Demyelinating diseases of the central nervous system

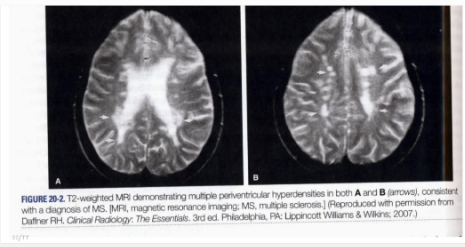
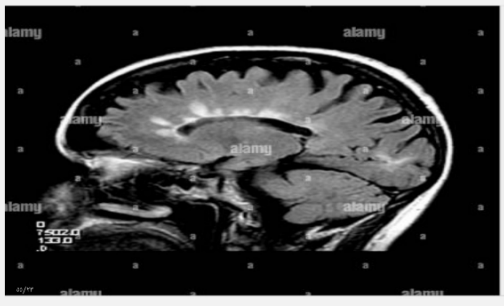


FIGURE 20-2. T2-weighted MRI demonstrating multiple periventricular hyperdensities in both **A** and **B** (arrows), consistent with a diagnosis of MS. [MRI, magnetic resonance imaging; MS, multiple sclerosis.] (Reproduced with permission from Daffner RH. *Clinical Radiology: The Essentials*. 3rd ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2007.)



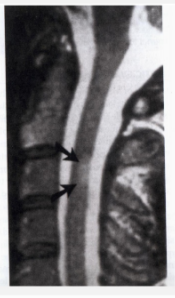


TABLE 20-1. Common Clinical Features of Multiple Sclerosis

Neurologic System	Clinical Sign or Symptom
Cranial nerves	Optic nerve dysfunction
	Visual acuity loss
	Red desaturation
	Papilledema or optic disc pallor
	RAPD
	Eye movement disorders
	Internuclear ophthalmoplegia
	Nystagmus
Motor system	Weakness
	Spasticity
	Reflex abnormalities
	Increased muscle stretch reflexes
	Babinski signs
	Clonus
Sensory system	Paresthesias
	Vibratory loss
	Joint position sense loss
	Lhermitte's sign
Cerebellar function	Ataxia
	Intention tremor
	Dysarthria
Autonomic system	Bladder dysfunction
Other	Fatigue
	Depression
	Uhthoff's phenomenon

RAPD, relative afferent pupillary defect.

MS

-affect young people, women ,monozygotic twins, and white more

- latent childhood viral infection may play a role in pathogenesis , migration after 15y will not change risk associated with childhood home

-multiple white matter lesions separated in space and time. (Not monophasic disease)

-In later stages, cognitive and behavioural abnormalities may occur

-Optic neuritis, common initial presentation

-painful loss of visual acuity in one eye (when eye moves), worse with heat (Uhthoff's phenomenon)

-could be mild blurriness, loss of color discrimination ,complete blindness

- optic disk could appear normal (retrobulbar ON) or swollen (papilledema)

-ON leads to : red desaturation, disk atrophy, RAPD

-Transverse myelitis ,affects particular tracts at the level of the lesion in a patchy way, bladder dysfunction

-Reflexes may be exaggerated below the lesion, and Babinski signs may be present

-pain around torso

-Internuclear ophthalmoplegia, dysfunction of the medial longitudinal fasciculus, inability to adduct one eye with nystagmus of the other eye ,convergence) is preserved

-Lhermitte' s sign electric sensation down the spine when the patient flexes the neck, and Uhthoff's phenomenon, a worsening of symptoms and signs in the heat

-types of clinical coarse : relapsing-remitting , primary progressive, secondary progressive,

progressive remitting
- good prognosis: young female , relapsing remitting, sensory symptoms
- On MRI new lesions: appear as T2-hyperintense areas , showed with FLAIR sequence , enhanced with gadolinium
-chronic lesions may become T1-hypointense, with a " black hole" appearance
CSF shows OCB

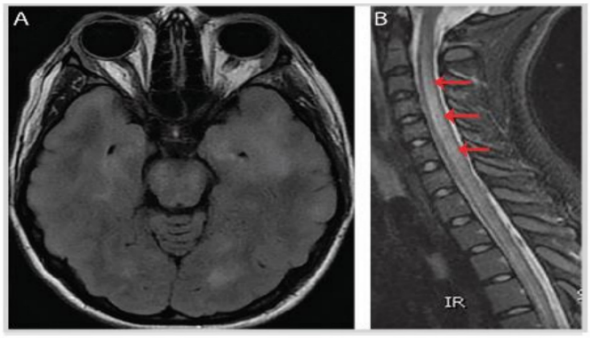


FIGURE 11-12 Imaging of the patient in **Case 11-2** with acute disseminated encephalomyelitis (ADEM). *A*, Axial fluid-attenuated inversion recovery (FLAIR) brain MRI shows poorly demarcated T2 hyperintensities in cortical, subcortical, and brainstem areas. *B*, Sagittal short tau inversion recovery (STIR) spinal cord MRI shows T2 hyperintense signal abnormality throughout the cervical cord (*arrows*).

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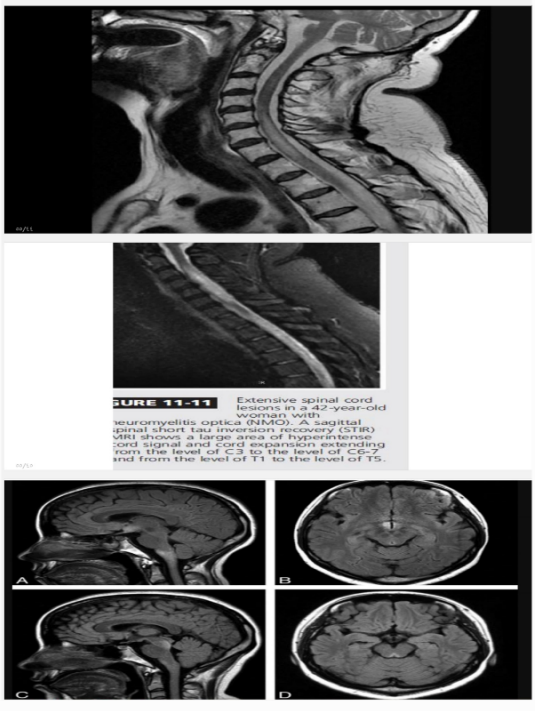
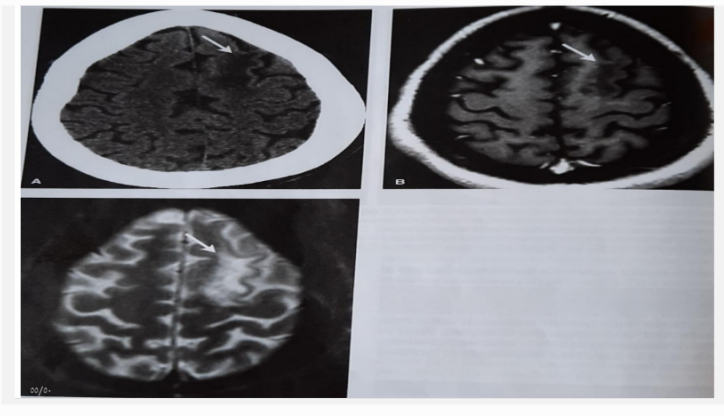
Acute disseminated encephalomyelitis (ADEM)

- all lesions are new and bilateral , acute after viral infection or vaccination
-cognitive and behavioral changes and seizures (which come late in MS)
-CSF show a lymphocytic pleocytosis, rare oligoclonal ab
-monophasic illness, steroids are given



Neuromyelitis optica (Devic disease)

-2 manifestations (transverse myelitis and optic neuritis)may be at same time or separated by 2 y
-Demyelination of the brain should be absent
- there is pain with more severe dificit than in MS
-CSF pleocytosis, sometimes with a neutrophilic pleocytosis
-antibodies to the aquaporin-4 channel(NMO Ab) or MOG Ab
-Acute treatment of NMO includes steroids and plasmapheresis
-Chemotherapeutic agents such as azathioprine, mycophenolate mofetil and rituximab are used to prevent recurrence



Progressive multifocal leukoencephalopathy (PML)

-dementia, focal cortical dysfunction, and cerebellar abnormalities
-patients with AIDS, leukemia, lymphoma, and MS patients treated with netalizumab
-JC virus infect oligodendrocytes
-CSF is normal