Autoimmune Glial Fibrillary Acidic Protein Astrocytopathy

- Rare but probably underdiagnosed autoimmune-related disease that affects the CNS, first described in 2016.
- Antibodies target the glial fibrillary acidic protein, which is an intermediate filament expressed by astrocytes.
- Although the origin of this disease is unknown, up to 20% of cases occur as a paraneoplastic syndrome (benign or malignant neoplasm)

Clinical Presentation

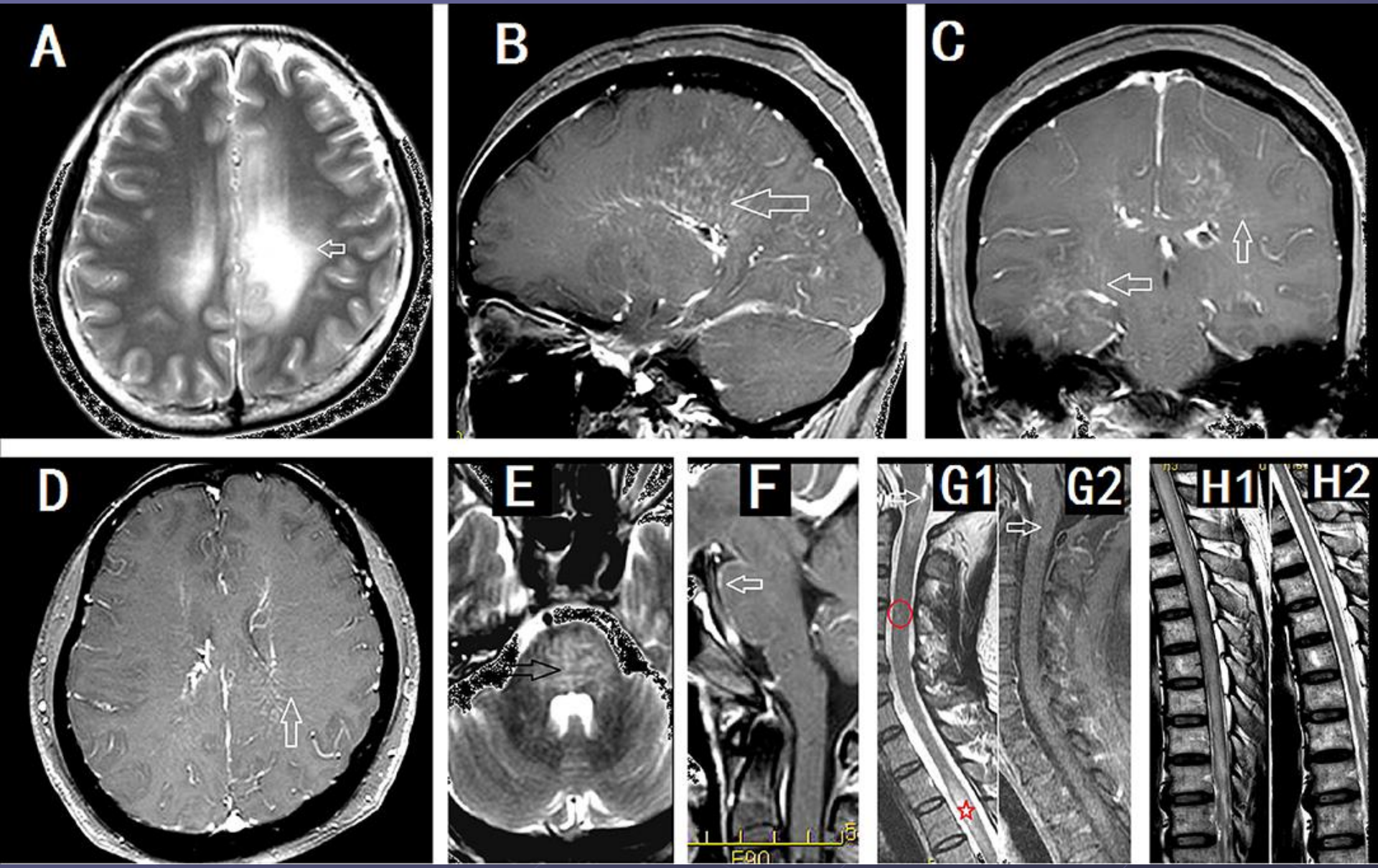
- Most patients present with an acute or subacute onset of meningo-encephalo-myelitis,
- main symptoms being
  - Headache
  - Encephalopathy
  - Involuntary movements
  - Ataxia,
  - Optic papillitis
  - Myelitis
- Early flu-like symptoms are often identified.

Key Diagnostic Features

- Diagnosis mainly relies on the detection of GFAP-IgG in CSF, along with imaging.
- Brain:
  - perivascular enhancement pattern (linear radial periventricular enhancement), T2- and FLAIR-hyperintensities in the supratentorial white matter, basal ganglia, and thalami
- Spinal cord:
  - longitudinally extensive “hazy” T2-hyperintensities, “speckle” intramedullary enhancement, periependymal and leptomeningeal enhancement

DDx:

- NMOSD and MOGAD: both entities can show similar spinal cord imaging findings as in GFAP astrocytopathy. However, supratentorial linear radial periventricular enhancement is uncommon in these diseases. Anti-aquaporin 4 and anti-MOG IgG are helpful when positive.
- CLIPPERS: perivascular enhancement predominantly involves brainstem and cerebellum; supratentorial structures are usually spared.
- CNS endovascular lymphoma: perivascular enhancement is accompanied by areas of restricted diffusion (cerebral infarcts) and hemorrhagic lesions.
- Infectious meningo-encephalo-myeliti: CSF analysis is the key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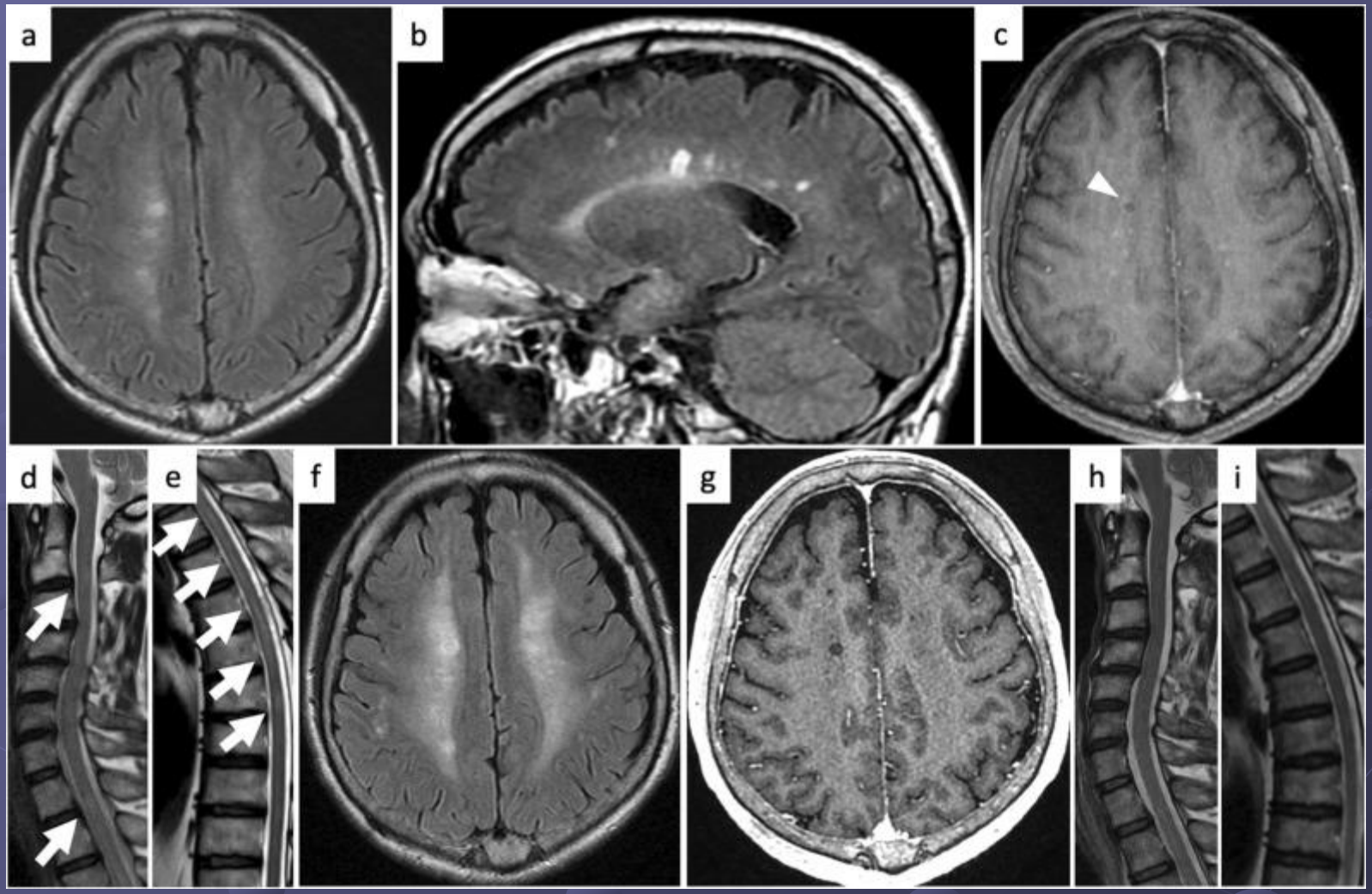
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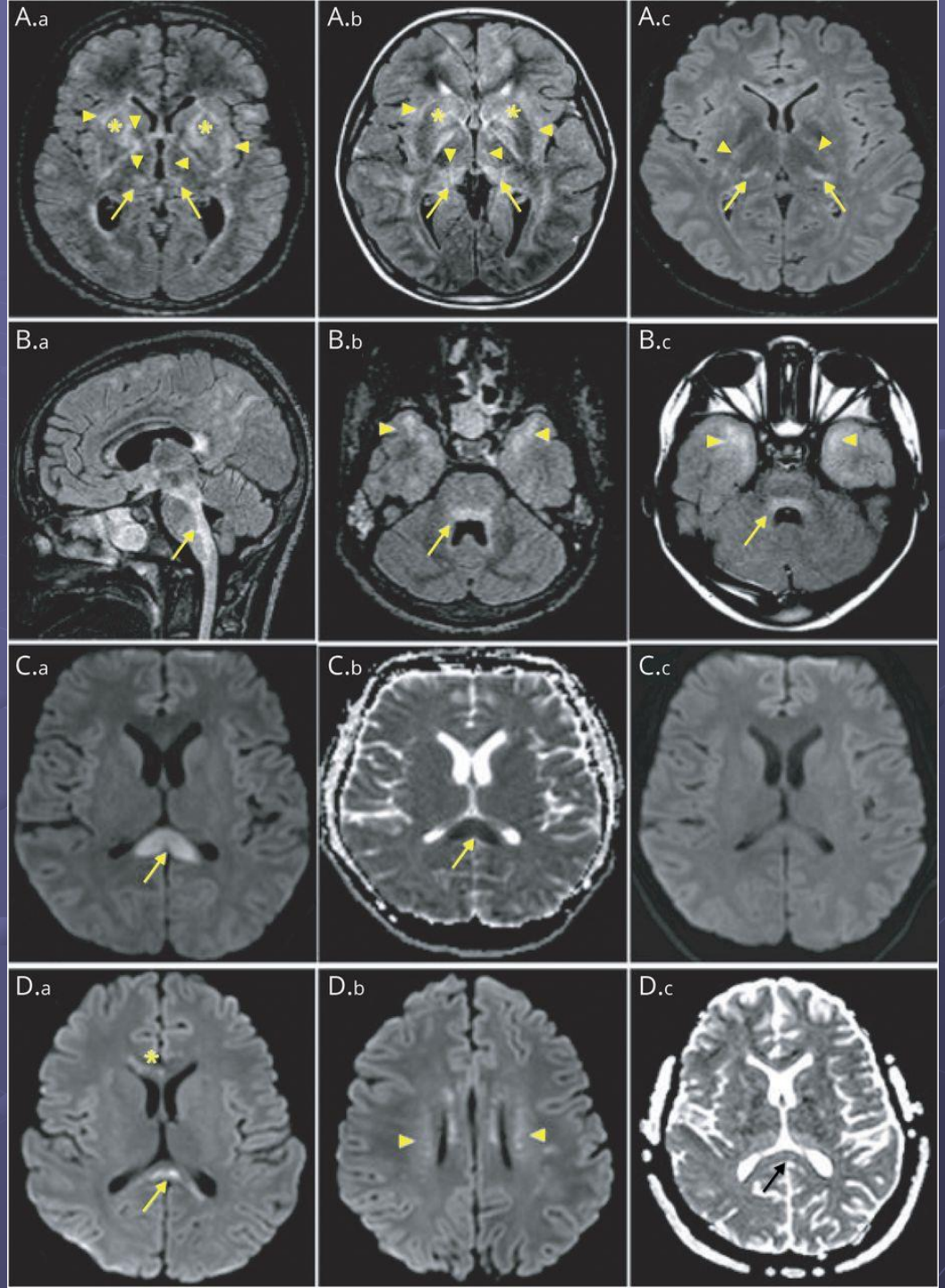
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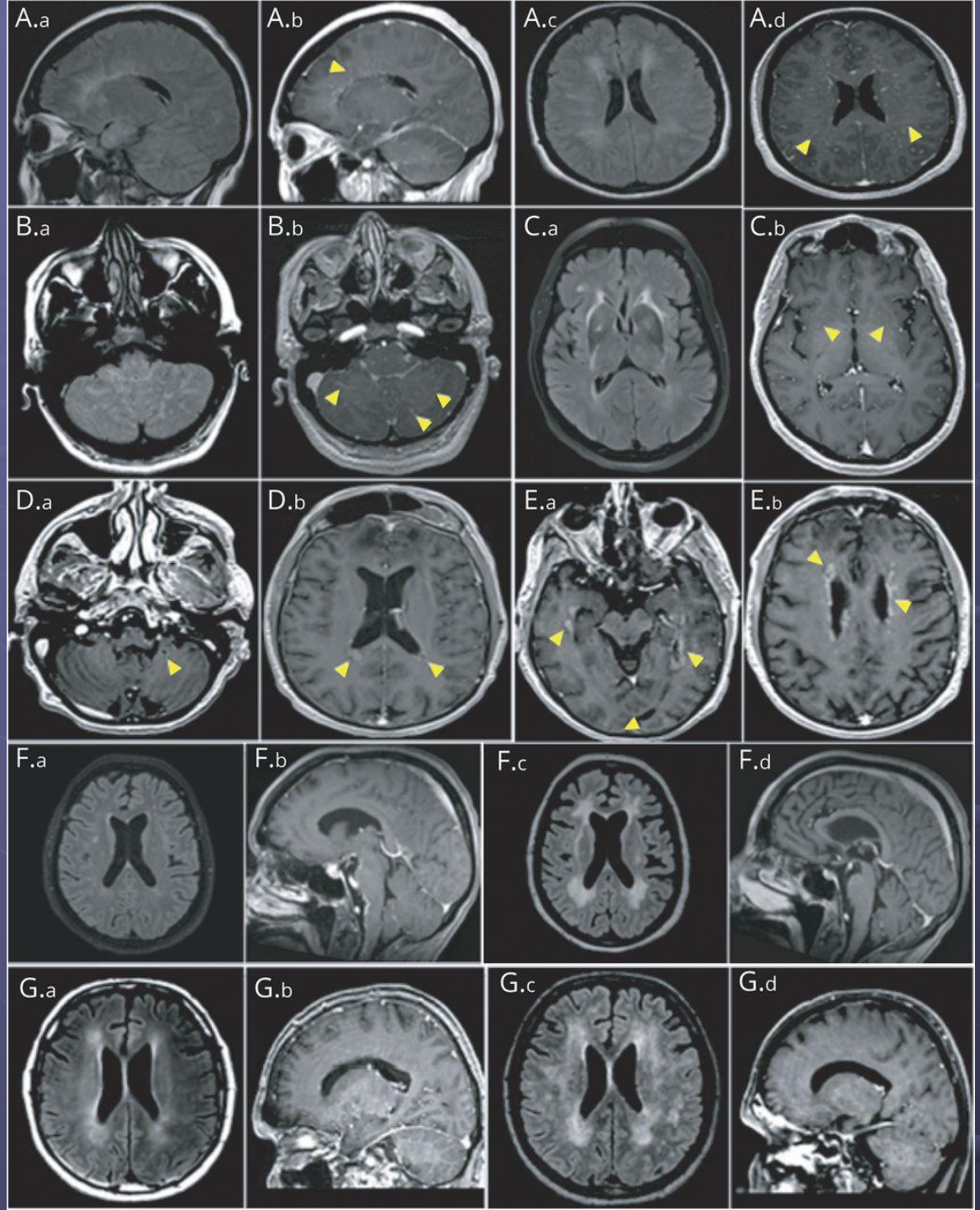
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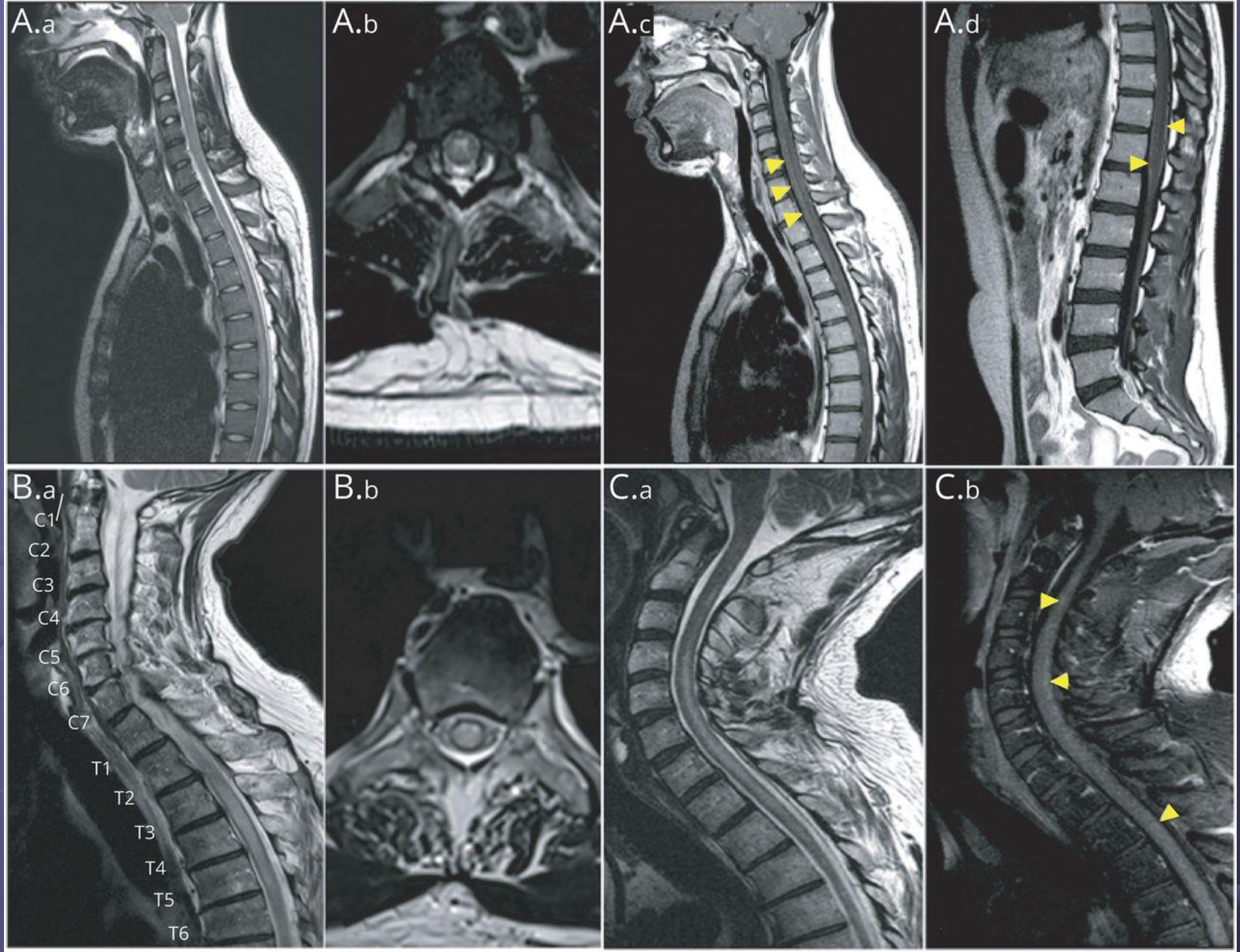
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Neurosarcoidosis

- Linear radial periventricular enhancement pattern on MRI brain is a hallmark of
- Autoimmune glial fibrillary acid protein (GFAP) astrocytopathy
- DDx:
  - Neurosarcoidosis
  - Intravascular lymphoma
  - CNS vasculitis