Case Report

Claude’s Syndrome: A Case Report of Rare Midbrain Syndrome from Bangladesh

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Abstract
Claude’s syndrome is a rare brainstem syndrome with ipsilateral third nerve palsy and contralateral ataxia. Here we present a case of a 60-year-old male with Claude’s syndrome due to cerebral infarct. The patient had left-sided pupil sparing third nerve palsy with right-sided ataxia. MRI of the brain revealed infarct in the left half of the tegmentum of the midbrain. MRA of the brain was unremarkable. The patient improved gradually and was almost asymptomatic after one month. Though the syndrome was first described a century ago, it warrants particular attention from clinicians due to its rarity and atypical features.

Key words: Claude’s syndrome, Midbrain syndrome, Brainstem syndrome, Bangladesh, Case report.

Introduction
The brainstem is a small but vital part of the brain containing important cranial nerve nuclei and other essential structures. Due to its complex and compact anatomical structures, brainstem syndromes create great interest among clinicians. Claude’s syndrome is one of the rare midbrain syndromes with very few reported cases since its first description from Henry Claude.¹ It has a unique presentation of ipsilateral third nerve palsy with contralateral cerebellar signs. This syndrome may occur due to multiple etiologies. Stroke is the most typical cause, but it may also occur due to neurocysticercosis, cryptococcal meningitis, or neurosyphilis.²⁻⁴ We are reporting this case due to its academic importance and rarity. To our best knowledge, this is the first reported case of Claude’s syndrome from Bangladesh.

Case Report
A 60-year-old male was admitted to the neurology ward of Combined Military Hospital, Dhaka, on February 17, 2021, with sudden onset drooping of left eyelid, difficulty in walking and slurring of speech for four days. The patient also had vertigo and sense of imbalance but had no limb weakness, dimness of vision or difficulty in swallowing. He needed assistance during walking due to tendency to fall. Also, he needed assistance while doing daily activities with his right hand due to incoordination. He was a known case of hypertension and diabetes mellitus for five years but did not take medications regularly. On examination, her pulse was 64/min and blood pressure 140/90mm of Hg. Other general parameters were unremarkable. He was conscious, oriented. The speech was slurred, but there was no definite scanning of speech. The patient had pupil sparing third nerve palsy on the left side. There was horizontal gaze-evoked nystagmus with direction towards the right side. Fundoscopy revealed pre-proliferative diabetic retinopathy. Other cranial nerves were intact. Muscle bulk, tone and power were normal. All jerks were diminished to absent. Cerebellar tests were positive on the right side, including intention tremor, dysdiadochokinesia, rebound phenomenon and heel-shin test. He also had sensory impairment of all modalities in gloves and stockings pattern. The gait was atactic with a tendency to fall towards the right side. Other systemic examinations were normal.

CT scan of the brain excluded hemorrhage and the patient was treated with aspirin, atorvastatin, ramipril and insulin. Risk factors evaluations were done, ECG & echocardiography were normal. Fasting blood sugar (12.9mmol/L, normal range: 3.33-6.11mmol/L) and 2 hours after breakfast sugar (17.3mmol/L, normal range: <7.8mmol/L) were uncontrolled. HbA1C was 10.7% (4.2-6.4%). Urine routine examination showed significant glycosuria and proteinuria. Fasting serum lipid profile revealed total cholesterol 197mg/dl (150-220mg/dl), triglycerides 229 mg/dl (<150mg/dl), HDL cholesterol 36mg/dl (>35mg/dl) and LDL cholesterol 115mg/dl (<130mg/dl). Other routine investigations were normal. MRI of the brain confirmed the diagnosis which revealed an acute infarct in the left half of the tegmentum of the midbrain (Figure-1,2,3,4). However, the MRA of the brain did not any significant stenosis in the posterior circulation (Figure-5). With treatment, he improved gradually. He was discharged with final diagnoses of

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Claude’s syndrome (left), hypertension, diabetes mellitus with diabetic retinopathy, nephropathy and neuropathy. After one month, third nerve palsy recovered completely and as ataxia improved significantly the patient could walk independently.

Figure-1: Axial diffusion-weighted (DW) MR image of the brain showing hyper intense signal in the left half of tegmentum of midbrain medial to the left red nucleus

Figure-2: Axial apparent diffusion coefficient (ADC) mapped MR image of the brain showing a drop of hyperintense signal found in the DW MR image (Figure-1) in the left half of tegmentum of midbrain medial to the left red nucleus, implying restricted diffusion

Figure-3: Axial T2 weighted MR image of the brain showing hyperintense signal in the left half of tegmentum of midbrain medial to the left red nucleus

Figure-4: Sagittal T2 weighted MR image of the brain showing hyperintense signal in the tegmentum of midbrain below red nucleus

Figure-5: MRA of the brain showing normal study
Discussion
Midbrain is a vital structure of the brain that may be affected by different brainstem syndromes with widespread clinical variations. Claude’s syndrome is one of these syndromes which is relatively rare with atypical presentations of ipsilateral third nerve palsy with contralateral ataxia. This syndrome was first described in 1912. It occurred due to various lesions involving the dorsomedial midbrain. Infarction is the commonest cause. It occurred mostly due to atherosclerosis of tiny arteries supplying the midbrain. Hypertension is the main risk factor leading to these small vessels occlusion. It was also reported that posterior cerebral artery occlusion might also lead to this syndrome.

Third nerve palsy occurs due to the involvement of oculomotor nuclei or fibers. It is one of the rare causes of oculomotor nuclear lesions. Due to complex structures of the oculomotor nuclei, presentations may vary from case to case. In nuclear lesions, contralateral partial third nerve palsy also occurs but maybe subtle and usually overlooked. The pupil may be involved due to the involvement of the Edinger Westphal nucleus. Other ocular cranial nerves like the trochlear nerve may also be affected due to the involvement of medial longitudinal fasciculus. In the cases of the fascicular lesions, only ipsilateral third nerve palsy was found. In this case, he had partial ptosis in the left side with involvement of other muscles supplied by the third nerve, sparing pupil, with the unaffected right eye indicating oculomotor fascicular lesion.

This syndrome is one of the rare clinical scenarios causing the positive contralateral cerebellar signs. Contralateral ataxia occurs due to the involvement of the red nucleus or superior cerebellar peduncle. Though lesions at the red nucleus level were considered the key for ataxia in Claude’s syndrome, now it is thought that lesion of the superior cerebellar peduncle just below and medial to the red nucleus causes ataxia in this syndrome. In this case also, the red nucleus was spared, involving only the superior cerebellar peduncle. Rarer variety like Claude’s syndrome without ptosis may also occur. Sometimes atypical presentation like bilateral Claude’s syndrome due to bilateral paramedian mesencephalic ischemic stroke was also reported. In case of a larger structural lesion, the patient may also present with Claude syndrome ‘plus’. Due to the complex arrangement of cerebellar fibers at this level, Claude’s syndrome with ipsilateral ataxia was also reported.

MRI is the investigation of choice, as CT scan may not reveal the lesion properly. Case series and literature review revealed a lesion in the medial tegmental area below the red nucleus causing this syndrome. We have also found infarct in the same area. MRA of the brain is usually normal like our case as the responsible anteromedial arteries from the interpeduncular fossa are not visualized. Rarely MRA abnormalities with larger artery involvement like posterior cerebral artery occlusion may be found.

Besides infarct, hemorrhage, neoplastic lesion, tuberculosis, neurocysticercosis, or neurosyphilis may also lead to this condition. Regardless of the etiologies, this syndrome usually has a good prognosis with complete or near-complete recovery in almost all cases. Our patient also recovered almost completely after one month. Still, it is an important clinical syndrome due to its unusual presentation and requires special attention for proper diagnosis.

Conclusion
Despite rare incidence, Claude’s syndrome represents a great challenge for clinicians due to its unusual clinical presentation generating confusion. It is essential to correlate anatomical, clinical and radiological skills for proper diagnosis and formulate an appropriate management approach.

References