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A Rare Pontine Neuro-ophthalmic Syndrome: Eight-and-a-Half Syndrome

Nadir Bir Pontin Nöro-oftalmolojik Sendrom: Sekiz Buçuk Sendromu

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Abstract

One-and-a half syndrome is seen in paramedian pontine lesions, and may also co-exist with cranial nerve paralysis. This clinical situation is called eight-and-a-half syndrome when facial nerve paralysis also accompanies this manifestation. A man aged 38 years was admitted with symptoms of sudden-onset binocular diplopia and dizziness. The patient had no known co-morbidities. Cranial magnetic resonance imaging showed a small-sized infarct in the left paramedian pontine tegmentum posterior area. In his neuro-ophthalmologic examination, total paresis of left eye horizontal movements, mild lateral deviation, and monocular nystagmus during abduction of the right eye were observed. The patient also had peripheral facial paralysis on the left side; therefore, no signs of motor deficit of his extremities were examined. Eight-and-a-half syndrome is a rare disorder that is seen in localized-small pons lesions, mostly accompanied by infarcts. This clinical manifestation is called eight-and-a-half syndrome and arises within lesions in both the parapontine reticular formation and the medial longitudinal fasciculus in the inferior pons tegmentum where horizontal eye movements are controlled, and facial axons adjacent to the nucleus of the sixth nerve. We wanted to present this case to emphasize this rare situation.

Keywords: One-and-a-half syndrome, facial palsy, pontin lesion

Öz

Paramedyan pontin lezyonlarında görülen bir buçuk sendromu, kraniyal sinir paralizileri ile birlikte de görülebilir. Fasiyal sinir paralizisi ile birlikte görülen klinik tablo sekiz buçuk sendromu olarak adlandırılır. Otuz sekiz yaşında erkek hasta, ani başlayan çift görme, baş dönmesi şikayetleriyle başvurdu. Bilinen komorbiditesi olmayan hastanın çekilen kraniyal manyetik rezonans görüntülerinde difüzyon ağırlıklı kesitlerinde sol paramedyan pontin tegmentum posterior kesimde küçük bir enfarkt alanı görüldü. Nöro-oftalmolojik muayenesinde sol göz horizontal göz hareketlerinde tam parezi, sağ göz primer pozisyonda hafif laterale deviasyon ile addüksiyon kısıtlılığı ve dışa bakış sırasında ortaya çıkan nistagmus görüldü. Ekstremitelerinde motor defisite rastlanmayan hastada solda periferik tipte fasiyal paralizi saptandı. Bir buçuk sendromu iyi lokalize küçük pons lezyonlarında, en sık olarak enfarktlar ile birlikte tanımlanmış nadir bir tablodur. Horizontal göz hareketlerinin düzenlendiği inferior pons tegmentumundaki paramedyan pontin retiküler formasyon, mediyal longitidunal fasikül ve altıncı kraniyal sinir nükleusu ile hemen komşuluğundaki fasiyal sinir aksonlarını tutan lezyonlar ile ortaya çıkan klinik tablo sekiz buçuk sendromu olarak adlandırılır. Literatürde nadir olarak bildirilmiş bu durumu vurgulamak için olguyu sunmak istedik.

Anahtar Kelimeler: Bir buçuk sendromu, fasiyal paralizi, pons lezyonu

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Introduction

The clinical picture that consists of one-and-a-half syndrome and peripheral facial paralysis is called eight-and-a-half syndrome. This definition was first made by Eggenberger (1) in 1998 (2). Very few cases have been reported to date. The lesion is localized to the paramedian tegmentum of lower pons. The most common cause of this syndrome is ischemic cerebrovascular disease. Our case is presented because it was a typical case of eight-and-a-half syndrome in terms of clinical picture, magnetic resonance imaging (MRI) findings, and etiology.

Case Report

A man aged 38 years was admitted to the emergency service with symptoms of sudden-onset double vision, and vertigo. His medical and familial histories were unremarkable; blood pressure was 120/70 mmHg, body temperature was 37 °C, and pulse rate was 72/minute. A neurologic examination revealed total paresis in left eye horizontal movements, mild lateral deviation in the primary position of the right eye, limitation of adduction and mild lateral deviation, and nystagmus during lateral gaze (Figure 1, 2). No motor deficit was detected in his extremities and he had peripheral-type left facial paralysis (Figure 3). Other neurologic examination findings were totally normal. Cranial MRI sections revealed an area of acute infarction in the posterior left paramedian pontine tegmentum, which showed limitation of diffusion in diffusion-weighted images (Figure 4). Other cranial MRI sections were totally normal. Acute cerebrovascular accident was diagnosed. In the etiologic examinations, electrocardiography (ECG) was at normal sinus rhythm, transthoracic echocardiography (ECHO), transesophageal ECHO, and a 24-hour Holter ECG recording were normal. MR angiography with lipid suppression was consistent with basilar artery fenestration (Figure 5). The examinations revealed normal results for protein C, S, antithrombin 3, anticardiolipin antibodies, anti-nuclear antibodies, anti dsDNA, and homocysteine. Factor 5 Leiden mutation and *MTHFR* gene mutations were not detected. Neurologic signs of patients treated with antiaggregant agents were improved near-totally.

Discussion

Horizontal gaze movements are regulated by connections between the paramedian pontine reticular formation (PPRF), 6th cranial nerve nucleus, and medial longitudinal fascicle (MLF). These structures are found in the medium and lower pontine tegmentum with an area of approximately 10 mm. Lesions in this area that involve the ipsilateral 6th cranial nerve, PPRF, and MLF together cause one-and-a-half syndrome. PPRF and MLF lesions may cause this syndrome without involvement of this nerve nucleus because the 6th cranial nerve and PPRF fibers converge (3,4). In one-and-a-half syndrome, horizontal eye movements are lost at the side of the lesion; when looking to the contralateral side, only the contralateral eye can abduct and nystagmus is seen in that eye.

One-and-a-half syndrome is rarely an isolated finding. Central facial paralysis (75%), hemiparesis or hemiplegia (30%), hemihypoesthesia (30%) are the most common accompanying findings (3,4). Peripheral facial paralysis is another clinical finding that accompanies one-and-a-half syndrome due to the anatomic structure of the nerve, as seen in our case. In this case, it is known as eight-and-a-half syndrome. The development of peripheral facial paralysis is defined as the following: facial motor nuclei are located on the ventrolateral side of the inferior pontine tegmentum. At the pontine tegmentum, facial motor axons first approach the midline towards the base of the fourth ventricle and then turn around the sixth cranial nerve nucleus (internal elbow) towards the pontocerebelar angle. Facial cranial nerve leaves the brain in front of the eighth cranial nerve at the pontomedullary junction. The internal elbow of the 5th nerve forms the facial colliculus at the base of the fourth ventricle, just in front of the stria medullaris (5). Peripheral facial paralysis occurs with lesions inside the pons or with involvement of the facial nerve nucleus, intrapontine axons (fascicules), or facial colliculus. Especially with inferior pontine paramedian tegmentum lesions, peripheral facial paralysis accompanies one-and-a-half syndrome and this clinical picture is known as eight-and-a-half syndrome (6). The etiology is generally due to ischemic cerebrovascular disease, although rarely demyelinating disease may be detected. Another interesting feature of this case was the detection of basilar artery fenestration in the etiology. Basilar artery fenestration is a rare congenital variant characterized by aneurysm and posterior circulation infarcts. Thrombus formation at the fenestration region due to turbulence is proposed to be responsible for the vascular

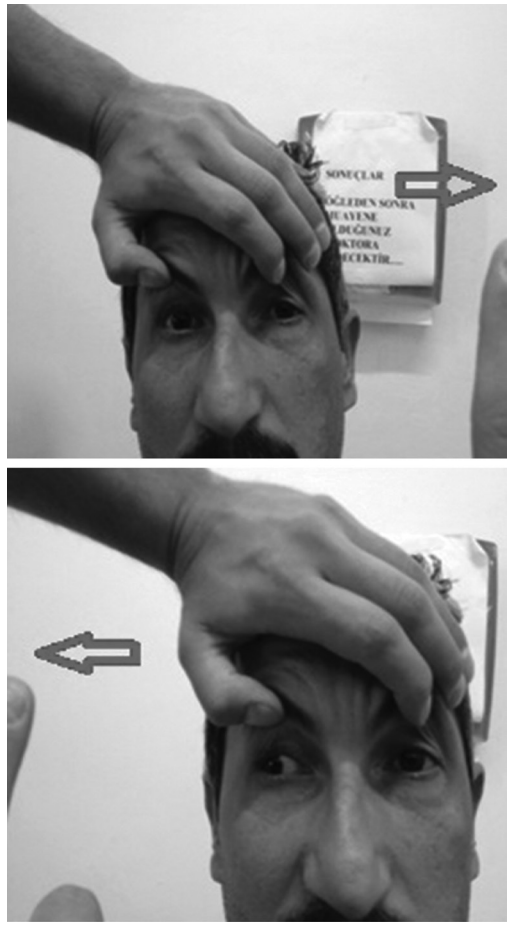

Figure 1, 2. Total paresis in left eye horizontal movements and limitation of adduction in the right eye

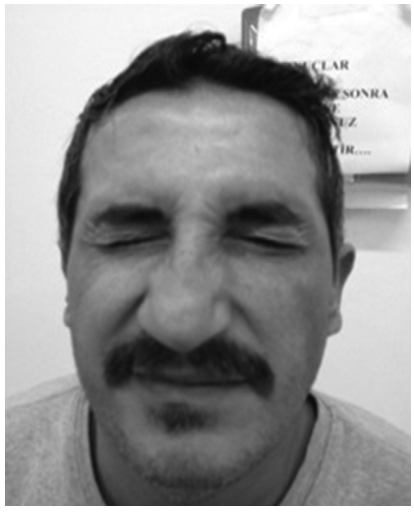

Figure 3. Peripheral facial paralysis on the right side.

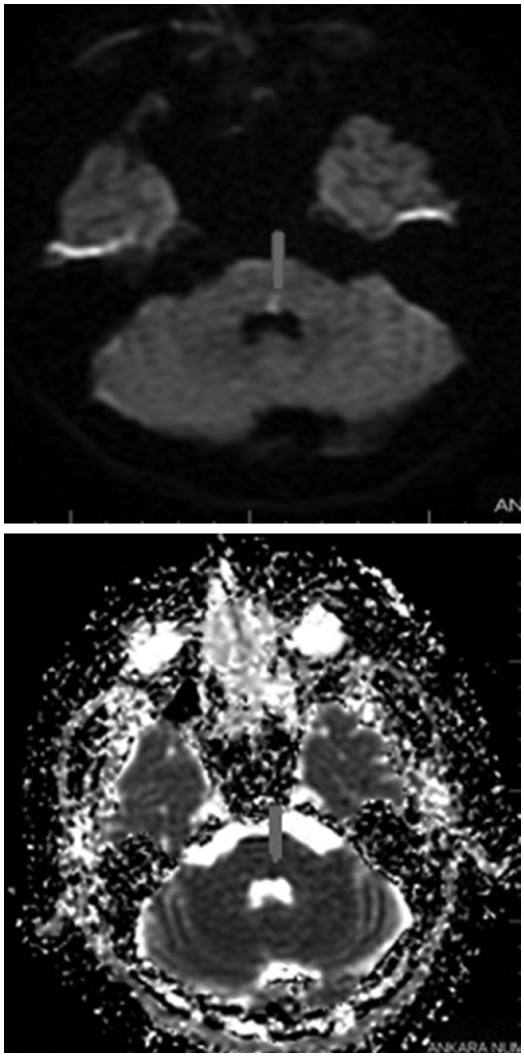

Figure 4. Restriction of of diffusion at posterior side of left paramedian pontine tegmentum.

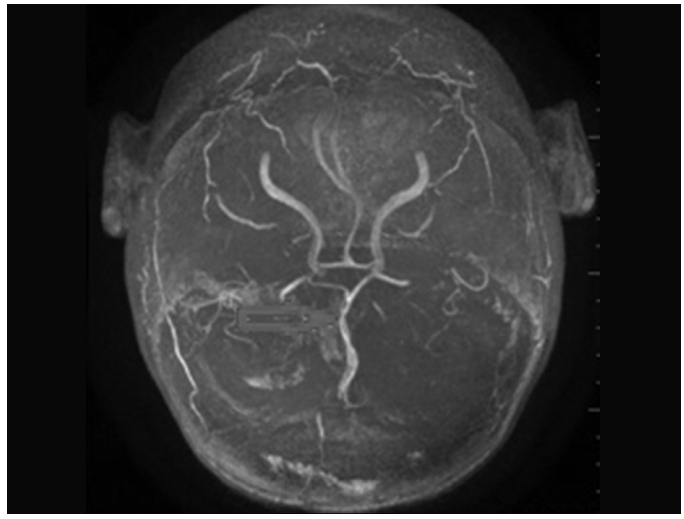

Figure 5. Image consistent with fenestration in a 5-6 mm segment of the proximal basilar artery.

accident (7,8). This rare diagnosis should be considered when comorbid eye movement limitation and peripheral facial paralysis are seen, and brain stem lesions should be searched in these patients.

Ethics

Informed Consent: Consent form was filled out by all participants.

Peer-review: Internally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: E.E., Ş.B., F.A, Concept: E.E., A.P.T., Design: E.E., M.M., Data Collection or Processing: E.E., T.A.T., Analysis or Interpretation: E.E., T.A.T., Ş.B., Literature Search: E.E., Writing: E.E.

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

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