Case Report

A Rare Case Report of Eight Syndrome Secondary to Syringomyelia Associated with Type I Chiari Malformation

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Abstract

Eight syndrome is defined as the combination of a unilateral conjugate gaze palsy and ipsilateral seventh cranial nerve palsy. It may occur as a result of demyelinating, vascular, infectious, or compressive lesions of the brainstem localized to the caudal pontine tegmentum. A 43-year-old woman was admitted to our clinic with complaints of headache, inability to look to the left, and weakness on the left side of her face. The complaints had begun abruptly about a month before her admission. Suboccipital decompression surgery for type I Chiari malformation had been performed 10 years earlier. Neuro-ophthalmological examination revealed left-sided horizontal gaze palsy and anisocoria. Cranial and cervical magnetic resonance images revealed cerebellar tonsillar herniation and syringomyelia, the latter of which was considered to be the cause of eight syndrome. No interventions were performed, and periodic follow-up was advised on neurosurgical consultation. Left gaze palsy and facial palsy recovered almost completely in three months, while the anisocoria persisted. Syringomyelia should be considered among the causes of horizontal gaze palsy plus ipsilateral seventh nerve palsy, termed as eight syndrome. Clinical suspicion and appropriate radiological examination can aid in the diagnosis.

Keywords: Chiari malformation, eight syndrome, horizontal gaze palsy, seventh cranial nerve palsy

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Introduction

Eight syndrome is described as the combination of a unilateral conjugated gaze palsy and ipsilateral seventh cranial nerve palsy. The syndrome and/or other variants may occur in demyelinating, vascular, infectious, or compressive lesions of the brainstem localized to the caudal pontine tegmentum. The brainstem structures primarily affected are the ipsilateral seventh cranial nerve and paramedian pontine reticular formation/sixth cranial nerve nucleus.[1]

Case Report

A 43-year-old woman presented to our clinic with complaints of headache, inability to look to the left, and weakness on the left side of her face. The complaints had begun abruptly about a month before her admission. Suboccipital decompression surgery for type I Chiari malformation (CM) had been performed 10 years earlier. No other known pre-existing systemic diseases or drug usage was present and her family history was unremarkable. On admission, complete physical examination including vital signs were normal. Neurological examination was unremarkable except for left seventh cranial nerve palsy (Figure 1A). Neuro-ophthalmological examination revealed left-sided horizontal gaze palsy (Figure 1B) and anisocoria. Other extraocular eye movements were within normal limits. Anisocoria was prominent in dim light (pupil diameter: 4 mm right, 3 mm left). However, ptosis was not noted. Pupillary light and near reflexes were normal. Pupillary dilation was not observed on the left side in dim light after instilling topical 0.5% apraclonidine (Iopidine, Alcon, Fort Worth, TX, USA). Cranial magnetic resonance imaging (MRI) performed to assess for a caudal pontine tegmental lesion revealed cerebellar tonsillar herniation and

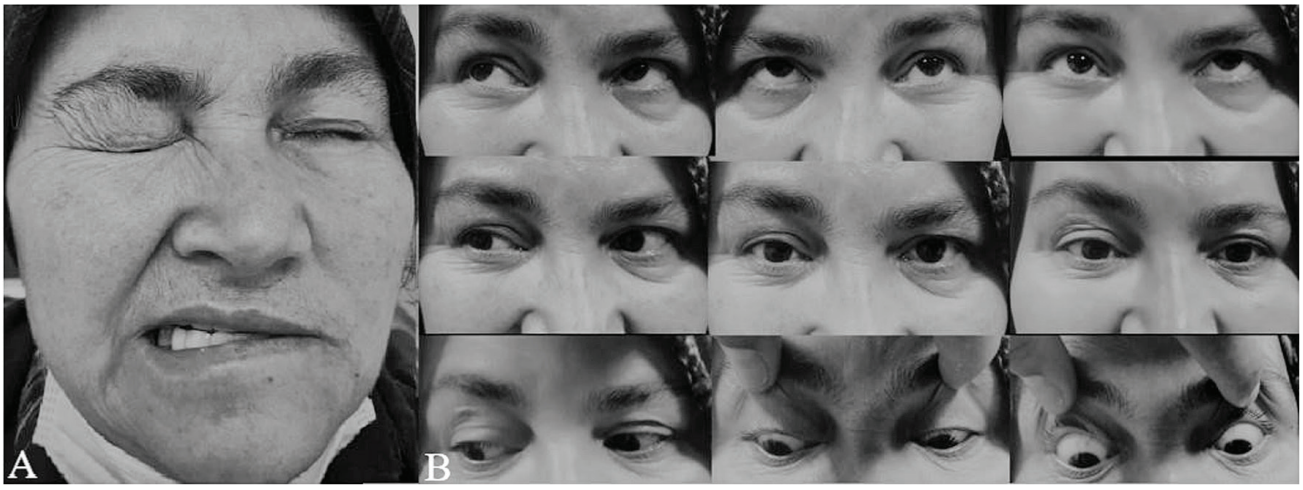

Figure 1. Image showing left peripheral seventh nerve palsy (A) and images of the patient in nine diagnostic gaze positions demonstrating left-sided horizontal gaze palsy (B)

syringomyelia descending from the caudal tegmental region of the pons to the second cervical vertebral level, which was better delineated on cervical MRI (Figure 2). This was considered to be the cause of eight syndrome. Intervention was not considered and periodic follow-up was advised on neurosurgical consultation. Left gaze palsy and facial palsy recovered almost completely in three months, while the anisocoria persisted.

Discussion

Type 1 CM is defined as herniation of the cerebellar tonsils into the upper cervical canal at the level of the foramen magnum. Syringomyelia, a rare neurological condition, often accompanies this craniocervical junction abnormality and is characterized by the presence of a fluid-filled cavity in the central canal of the spinal cord or within its parenchyma.[2,3,4] The prevalence of syringomyelia ranges from 8.4/100,000 to 0.9/10,000 and is commonly observed in patients aged 20 to 50 years.[3]

Apart from type I CM, it can develop as a post-inflammatory or post-traumatic condition, and spinal cord tumors and secondary myelomalacia are among the other known causes.[3,4] Although various theories have been put forward to explain the pathophysiological process, the most valid explanation is impairment of cerebrospinal fluid (CSF) circulation caused by obstruction of the subarachnoid space.[4]

Patients with syringomyelia may present with a wide variety of non-specific symptoms and/or findings depending on the size, location, and extent of the cyst within the spinal cord and/or brainstem. However, some cases are completely asymptomatic and incidentally discovered on radiologic evaluation.[1,2,3,4,5]

Diagnosis is made with clinical suspicion based on symptoms and/or signs. MRI is currently the most widely preferred imaging modality for diagnosis and follow-up. The fluid-filled cavities appear hyperintense on T2-weighted images but remain hypointense on T1-weighted images. In addition to its use in diagnosis and follow-up, MRI also reveals secondary causes such as tumors and type 1 CM that may be associated with syringomyelia.[5]

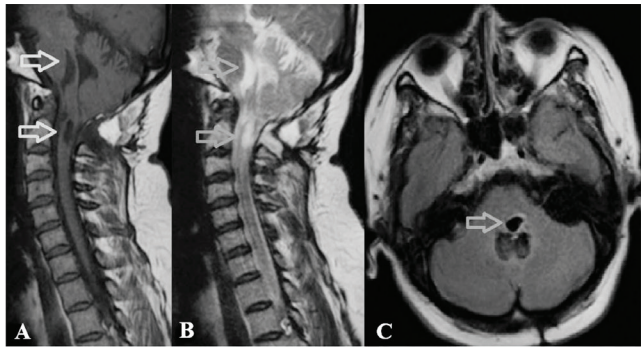

Figure 2. Magnetic resonance imaging (MRI) of the brainstem and spinal cord. T1-weighted (A) and T2-weighted (B) sagittal MRI of the brainstem and spinal cord revealing tonsillar herniation and hypointense (arrows) and hyperintense (arrows) cystic cavity corresponding to syringomyelia, respectively. T1-weighted axial MRI of the brainstem (C) showing the hypointense (arrow) cystic cavity corresponding to syringomyelia in the caudal tegmental region of the pons

Surgical treatment of type 1 CM-related syringomyelia aims to restore normal CSF circulation at the level of foremen magnum, reduce the syrinx, and eliminate the compression exerted by the cerebellar tonsil on the brainstem.[2,3,4,5]

To the best of our knowledge, here we describe the first patient with eight syndrome due to syringomyelia involving the brainstem in the literature. Interestingly, although the patient did not undergo any medical or surgical intervention and the cyst did not change in size, her clinical findings improved during follow-up. We cannot fully explain the abrupt onset and spontaneous clinical improvement in this case. The most plausible explanation may be the fluctuations in cyst volume with alterations in the CSF circulation caused by tonsil herniation.

Syringomyelia should be kept in mind among the causes of horizontal gaze palsy plus ipsilateral seventh nerve palsy, termed as eight syndrome. Clinical suspicion and appropriate radiological examination can aid in the diagnosis.

Ethics

Informed Consent: Informed consent was obtained from the patient for the publication of this report.

Peer-review: Externally peer reviewed.

Authorship Contributions

Surgical and Medical Practices: D.T.K., Concept: D.T.K., Ö.K., N.Ç., Design: D.T.K., Ö.K., N.Ç., Data Collection or Processing: D.T.K., Analysis or Interpretation: D.T.K., P.K., Literature Search: P.K., Writing: D.T.K., P.K.

Conflict of Interest: No conflict of interest was declared by the authors.

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