CASE REPORT

Rare occurrence of eight-and-a-half syndrome as a clinically isolated syndrome

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SUMMARY

Eight-and-a-half syndrome is a rare condition that is described as a combination of one-and-a-half syndrome and an ipsilateral facial nucleus lesion. We present a clinical case of occurrence of eight-and-a-half syndrome that was caused by a demyelinating lesion in the dorsal pontine tegmentum. A 44-year-old man presented to the hospital with a subacute onset of horizontal diplopia and left-sided facial weakness. MRI revealed a T2 hyperintense lesion in his dorsal pons, which was consistent with a demyelinating pathology. Treatment with intravenous steroids showed significant improvement in his symptoms. In our case, it occurred due to a suspected demyelinating lesion that was this patient's first and only demyelinating event, leaving him with a diagnosis of clinically isolated syndrome. His responsiveness to steroids represents the first case report of an adult patient presenting with an eight-and-a-half syndrome secondary to a suspected demyelinating pathology.

BACKGROUND

Horizontal conjugate gaze or the ability to maintain an object in focus across a horizontal plane, requires extremely precise coordination between multiple cranial nerves (CNs) including CN III, CN IV, CN VI as well as a complex interneuronal pathway called the medial longitudinal fasciculus (MLF) and the paramedian pontine reticular formation.¹

Figure 1 Patient having decreased wrinkling of forehead on the left side.

Figure 2 Patient is unable to completely close his left eye.

Internuclear ophthalmoplegia is a clinical syndrome attributed to a lesion of the MLF pathway residing within the dorsomedial pontine or midbrain tegmentum and prevents affected individuals from adducting the affected side during horizontal conjugate gaze. The MLF pathway runs near the paramedian pontine reticular formation and the abducens nucleus in the dorsomedial pons, which allows a single lesion to occasionally affect either of these pathways in addition to the MLF creating a specific clinical pattern termed 'one-and-a-half syndrome', first described by Fisher in 1967.² This syndrome causes complete gaze palsy on the ipsilateral side of the lesion making the 'one' within the 'one-and-a-half' syndrome and additionally prevents adduction of the ipsilateral eye making the 'half' portion of the syndrome.² In addition to these pathways being in close proximity to one another, cranial nerve VII nucleus resides in the lateral pontomedullary junction with the peripheral portion of the nerve wrapping around the nucleus of cranial nerve VI on its trajectory out of the lateral pons.¹ On rare occasions an individual lesion, often in the dorsal pontine tegmentum, can cause a 'one-and-a-half syndrome' as well as causing a cranial nerve 7 palsy, which has been termed an 'eight-and-a-half-syndrome' by Eggenberger.³

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Figure 3 Patient looking to the left, showing a complete left lateral gaze palsy.

Figure 4 Patient looking to the right showing impaired adduction of the left eye with preserved abduction in the right eye.

Figure 5 Axial T2 FLAIR showing a hyperintense lesion in the dorsal pontine tegmentum. FLAIR, fluid-attenuated inversion recovery.

In review of the current published cases, this syndrome has been mostly reported in adult patients secondary to ischaemic injury, with only a couple cases published related to demyelinating injury found in paediatric or young adult patients.

CASE PRESENTATION

A 44-year-old right-handed man with history of hypertension (not on any medications) presented with 2-week progression of horizontal diplopia that was worse on left lateral gaze. Six weeks prior to the occurrence of diplopia, the patient had experienced a sudden loss in taste sensation. He had also noticed a subtle left facial weakness as he said that he was spilling coffee from the left side of his mouth. On examination, clinically he was found to have a mild facial palsy, involving both his upper and lower face with decreased wrinkles in his left forehead and asymmetric smile (figures 1 and 2) and a complete conjugate gaze palsy to the left and impaired adduction in his left eye (figures 3 and 4).

INVESTIGATIONS–

MRI of the brain with and without contrast was obtained which showed a T2 hyperintense lesion in the medial periventricular

Figure 6 Coronal T1 with gadolinium showing an enhancing lesion in the dorsal pons.

Figure 7 Axial T1 with gadolinium showing an enhancing lesion in the dorsal pons.

region concerning for a demyelinating injury in the pons with subtle contrast enhancement (figures 5–7). This lesion was also hyperintense on diffuse-weighted imaging (figure 8) but lacked concurrent hypointensity on apparent diffusion coefficient (figure 9), overall suggesting a demyelinating pathology. MRI cervical spine and CT, angiography intracranial and extracranial were done and were all unremarkable. A lumbar puncture was performed, with cerebral spinal fluid (CSF) showing elevated protein at 51 with a completely normal cell count and differential. CSF studies included cell culture, cryptococcal antigen, rapid plasma regain, lyme serology, herpes simplex virus PCR, myelin basic protein, multiple sclerosis panel, flow cytometry, ACE and aquaporin-4 antibodies were all normal. Serum aquaporin-4 antibody was negative as well.

Figure 8 Diffusion-weighted imaging showing a hyperintense lesion in the dorsal pons.

Figure 9 ADC imaging that is lacking the concurrent hypodensity in the dorsal pons. ADC, apparent diffusion coefficient.

DIFFERENTIAL DIAGNOSIS

- ▶ Demyelinating lesion.
- ▶ Vascular lesion.
- ▶ Neoplasm.

TREATMENT

He was treated with 1 g of intravenous Solu-Medrol daily for 5 days and showed significant improvement, as he was able to adduct the left eye by the time of discharge.

OUTCOME AND FOLLOW-UP

He continued to improve at his follow-up appointment 2 weeks later, still showing dysconjugate gaze, specifically during left lateral gaze but had a nearly normal right lateral gaze.

He underwent repeat MRI brain and C spine 6 months after his initial episode. His repeat MRI brain done with and without

Figure 10 Axial T2 FLAIR (6 months from initial visit) showing hyperintense lesion in the dorsal pons. FLAIR, fluid-attenuated inversion recovery.

Figure 11 Axial T1 with gadolinium (6 months from the initial visit) shows resolution of the previously seen enhancement.

contrast continued to show the T2 hyperintense lesion in the dorsal pons with no other additional changes (figures 10–12).

DISCUSSION

Eight-and-a-half syndrome has been reported in the past, in association with multiple sclerosis, infarcts and tumours.^{4–6} In a review of the current published cases of eight-and-a-half syndrome, only four cases were described secondary to demyelinating lesions, and all of those cases were in a more typical demographic patient, young adult or paediatric patient.^{7–10} In our case report, the occurrence of this syndrome was attributed

Figure 12 Coronal T1 with gadolinium (6 months from the initial visit) shows resolution of the previously seen enhancement.

to the presence of a demyelinating lesion that can be labelled as a clinically isolated syndrome given the lack of any previous neurological episodes. Clinically isolated syndrome has been described as the occurrence of a first clinical demyelinating event in a person with no prior history of demyelination.¹¹ In most instances, the symptoms and signs indicate a lesion of the spinal cord, optic nerve or brainstem.¹²

This case highlights the importance of the bedside examination and ability to correlate the constellation of clinical findings to known brainstem neuroanatomy to help focus the work-up. Understanding this clinical pattern from a neuroanatomical standpoint confirmed by neuroimaging, helped to aid in diagnosis and prompt management of this reversible, steroid responsive and lesion.

Learning points

- Eight-and-a-half syndrome is a rare condition that is described as a combination of one-and-a-half syndrome and an ipsilateral facial nucleus lesion.
- Clinical syndrome showing a neuroanatomical correlate of a lesion at the dorsal pontine tegmentum.
- Rare unusual presentation of clinically isolated syndrome.

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