Demyelinating Diseases of the Brain

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T1-Weighted Images
Normal White Matter

Contents

- Axons with envelope of myelin
- Neuroglia - Astrocytes & Oligodendrocytes
- Vascular structures
- Interstitial space
CNS Myelin

- Produced & maintained by oligodendrocytes
- Functions as insulator of axons
- Facilitates transmission of neural impulses
- Relatively short T1 and T2 relaxation times
- Demyelinating disease
  - Less myelin & more water
  - Prolongation of both T1 and T2
White Matter Diseases

- Effects of aging
- Multiple sclerosis
- Infection & Inflammation
- Radiation injury
- Toxic & Metabolic disorders
- Hereditary disorders
History: 26 y/o female with migraine headaches
Dx:
History: 59 y/o woman with frequent falls

Dx:
History: 77 y/o man with hearing loss
History: 55 y/o man with cognitive decline
Dx: Etat crible
History: 89 y.o. man with multiple stroke-like episodes
Dx:
White Matter Ischemia

- Pathophysiology
- Functional significance
- Associated with aging
- What do we call it?
- MR findings
- Differential diagnosis
Deep White Matter Ischemia

Imaging Features

- Subcortical & periventricular location
- Hyperintense on T2 & FLAIR images
- Mildly hypointense on T1 images
- No enhancement
- Sparing of subcortical U-fibers & corpus callosum
History: 62 y.o. male with history of poorly controlled hypertension
History: 76 y.o. male with vertebral-basilar TIA's
Vertebral-basilar TIA's
History: 79 y/o man with left leg weakness
Leg weakness {Page 2}
Deep White Matter Ischemia
Periventricular Disease
Dx:
History: 55 y/o female with motor weakness, double vision, & emotional lability
Motor weakness

{Page 2}
Dx: Chronic multiple sclerosis; Incidental sebaceous cyst left posterior scalp.
Multiple Sclerosis
Clinical Features

- Chronic inflammatory disease of myelin
- Patients between 20 and 50 years old
- Etiology unknown, but probably a post-viral autoimmune reaction
- Acute, transient attacks of neurologic dysfunction
- Dx by history, physical exam, lab & imaging
History: 35 y/o man with bilateral facial pain
History: 50 y/o woman with a right hemiparesis
Dx: MS – subcallosal striations
History: 28 y/o woman referred for brain tumor
History: 45 y/o woman with arm & leg weakness, sensory changes, & bilateral visual loss
Dx: Neuromyelitis optica (Devic's disease)
History: 25 y/o woman with right sided weakness
Dx: Balo's concentric sclerosis
History: 23 y/o man with intermittent and migratory numbness & paresthesias for several months
Dx: Chronic progressive MS
## Differential Features

<table>
<thead>
<tr>
<th></th>
<th>WM Ischemia</th>
<th>Multiple Sclerosis</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Age</strong></td>
<td>&gt; 60 years</td>
<td>20 - 50 years</td>
</tr>
<tr>
<td>Corpus callosum</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>Subcortical U-fibers</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>Enhance</td>
<td>No</td>
<td>Yes (acute)</td>
</tr>
</tbody>
</table>
History: 62 y.o. man with episodes of weakness which was poorly localized
Episodes of weakness
{Page 2}
Dx: Multiple sclerosis
History: 53 y/o woman with 4 days of headache, fatigue, ataxia and poor memory
Dx: Metastasis – Adeno CA Lung & chronic MS
White Matter Diseases

Infectious & Inflammatory Disorders

- Progressive multifocal leukoencephalopathy
- Postinfectious encephalomyelitis
- HIV encephalitis
History: 31 y/o male with multiple neurological deficits
Dx: AIDS - PML
History: 48 y/o HIV+ man with fever, mutism, ataxia & dysphagia
Ataxia & dysphagia
{Page 2}

Dx:
History: 8 year old boy with 1-2 week history of left eye ptosis, limited EOMs, and ataxia.
Dx:
Acute Disseminated Encephalomyelitis (ADEM)

- A post-infectious encephalitis
- Immune-mediated reaction
- Viral agents: measles, chickenpox or vaccination for rabies or smallpox
- Acute demyelinating disease
- Mostly a monophasic illness
History: 67 y/o woman with progressively altered mental status over several days
Dx: Probable ADEM

Bronchitis 3 weeks earlier
History: 61 y/o woman with diabetes & hypertension presented with aphasia
Dx: Post-Infectious encephalitis (Acute disseminated encephalomyelitis)
History: 35 y/o male with fever & ataxia
Dx: HIV encephalitis
Radiation Injury

Imaging Features

- Latent period of 6-8 months
- Symmetric white matter involvement
- Initial sparing of corpus callosum and subcortical U-fibers
- Scalloped outer margins
- Progression over 2-3 years
History: 51 y/o woman with a breast mass
4 months post radiation
Dx: Radiation change

14 months post radiation
History: 52 y/o women had radiation therapy for a glioma 1 year ago
Dx: Radiation injury
White Matter Diseases

Acquired Toxic & Metabolic Disorders

- Effects of chemotherapeutic agents
- Immunosuppressant therapy (PRES)
- Central pontine myelinolysis
- Organic Compounds and Drugs
Neurotoxic Chemotherapeutic Agents

Methotrexate, *cis*-platin, ARA-C, BCNU, Thiotepa

- **Acute effects:**
  - Develop during therapy
  - Transient diffuse WM hyperintensity
- **Delayed effects:**
  - Onset within few weeks or months
  - Patchy or confluent WM pattern
  - Little mass effect or enhancement
  - Calcification in children
  - Synergistic effect of chemo & radiation
History: 67 y/o female with lung CA

Dx:
History: 19 y/o woman with systemic lupus erythematosus, candida pneumonia & increasing confusion
Treated with cytoxan 2 weeks earlier
Dx: Toxic encephalopathy from cytoxan therapy
History: 47 y/o woman with myelodysplasia developed seizures and mental confusion
s/p bone marrow transplantation
Dx: Cyclosporine toxicity
Immunosuppressant Therapy

- Cyclosporines most widely used
- Neurotoxic effects in 20%
- High signal lesions on T2WIs
- Predilection for occipital white matter
- Gray matter damage also occurs
- White matter injury often reversible
History: 30 y/o man with headache, left HH, & left HP
Renal transplant rejection while on cyclosporine; Bp = 220/120
Tx: anti-hypertensives & steroids 2 months later

Dx: PRES (Cyclosporine & acute hypertension)
History: 53 y/o man admitted with liver failure & hyponatremia
Dx: Central pontine myelinolysis
History: 25 y/o male found comatose with decerebrate posturing
Dx: Toxic encephalopathy – Benzodiazepine & Heroine
Metabolic Diseases

- Inherited disorders
- Specific enzyme deficit
- Early infancy or childhood
- Prominent white matter involvement, but gray matter also affected
History: 14 y.o. boy with recent onset of disorientation and confusion
Disorientation & confusion
Dx: Adrenoleukodystrophy