Mystery Case: Bilateral Claude syndrome

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Figure MRI of the brain

Bilateral paramedian mesencephalic stroke, hyperintense on diffusion-weighted imaging (A.a and B.a, with magnification in A.b and B.b) and hypointense in corresponding areas in the apparent diffusion-coefficient maps (C, D). Lesions involve the third nerves, Edinger-Westphal, and red nuclei, superior cerebellar peduncle, and the reticular activating system, sparing the crura cerebri and tectum.

A 59-year-old man with hypertension, hyperlipidemia, and type 2 diabetes presented with sudden-onset, bilateral ptosis, ataxia, and confusion. Examination showed bilateral ophthalmoparesis sparing minimal adduction and abduction, poorly reactive and dilated pupils, and global ataxia (video). The oculoccephalic reflex revealed normal bilateral abduction. Bell phenomenon and convergence were absent. MRI brain showed bilateral paramedian mesencephalic ischemic stroke (figure). Here in a bilateral variant, clinical presentation and imaging closely follow the description of Henri Claude's first case,¹ where pathologic examination revealed a paramedian midbrain stroke involving the superior cerebellar peduncle, red nucleus, and medial longitudinal fasciculus (MLF).¹

Third nuclei and MLF lesions likely cause ptosis and vertical and adduction deficits, while injury to fronto-pontine horizontal gaze pathways passing through the midbrain may explain the

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Table Historical overview of brainstem syndromes

	Syndrome				
	Weber	Claude	Nothnagel	Benedikt	Parinaud
Author describing syndrome, year of publication	Hermann David Weber, 1863	Henri Claude, 1912	Carl Wilhelm Hermann Nothnagel, 1879	Moriz Benedikt, 1889	Henri Parinaud, 1883
Clinical presentation	Oculomotor palsy, contralateral hemiparesis	Ptosis, ophthalmoplegia, contralateral dysdiadochokinesis/ataxia	Bilateral oculomotor palsies, ataxia, nystagmus	Oculomotor palsy, contralateral hemiparesis, tremor	Paralysis of upward gaze and accommodation, fixed pupils
Anatomical structures involved	Third nerve nucleus, corticospinal tract (crus cerebri)	Third nerve fascicle, red nucleus, superior cerebellar peduncle	Superior and inferior colliculi	Third nerve fascicle, red nucleus, cerebral peduncle	Dorsal midbrain

impaired abduction.² Eponyms of brainstem syndromes are rarely used, because of overlap among the historical descriptions (table).

Author contributions

J. Witsch: clinical care of the patient, study concept, making of figure, editing of video, writing of the manuscript. R. Narula: clinical care of the patient, making of video, revision of the manuscript. H. Amin: clinical care of the patient, revision of the manuscript. J. Schindler: clinical care of the patient, study concept, acquisition of data, revision of the manuscript.

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Mystery Case responses

The Mystery Case series was initiated by the *Neurology*[®] Resident & Fellow Section to develop the clinical reasoning skills of trainees. Residency programs, medical student preceptors, and individuals were invited to use this Mystery Case as an educational tool. Responses were solicited through

a group email sent to the American Academy of Neurology Consortium of Neurology Residents and Fellows and through social media.

Forty-six percent of respondents correctly localized the lesion to the paramedian mesencephalic region. Similarly, 48% of respondents correctly identified the nuclei involved to cause the particular constellation of symptoms described. Only 8% of respondents correctly identified large artery disease as the most common cause of mesencephalic stroke.¹ Four respondents, including 1 medical student, answered all 3 questions correctly.

This mystery case illustrates Claude syndrome, which is a brainstem stroke syndrome characterized by ptosis, ophthalmoplegia, dysdiadochokinesis, and ataxia first described secondary to damage to the third nerve fascicle, red nucleus, and superior cerebellar peduncle. As the authors point out, historical eponyms for brainstem strokes can be confusing due to overlapping clinical presentations. The similarities among syndromes arise from the close anatomical location of these nuclei within the brainstem. Thus, lesion localization within the brainstem requires interpretation of both clinical symptoms and imaging findings combined with detailed knowledge of cranial nerve pathways.

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