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Case Report

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Isolated eight-and-a-half syndrome associated with pontin hemorrhage: a rare clinical presentation with topological significance

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ABSTRACT

The eight-and-a-half syndrome is a critical clinical condition defined by peripheral facial palsy and the ocular symptoms mostly linked to ischemic cerebrovascular disease affecting the pontin tegmentum. We present a case of isolated eight-and-a-half syndrome with a pontine hemorrhage and discuss it's topological implications in context of neurological practice. This is the one of few cases presenting with eight-and-a-half syndrome following the pontine hemorrhage. Despite showing similar clinical features and clinical prognosis the our case is intriguing due to its unique topological anatomy demonstrating an initial improvement in ocular symptoms.

Keywords: Pontin hemorrhage, opthalmoplegia, eight-and-a-half syndrome

INTRODUCTION

The eight-and-a-half syndrome is a clinical condition defined by complete absence of horizontal eye movements in one direction, loss of inward eye movement in the other direction, involuntary eye movements during outward eye movement, and facial paralysis along with these symptoms. The syndrome, initially identified by Eggenberger in 1998, is characterized by peripheral facial palsy and the ocular symptoms as described by Fischer.[1] The dorsal tegmentum of the caudal pons is a critical region of the brainstem involving the median longitudinal fasciculus (MLF) and parapontine reticular formation (PPRF) with the nucleus and/or the fasciculus of the facial nerve. Within that context, horizontal eye movements are accomplished by connections between the PPRF, the nucleus of the sixth cranial nerve, and the MLF. There have been several etiologies leading to eight-and-a-half syndrome in the pontin region mostly related to the impairment of the dynamic interaction between PPRF, MLF and the facial nucleus and its fasciculus. Among them, the eight-and-a-half syndrome is mostly linked to ischemic cerebrovascular disease (CVD), specifically affecting the pontin tegmentum.[2] It is also worth mentioning that there are also exceptional cases of this syndrome related to some demyelinating disorders, vasculitis and some specific brainstem tumors, such as, brainstem tuberculoma.[1-5]

Here, we present a case of isolated eight-and-a-half syndrome with a pontine hemorrhage and discuss it's topological implications in context of neurological practice.

CASE

A 50 years old-male patient was admitted to the emergency services experiencing intense headaches, and fainting in the last 24 hours. His examination in the emergency room revealed stabil vital signs with the exception of elevated blood pressure of 180/120 mmHg. The patient's neurological examination revealed restricted abduction and adduction in the left eye, nystagmus in the right eye, and peripheral facial paralysis on the left side. The routine blood tests revealed no clinical symptoms, except for a slight elevation in blood urine levels. The brain computed tomography (CT) scan detected a hypertensive hemorrhage with the size of 16×20 mm in the posterior pontine region. Also gradient-echo T2-weighted magnetic resonance imaging (MRI) confirmed the bleeding in the left paramedian tegmentum of the pons while the CT angiography revealed no notable abnormalities except for the presence of plaque, resulting in a 50% narrowing of the left internal carotid artery (ICA). Treatment for pons hemorrhage targeting high blood pressure and brain swelling

has been started immediately after the diagnosis of pontin hemorrhage. The patient responded well to the treatment and on the fourteenth day of recovery, there was a partial but considerable resorbtion of the hemorrhage associated with partial improvement in clinical symptoms including mainly improvement in eye movements that was prominent after one month of the cerebrovascular accident with a sequele peripheral facial palsy that was only partially improved.

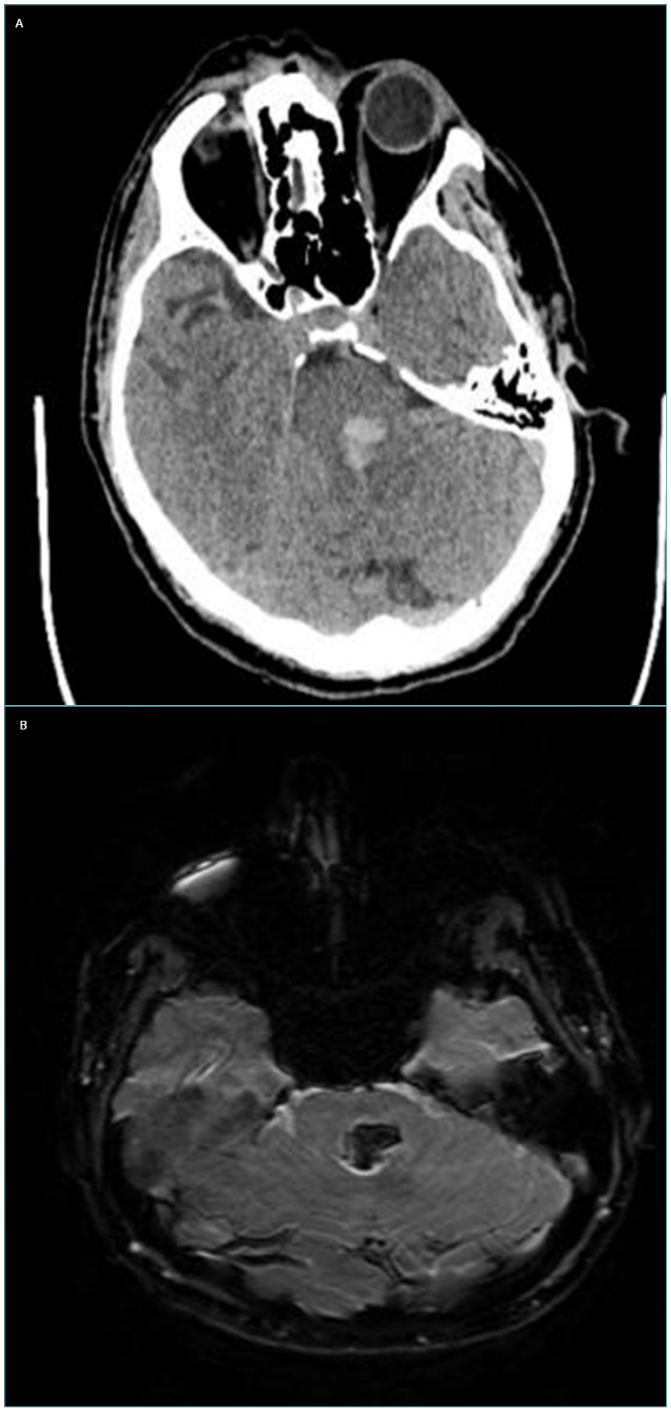

Figure 1a. Computarized Tomography (CT) consisted with pontin hemorrhage, **1b.** Hipointense area consisted with pontin hemorrhage on gradienr-echo T2 weighted MRI

DISCUSSION

The dynamic interaction between the MLF and PPRF are critical in regulating the horizontal gaze movements. However these regulatory structures are tightly located in the lower pontine tegmentum in a very small area leading to an accellerated functional impairment beyond the size of the lesion. Herein, the eight-and-a-half syndrome is a rare entity holding topological significance with its diverse clinical manifestation. For instance, eight-and-a-half syndrome might be also accompanied with some atypical clinical manifestations, such as vertical gaze palsy, however the typical clinical manifestation is a horizontal gaze palsy, central facial paralysis, hemiparesis and hemihypoesthesia defined in most of the cases, as was also described in our case. In this respect the most interesting ophthalmological finding is related to the involvement of PPRF and MLF leading to a clinical picture of a gaze palsy when looking to the contralateral side, where the nystagmus is seen in that eye and only the contralateral eye can abduct.[6,7]

Although ischemic stroke and demyelination are common etiologic factors, pontin hemmorrhage is an uncommon factor manifesting as eight-and-a-half syndrome.[3-5] Herein, there have been totally 21 cases presenting with eight-and-a-half syndrome in the literature related mostly to ischemic stroke and demyelinization with only few cases presenting with eight-and-a-half syndrome related to brainstem hemorrhage.[8] Similar to our case, a patient of pontine hemorrhage exhibited symptoms of left hemiparesis, right facial paralysis, and Fischer syndrome that improved within one month fitting well with the recovery pattern of our patient showing a considerable improvement in the clinical picture within the same time-frame.[9] Similarly, Yadegari et al.8 showed two patients presenting with eight-and-a-half syndrome related to right and posterior pontin ICH with a near identical clinical presentation characterized with an isolated eight-and-a-half syndrome and additional left hemiparesis. Our case's favorable recovery pattern align well with previous literature showing an earlier improvement in eye findings (i.e opthalmoplegia) defined also in previous case reports presenting with eight-and-a-half syndrome.[10]

To the best of our knowledge this is the one of few cases presenting with eight-and-a-half syndrome following the pontine hemorrhage showing similar clinical features and clinical prognosis as was previously reported by Xia and others.[9]

CONCLUSION

The eight-and-a-half syndrome is intriguing clinical syndrome due to its unique topological anatomy and clinical presentation demonstrating an initial improvement in ocular symptoms. Our case suggest a very rare diagnosis that should be considered if a patient is presented when gaze palsy is simultaneously seen with a facial paralysis especially when it comes to older patients with considerable vascular risk factors. Such an clinical appoach might require a special focus in these patients given the tight topological location of these regulatory centers in the pontin region.

ETHICAL DECLARATIONS

Informed Consent

All patients signed the free and informed consent form.

Referee Evaluation Process

Externally peer-reviewed.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

Financial Disclosure

The authors declared that this study has received no financial support.

Author Contributions

All of the authors declare that they have all participated in the design, execution, and analysis of the paper, and that they have approved the final version.

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