

# Claude's Syndrome Associated with Supranuclear Horizontal Gaze Palsy Caused by Dorsomedial Midbrain Infarction

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**Abstract-** Claude's syndrome caused by dorsal midbrain lesion is characterized by ipsilateral third nerve palsy and contralateral ataxia. To date, reports in the literature concerning Claude's syndrome associated with the midbrain paresis of horizontal gaze are rare. A 62-year-old man suddenly developed left third cranial nerve palsy, right lateral gaze palsy, and right ataxia. Intact Bell's phenomenon and preserved right horizontal oculocephalic reflex suggested the lateral gaze palsy in the right eye was supranuclear in nature. Magnetic resonance imaging (MRI) revealed an infarction in the left dorsomedial midbrain. Although the red nucleus has often been suggested as the lesion site responsible for Claude's syndrome, a lesion of the superior cerebellar peduncle just below and medial to the red nucleus could be responsible for this syndrome. This case demonstrates neurological heterogeneity of midbrain infarction.

**Key Words:** Midbrain infarction, Ophthalmoplegia, Magnetic resonance imaging

*Acta Neurol Taiwan 2005;14:147-150*

## INTRODUCTION

Midbrain lesions may give rise to complex eye movement disorders<sup>(1)</sup>. Three main types of dysfunction are delineated. First, a fascicular syndrome of the third nerve is indicated when the peripheral type of third cranial nerve palsy is associated with some specific neurological disturbance (syndrome of Weber, Benedict, or Claude). Second, a nuclear syndrome is suggested when bilateral ptosis, bilateral mydriasis, bilateral or contralateral superior rectus weakness are present. And, third, a supranuclear syndrome usually includes vertical gaze palsy, skew deviation, sea-saw nystagmus, and ver-

tical one-and-a half syndrome. Claude's syndrome is a well-known midbrain syndrome characterized by ipsilateral oculomotor nerve palsy and contralateral cerebellar ataxia. This syndrome is very rare and only a few cases<sup>(2-5)</sup> have been reported since 1912<sup>(6)</sup>. A patient presenting with Claude's syndrome and supranuclear horizontal gaze palsy is described herein.

## CASE REPORT

On January 23, 2004, a 62-year-old male farmer was admitted for sudden onset of dizziness and inability to open his left eye. Before the onset of these symptoms,

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Received January 31, 2005. Revised March 2, 2005. Accepted May 25, 2005.

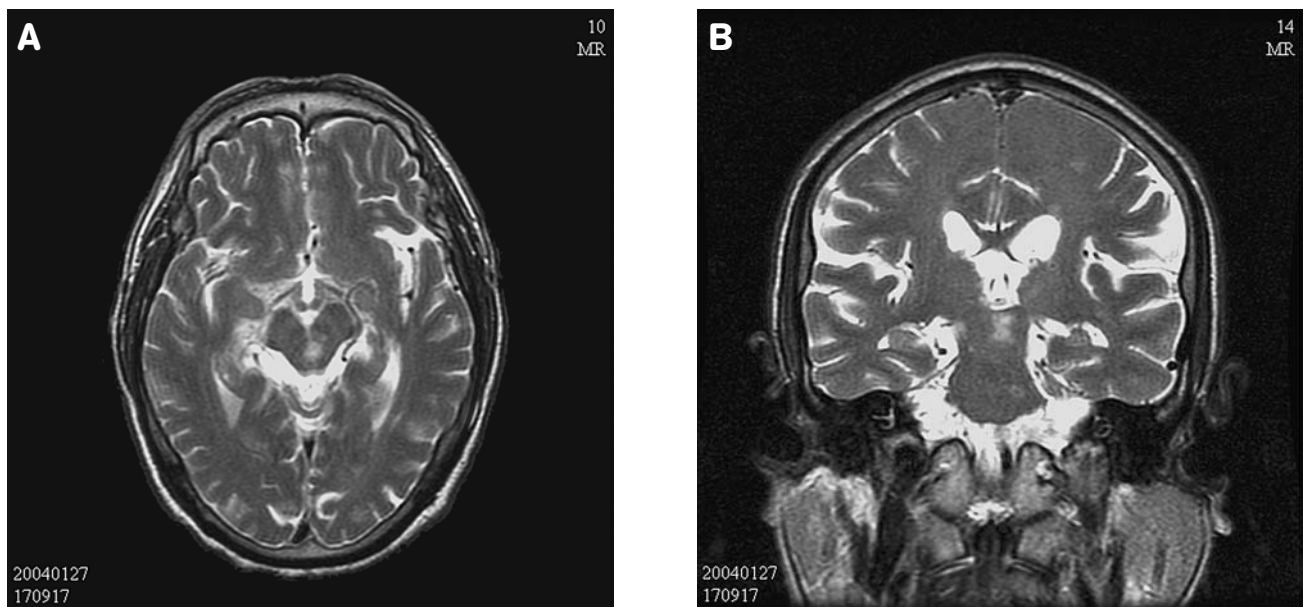
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he was in good health except for uncontrolled hypertension and diabetes for 6 years. He had no prior history of strokes.

On admission, the general physical findings were normal. No carotid bruit was heard. Blood pressure was 185/100 mmHg and pulse rate was 92 beats per minute. On neurological examination, the patient was found to be alert, and his language function was intact. His visual acuity and visual fields were normal. There was ptosis of the left eye. Pupillary diameter was 3 mm on the right and 5 mm on the left. Reaction to light stimuli was brisk on the right and absent on the left. In the primary position the right eye was orthophoric and the left was exotropic. Adduction, elevation, and depression of the left eye were limited, but abduction was full. Abduction of the right eye was limited, but adduction, elevation and depression were intact. Intortion of bilateral eyes on attempting adduction and depression suggested the fourth cranial nerve was intact. Oculocephalic maneuver elicited abduction responses in the right eye, but failed to improve eye movement in the left. Bell's phenomenon was shown in the right eye, but absent in the left. Neither positional nor gaze-evoked nystagmus was present.

Muscle power was grade 5. The Babinski response was flexor. There was no sensory disturbance. There was dysmetria of the right side limbs. The patient's gait was wide-based, and he swerved to the right when attempting to walk a straight line. Fasting blood sugar level was 233 mg/dL. Liver, renal function, lipid study, complete blood count, urinalysis, coagulation profile, and electrolyte test results were all within reference ranges. Electrocardiographic and echocardiographic findings revealed no cardiac source of emboli. T2 weighted MRI demonstrated a high signal intensity lesion over the left dorsomedial midbrain tegmentum (Fig. 1). The rest of the brain stem seemed normal. The carotid duplex showed increased thickness of the intima media of the left common carotid artery. Transcranial Doppler findings indicated bilateral poor temporal windows, and normal velocity in the bilateral vertebral and basilar arteries through transforaminal window. Magnetic resonance angiography showed patent vertebral, basilar and posterior cerebral arteries.

On the days following initial assessment, the right lateral gaze palsy was partially resolved; however, the left third cranial nerve palsy persisted. He was discharged on January 28, 2004.



**Figure 1.** (A) Axial T2-weighted MRI depicts an area of high signal intensity in the left midbrain tegmentum medial to the red nucleus; (B) Coronal T2-weighted MRI demonstrates a focal high signal lesion in the tegmental area caudal to the red nucleus.

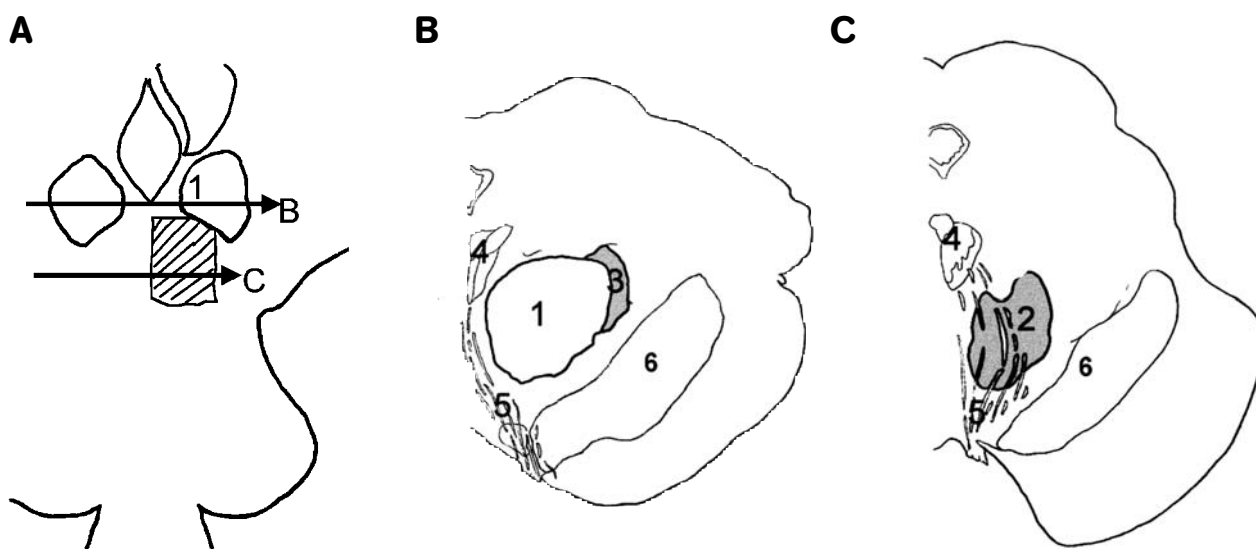
## DISCUSSION

Monocular palsy of elevation, depression, and adduction associated with large pupil and ptosis in the left eye may correspond to involvement of the third cranial nerve fascicle or nucleus. Due to sparing of the contralateral lid levator and the pupil constrictor, nuclear involvement of the third cranial nerve was unlikely. Failure of oculocephalic maneuver or Bell's phenomenon to overcome the limitation of the left eye palsy also indicated a fascicular lesion of the third cranial nerve in our patient. The clinical feature of complete ipsilateral oculomotor nerve paralysis and contralateral cerebellar ataxia is consistent with Claude's syndrome.

The principal localization of Claude's syndrome has previously been attributed to a lesion of the red nucleus, since the cerebellar efferent fibers (dentatothalamic fibers) and oculomotor nerve fascicles meet at the level of the red nucleus<sup>(4)</sup>. Information concerning the precise pathway of the cerebellar efferent fibers in the midbrain is incomplete. Dentatothalamic fibers do not pass through the red nucleus, but ascend directly toward the

lateral thalamus, sending a few branches to the red nucleus (Fig. 2). Fibers of the superior cerebellar peduncle are found medial to the red nucleus at the level of the caudal end of the red nucleus<sup>(7)</sup>. The MRI in our patient showed that Claude's syndrome occurred because of a lesion in the superior cerebellar peduncle just below and medial to the red nucleus. This patient's findings are compatible with previous reports that Claude's syndrome is resulted from the lesions on the tegmental area below the red nucleus, where the superior cerebellar peduncle is located<sup>(2-5,7)</sup>. Only a few reported cases indicated additional lesions in the red nucleus<sup>(7,8)</sup>. These findings strongly suggest that the superior cerebellar peduncle is responsible for this syndrome and that the red nucleus contributes little to the syndrome.

In Claude's syndrome, the oculomotor nerve fascicles, as well as the cerebellar efferent fibers, are involved concurrently. At the red nucleus level, the oculomotor nerve fascicles do not meet the cerebellar efferent fibers; since the oculomotor nerve fascicles pass medially and the dentatothalamic fibers ascend laterally to the red nucleus. However, at the level of the caudal



**Figure 2.** (A) Coronal diagram shows the localization of Claude's syndrome. The hatched area represents the location of the lesion in our patient which was demonstrated on MRI. (B) (C) Axial diagrams show cerebellar efferent fibers (gray colored-area; the superior cerebellar peduncle and the dentatothalamic tract), the red nucleus, and the oculomotor nerve fascicles in the corresponding midbrain levels. 1: red nucleus; 2: superior cerebellar peduncle; 3: dentatothalamic tract; 4: oculomotor nuclei; 5: oculomotor nerve fascicles; 6: substantia nigra.

end of the red nucleus, the oculomotor nerve fascicles traverse the midbrain divergently and run across the superior cerebellar peduncle. Hence, a small lesion at this level can produce all of the symptoms of Claude's syndrome. These anatomic findings support the observations made in our patients.

Neurons related to the horizontal gaze are located in the frontal eye field (FEF, Brodmann area 8). Impulses from FEF proceed caudally in a pathway that runs in the internal capsule and medial portion of the cerebral peduncle, decussates at the pontomesencephalic junction, and ends in the contralateral paramedian pontine reticular formation (PPRF). Knowledge of midbrain control of horizontal gaze is scanty<sup>(9,10)</sup>. In monkeys, unilateral stimulation in the paramedian region of the midbrain tegmentum elicited contralateral deviation of the eye<sup>(11)</sup>. Zackon and Sharpe<sup>(9)</sup> demonstrated that mesencephalic reticular formation contained supranuclear pathways for horizontal eye movements. In our patient, damage to the left midbrain tegmentum caused paresis of the contralateral saccades, evidenced by limited abduction in the right eye.

The vascular supply to the midbrain is complex<sup>(12)</sup>. The basilar artery supplies the ventral paramedian region of the midbrain, the superior cerebellar artery supplies the lateral-dorsal region of the caudal two-thirds of the midbrain, and the posterior cerebral artery (PCA) supplies the upper half of the midbrain. The PCA contribution is divided into three components, the first from the P1 segment (paramedian thalamomesencephalic artery supplying the dorsal paramedian part of the upper midbrain and paramedian part of the thalamus), and two others from the P2 segments (the peduncular artery and the posterior choroidal artery supplying the cerebral peduncle, upper midbrain tegmentum and superior colliculus). The lesions shown on MRI from this patient were consistent with infarction of the territories of the paramedian thalamomesencephalic artery.

In conclusion, our patient presented with left complete oculomotor nerve palsy, right cerebellar ataxia, and right horizontal gaze palsy. Claude's syndrome associat-

ed with supranuclear horizontal gaze palsy in the same patient has seldom been reported. The MRI findings in our patient emphasize the notion that Claude's syndrome occurs because of a lesion of the superior cerebellar peduncle just below and medial to the red nucleus.

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