

Two Rare Presentations of Pontine Lesion: One-and-a-half Syndrome and Eight-and-a-half Syndrome

Pons Lezyonunun İki Nadir Prezantasyonu: Bir Buçuk ve Sekiz Buçuk Sendromu

İD Turgay Dölek, İD Muammer Korkmaz, İD Ayşe Özlem Akgün,
İD Semai Bek, İD Gülnihal Kutlu Günergin

Muğla University, Medical Faculty, Training and Research Hospital, Department of Neurology, Muğla/Türkiye

ABSTRACT

One-and-a-half syndrome refers to a unilateral lesion involving the ipsilateral medial longitudinal fascicle and the 6th cranial nerve nucleus and/or the paramedian pontine reticular formation. Eight-and-a-half syndrome is a syndrome in which one-and-a-half syndrome and ipsilateral 7th cranial nerve palsy are seen together. These rare conditions result from limited lesions of the pontine tegmentum. We presented two cases with one-and-a-half and eight-and-a-half syndromes with ischemic infarction and hemorrhage in pons, respectively.

ÖZET

Bir buçuk sendromu, ipsilateral medial longitudinal fasikül ve 6. kranial sinir çekirdeği ve/veya paramedian pontin retiküler oluşumunu içeren tek taraflı bir lezyonu ifade eder. Sekiz buçuk sendromu ise bir buçuk sendromu ile ipsilateral 7. kranial sinir felcinin birlikte görüldüğü bir klinik durumdur. Bu nadir tablolar ponsun tegmentumuna yerleşik lezyonlardan kaynaklanır. Burada sırasıyla iskemik infarktüs ve kanaması olan bir buçuk ve sekiz buçuk sendromlu iki olguyu sunduk.

Keywords:

Brainstem
Infarct
Pons
Hemorrhage

Anahtar Kelimeler:

Beyinsapı
İnfarakt
Pons
Hemoraji

INTRODUCTION

The brainstem is a complex structure that includes the cranial nerves, their nuclei, descending and ascending pathways and provides the connection between the brain and the spinal cord. Brainstem lesions can cause critical clinical processes because of the presence of important structures in a restricted area. At the same time, lesions of these structures may cause clinical presentation which is rare and has localization value.

CASE 1

A 65-year-old male patient was admitted with complaints of weakness in his left arm and leg and diplopia. He had history of smoking, hypertension (HT), hyperlipidemia (HL) and left thalamic ischemic infarction one and a half months ago (healed without sequelae). He was taking acetylsalicylic acid, atorvastatin and amlodipine. Neurological examination revealed complete rightward gaze paralysis in the horizontal plane, limited adduction in the right eye and 3/5 left-sided hemiparesis (Figure 1). Abduction of the left eye was normal. Patient had one-and-a-half syndrome and diffusion-weighted imaging (DWI) revealed a hyperintensity lesion extending to the tegmentum in the right of the pons and a mildly hypointensity corresponding to the same area on the apparent diffusion coefficient (ADC) (Figure 2), [Siemens Magnetom Skyra 3T magnetic resonance imaging (MRI)]. Clopidogrel was added to treatment. No cardiac embolism

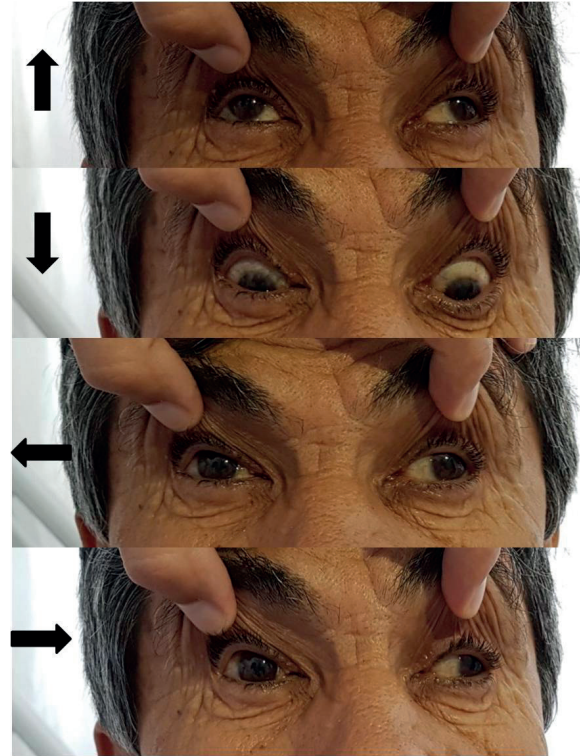


Figure 1: There are complete rightward gaze paralysis in the horizontal plane and limited adduction in the right eye.

Correspondence: Turgay DÖLEK, Muğla Üniversitesi Eğitim ve Araştırma Hastanesi Nöroloji A.D. Muğla Türkiye. E-mail: turgaydolek@gmail.com

Cite as: Dölek T, Korkmaz M, Akgün AÖ, Bek S, Günergin GK. Two Rare Presentations of Pontine Lesion: One-and-a-half Syndrome And Eight-and-a-half Syndrome. Phnx Med J. 2023;5(1):49-52.

Received: 01.06.2022

Accepted: 09.12.2022



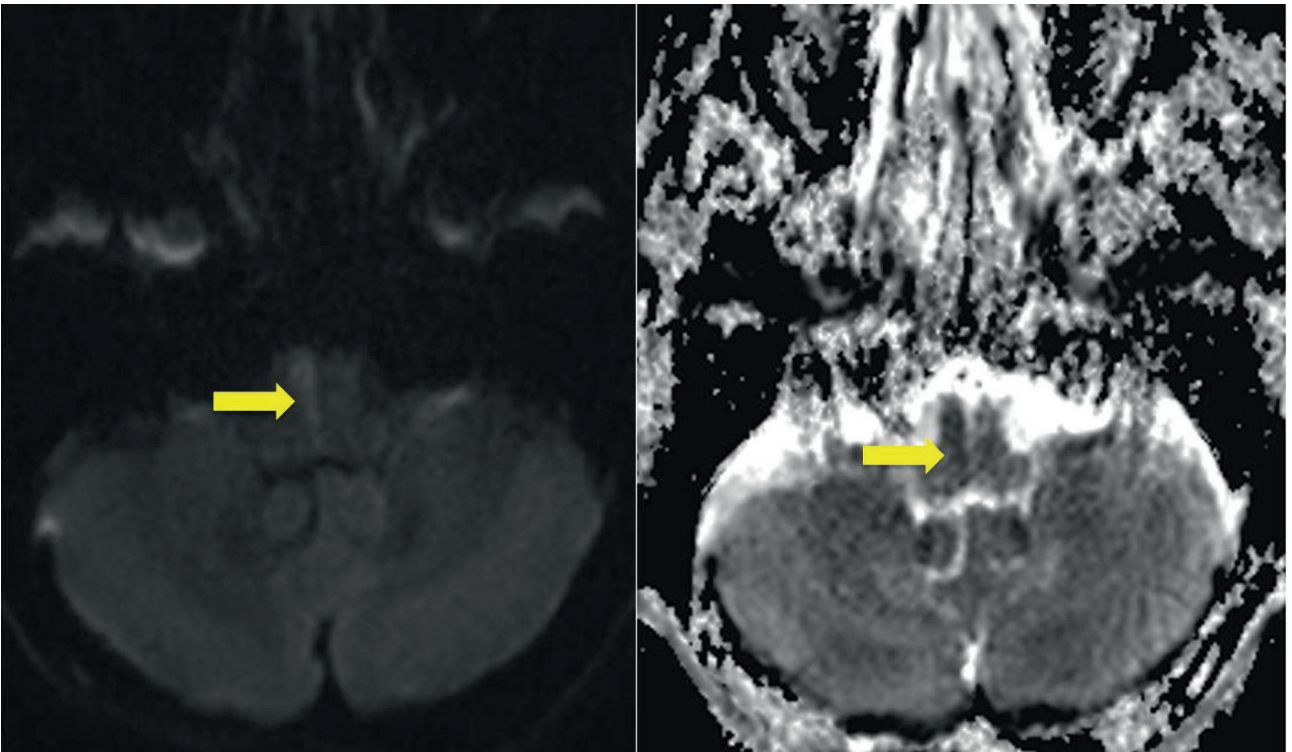


Figure 2: Paramedian pontine infarct. The lesion extends from the right paramedian pontine base to the tegmentum, there is hypointensity in the same area on the ADC map (arrows).



Figure 3: There are complete rightward gaze paralysis in the horizontal plane, limited adduction in the right eye and facial paralysis on the right side of face.

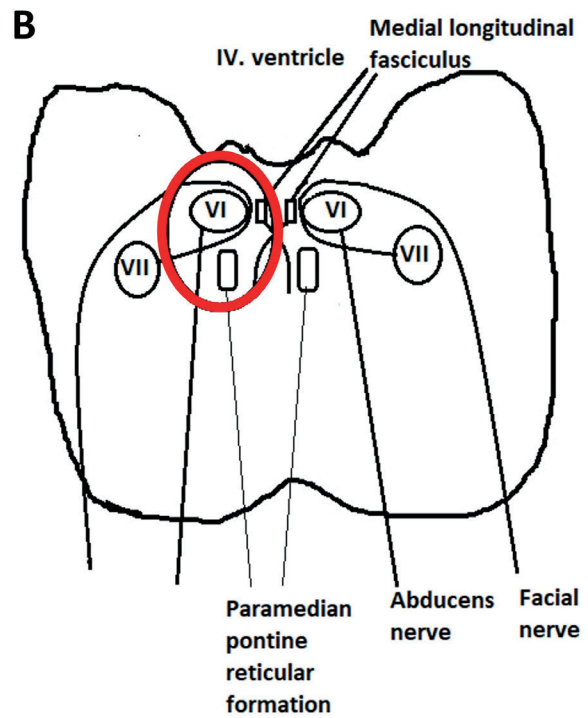
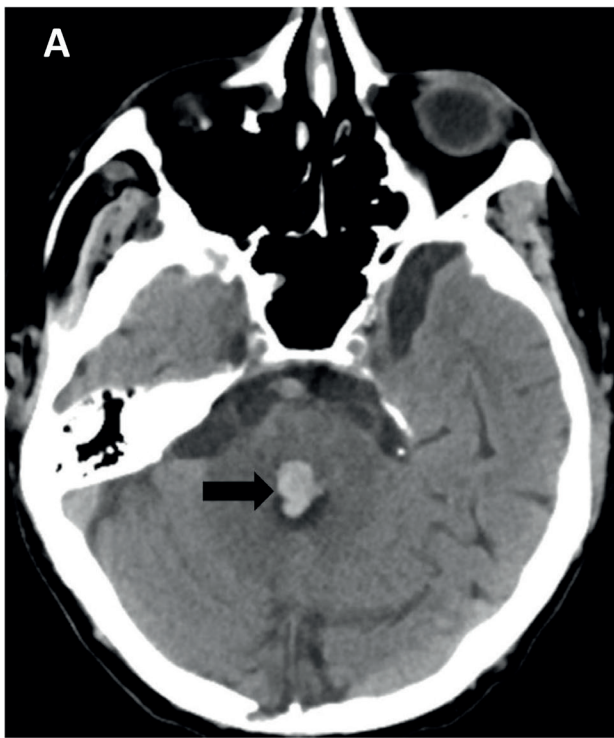


Figure 4: There is hematoma in the tegmentum of pons on CT (arrow, A) and the cross section of pons at the level of the abducens and facial nucleus and the responsible lesion for eight-and-a-half syndrome (B).

was found in the evaluation for the etiology of ischemic stroke. Rhythm holter which was applied two times for each 24 hours was observed as normal. No pathological stenosis was detected on brain/neck computed tomography angiography. On the fourth day of follow-up, his diplopia and bilateral eye movements completely recovered. Left-sided hemiparesis persisted. Dual anti-aggregant therapy was recommended to the patient.

CASE 2

A 78-year-old male patient was admitted with complaints of imbalance and diplopia. He was on medical treatment for HT, type 2 diabetes mellitus (DM) and ischemic cerebrovascular diseases two years ago before the date of admission. He had healed without sequelae. He was taking acetylsalicylic acid for ischemic stroke prophylaxis as well as medications for HT and DM. In neurological examination, there was limited adduction in the right eye, complete rightward gaze paralysis and facial paralysis on right side of the face and significantly dysarthria was detected (Figure 3). No limb weakness was detected. The patient who was considered eight-and-a-half syndrome had a hematoma in the right posterior of the pons which extends to the 4th ventricle on cranial computed tomography (CT) (Figure 4) and anti-aggregant medication was stopped. No abnormality was detected in cranial MR angiography. He was closely monitored due to his resistant HT. His blood pressure was controlled with quadruple anti-hypertensive therapy (carvedilol, benidipine, doxazosin, spironolactone). No stenosis was detected in examination of the renal arteries by using the doppler ultrasound for the etiology of resistant HT. No improvement in eye movements was observed in the patient's 3rd month follow-up.

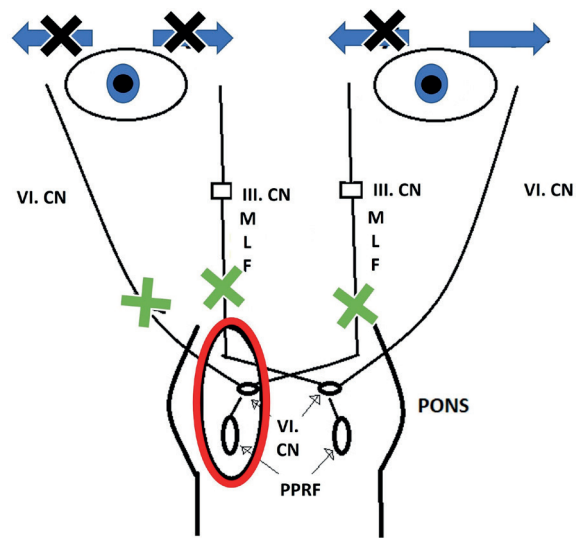


Figure 5: A schematic explanation of one-and-a-half syndrome. A lesion is shown affecting the right PPRF (paramedian pontine reticular formation) and right abducens nucleus causing an ipsilateral complete conjugate horizontal gaze palsy and affecting the right MLF (medial longitudinal fasciculus) causing an ipsilateral internuclear ophthalmoplegia (red ring). Contralateral abduction is normal. III. CN: Oculomotor nerve, VI. CN: Abducens nerve.

DISCUSSION

One-and-a-half syndrome defined by Fisher refers to involvement of the pontine paramedian reticular formation and the medial longitudinal fasciculus as schematized in Figure 5 (1). The facial nerve fibers travel around the nucleus of the abducens and a lesion in this area causes lower motor neuron palsy in the facial nerve along with the signs

of one-and-a-half syndrome (Figure 4). This syndrome was named eight-and-a-half syndrome by Eggenberger in 1998 based on three cases of ischemic stroke (2). In both of these situations, there is pathology in the tegmentum of the pons. Pontine tegmentum is most frequently supplied by the anterior inferior cerebellar artery or paramedian pontine perforators of the basilar artery. The most common cause of these syndromes are ischemic stroke but different etiologies have been reported such as multiple sclerosis, vasculitis, brainstem hemorrhage, tumor metastases and

brainstem tuberculoma (3,4). Many variants have been described such as nine syndrome, thirteen-and-a-half syndrome, fifteen-and-a-half syndrome. These diagnostic definitions are important because the lesion may be too small to be seen on conventional MRI. We presented two patients with one-and-a-half syndrome and eight-and-a-half syndrome that were found ischemic infarction and hemorrhage in the pons, respectively, because they are rare and provide diagnostic value when detected.

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

Ethics: The patient informed consent form was obtained.

Funding: There is no financial support of any person or institution in this research.

Approval of final manuscript: All authors.

REFERENCES

1. Fisher CM. Some neuro-ophthalmological observations. *J Neurol Neurosurg Psychiatry*. 1967;30:383–392. doi:10.1136/jnnp.30.5.383.
2. E Eggenberger. Eight-and-a-half syndrome: one-and-a-half syndrome plus cranial nerve VII palsy. *J Neuroophthalmol*. 1998;18(2):114-6.
3. Wei Li, Deren Hou, Yanyao Deng, Yi Tian. Eight-and-a-Half Syndrome: A Combination of Intracranial Capillary Telangiectasia and Hematencephalon. *J Stroke Cerebrovasc Dis*. 2018;27(7):e135-e137. doi: 10.1016/j.jstrokecerebrovasdis.2018.02.020
4. Xia NG, Chen YY, Li J, Chen X, Ye ZS, Chen SY, et al. Eight-and-a-half syndrome caused by a pontine haemorrhage: a case report and review of the literature. *Int J Neurosci*. 2018;128(8):746-750. doi: 10.1080/00207454.2017.