

Eight-and-a-Half Syndrome as an Initial Manifestation of Multiple Sclerosis

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Introduction

- Eight-and-a-half syndrome classically presents ipsilateral conjugate horizontal gaze palsy, internuclear ophthalmoplegia (INO), and an ipsilateral facial nerve palsy.
- The syndrome localizes to the dorsal tegmentum of the caudal pons involving lesions in the paramedian pontine reticular formation or abducens nucleus, the medial longitudinal fasciculus, and cranial nerve VII¹⁻⁴.
- Although ischemic stroke is the most common cause, multiple sclerosis (MS) is a rare etiology.

Objectives

- We describe eight-and-a-half syndrome as an atypical initial manifestation of MS.
- We discuss the clinical presentation, neuroimaging findings, and management associated with this syndrome.

Case Presentation

History of Present Illness:

- A 34-year-old male presented to our facility with headache, acute right facial weakness and diplopia.
- No inciting event, associated trauma, or prior history of stroke-like symptoms.
- No other constitutional, cardiovascular, pulmonological, GI, or MSK symptoms; review of systems otherwise negative.

Past Medical History:

- Recently diagnosed hyperthyroidism.

Family History:

- Denies family history of autoimmune diseases.

Social History:

- Denies history of alcohol use, tobacco use, or illicit drug use.

Medications:

- Denies medications.

Physical Exam:

- Hemodynamically stable; Vital Signs: HR: 74 bpm, RR: 16 br/min, BP: 114/70 mmHg, O2 saturation: 98% on RA
- Exam: Alert and oriented to person, place, time, and situation. Neurological exam significant for right lower motor neuron facial palsy, right horizontal gaze palsy, and right INO. Cranial nerves, strength elsewhere, cerebellar function, and gait within normal limits. Otherwise, physical exam normal.

Laboratory studies:

- Unremarkable for inflammatory, autoimmune, and infectious etiologies.

Results

- CT/CTA/CTV: No acute intracranial hemorrhage, mass effect, or significant stenosis. CTV Head with questionable dural venous thrombosis, but subsequent MRV Head was unremarkable.
- MRI C Spine/T Spine wwo: Minimal degenerative changes. No additional demyelinating lesions.
- MRI Brain wwo: Enhancing right pontine lesions with numerous non enhancing ovoid periventricular FLAIR hyperintensities perpendicular to the ventricles.

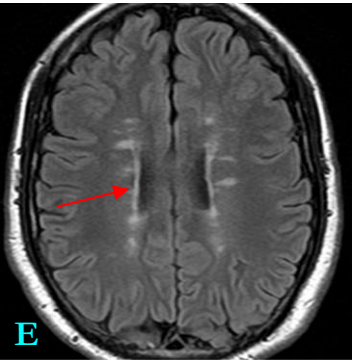
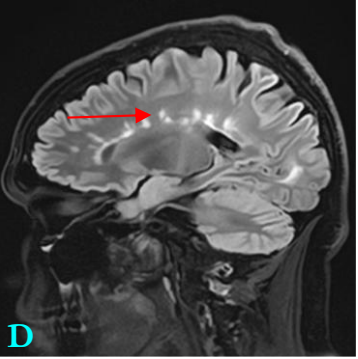
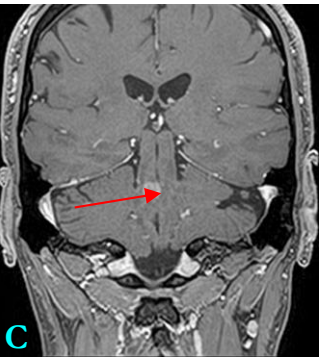
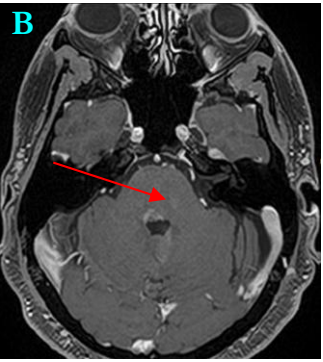
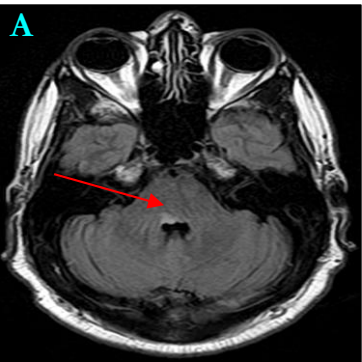


Figure 1: MRI brain with and without contrast (A) Axial FLAIR showing right dorsal pontine hyperintensity; (B, C) Axial and coronal, respectively, post-contrast T1 sequence showing an enhancing right dorsal pontine lesion concerning for demyelinating disease; (D, E) Sagittal and axial, respectively, FLAIR sequences showing ovoid periventricular hyperintensities perpendicular to the ventricles suggestive of demyelinating disease.

Clinical Course

Clinical Course:

- The patient's clinical neurological deficits and the presence of both an enhancing right pontine lesion and non enhancing periventricular lesions satisfied dissemination in space and dissemination in time under the 2024 McDonald diagnostic criteria supporting MS as the likely etiology.
- The patient was treated on a three day course of intravenous steroids with significant improvement and discharged.
- At six months follow-up with neuro-ophthalmology, symptoms resolved with no recurrence.
- A repeat MRI of the brain and orbits was stable demonstrating no active lesions; repeat CTA head and neck was also negative.
- He deferred disease modifying therapy and remained clinically stable with annual surveillance.

Conclusions

- A limited number of adults and pediatric cases have reported eight-and-a-half syndrome as the initial presentation of MS particularly in patients with acute brainstem symptoms⁵⁻⁸.
- Although rare, eight-and-a-half syndrome should be recognized as a possible presenting manifestation of MS and should prompt evaluation for a demyelinating etiology.
- While INO is the most common cause of diplopia for MS, a larger demyelinating pontine lesion that involves adjacent brainstem structures produces the constellation of findings in eight-and-a-half syndrome.
- The presence of an enhancing pontine lesion and non enhancing periventricular lesions, significant improvement with high-dose corticosteroids, and follow-up radiographic resolution of active enhancement supported a diagnosis of MS.
- Recognition of this syndrome may lead to earlier diagnosis of MS and timely initiation of disease-modifying therapy if necessary.

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References

