Claude syndrome

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A 53-year-old man presented with drooping of the right eyelid and swaying during walking since morning. He had difficulty in reaching for objects with the left hand. He noticed diplopia when he opened his right eye. There was no history of dysarthria, weakness, or numbness. There was no history of headache, vomiting, fever, breathlessness, palpitation, or pedal edema. He has been a diabetic for the past 8 years. He had acute coronary syndrome 2 days before the current symptoms and underwent coronary angiography with stenting 1 day before his neurological symptoms. On examination, blood pressure was 110/80 mmHg and pulse rate was 80/min and was regular. Neurological examination showed complete right ptosis with impaired elevation, depression, and adduction in the right eye. Right eye intorsion was preserved. There was no nystagmus. Pupils were equal and reactive to light and accommodation. The left eye movements were normal. There was no facial deviation or dysarthria and motor power in both upper and lower limbs was normal. His deep tendon reflexes were sluggish bilaterally. The sensory system was normal including vibration and joint position sense. He had left-finger nose incoordination and toe-finger incoordination. He swayed to the left while walking. His right fascicular third nerve palsy with left cerebellar signs suggested Claude syndrome. His electrocardiogram showed ST-T changes and his echocardiogram showed regional wall motion abnormalities with normal ejection fraction. There was no atrial or ventricular thrombus. Magnetic resonance imaging (MRI) of the brain showed acute infarct in the right ventral paramedian midbrain sparing cerebral peduncle (Figs. 1 and 2). Cerebral magnetic resonance angiogram showed right proximal posterior cerebral artery (P1) stenosis (Fig. 3).

Claude's syndrome is a very rare midbrain syndrome characterized by ipsilateral oculomotor nerve palsy and contralateral cerebellar ataxia. Henri Claude described a house painter who developed right third cranial nerve palsy with contralateral ataxia. The pathological examination revealed a paramedian mesencephalic infarction on the right involving the superior cerebellar peduncles and the medial half of the red nucleus [1]. Only a few cases have been reported since Claude's original description in 1912. The condition occurs by simultaneous involvement of the cerebellar efferent fibers to the thalamus and the oculomotor nerve fascicles. The red nucleus often has been suggested to be the lesion site responsible for Claude's syndrome, but the precise localization of the syndrome is uncertain [2]. Recent MRI-based studies showed that the lesion responsible for Claude's syndrome is in the superior cerebellar peduncle, below the red nucleus [3]. Oculomotor nerve palsy is often partial. The oculomotor nerve fascicles are widely separated in the midbrain, both rostral to caudal and medial to lateral, before they exit at the interpeduncular fossa. Therefore, it is not surprising that a small lesion involving the superior cerebellar peduncle produces a partial involvement of the fascicles. The invariable appearance of medial rectus palsy indicates that the fibers of the medial rectus pass through the superior cerebellar peduncle just below the red nucleus. Pupillary fibers run the most rostral course and are often spared [4].

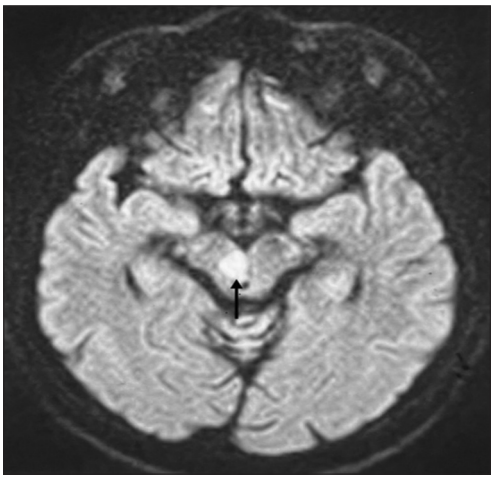

Figure 1: Magnetic resonance imaging brain axial diffusion-weighted image showing restricted diffusion in right ventral paramedian midbrain sparing cerebral peduncle

The most common cause of Claude's syndrome is cerebrovascular disease and malignancy. Claude's syndrome is most often related to occlusion of perforating arteries derived from the pre-communal segment (P1) of the posterior cerebral artery, in the setting of a small vessel disease. It is also described

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Table 1: Fascicular III nerve syndromes

Fascicular III nerve syndromes	Structures involved	Clinical findings
Weber syndrome	III nerve, cerebral peduncle	Ipsilateral III nerve palsy, contralateral hemiparesis
Benedict syndrome	III nerve, red nucleus, cerebral peduncle	Ipsilateral III nerve palsy, contralateral hemiparesis, contralateral cerebellar ataxia or Holmes tremor or choreoathetosis
Claude syndrome	III nerve, superior cerebellar peduncle	Ipsilateral III nerve palsy, contralateral ataxia

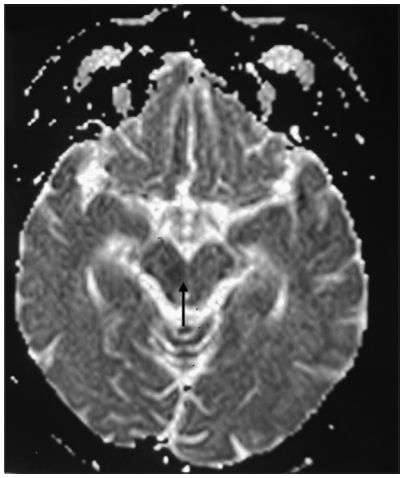

Figure 2: Magnetic resonance imaging brain axial apparent diffusion coefficient image showing hypointensity in right paramedian midbrain

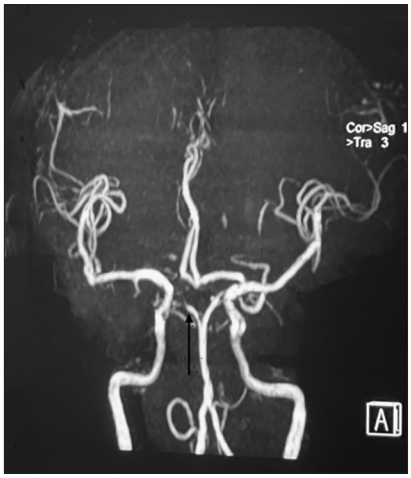

Figure 3: Cerebral magnetic resonance angiogram showing right P1 stenosis

in cardioembolic stroke and large vessel disease [5]. It can also occur in persons with low atherosclerotic cardiovascular disease scores [6]. It is also reported in neurosarcoidosis [7], neurocysticercosis [8], and neurosyphilis [9]. Eponymic oculomotor fascicular syndromes include Weber syndrome, Benedikt syndrome, and Claude syndrome [Table 1]. Other midbrain syndromes are Nothnagel syndrome, Parinaud syndrome, and Wernekink commissure syndrome.

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