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Case Report
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Loss of Vision due to Bilateral Ptosis in a 96-Year-Old Woman – A Case Report

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Abstract

Objectives: Bilateral ptosis is a very seldom presentation and an interesting clinical challenge because of the wide range of possible differential diagnoses of this neurological sign. We report a case of nuclear oculomotor nerve syndrome due to isolated symmetric midbrain ischemia as a rare cause. Through this case report, we aim to determine the difference between a bilateral ptosis with lesion in the oculomotor nucleus, ocular myasthenia gravis or Miller Fischer (MF) syndrome with difficulty in initiating the act of lid elevation, in spite of adequate understanding, motor control and cranial nerve pathways.

Methods: The case report of a 96-year-old woman presenting bilateral ptosis without relative afferent pupillary defect and ischemic lesion in the midbrain.

Results: Our patient suffered a midbrain stroke caused by paroxysmal atrial fibrillation.

Conclusion: Our case report confirms multiple differential diagnoses in bilateral ptosis and the importance of clinical examination in spite of good neurological imaging.

Keywords: Embolism; Stroke; Clinical neurology examination; MRI; Case report

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Abbreviations

AChR: Acetylcholine Receptor Antibodies; ADC: Apparent Diffusion Coefficient; CSF: Cerebrospinal Fluid; DWI: Diffusion-Weighted Imaging; Ewpg: Edinger-Westphal nucleus; FLAIR: Fluid-Attenuated Inversion Recovery; D1b: Anti-GD1b Antibodies; GM1: Anti-GM1 Antibodies; GQ1b: Anti-GQ1b Antibodies; MRI: Magnetic Resonance Imaging; MuSK: Muscle Specific Tyrosine Kinase Antibodies; pAF: Paroxysmal Atrial Fibrillation; RNS: Repetitive Facial Nerve Stimulation

Introduction

Acute loss of vision is a neurological emergency typically caused by an affection of the eyes, optic nerves or central visual pathways. An alternative cause is a lesion of the third cranial nerve or its nucleus in the midbrain resulting in a complete ptosis, paresis of extraocular muscles and a mydriasis of one eye [1]. A bilateral ptosis has a broad, preferentially neuromuscular differential diagnosis but typically does not cause visual loss, because the pupils are only partially covered by the eyelids [2].

By contrast, the simultaneous occurrence of bilateral midbrain lesions affecting the oculomotor nucleus but sparing the immediately adjacent parasympathetic Edinger-Westphal nucleus [3] as well as other brainstem functions is highly unlikely from an anatomical and pathophysiological perspective. We report a rare patient with a bilateral nuclear oculomotor nerve syndrome due to symmetrical lacunar midbrain ischemia.

Case Presentation

A 96-year-old woman presented with sudden bilateral loss of vision that had occurred 3 days before admission. There was no daytime dependent fluctuation, no generalized fatigue, dysphagia, dyspnea, neck or proximal muscle weakness.

The patient's medical history included arterial hypertension, left ventricular dysfunction and chronic venous insufficiency with bilateral crural ulcer as well as bilateral glaucoma, impairing

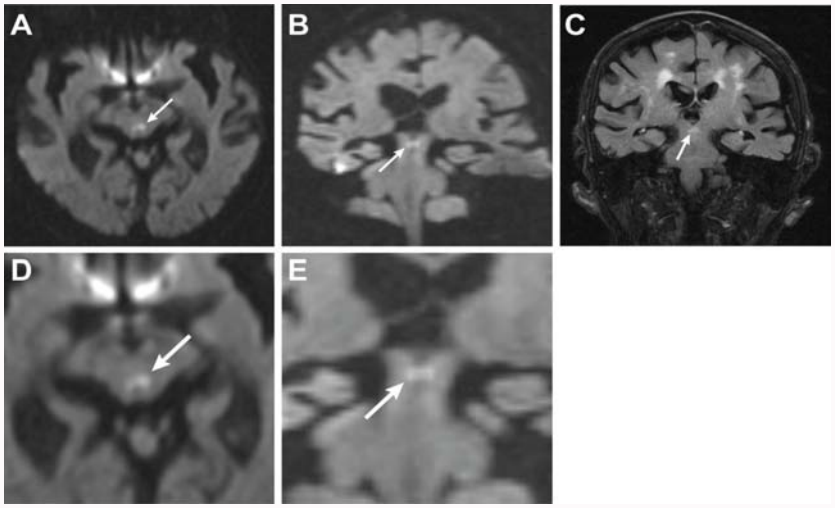

Figure 1: Transversal (A) and coronary (B) Diffusion-Weighted Imaging (DWI) shows a hyperintense bilateral midbrain lesion in location of the ophthalmic nerve nucleus (arrow). (C) Coronary Fluid-attenuated inversion recovery image reveals demarcated ischemia as hyperintense signal in the superior midbrain (arrow). Multiple hyperintense areas in the periventricular white matter and centrum semiovale are also visible and were interpreted as cerebral small vessel disease. (D) Closeup of picture (A) to show bilateralism of ischemic lesion, (E) closeup of picture (B).

vision. There was no history of diabetes mellitus or a family history of neurological diseases.

Neurological examination revealed a bilateral complete ptosis and a complete external ophthalmoplegia (the patient was unable to depress, adduct, abduct or elevate her eyeballs). The pupils were isocore showing a medium diameter and a slowed direct and indirect light reflex.

Furthermore, she had an areflexia for patellar tendon reflex, ankle-jerk reflex and a bilateral pallanesthesia at her ankles. There was no evidence of other focal neurological deficits. The Simpson test was not applicable.

Investigations

Imaging

Magnetic Resonance Imaging (MRI) revealed a symmetric lacunar hyperintense midbrain lesion on Diffusion Weighted Imaging (DWI) (Figure 1A), a hypointensity in the Apparent Diffusion Coefficient (ADC) image and a corresponding signal alteration in the Fluid-Attenuated Inversion Recovery (FLAIR) image (Figure 1C). T2-weighted images showed hyperintensity surrounding the lesion (Figure 2B), indicating postischemic vasogenic edema. There was no contrast agent enhancement of the lesion (Figure 2A) arguing against a tumor infiltration or an inflammatory disease as seen for instance in midbrain tuberculoma [4]. In addition, the MRI showed a remarkable cerebral microangiopathy (Figure 1C).

Cardiovascular diagnostics

Doppler and color-coded duplex sonography of the cerebral arteries revealed atherosclerotic plaques with a 40% stenosis of the left internal carotid artery. In her 24-h electrocardiogram paroxysmal Atrial Fibrillation (pAF) was detected. The 24 h-blood pressure measurement was normal with a mean of 126/80 mmHg.

Laboratory findings

Besides unremarkable routine laboratory studies, including normal HbA1c, extended serological investigations revealed negative antibodies directed against the Acetylcholine Receptor (AChR), Muscle-Specific Kinase (MuSK) and titin, as well as GQ1b, GM1, and GD1b. Cerebrospinal Fluid (CSF) analyses revealed no pleocytosis, total protein elevation or oligoclonal bands, and antibody indices for borrelia burgdorferi as well as neurotropic viruses were negative.

Furthermore, 3 Hz Repetitive Facial Nerve Stimulation (RNS) of the orbicularis oculi muscles did not show a decrement.

Treatment

Due to pAF and a CHA(2)DS(2)-VASc score of 5 points, we initiated anticoagulation with apixaban. Furthermore, we started a treatment with atorvastatin because of an elevated low-density lipoprotein cholesterol level of 129 mg/dl. During the 5-day hospital stay the bilateral ptosis and oculomotor dysfunction improved only slightly.

Discussion

We here report a bilateral nuclear oculomotor nerve syndrome caused by an isolated symmetric, lacunar-like midbrain ischemia. Relevant differential diagnoses such as ocular myasthenia gravis or Miller Fisher syndrome were ruled out by extended serological, CSF, and electrophysiological investigations. While unilateral oculomotor nerve palsy with pupil sparing is not uncommon as an isolated manifestation of midbrain ischemia, bilateral nuclear oculomotor nerve affection is usually caused by large brainstem lesions going along with other neurological deficits as well as pupillomotor impairment [3,5]. A bilateral ischemic affection of the oculomotor nucleus without involving other structures of the brainstem, as reported in this case, is highly unlikely. In particular the absence of a parasympathetic pathway dysfunction indicates isolated impairment of the ophthalmic nerve nucleus, rather than supranuclear or premotor eyelid control disorders or dysfunction of the central caudal nucleus of the third nerve complex. It is remarkable that the ischemic lesion did not affect the nearby preganglionic Edinger-Westphal nucleus (EWpg), and accordingly a relative afferent pupillary defect was lacking [6]. Treatment of her cardiovascular risk factors to prevent further ischemic strokes in consideration of the patient's age was initiated.

The infarct responsible for the above syndrome is placed in the territory of long mesencephalic median and paramedian perforators

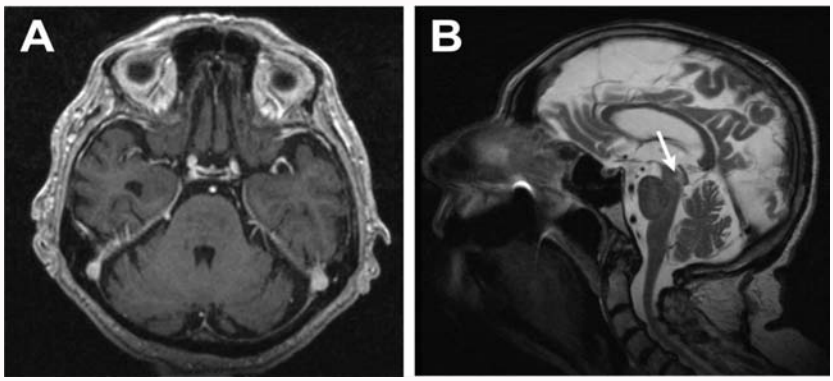

Figure 2: (A) Postcontrast transversal T1 scan does not show a gadolinium enhancement of the midbrain lesion. (B) T2 image shows hyperintense signal as sign of the extended perifocal edema.

(perforating small arteries) arising from the top of the basilar artery. Simultaneous bilateral infarcts can be explained by bilateral small vessel occlusive disease in our patient of old age, possibly by a rare anatomical variant with a common trunk of the left and right perforators (Percheron type of artery [7,8]), or, by a transient embolic occlusion of the top of the basilar artery due to a cardiogenic embolus.

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