Sixteen syndrome, a new pontine ophthalmo-neurological syndrome within the one and a half syndrome spectrum of disorders in a diabetic patient: A case report

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Abstract

A 52-year-old diabetic male presented with dizziness, diplopia, salivation, and mild weakness in the right limb for 3 days. Neurological examination revealed complete absence of adduction of the left eye and incomplete mild abduction. There was bilateral facial palsy. The right upper and lower limbs were mildly weak with abnormal Babinski reflex. Laboratory tests showed an increase in fasting blood glucose. Magnetic resonance diffusion-weighted imaging and apparent diffusion coefficient showed bilateral pontine tegmentum and pars basilaris infarction. He was discharged after 10 days with significant improvement in dizziness and limb weakness. At one month of follow-up, diplopia disappeared leaving only a slight peripheral facial paralysis on the left side. This rare case further expands the landscape of ponto-opthalmic disorders.

INTRODUCTION

Due to their predilection for vascular anomalies, diabetic patients are considered high risk for rare vascular lesions. One and half syndrome is a term originally proposed by Fisher to describe a syndrome with ocular dyskinesia, exhibiting ipsilateral conjugate gaze palsy and ipsilateral internuclear ophthalmoplegia (INO). The pathological lesion is frequently at the level of unilateral tegmentum affecting the paramedian pontine reticular formation (PPRF), abducens nucleus and the median longitudinal fasciculus (MLF). In majority of the cases, the lesion is caused due to cerebrovascular disease of smaller vessels leading to ponto-cerebral infarction. A number of numbered ophthalmo-pontine neurological syndromes have been reported with the clinical features of one and a half syndrome preserved in all the reports. We report here a case of sixteen syndrome, the first case in the literature to the best of our knowledge, in a diabetic patient. An informed consent was obtained from patient for publication.

CASE REPORT

A 52-year-old diabetic, male presented with dizziness, diplopia, salivation, and mild weakness in the right limb for 3 days. He had poorly controlled hypertension and diabetes, with history of smoking and drinking alcohol for nearly 30 years. Blood pressure at hospital admission was 162/88mmHg. Neurological examination revealed complete absence of adduction of the left eye and incomplete mild abduction. There was bilateral facial palsy. The right upper and lower limbs were mildly weak with abnormal Babinski reflex. Laboratory tests showed an increase in fasting blood glucose (10.02 mmol/L) and glycated hemoglobin (8.1%). Urine routine glucose (4+), low-density lipoprotein cholesterol (2.49 mmol/L), and homocysteine levels were normal. Magnetic resonance diffusion-weighted imaging (DWI) and apparent diffusion coefficient (ADC) showed bilateral pontine tegmentum and pars basilaris infarction. He was discharged after 10 days with significant improvement in dizziness and limb weakness. At one month of follow-up, diplopia disappeared leaving only a slight peripheral facial paralysis on the left side. This rare case further expands the landscape of ponto-opthalmic disorders.
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basilaris intrusion, as evident from the distinct abnormal signals, along the arrow pointing continuous linear anomaly signal (Figure 1A, 1B). Magnetic resonance angiography (MRA) did not show evidence of significant intracranial lesions.

After admission, he was given dual antiplatelet, statin, antihypertensive and antidiabetic therapy. After 2 days, the right limb weakness completely recovered, but abnormal Babinski reflex continued. He was discharged after 10 days with significant improvement in dizziness but the peripheral facial paralysis and diplopia remained. At one month of follow-up, diplopia disappeared leaving only a slight peripheral facial paralysis on the left side. This persisted at three and six months follow-up.

DISCUSSION

We named the syndrome as sixteen syndrome keeping in line with the other named numerical neuro-opthalmological syndromes. In this case, the axons of internuclear neurons from the abductor nucleus were affected bilaterally as they pass through the midline through the medial longitudinal bundle (MLB) rising up to the level of the oculomotor sub-nucleus that controls the medial rectus. Hence the patient’s both eyeballs had no adduction. Further, the left abductor nerve was slightly damaged affecting the abduction of the left eye which was incomplete. This constitutes the one-and-a-half syndrome (1.5). Also, the bilateral facial nerve nucleus was damaged (7 +7), resulting in bilateral peripheral facial paralysis. The mild weakness of the right limb (0.5)\(^2\), lasted 5 days with abnormal Babinski reflex.

Clinically similar fifteen and a half syndrome may be caused by dolichoectasia of basilar artery and long anterior inferior cerebellar arteries (AICAs), suggestive of vascular damage. However, there was no previous imaging evidence. In our case, the DWI and ADC in the MRI of the patient indicates that the paramedian artery as origin of the lesions, thus a pathology of the small penetrating artery. Patients with 15.5 syndrome were associated with hypertension and diabetes, supportive of vascular disease. This is similar in our patient. Our case is extremely rare and it further expands the landscape of ponto-opthalmic disorders. Knowledge of the clinical features will help in identifying the lesion locus which may improve the clinical care.

DISCLOSURE

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REFERENCES