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KEY FACTS

TERMINOLOGY

- Autoimmune-mediated white matter (WM) demyelination of brain &/or spinal cord, usually with remyelination

IMAGING

- Best diagnostic clue: Multifocal WM and deep gray lesions days to weeks following infection/vaccination
- May involve both brain and spinal cord; WM > gray matter, but usually both affected
- Deep/juxtacortical WM > periventricular WM
- Both supratentorial and infratentorial lesions
- Multifocal punctate to large flocculent FLAIR hyperintensities
- Do not usually involve callososeptal interface
- Most lesions show increased signal on DWI (T2 shine through)
- Punctate, ring, incomplete ring, peripheral enhancement
- Absence of enhancement does not exclude diagnosis
- MRS: ↓ NAA within lesions; may see ↑ Cho, ↑ lactate

TOP DIFFERENTIAL DIAGNOSES

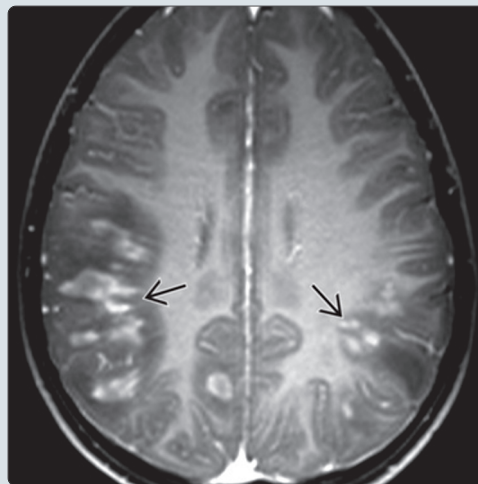
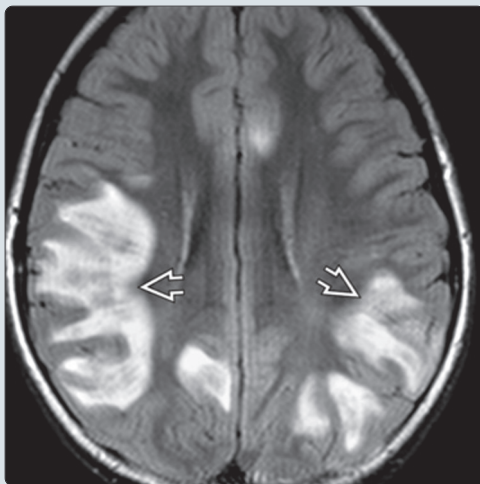
- Multiple sclerosis (MS)
 - Incomplete rim enhancement most common in ADEM
- Neuromyelitis optica
- Autoimmune-mediated vasculitis
- Acute hypertensive encephalopathy, PRES
- Fabry disease
- Behçet disease



PATHOLOGY

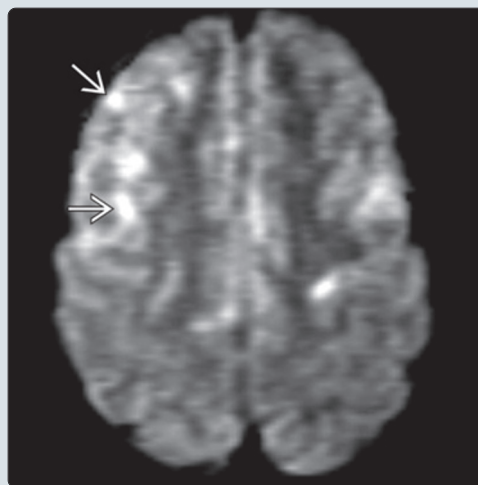
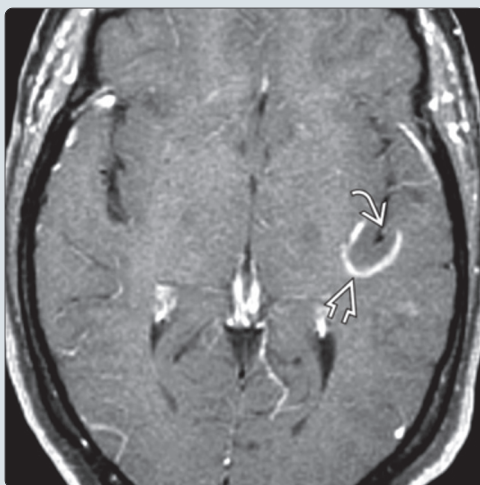
- > 30 different infectious agents & immunizations reported




CLINICAL ISSUES

- Mean age is 5-8 years but can occur at any age
- Male predominance (M:F = 1.0:0.6-0.8), unlike MS
- Usually monophasic, self-limited
- Complete recovery within 1 month: 50-60%
- Mortality: 10-30%



(Left) Axial FLAIR MR shows peripheral, confluent areas of hyperintensity  predominantly involving the subcortical white matter in this child with ADEM. The bilateral but asymmetric pattern is typical of ADEM. (Right) Axial T1WI C+ MR in the same patient shows marked, irregular enhancement of nearly all of the lesions . Because ADEM is usually a monophasic illness, enhancement of the majority of lesions is typical, as the lesions all have a similar time course. Enhancement of MS lesions is more variable.



(Left) Axial T1WI C+ MR shows an incomplete ring or "horseshoe" of peripheral enhancement  with open ends pointing toward the cortex , classic for a demyelinating process (ADEM > MS). (Right) Axial DWI MR in a patient with ADEM shows multiple foci of increased signal . The foci were hypointense on ADC images (not shown), indicating diffusion restriction. White matter, gray matter involvement is present. Diffusion restriction is an uncommon finding, associated with a worse prognosis.