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Case Report

Eight-and-a-half syndrome: A unique manifestation of pontine infarction

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Abstract

This report details a case of eight-and-a-half syndrome characterized by conjugate horizontal gaze palsy, ipsilateral internuclear ophthalmoplegia, and ipsilateral lower motor neuron-type facial palsy. The patient, a 42-year-old male with a history of poorly controlled hypertension, presented with right-sided hemiparesis and restricted extraocular movements. Ocular examination revealed right eye exotropia with absent adduction, while the left eye showed absent abduction and adduction, left horizontal gaze palsy, gaze-evoked nystagmus, and left facial weakness. Magnetic resonance imaging of the brain indicated acute to subacute infarction of the left corona radiata and left centrum semiovale. Eight-and-a-half syndrome commonly arises due to a lesion affecting the paramedian pontine reticular formation, the medial longitudinal fasciculus, and the facial nerve fascicle on one side. Common etiologies include infarction or demyelination at the level of the pons. Recognizing the clinical features of this rare syndrome is essential for conducting appropriate diagnostic examinations, accurately localizing the lesion, and formulating an individualized treatment plan.

Introduction

Eight-and-a-half syndrome is a distinctive neurological disorder resulting from a localized infarction in the pons, characterized by a rare combination of ophthalmoplegic and facial nerve palsy manifestations. It represents a unique amalgamation of symptoms seen in one-and-a-half syndrome, which includes ipsilateral horizontal gaze palsy and internuclear ophthalmoplegia (INO), with the additional feature of ipsilateral facial muscle weakness. INO is characterized by inadequate adduction of the eye on the affected side and ataxic horizontal jerky nystagmus of the contralateral eye on abduction, while gaze toward the side of the lesion remains normal. INO arises from a lesion in the medial longitudinal fasciculus (MLF), a pair of crossed axonal tracts near the brainstem. This condition receives signals from the contralateral paramedian pontine reticular formation (PPRF) and sends signals to its ipsilateral oculomotor nerve, coordinating conjugate eye movements. Lesions leading to INO can include stroke, space-occupying lesions, and demyelination. Lesions affecting both the PPRF and MLF on one side lead to the formation of one-and-a-half syndrome, a condition first described by Fisher. This syndrome consists of ipsilateral conjugate horizontal gaze palsy, referred to as the "one," and ipsilateral INO, referred to as the "half," with the only remaining ocular movement being the abduction of the contralateral eye. When a lesion additionally involves the ipsilateral facial nerve fascicle around the area of the facial colliculus, it results in eight-and-a-half syndrome, which includes the ocular deficits of one-and-a-half syndrome plus ipsilateral facial muscle weakness.

Eight-and-a-half syndrome was first described by Eggenberger in 1998, with magnetic resonance angiography revealing it to be caused by vertebral basilar disease.[1] Subsequent studies by Kakar *et al*., Sampath Kumar *et al*., Nandhagopal and Krishnamoorthy, Bocos-Portillo *et al*.[2-5] demonstrated that pontine infarctions frequently lead to this syndrome, as confirmed by magnetic resonance imaging. Demyelinating conditions at the level of the pons, such as multiple sclerosis, have also been shown to cause eight-and-a-half syndrome, as demonstrated by Skaat and

Huna-Baron, Mortzos *et al.*[6,7] In rare cases, space-occupying lesions like tuberculomas can initiate the syndrome, as reported by Van Toorn *et al.*[8] Given its rarity and specific clinical and radiological features, eight-and-a-half syndrome holds a significant interest in neurology and ophthalmology. This case report explores the clinical presentation, diagnostic strategies, and therapeutic considerations in a patient with eight-and-a-half syndrome secondary to pontine infarction, contributing valuable insights to the evolving literature on this intriguing neurological condition.

Case Description

A 42-year-old male with a history of poorly controlled hypertension presented with sudden onset of right upper and lower limb weakness, slurred speech, partial closure of the left eyelid, and deviation of the angle of the mouth to the right side. On examination, his visual acuity was 6/24 in the right eye (OD), improving to 6/18 with pinhole correction, and 6/12 in the left eye (OS), improving to 6/9 with pinhole correction. Normal color vision in both eyes, with no relative afferent pupillary defect. Examination of extraocular movements showed horizontal gaze palsy with absence of adduction and abduction in OS [Figure 1]. OD exhibited an absence of adduction with gaze-evoked nystagmus. The patient demonstrated left peripheral facial nerve palsy, characterized by 2 mm of lagophthalmos, inability to wrinkle the forehead, a shallow nasolabial fold, and right-sided deviation of the angle of the mouth [Figure 2]. The remainder of the neurologic examination was unremarkable [Figure 3]. Slit lamp examination was essentially

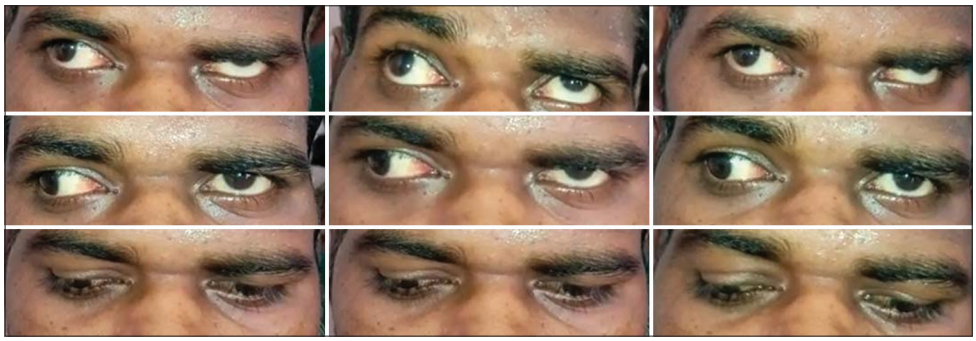

Figure 1: Image showing the nine cardinal positions of gaze

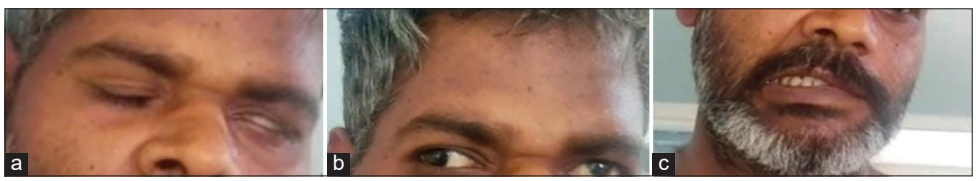

Figure 2: Image showing (a) 2 mm of lagophthalmos in the left eye, (b) Inability to wrinkle the forehead, and (c) Shallow nasolabial fold and deviation of angle of mouth to right side

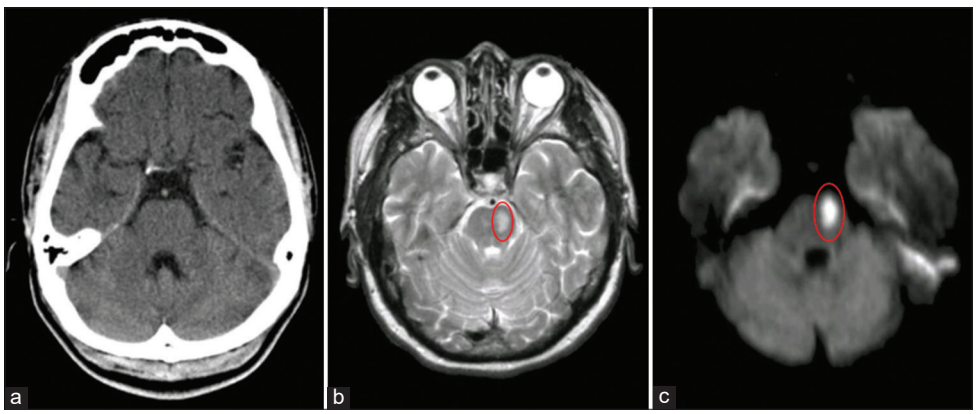

Figure 3: (a) Non-contrast computed tomographic of the patient's brain. (b) T2-weighted magnetic resonance imaging (MRI) demonstrating infarct (red circle) at the level of pons. (c) Diffusion-weighted MRI demonstrating infarct (red circle) at the level of pons

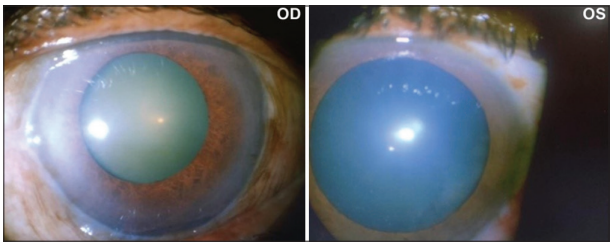

Figure 4: Slit lamp examination of the right eye and the left eye, respectively, revealing normal findings, with the exception of immature cataracts

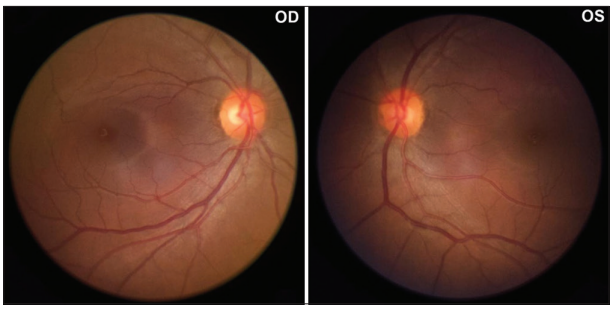

Figure 5: Normal fundus examination of the right eye and left eye

normal, except for immature cataracts [Figure 4]. Fundus examination was within normal limits in both eyes [Figure 5]. The patient was treated with a hyperosmotic agent as part of anti-edema management. Additional treatments included hypolipidemic drugs, antiplatelet therapy, antihypertensives, and physiotherapy.

Discussion

Eight-and-a-half syndrome, while rare, is a clinically significant entity that combines the features of one-and-a-half syndrome with ipsilateral facial nerve palsy. Our case illustrates the classic presentation of this syndrome, with the patient displaying a distinctive combination of horizontal gaze palsy, INO, and peripheral facial nerve palsy, secondary to a pontine infarction. The strength of this case report lies in the thorough clinical documentation and detailed examination findings that align with previously reported cases of eight-and-a-half syndrome. The clear correlation between the clinical signs and neuroanatomical pathways provides a robust framework for understanding the pathophysiology of this condition.

The primary concern in managing eight-and-a-half syndrome is addressing the underlying cause, which, in this case, was a pontine infarction likely due to uncontrolled hypertension. This case emphasizes the critical need for effective blood pressure management and secondary stroke prevention strategies. The integration of a multidisciplinary approach, including antihypertensives, antiplatelet therapy, and physiotherapy, was pivotal in the patient's management. Future studies should aim to include larger cohorts to better understand the prevalence and varied presentations of eight-and-a-half syndrome. Longitudinal studies tracking patient outcomes over time could provide valuable insights into the long-term prognosis and efficacy of different therapeutic interventions.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflict of interest

There are no conflicts of interest.

Conclusion

Eight-and-a-half syndrome is a rare clinical presentation characterized by conjugate horizontal gaze palsy, ipsilateral INO, and ipsilateral peripheral facial nerve palsy. This case is notable for its rarity, wide clinical spectrum, and the diagnostic challenges it presents. Early recognition of this syndrome is crucial for precise localization of the lesion, enabling timely intervention and improving prognosis.

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