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Case report

# Pupil-sparing third nerve palsies and hemiataxia: Claude's and reverse Claude's syndrome

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#### ABSTRACT

We report two patients with midbrain infarction with pupil-sparing third nerve palsies and hemiataxia: one with contralateral ataxia (Claude's syndrome) and one with ipsilateral ataxia (which we refer to as reverse Claude's syndrome). We highlight the importance of a thorough neurologic evaluation with partial oculomotor palsies and describe, to our knowledge, the fourth account in the literature of a pupil-sparing third nerve palsy with ipsilateral cerebellar ataxia.

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#### 1. Introduction

Isolated pupil-sparing third nerve palsies are frequently attributed to diabetic or hypertensive cranial nerve ischemia. The topographic organization of the third cranial nerve is such that the pupillomotor fibers are located on the periphery of the nerve and ischemic lesions (that is, those seen in hypertensive and diabetic ischemic neuropathies) typically result in pupil-sparing third nerve palsies that range from partial to complete ipsilateral third-nerve palsies [1].

#### 2. Case reports

#### 2.1. Patient 1

A 74-year-old woman presented to our hospital emergency department with the acute onset of speech, ambulation, and balance difficulties. She had left arm and leg ataxia. She had intact pupillary responses to light bilaterally, right impaired superior rectus, inferior rectus, and medial rectus function and left impaired superior rectus function. MRI showed an acute infarction of the right midbrain and pons (Fig. 1A, B).

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#### 2.2. Patient 2

An 88-year-old woman developed horizontal diplopia and balance problems with falling to the right. She presented to her primary care physician's office after 2 days of symptoms. She was referred for neurological consultation after discovery of a right midbrain infarction (Fig. 1C). She had mild ataxia of the right upper extremity, normal pupillary responses to light bilaterally and impaired adduction of the right eye.

#### 3. Discussion

Isolated pupil-sparing third nerve palsies are frequently attributed to diabetic or hypertensive cranial nerve ischemia. In both patients presented here the presence of hemiataxia indicating a unilateral lesion to the brainstem or cerebellum was difficult to reconcile with a pupil-sparing third nerve palsy traditionally associated with peripheral lesions.

Claude's original description of his now eponymous syndrome included a patient with a full third nerve lesion including a fixed and dilated pupil in addition to contralateral ataxia [2]. At autopsy this patient was discovered to have an infarction involving the right paramedian midbrain with involvement of the red nucleus, third nerve fascicle, and the superior cerebellar peduncle [3,4].

Seo et al. [5] reviewed the known cases of Claude's syndrome. They argue that, while the classic description of localization is

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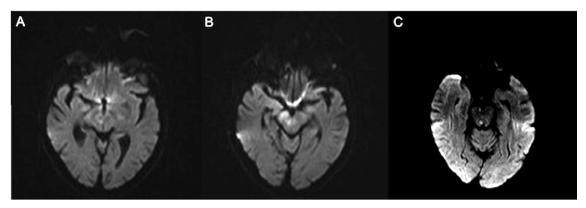
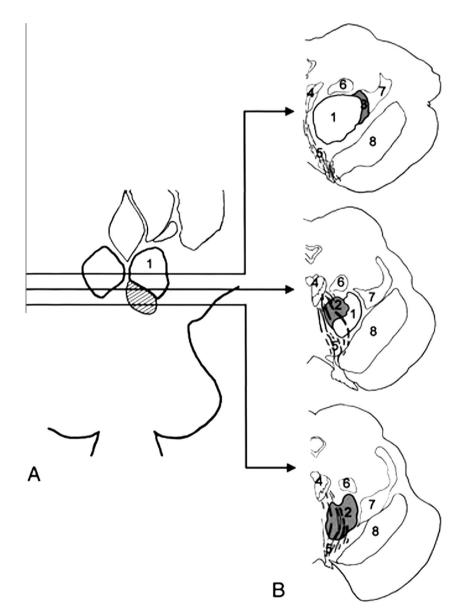


Fig. 1. Axial diffusion weighted MRI. (A, B) Patient 1. Images through the level of the red nucleus (A) and immediately below, showing the involvement of the superior cerebellar peduncle (B). (C) Patient 2. Image through the level of the caudal midbrain, below the red nucleus.



**Fig. 2.** Relationship of the oculomotor fascicles to the superior cerebellar peduncles. (A) Coronal diagram with the localization of Claude's syndrome (hatched area). (B) Axial diagrams show cerebellar efferent fibers (grey colored-area). Red nucleus (1), superior cerebellar peduncle (2), dentatothalamic tract (3), oculomotor nuclei (4), and oculomotor nerve fascicles (5). Reproduced with permission from Seo et al. Neurology 2001;57:2304–7.

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the red nucleus and third nerve fascicle, it is neither necessary nor sufficient for the red nucleus to be involved to result in the clinical abnormalities associated with the syndrome. Most examples of Claude's syndrome, they note, involve lesions of the postdecussation superior cerebellar peduncle immediately inferior to the red nucleus (Fig. 2). Furthermore, these lesions, more often than not, spare the more rostral pupillomotor fibers, as did the lesion in our patients (Fig. 2A). Tokunaga et al. [6] report a case similar to our Patient 2 and only found two additional cases in the literature of a Claude's syndrome with ipsilateral ataxia. They used tractography to determine that their patient's lesion was at the "rostral extremity of the decussation touching the dorsal side" which would lead to damage of fibers prior to their decussation leading to the ipsilateral ataxia (Fig. 2B). The pupillary light response was normal in both additional cases.

Several important points can be learned from these two patients. Though most pupil-sparing third nerve lesions are peripheral, a midbrain lesion should be considered. Superior rectus involvement of the contralateral eye in Patient 1 indicates nuclear involvement as the superior rectus subnuclei send afferent axons contralaterally and through the contralateral superior rectus subnuclei leading to bilateral deficits in the superior recti in unilateral midbrain lesions. In both cases the presence of hemiataxia and oculomotor deficits suggested localization to the midbrain, but the lack of pupillary involvement led to initial diagnostic uncertainty. We present, to our knowledge, the fourth report of what we refer to as a reverse Claude's syndrome: oculomotor palsy with ipsilateral cerebellar ataxia.

#### **Conflicts of Interest/Disclosures**

The authors declare that they have no financial or other conflicts of interest in relation to this research and its publication.

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