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An eight-and-a-half syndrome case: a rare presentation of pontine lacunar infarction

Bir sekiz buçuk sendromu olgusu: pontin laküner infarktın nadir bir prezentasyonu

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Abstract

The eight-and-a-half syndrome is a one-and-a-half syndrome [conjugated horizontal gaze palsy and internuclear ophthalmoplegia (INO)], plus an ipsilateral cranial nerve seventh (CN VII) palsy. This rare syndrome is caused by the pontine tegmentum's lesions involving the abducens nucleus, the ipsilateral medial longitudinal fasciculus (MLF), and the facial colliculus. In this case, we report a 68-year-old male patient admitted to our hospital with binocular horizontal diplopia later diagnosed with the eight-and-a-half syndrome. Although the eight-and-a-half syndrome is rare, it should be kept in mind with patients presenting diplopia and peripheral facial palsy simultaneously.

Keywords: Diplopia, internuclear ophthalmoplegia, facial palsy, abducens nucleus, nystagmus

Özet

Sekiz buçuk sendromu, bir buçuk sendromu [konjuge yatay bakış felci ve internükleer oftalmopleji (INO)], artı ipsilateral kraniyal sinir yedinci (CN VII) felcidir. Bu nadir sendrom, abdusens nükleusu içeren, ipsilateral medial longitudinal fasciculus (MLF) ve fasiyal kollikulusu tutan pontin tegmentum lezyonları sebebiyle meydana gelir. Bu olguda binoküler horizontal diplopi ile hastanemize başvuran ve sonrasında sekiz buçuk sendromu tanısı alan 68 yaşında bir erkek hastayı sunmaktayız. Sekiz buçuk sendromu nadir olmakla birlikte, aynı anda hem diplopi hem de periferik yüz felci olan hastalar da akılda tutulmalıdır.

Anahtar Kelimeler: Diplopi, internükleer oftalmopleji, fasiyal palsi, abdusens nükleusu, nistagmus

1. Introduction

The eight-and-a-half syndrome is a one-and-a-half syndrome in addition to the ipsilateral peripheral CN VII palsy. This is an uncommon syndrome caused by lesions of the pontine tegmentum involving the abducens nucleus, the ipsilateral medial longitudinal fasciculus (MLF), and the adjacent facial colliculus. On examination, ipsilateral one-and-a-half syndrome and lower motor neuron-like facial palsy, contralateral dissociated nystagmus were detected. Vertical ocular movements from the primary position were normal. The only remaining ocular movement is the abduction of the left eye [1]. The eight-and-a-half syndrome is rare but, we recommend that all medical doctors keep this uncommon syndrome in mind for patients presenting diplopia.

2. Case report

A 68-year-old male patient was admitted to the emergency department with a sudden onset of binocular horizontal diplopia. Then he was consulted with our neurology department. He had a history of type 2 diabetes mellitus and primary hypertension. He had no history of using anticoagulant or antiaggregant medications or any neck injuries. On examination, ipsilateral one-and-a-half syndrome and lower motor neuron-like facial palsy, contralateral dissociated nystagmus were

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detected. Vertical ocular movements from the primary position were normal. The only remaining ocular movement was the abduction of the left eye. Therefore, the neurological examination suggested a right-sided (ipsilateral) eight-and-a-half syndrome. The assessment of the patient's brain CT scan indicated no significant lesions. The brain diffusion-weighted MRI (Fig.1) and apparent diffusion coefficient (ADC) map showed a millimetric diffusion restriction in a focal area in the posterior half of the pons (Fig. 2). No abnormality was found in the routine blood examination, the hepatic and renal function, electrolyte, or the homocysteine level. Oral clopidogrel of 75 milligrams per day (mg/d) and acetylsalicylic acid (ASA) of 100 mg/d were initialized, and he was hospitalized in our neurology department. His glycosylated haemoglobin (HbA1c) level was 7.9%, his low-density lipoprotein (LDL) level was 170 milligrams per deciliter (mg/dL), which were high. In contrast, his B12 vitamin level was lower than 200 picograms per milliliter (pg/mL). Thus, we initiated a B12 vitamin replacement and an atorvastatin 10 mg/d treatment. The electrocardiogram showed sinus rhythm. The cardiology department performed the cardiac examination and the transthoracic echocardiogram (TTE) and holter ECG. No significant abnormality was found. Therefore, oral use of clopidogrel 75 mg/d and ASA 100 mg/d was continued. The patient did not show any additional neurological deficits; his diplopia complaint also gradually decreased. Until MR angiography and doppler were taken, the patient was given dual antiaggregant for possible excess carotid plaques. The assessment of intracranial and carotid-vertebral arteries via MRA indicated no vascular stenosis. Because these imagings were normal and the risk of gastrointestinal bleeding was lower, treatment with clopidogrel 75 mg/d was continued. An informed consent form was signed by the patient.

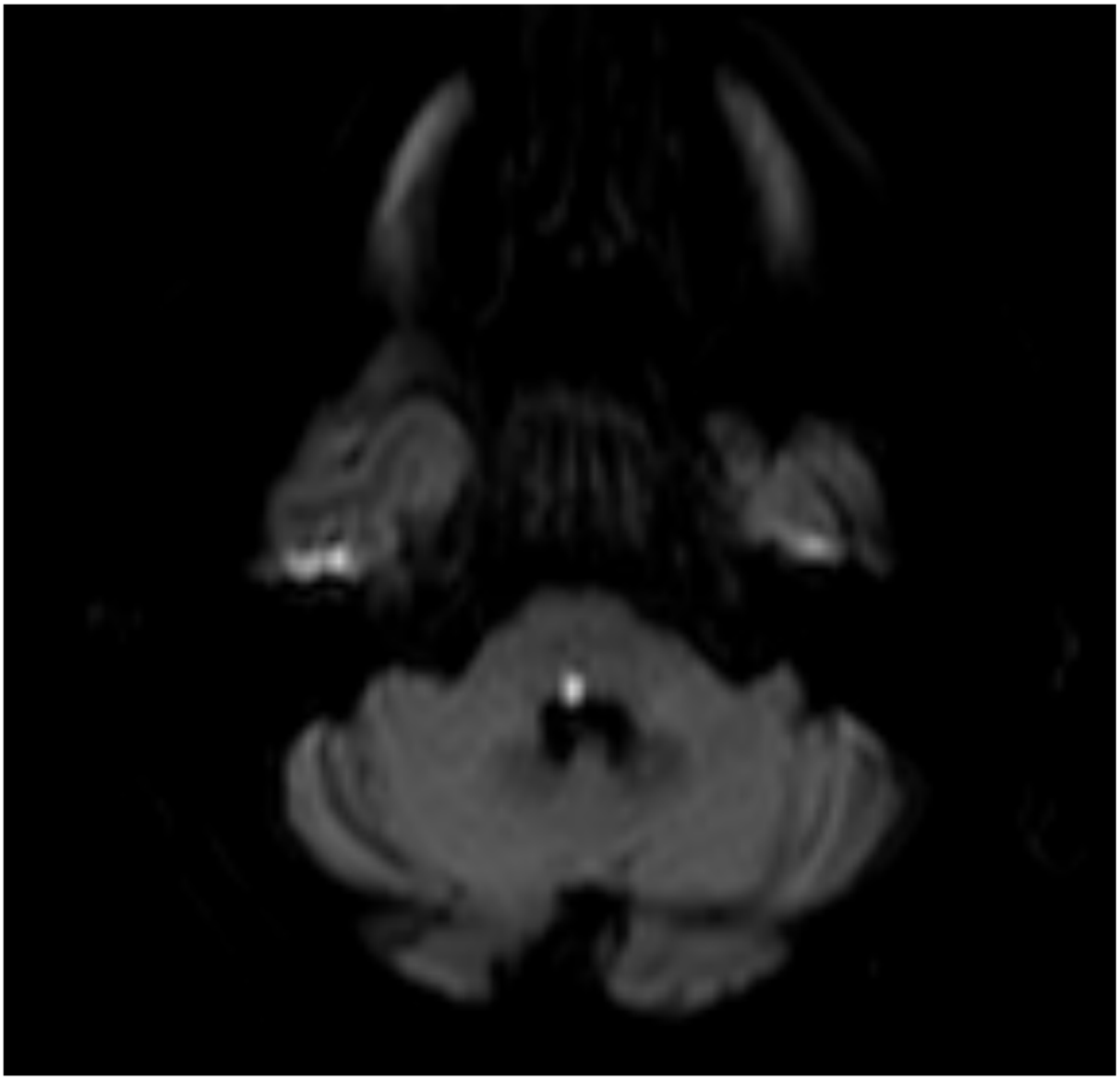

Figure 1. Diffusion-weighted imaging

3. Discussion

The eight-and-a-half syndrome is a one-and-a-half syndrome with ipsilateral lower motor neuron-like type CN VII palsy. This syndrome usually occurs due to lesions of the paramedian pontine reticular formation (PPRF), the ipsilateral MLF, and the adjacent facial nerve fascicle [1]. To start the horizontal eye movement, a signal is produced and then released from the frontal eye field (FEF) contralateral to the gaze direction. This initial signal goes to the contralateral abducens nucleus in pons through the PPRF. The abducens nucleus via nervus (n.) abducens sends signals to the ipsilateral lateral rectus muscle through interneurons, which cross to the other side and later create the MLF. The interneurons of the MLF are connected to the contralateral mesencephalic medial rectus subnucleus of n. oculomotorius nucleus, which innervates the contralateral medial rectus muscle [2]-[4]. A mesencephalic or pontine lesion of the MLF might cause the ipsilateral medial rectus subnucleus of the n. oculomotorius nucleus to receive no more signals from the contralateral n. abducens nucleus, therefore causes an ipsilateral adductional gaze limitation [2]-[3], [4]. Hering's law might explain the disassociated jerky abductional nystagmus: the weak medial rectus muscle triggers the cortex to escalate innervation to the non-affected abducting eye. It results in exaggerated ataxic horizontal jerky abductional nystagmus [2], [4].

4. Conclusion

In conclusion, we suggest that this rare eight-and-a-half syndrome should be kept in mind, especially with patients with diplopia and peripheral facial palsy.

5. Author contribution statement

Idea/Concept: Ufuk Çınkır; Design: Ufuk Çınkır; Control/Supervision: Ufuk Çınkır, Saltanat Mert, Ayhan Köksal; Data Collection and/or Processing: Ufuk Çınkır, Saltanat Mert Analysis and/or Interpretation: Ufuk Çınkır, Saltanat Mert; Literature Review: Ufuk Çınkır, Saltanat Mert, Ayhan Köksal; Writing the Article: Ufuk Çınkır, Saltanat Mert, Ayhan Köksal; Critical Review: Ufuk Çınkır, Saltanat Mert, Ayhan Köksal.

6. Ethics committee approval and conflict of interest statement

An informed consent form was signed by the patient. Authors declare no conflict of interest.

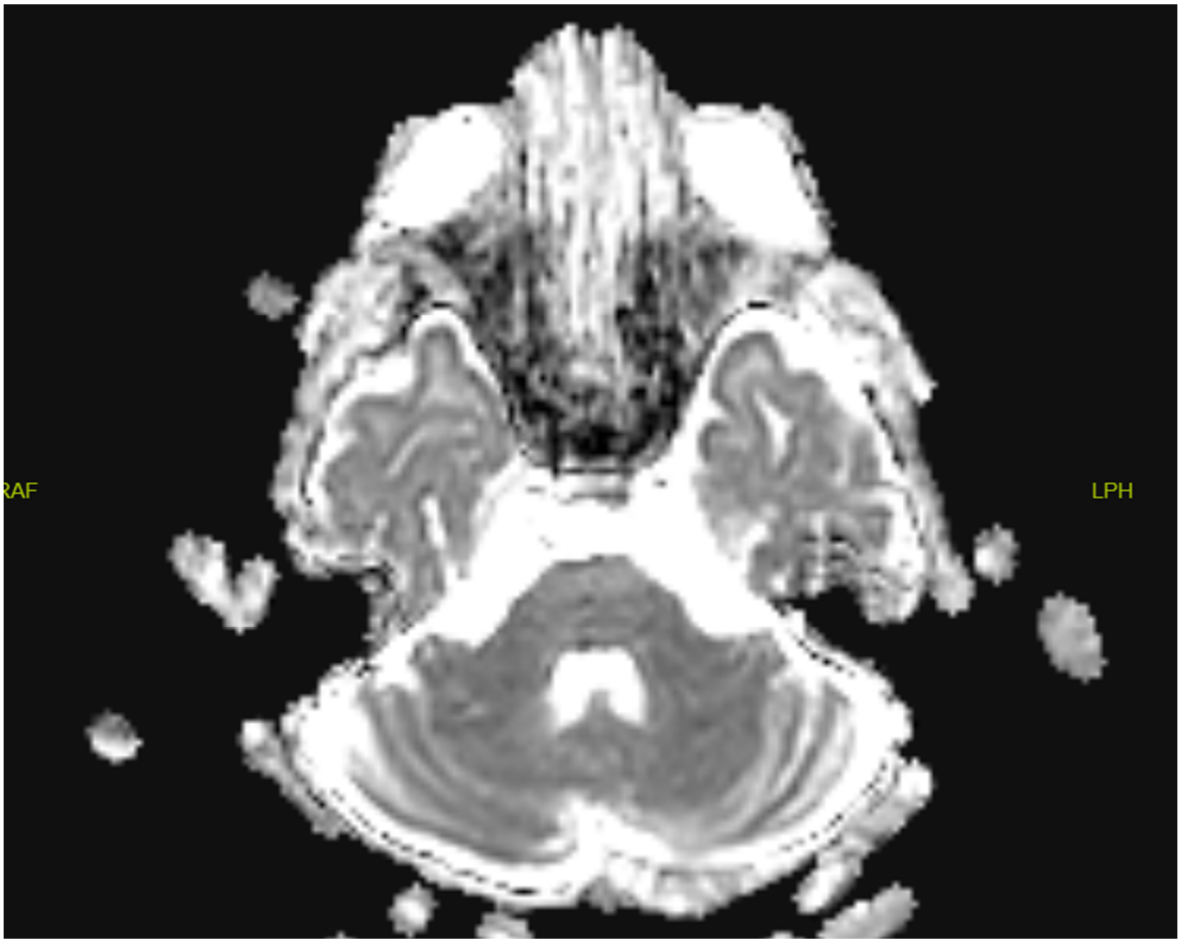

Figure 2. Apparent diffusion coefficient

7. References

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