

Plus Minus Lid Syndrome with Ataxia

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Abstract

Plus minus lid syndrome is an ocular syndrome characterized by unilateral ptosis and contralateral lid retraction. Also when the ipsilateral lid is raised manually, the contralateral retracted lid does not revert. This patient presented with features of plus minus lid syndrome with ataxia due to a vascular mesencephalic lesion. ©

INTRODUCTION

Plus minus lid syndrome is an acquired neurological abnormality of eyelid position, which concerns the association of unilateral ptosis with contralateral eyelid retraction.¹ This association has been previously described in ocular myasthenia after lesions of ocular motor nerve, ocular myositis and paramedian mesencephalic – diencephalic lesions.²

CASE REPORT

A 45 yrs old male presented with a 24-hour history of acute onset imbalance on walking with a tendency to fall on the right side associated with a partial drooping of the left eyelid. There was no history of weakness, sensory impairment, sphincteric disturbances or worsening of imbalance in the dark. There was no diplopia, dysphagia or nasal regurgitation to suggest other cranial nerve involvement. Patient was non-diabetic and non-hypertensive without any history of addiction or high-risk sexual behavior. Physical examination revealed a pulse of 82 beats per minute and blood pressure of 124/76 mmHg. On neurological examination, the higher functions including speech were normal. There was partial ptosis of the left eye and lid retraction of the right eye (Fig. 1). The retraction however, did not reduce or revert when the ptotic left eyelid was manually raised. However the pupils were bilaterally equal, reacting to light with normal and complete extraocular movements and no visual field defects were noted. Examination of other cranial nerves as well as motor and sensory systems was unremarkable except for right-sided cerebellar signs (dysmetria and



Fig. 1 : Clinical presentation: showing ptosis of the left eyelid and retraction of the right eyelid.

dyssynergia). Both the plantars were flexors. Examination of other systems was normal.

Isolated unilateral ptosis with contralateral lid retraction of sudden onset made ocular myopathies or myasthenia unlikely. Also the ataxia supported the possibility of a central vascular lesion. So an urgent CT scan was requested at admission, but failed to detect the lesion. However a subsequent MRI revealed a small left ventral and paramedian infarct involving the midbrain (Figs. 2 & 3). Further investigations for risk factor evaluation were normal except for hypertriglyceridemia. The patient was started on anti platelet drugs and followed up over a period of 4 weeks, during which remarkable improvement occurred.

DISCUSSION

Unilateral ptosis and contralateral eyelid retraction originally described as palpebral plus-minus syndrome by Gaymard *et al*¹ can occasionally occur due to lesions involving the III rd nerve fascicles. Two muscles are involved in upper eyelid elevation; the tarsal smooth muscle of Muller that has a role limited to tonic control

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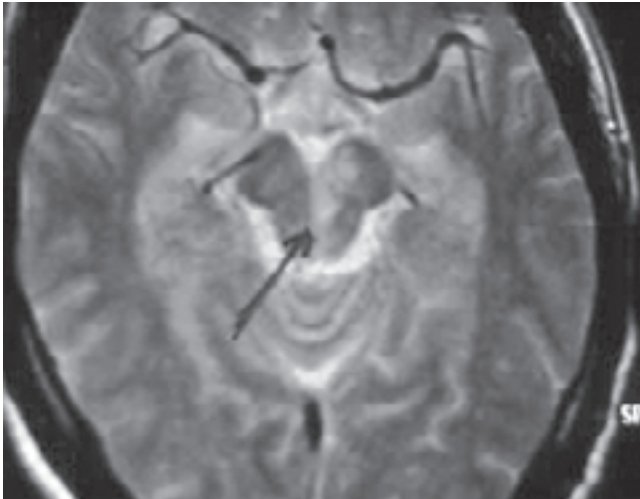


Fig. 2 : MRI brain: (arrow) showing left sided paramedian infarct in the midbrain.

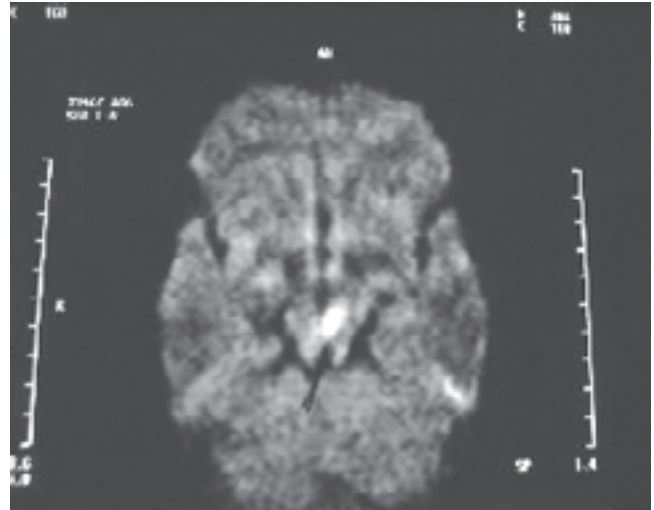


Fig. 3 : MRI brain (diffusion scan): (arrow) showing left sided paramedian infarct in the midbrain.

of eyelid position and the levator palpebrae, a skeletal muscle innervated by fascicles of the oculomotor nerve. These fascicles arise from a single medial nucleus called the central caudal nucleus, a subdivision of the oculomotor nuclear complex.³ Nuclear lesions therefore lead to bilateral ptosis; however fascicular lesions can cause unilateral ptosis.⁴ Another structure, the nucleus of the posterior commissure provides inhibitory inputs to the central caudal nucleus. Each nucleus of the posterior commissure is connected with its contralateral counterpart through the posterior commissure but does not project directly on the central caudal nucleus. It projects on the levator palpebrae motor neurons in the supraoculomotor area, located dorsolaterally to the oculomotor nucleus, within the periaqueductal gray mater.³

Our patient had a left paramedian infarct in the midbrain, ptosis of the left eye and lid retraction of the right eye. He did not have other signs of oculomotor nerve involvement. Ipsilateral ptosis was due to involvement of the levator palpebrae fascicles as they emerge from the central caudal nucleus (the nucleus itself being spared). The eyelid retraction was possibly due to failure of inhibition of the contralateral levator palpebrae (by the nucleus of the posterior commissure) resulting in its overaction.³ Contralateral retraction may also occur as a mechanical effect secondary to ipsilateral ptosis (due to Hering's law); in such case however, on raising the ptotic lid manually the contralateral retraction corrects itself.⁵ It may thus be inferred that inhibitory connections between the nucleus of posterior commissure and the central caudal nucleus (through the supraoculomotor area) are unilateral and crossed.³ Cases have been reported in which mesencephalic lesions have led to contralateral ptosis and ipsilateral lid retraction.² Thus, a crossed pattern may also exist for

excitatory afferents to the central caudal nucleus.³

Also our patient had ataxia on the right side. This was probably due to concomitant damage to the fibres of superior cerebellar peduncle causing contralateral ataxia.⁶ To the best of our knowledge such presentation of plus minus syndrome with contralateral ataxia has not yet been reported from Asia.

To conclude, this combination of ipsilateral ptosis with contralateral lid retraction even in the absence of other brainstem or long tract signs, though rare has been documented to be due to central mesencephalic lesions and investigations to confirm the same are justified prior to other tests for isolated lid dysfunction.

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