was made with advice of CSF analysis of anti-measles antibody analysis which was positive in all three cases. One case was a follow up case of a girl child of 12 year age. She was on anti viral and immunomodulator treatment and showed decrease in lesion size and site involvement on follow up imaging. 3 of 40 (7.5%) cases were of MLD. Most of them were of juvenile and one young adult age group. Most common clinical presentation were gait abnormality, loss of vision followed by convulsion and muscle rigidity. All cases (100%), on imaging showed typical b/l symmetrical confluent periventricular deep white matter hyperintensities on T2/FLAIR with subcortical U-fibre and perivenular sparing. No one showed enhancement. MRS showed reduced NAA and some peak at myoinositol and lactate level. 1 out of 40 cases were seen in ALD (fig.1 & 3), Leigh, tumefactive demyelination and autoimmune encephalitis with prevalence of 2.5% each.

ALD case found was in male child of age 5 year who presented with walking difficulty and history of frequent fall. MRI showed involvement of symmetrical parieto-occipital white matter, splenium of corpus callosum, corticospinal tract and cerebellar white matter projecting areas with typical three zonal visualisation on T1/T2 and peripheral serpiginous enhancement. MRS showed decreased NAA and raised lactate peak. Plasma VLCFA level assessment were advised which was elevated. Leigh was noted in 1 year male child who presented with psychomotor delay, ataxia and ophthalmoplegia. MRI findings were b/l putamen and caudate nucleus involvement. Elevated lactate and choline peak with low NAA peak was seen on MRS. CSF examination for lactate was raised.

One case of tumefactive demyelination presented with neurological deficit. On MRI, two lesion with typical open ring enhancement noted in deep white matter without any perilesional edema nor mass effect. One case of autoimmune encephalitis presented with altered mental status and seizure. MRI showed atypical findings with involvement of fronto-temporal juxtacortical involvement without enhancement and subtle patchy restriction. This was paraneoplastic autoimmune encephalitis on follow up.



**Discussion**

MRI is a non-invasive specific and sensitive imaging tool for identification of small lesion of subclinical and clinical cases, disease extent, and disease progression and in their follow up

due to its multiplanar and high resolution images. The pattern of myelination and chronic micro hemorrhages in developmental cases can be identify on CT; but in subclinical or idiopathic cases MR is highly sensitive to identify disease than CT due to its limitations. In our study, we found that FLAIR sequences had a better sensitivity for subtle white matter hyperintensity. PRES<sup>7,8</sup> is a neurotoxic condition which results from raised systemic blood pressure and cerebral hyperperfusion mostly in the posterior region. In our case, all 16 cases (100%) showed parieto-occipital involvement in addition to 62.54%temporal, 25% frontal involvement. No cerebellar involvement seen as compared to Donmez study in which 33.3% was cerebellar involvement.

MS is an autoimmune demyelinating condition<sup>10,12</sup>.In follow up cases MRI plays important role for dissemination in time (DIT) and dissemination in space (DIS) lesion. The most common site of lesion in our study was the periventricular area with female preponderance. All lesions showed faint peripheral and nodular enhancement. ADEM is an immune mediated demyelination<sup>4,5,6</sup> which occurs five days to two weeks following a viral illness or immunization.

Imaging shows asymmetrical subcortical white matter with or without gray matter involvement along with spinal cord involvement.In our study,100% subcortical ,80%b/l basal ganglia,40%brainstem &20%periventricular involvement is seen. Osmotic myelinolysis<sup>9</sup> results from rapid correction of blood low sodium level. Imaging shows pons, midbrain, thalamus and deep white matter involvement. SSPE or Dawson disease<sup>15</sup> is chronic progressive encephalitis of child and young adult age group, caused by persistent infection of immune resistant measles virus. MLD is an AR dysmyelination in which accumulation of sulfatides is seen. Imaging shows B/L symmetrical and confluent periventricular , peri-atrial involvement with perivenular and U fibre sparing.

Our all 3 (100% cases) showed all typical findings. ALD is an X-linked leukodystrophy of children and adults. Loes et al. described five MRI patterns of ALD based on anatomic locations. deep white matter in the parieto-occipital lobes and splenium of the corpus callosum (66% of cases, chiefly in children),frontal lobe or genu of the corpus callosum (15.5%, mostly in adolescents), front pontine or corticospinal projection fibres (12%, mostly in adults), cerebellar white matter (1%, mostly in adolescents) and combined parieto-occipital and frontal white matter (2.5%, mostly children).Except corpus callosum; rest all four findings we noted in a single case of ALD . Leigh syndrome is a mitochondrial disorder with progressive neurodegeneration that invariably leads to death, usually in childhood. our case showed b/l putamen and basal ganglia involvement.

We noted Paraneoplastic related non-specific findings of single case of autoimmune encephalitis.

Thus, MRI is a gold standard modality for early lesion detection, disease progression and in follow up cases in both subclinical and clinical settings. There fore, early management of cases is possible for better patient cure as well as better rehabilitation in chronic cases .

**Table 1: Prevalence of white matter lesions (n=40)**

DISEASE	TOTAL NO. OFCASES	PERCENTAGE
MS	5	12.5

ADEM	5	12.5
PRES	16	40
SSPE	3	7.5
LEIGH	1	2.5
MLD	3	7.5
ALD	1	2.5
OSMOTIC MYELINOLYSIS	4	10
TUMEFACTIVE DEMYELINATION	1	2.5
AUTOIMMUNE ENCEPHALITIS	1	2.5

**Table 2: Site of lesion & prevalence in ADEM (N=5)**

Site of involvement	No. of cases	Percentage
Sub cortical white matter	5	100
BG	4	80
BS and /or CB	2	40
Periventricular	1	20

**Table 3: Site of lesion & prevalence in MS(N=5)**

Site of involvement	No. of cases	Percentage
Periventricular white matter(Dawson finger)	5	100
Deep white matter	3	60
Spinal cord	1	20
Optic nerve	1	20
Juxtacortical lesion	2	40

**Table 4: Site of lesion & prevalence in PRES (N=16)**

Site of involvement	No. of cases	Percentage
Posterior periventricular(parieto-occipital deep white matter region)	16	100
Temporal lobe	10	62.54
Frontal lobe	4	25
Hemorrhagic foci on gradient sequence	2	12.5

**Table 5: Site of lesion & prevalence in Acquired demyelination(osmotic myelinolysis)(N=4)**

Site of involvement	No. of cases	Percentage
Pons	2	50
Basal ganglia	2	50
Subcortical white matter	2	50
Thalamus	1	25
Diffusion restriction	3	75

## Conclusion

MRI is the modality of choice for early lesion detection, disease progression and in follow up cases in both subclinical and clinical settings due to its multiplanar images and better resolution. Therefore, early management for better patient cure is possible.

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