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# Pitfalls in the diagnosis of pupil-sparing oculomotor nerve palsy without limb ataxia: A case report of a variant of Claude's syndrome and neuroanatomical analysis using diffusion-tensor imaging

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ABSTRACT

Midbrain infarction causing oculomotor nerve palsy with contralateral ataxia is named Claude's syndrome. Herein we report the case of a variant of Claude's syndrome, which shows pupil-sparing oculomotor nerve palsy without the accompanying neurological deficits other than subtle truncal ataxia. MRI and Diffusion Tensor Imaging revealed that midbrain infarction was located rostrally above the decussation of the superior cerebellar peduncle (SCP) and might have partially destructed the tectospinal tract, which resulted in the absence of limb ataxia and presence of subtle truncal ataxia. In this variant of Claude's syndrome, we should carefully assess truncal ataxia to avoid misdiagnosing it as isolated pupil-sparing oculomotor nerve palsy because the patient showed apparently normal gait and truncal ataxia was only revealed by unstable tandem gait.



## 1. Introduction

Isolated pupil-sparing oculomotor nerve palsy is caused by diabetic or hypertensive oculomotor nerve ischemia, because end-arterial ischemia tends to cause more severe damage to centrally located extraoculomotor fibers than to peripherally located pupillomotor fibers [1–3]. Since oculomotor nerve palsy caused by midbrain infarction usually accompanies other neurological abnormalities [4] and oculomotor nerve palsy without other neurological findings has been considered to be rare [5], isolated pupil-sparing oculomotor neve palsy is usually attributed to peripheral oculomotor nerve ischemia.

## 2. Case presentation

A 69 year-old man who took carvedilol and warfarin for hypertension and chronic atrial fibrillation experienced sudden-onset diplopia and came to our hospital 3 h after onset.

On examination, he had impaired function of the right superior, inferior, and medial rectus muscles with intact pupillary responses to light and no ptosis bilaterally. Nose-finger-nose, heel-shin, knee-pat, and hand pronation and supination tests were bilaterally normal, thus not suggesting limb ataxia. He had no paralysis, extrapyramidal signs, dysarthria, or sensory disturbance. Even though his gait was apparently normal and did not include ataxic features, subtle truncal ataxia was revealed by unstable tandem gait and Mann's posture. Blood examination revealed neither diabetes mellitus nor dyslipidemia, and the prothrombin time-international normalized ratio (PT-INR) was 2.44, within therapeutic range.

Axial diffusion-weighted imaging (DWI) at 1.5-Tesla MRI showed no definite lesion; however, coronal DWI depicted a slightly hyperintense area, suggesting an acute ischemic lesion in the right para-median midbrain, which was located between the two axial sections (Fig. 1A). Two days later, although no neurological signs progressed, the lesion became more evident (Fig. 1B). MR angiography findings were within normal limits. Consequently, acute lacunar infarction was diagnosed. His diplopia improved gradually, and he had almost no visual disturbance 1 month later. The subtle truncal ataxia disappeared as well.

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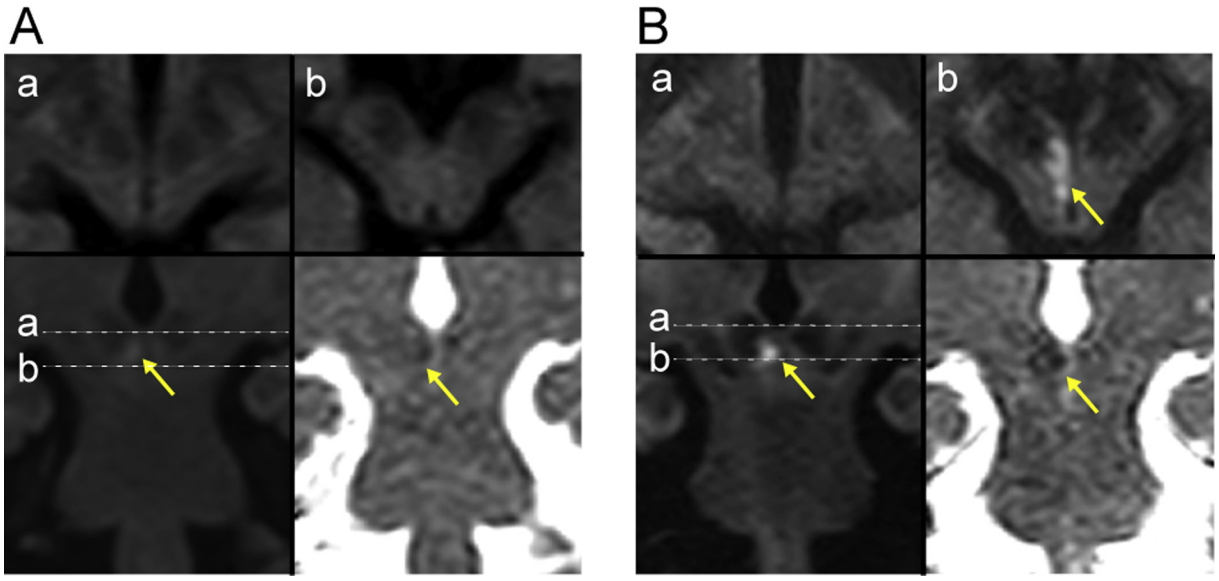


**Fig. 1.** An acute ischemic lesion in the right para-median midbrain with restricted diffusion on diffusion-weighted imaging (DWI) and apparent diffusion coefficient (ADC) map. Yellow arrows indicate the lesion (upper figures: axial images, each corresponding to the axial section indicated by the dotted line in the lower left-hand side figure. Lower figures: coronal images; left, DWI; right, ADC map). (A) On admission, no definite lesion was detected on axial DWI; however, coronal DWI depicted a slightly hyperintense area suggestive of acute ischemic lesion in the midbrain. The coronal ADC map also showed a slightly hypointense area. Note that the lesion is located between the axial sections. (B) Two days later, the lesion became more evident even though no neurological signs progressed during the 2 day period. The lesion is also depicted on axial DWI.

## 3. Discussion

In the present case, the patient showed intact pupillomotor function and no ptosis because the ischemic lesion was so small that parasympathetic fibers were spared and oculomotor fibers were not damaged completely. This case is characterized as pupil-sparing oculomotor nerve palsy without limb ataxia due to midbrain infarction, which is located below and medial to the red nucleus: a variant of Claude's syndrome that differs from the typical type in that it has no limb ataxia.

Claude's syndrome is well known as midbrain infarction causing oculomotor nerve palsy with contralateral ataxia. Seo et al [6] demonstrated that the localization of Claude's syndrome involves a lesion of the superior cerebellar peduncle (SCP) just below and medial to the red nucleus. Thereafter, ataxia in Claude's syndrome has been attributed to the destruction of the SCP, which contains the efferent fibers from the cerebellum. Tokunaga et al. [7] reported a case of midbrain infarction causing oculomotor nerve palsy with ipsilateral limb ataxia, a variant of Claude's syndrome with ipsilateral ataxia, and discussed the findings of diffusion-tensor imaging (DTI). They argued that the lesion located at the rostral extremity of the decussation of the SCP caused ipsilateral ataxia, and the destruction above the decussation resulted in the contralateral ataxia.

To identify the anatomical relationship between the ischemic lesion and the SCP, we performed DTI using a 3-Tesla MRI 27 days after the onset (Fig. 2). It revealed that the lesion was located rostrally above the decussation of the SCP and medial to the red nucleus. Because the lesion was localized adjacent to the midline and did not extend laterally and caudally, the SCP that ascends laterally after the decussation seemed to be spared. This anatomical feature explains the absence of limb ataxia in our case.

In addition, our case is characterized by the presence of subtle truncal ataxia. It is hypothesized that the ischemic lesion involved the decussation of the tectospinal tract that locates medially to the red nucleus and the SCP [8] and that the destruction of the tectospinal tract might cause the dysfunction of postural change control. The tectospinal tract originates from the superior colliculus, decussates in the dorsal tegmental decussation medial to the red nucleus, and descends ventrolaterally to the medial longitudinal fascicle [8]. Even though the anatomy and the function of the tectospinal tract in humans largely remains undiscovered, the tectospinal tract is considered to be responsible for controlling the movement of the head and postural changes in response to the visual information [9]. Therefore, the subtle truncal ataxia in our case might be explained by the strictly localized destruction of the tectospinal tract.

In particular, in this variant of Claude's syndrome, we should carefully assess truncal ataxia to avoid misdiagnosing it as isolated pupil-sparing oculomotor nerve palsy because the patient showed apparently normal gait and truncal ataxia was only revealed by unstable tandem gait. As shown in our case, to properly diagnose pupil-sparing oculomotor nerve palsy caused by midbrain infarction, we need more careful neurological examination and additional coronal DWI because it is often difficult to detect radiologically a small midbrain infarction on axial DWI alone [10].

In summary, we have presented the case of a variant of Claude's syndrome, which shows pupil-sparing oculomotor nerve palsy without the accompanying neurological deficits other than subtle truncal ataxia. Based on the DTI, midbrain infarction was located rostrally above the decussation of the SCP and might have partially destructed the tectospinal tract.

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