AMSER Case of the Month February 2025

40 y.o presenting with neck pain and progressive weakness of all 4 extremities

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Patient Presentation

- 40 y.o with PMH of fibromyalgia presented to her PCP with neck pain and worsening weakness in all four extremities, worse on the right side. The patient also complained of decreased depth perception in both eyes, intermittent headaches, and upper extremity tingling. She denied any episodes or fever.

Pertinent Labs

- CSF
  - 0 WBC
  - 59 Glucose
  - 56 Total protein
  - West Nile/HSV Ab - negative
  - AQP-4 antibodies – positive/detected
- Blood
  - WBC 6.9 K/uL
  - ESR 4mm/Hr

What Imaging Should We Order?

Select the ACR Appropriateness Criteria

Variant 1: **Acute onset myelopathy. Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
MRI spine area of interest without and with IV contrast	Usually Appropriate	O
MRI spine area of interest without IV contrast	Usually Appropriate	O
CT myelography spine area of interest	May Be Appropriate	Varies
CT spine area of interest with IV contrast	May Be Appropriate	Varies
CT spine area of interest without IV contrast	May Be Appropriate	Varies
Arteriography spine area of interest	Usually Not Appropriate	Varies

Variant 7: **Headache with one or more of the following "red flags": increasing frequency or severity, fever or neurologic deficit, history of cancer or immunocompromise, older age (>50 years) of onset, or posttraumatic onset. Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
MRI head without and with IV contrast	Usually Appropriate	O
MRI head without IV contrast	Usually Appropriate	O
CT head without IV contrast	Usually Appropriate	☢☢☢
Arteriography cervicocerebral	Usually Not Appropriate	☢☢☢

These studies were ordered by the PCP

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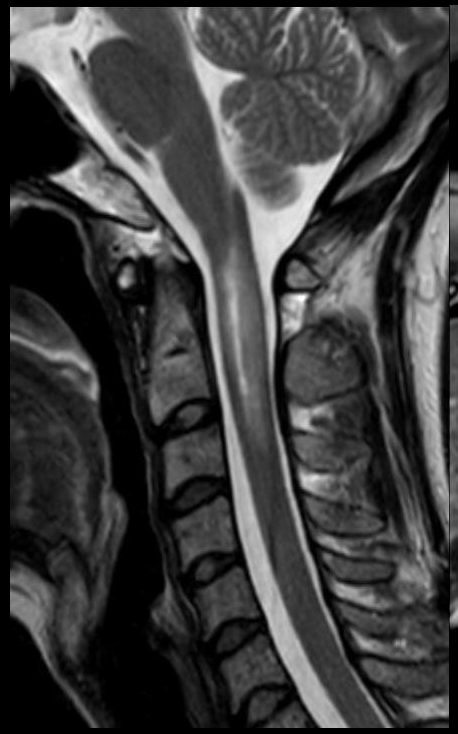
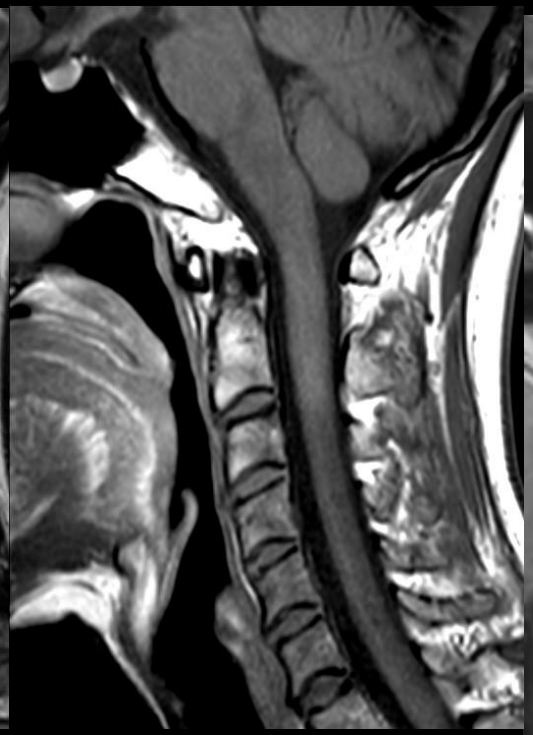
Findings: (unlabeled)

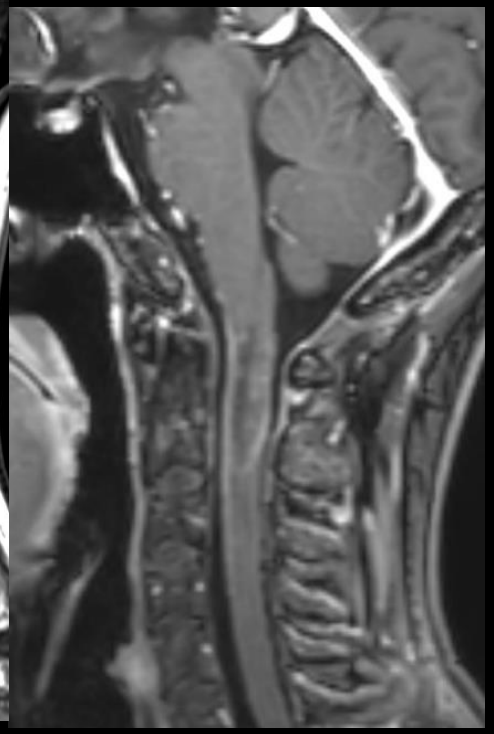
Sag T2



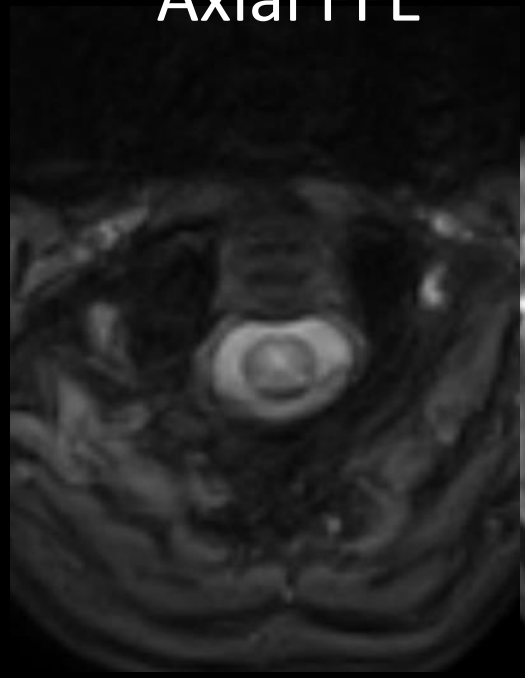
Sag T1



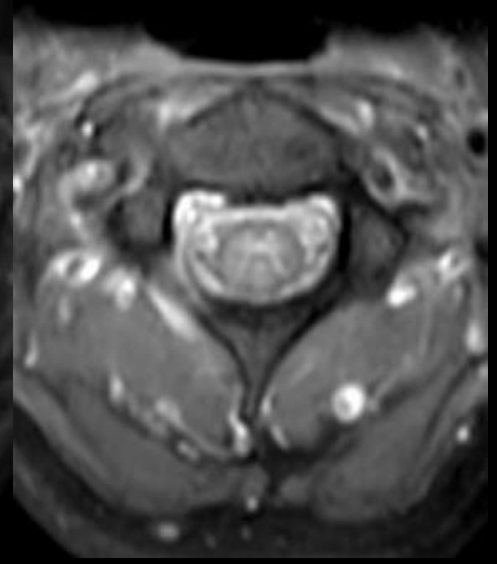
Sag T1 +C



Axial FFE



Axial T1 +C



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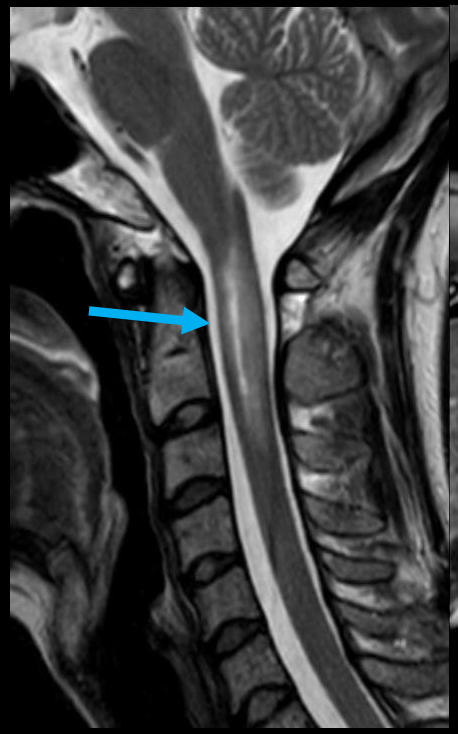
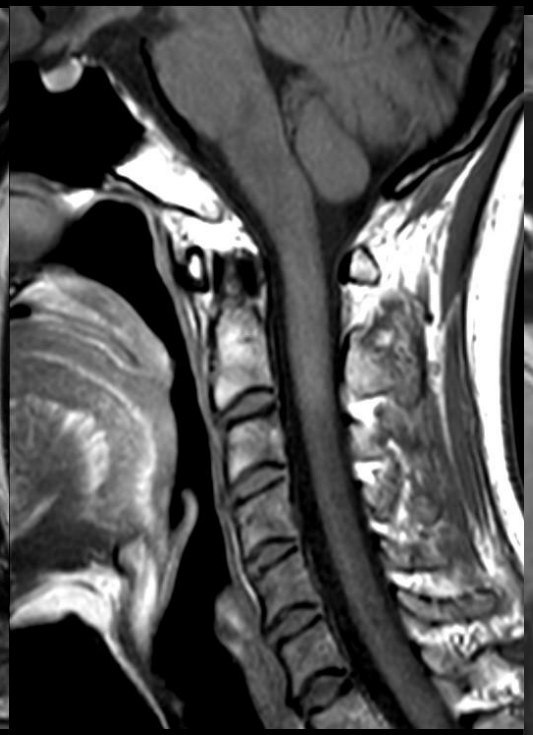
Findings: (labeled)

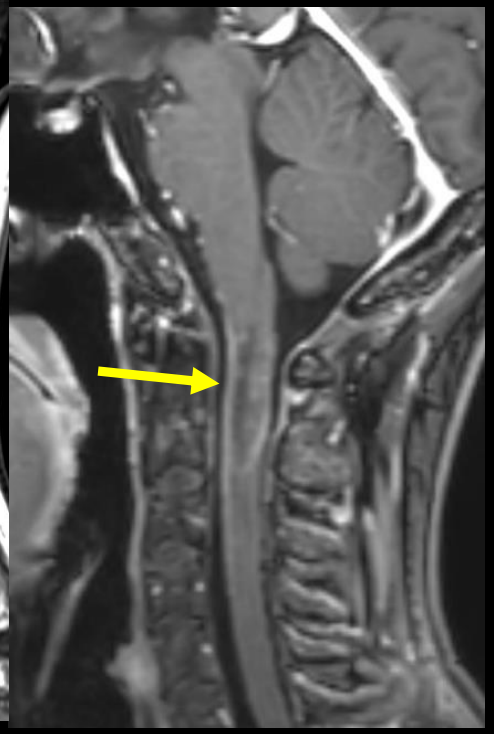
Sag T2



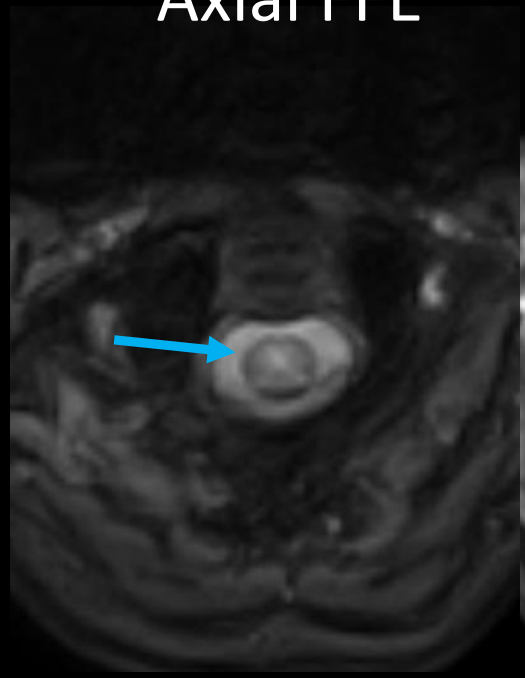
Sag T1



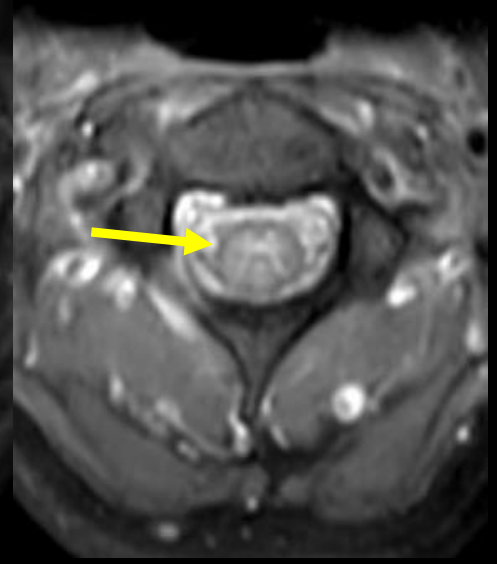
Sag T1 +C



Axial FFE



Axial T1 +C



Expansile T2 hyperintensity centered in central spinal cord from C1 to C3 with patchy peripheral and central enhancement.

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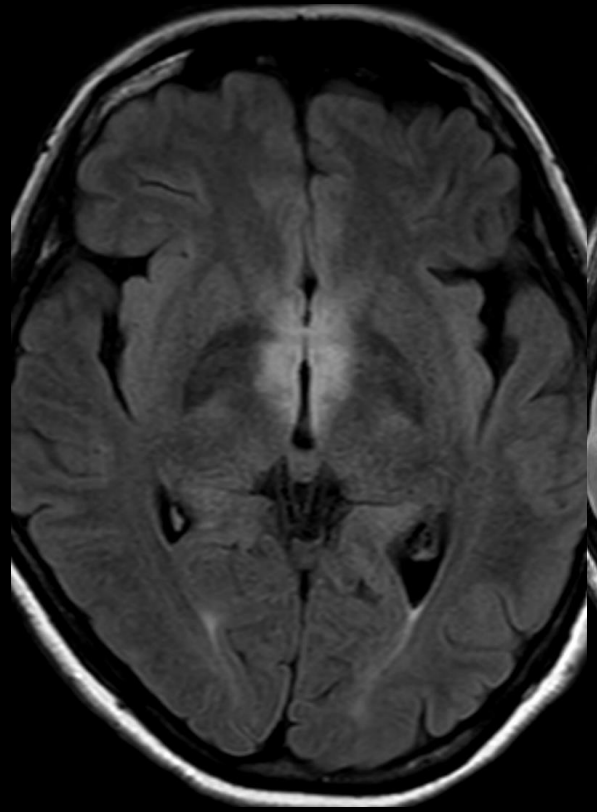
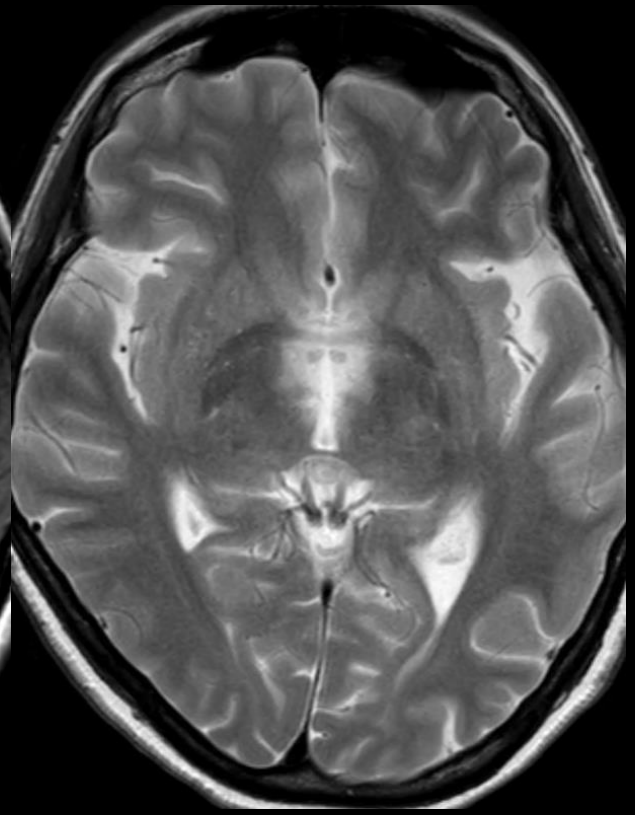
Findings (unlabeled)

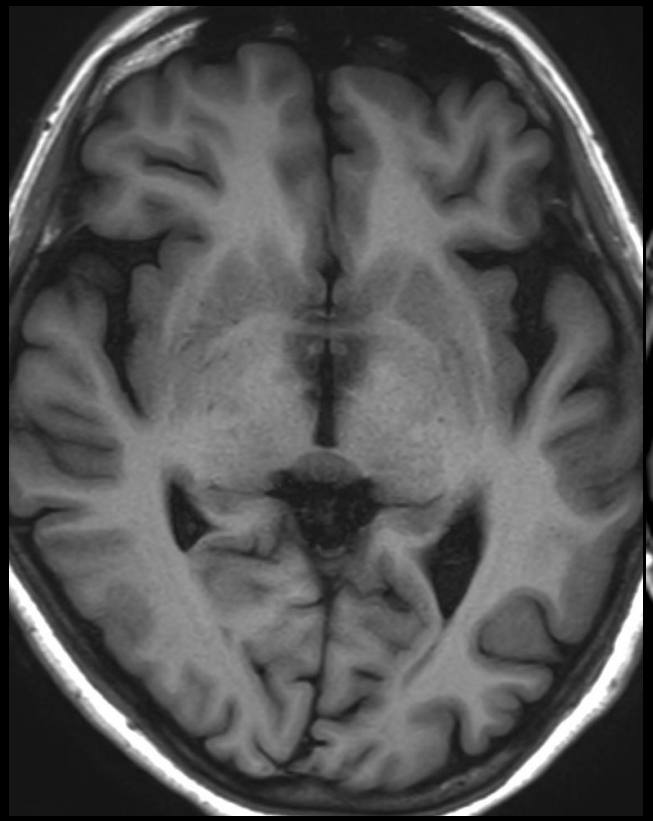
Axial T2-FLAIR



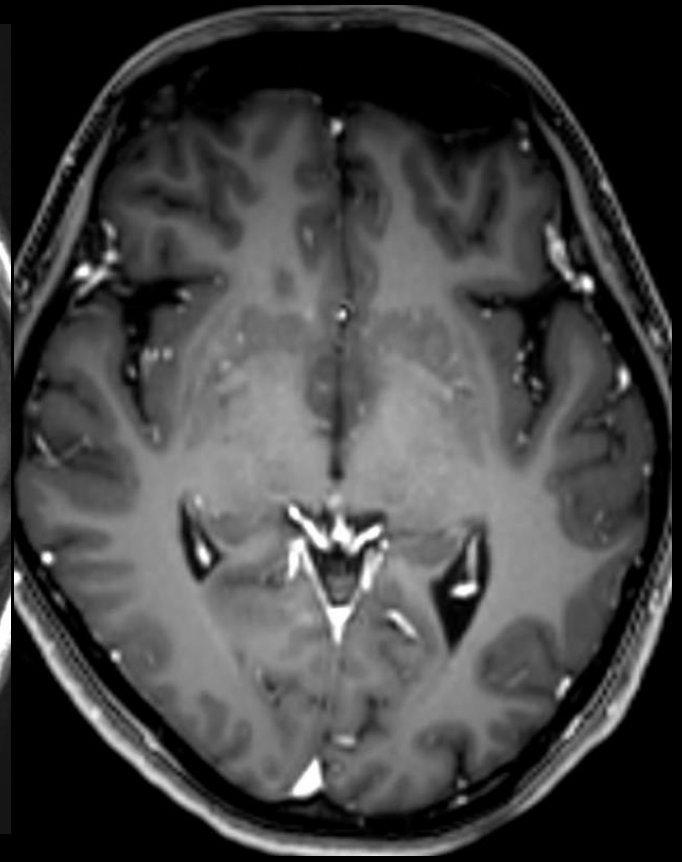
Axial T2



Axial T1



Axial T1+C



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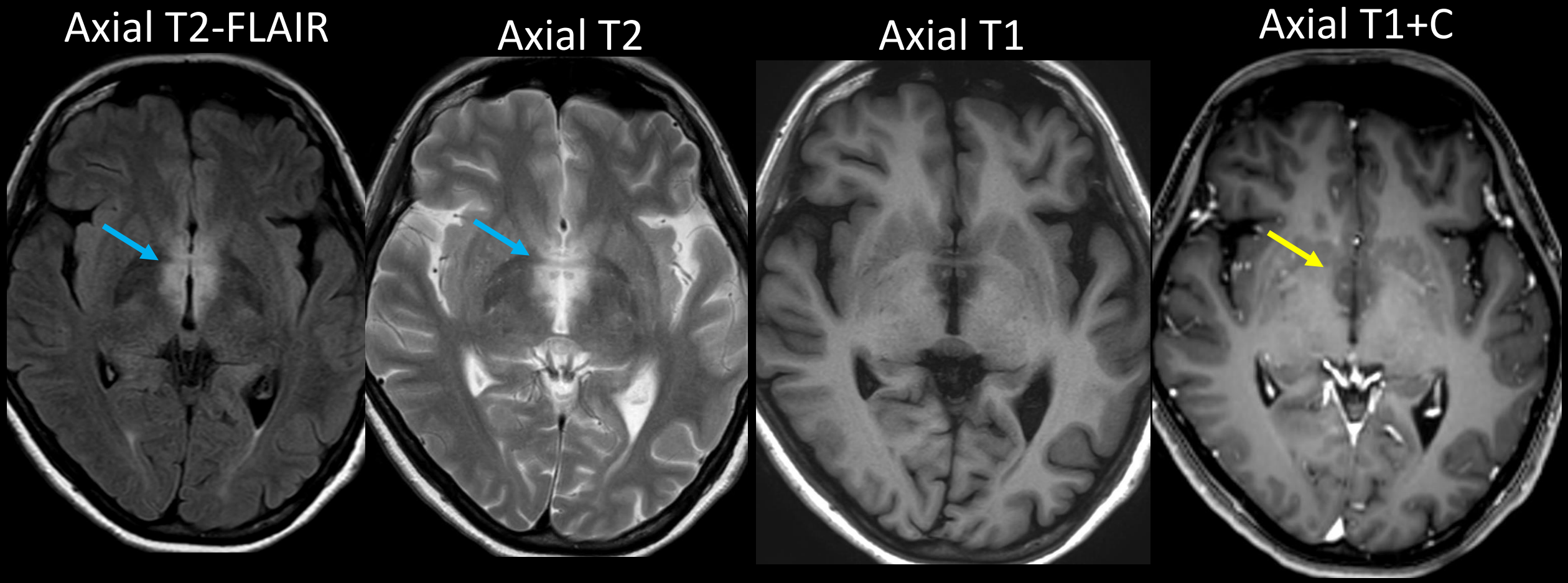
Findings (labeled)



Hyperintensity centered in hypothalamus, medial thalami, and anterior commissure on FLAIR and T2WI. No visible enhancement.

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Final Dx:

Neuromyelitis Optica

Case Discussion

- Background
  - Neuromyelitis optica (NMO) is an autoimmune inflammatory demyelinating disorder caused by aquaporin-4 antibodies (AQP4-IgG) against astrocyte water channels in the central nervous system (CNS).[1]
  - High female to male predilection (9:1) with average age of onset around 40 years old.[2,3]
  - NMO often co-exists with other autoimmune disorders (Lupus and Sjogren's).
- Pathology
  - AQP4-IgG biomarker provides distinction from other demyelinating disorders (i.e. multiple sclerosis) and broadens the recognized sites of CNS involvement (e.g., optic nerve, spinal cord, diencephalon, brainstem, and cerebrum).[1]

Case Discussion

Clinical Presentation

- Classic triad of positive AQP4-IgG test, optic neuritis, and myelitis.
  - Often present with blindness and paraplegias (not always simultaneous).
- 2015 International Panel for NMO Diagnosis created criteria as follows:

  1) Presence of one or more core clinical characteristics:
     - optic neuritis, acute myelitis, area postrema syndrome, acute brainstem syndrome, symptomatic narcolepsy, acute diencephalic clinical syndrome with NMOSD-typical lesions on MRI, symptomatic cerebral syndrome with NMOSD-typical brain lesions

  2) positive AQP4-IgG test

  3) exclusion of alternative diagnoses[3]
- Additional diagnostic criteria were also generated for NMO without AQP4-IgG antibodies or NMO with an unknown AQP4-IgG status[3]

Case Discussion

- Typical radiologic findings
  - Bilateral optic neuritis (involvement to the chiasm is suggestive of NMO).[1,4]
  - Transverse myelitis spanning multiple segments (> 3 vertebra), follows aquaporin-4 channels of gray matter around central canal of spinal cord.[1,4]
    - Variable enhancement, bright T2/STIR hyperintensity
  - Brain lesions follow location of aquaporin-4 channels (periependymal regions)[1,4]
    - Periventricular - sessile lesions (not like Dawson fingers of multiple sclerosis)
    - Periaqueductal gray matter
    - Corpus callosum
    - Hypothalamus, medial thalamus, dorsal pons, and medulla

Case Discussion

- Treatment
  - Acute attacks require high dose glucocorticoids or apheresis therapy.[5]
  - Long-term management with immunosuppressant therapy, various therapeutic antibodies, or IVIG.[5]

- Differentiating NMO from multiple sclerosis is important as the disease-specific therapies are different and the use of MS medications in NMO have reportedly exacerbated the disease
  - Main differences: NMO has longer segment involvement of spinal cord, preferential posterior segment involvement of optic nerves, more sessile type lesions in periventricular white matter, and lacks open ring enhancement[1]

References:

1. Dutra BG, da Rocha AJ, Nunes RH, Maia ACM Júnior. Neuromyelitis Optica Spectrum Disorders: Spectrum of MR Imaging Findings and Their Differential Diagnosis [published correction appears in Radiographics. 2018 Mar-Apr;38(2):662. doi: 10.1148/rg.2018184002]. *Radiographics*. 2018;38(1):169-193. doi:10.1148/rg.2018170141
2. Hor JY, Asgari N, Nakashima I, et al. Epidemiology of Neuromyelitis Optica Spectrum Disorder and Its Prevalence and Incidence Worldwide. *Front Neurol*. 2020;11:501. Published 2020 Jun 26. doi:10.3389/fneur.2020.00501
3. Flanagan EP, Cabre P, Weinshenker BG, et al. Epidemiology of aquaporin-4 autoimmunity and neuromyelitis optica spectrum. *Ann Neurol*. 2016;79(5):775-783. doi:10.1002/ana.24617
4. Wingerchuk DM, Banwell B, Bennett JL, et al. International consensus diagnostic criteria for neuromyelitis optica spectrum disorders. *Neurology*. 2015;85(2):177-189. doi:10.1212/WNL.0000000000001729
5. Kümpfel T, Giglhuber K, Aktas O, et al. Update on the diagnosis and treatment of neuromyelitis optica spectrum disorders (NMOSD) - revised recommendations of the Neuromyelitis Optica Study Group (NEMOS). Part II: Attack therapy and long-term management [published correction appears in J Neurol. 2024 Jun;271(6):3702-3707. doi: 10.1007/s00415-024-12288-2]. *J Neurol*. 2024;271(1):141-176. doi:10.1007/s00415-023-11910-z

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