CLAUDE SYNDROME: A CASE REPORT

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ABSTRACT

Introduction: **Detailed description of infarction of the midbrain is sparse likely due to the complex arterial supply of this region of the brain. Among the ventral midbrain syndromes reported is Claude syndrome. This syndrome caused majorly by a vascular insult to the ventromedial midbrain, characteristically presents with ipsilateral third cranial nerve palsy and contralateral hemiataxia. It is a rare syndrome and only a few cases have been reported since 1912 when it was first described by Henri Claude.**

Case presentation: **A 45-year-old male, who developed sudden onset dysarthria, right third cranial nerve palsy and left sided ataxia. An infarct in the right ventromedial midbrain was revealed on magnetic resonance imaging of the brain.**

Conclusion: **We describe a case report of a middle-aged man with minimal vascular risk factors (ASCVD = 1.3%) for stroke, who presented with features suggestive of Claude syndrome.**

Keywords: Claude syndrome, Midbrain infarction, ASCVD

INTRODUCTION

The midbrain also known as the mesencephalon is the division of the brainstem linking the pons and cerebellum with the forebrain and is divided into a ventral part, the tegmentum, and a dorsal part, the tectal or quadrigeminal.[1] Midbrain infarction though rare can be caused by the obstruction of its blood supply and is responsible for about 0.6% of the total patients admitted. Most case are described in individuals with high cardiovascular risk (ASCVD >7.5%)with ischemic stroke.[2] The midbrain receives its blood supply from the paramedian mesencephalic branches (basilar), peduncular perforating branches (posterior cerebral artery) as well as from the superior cerebellar artery and the choroidal arteries..[3] Studies have shown that infarction of the midbrain is 10-fold more likely to be accompanied by ischemia of neighbouring structures[4] The constellation of signs and symptoms caused by the lesion of these structures give rise to syndromes of both ventral and dorsal midbrain origins. Among the ventral midbrain syndromes is Claude syndrome. This syndrome comes in various forms which include affectation of the fibers from CN III, the rubrodentate fibers, corticospinal tract fibers and corticobulbar fibers.[5] The manifestation of this syndrome depends on the extent of the lesion to the structures involved and can present as ipsilateral CN III palsy, contralateral ataxia with or without contralateral hemiplegia of lower facial muscles, tongue, shoulder, upper and lower limb along. The blood supply involved is from the posterior cerebral artery.[6] Detailed description of this syndrome is sparse, with few studies on the imaging findings of the site of lesion and the whole spectrum of clinical manifestation involved. Other ventral midbrain syndromes described are Benedikt, Weber, Nothnagel syndrome and Parinaud syndrome which is a dorsal midbrain syndrome

Benedikt syndrome, also called Paramedian midbrain syndrome is caused by an insult in the tegmentum of the midbrain and cerebellum. It is characterized by the presence of oculomotor nerve palsy and cerebellar ataxia including tremor and involuntary chore athetotic movements.

Weber syndrome presents with ipsilateral oculomotor nerve palsy with contralateral hemiparesis. It occurs due to the involvement of oculomotor fascicles in the interpeduncular cisterns and cerebral peduncle

Nothnagel syndrome is a rare midbrain syndrome that involves the tectum of the midbrain and superior cerebellar peduncles. Classically, the syndrome involves the oculomotor nerve fascicles and superior cerebellar peduncle, leading to ipsilateral CN III palsy and limb ataxia respectively[7]

Parinaud syndrome a dorsal midbrain syndrome is classically described by the triad of impaired upward gaze, convergence retraction nystagmus, and pupillary hyporeflexia.[8]

We herein describe a case of a patient with low ASCVD score who presented with clinical features and neuroimaging findings suggestive of Claude syndrome.

CASE PRESENTATION

We report the case of a 45-year male with minimal vascular risk factor who presented in our facility about a year and six months ago with sudden onset of dysarthria, double vision and in coordination of the left side of the limb. Before the onset of these symptoms, he was in good health and was not previously known to be hypertensive or diabetic.

He was taking a walk on the faithful day and noticed sudden onset unsteadiness of gait and in coordination of the left hand which was accompanied with double vision and slurring of speech as he attempted to talk. He described a dizzy feeling just prior to the onset of the symptoms and was subsequently brought to our facility with the help of some attendants following the incident. He also noticed that the double vision progressed over a couple of hours to drooping of the right eye. There was no history of associated pain, redness or swelling over right eye. No history of trauma prior to the onset of symptoms. Symptoms were not accompanied by headache, facial deviation, sensory symptoms, dysphagia or hemi body weakness. No constitutional symptom or major organ decompensation was noted. Patient does not smoke or drink or use illicit substances.

On examination, he was alert, conscious and oriented to time, place and person. BP was 130/80 mm of Hg. On neurological examination, normal higher mental functions and absent meningeal signs were noted. He was dysarthric, and he swerved to the left when attempting to walk. Cranial nerves examination revealed right sided 3rd nerve palsy manifesting as right eye ptosis and anisocoria (right pupil was 5mm and left pupil of 2mm). Other cranial nerves were normal. Motor system revealed normal muscle bulk and tone; no pyramidal weakness in all limbs and no involuntary movements. Reflexes were normal with bilaterally flexor plantar response. No sensory abnormality was noted. Cerebellar signs were present on left characterized by dysmetria, dysdiadochokinesia/ heel to knee test and tendency to fall to the left. Fundus examination was normal.

Calculated ASCVD score was 1.3%.

Fasting blood sugar level was 110 mg/dl, HbA1c 5.9%, HbA1c 1FCC 40.9. Lipid profile: total cholesterol-150mg/dl, triglyceride-160mg/dl, HDL-50mg/dl, LDL-70mg/dl. The Liver, renal function, lipid study, complete blood count, urinalysis, coagulation profile, and electrolyte test results were all within reference ranges. Electrocardiographic and echocardiographic findings revealed no abnormality. T2 weighted Magnetic Resonance Imaging (MRI) Brain, revealed an acute infarct in the right ventromedial area of the midbrain.

Figure 1: Extraocular muscle involvement in the subject with claude syndrome

After confirmation of the lesion on MRI. The patient was administered aspirin 300mg in the emergency department and atorvastatin for secondary prevention The patient was discharged home after a 7-day hospital stay with improvement of clinical symptoms and signs

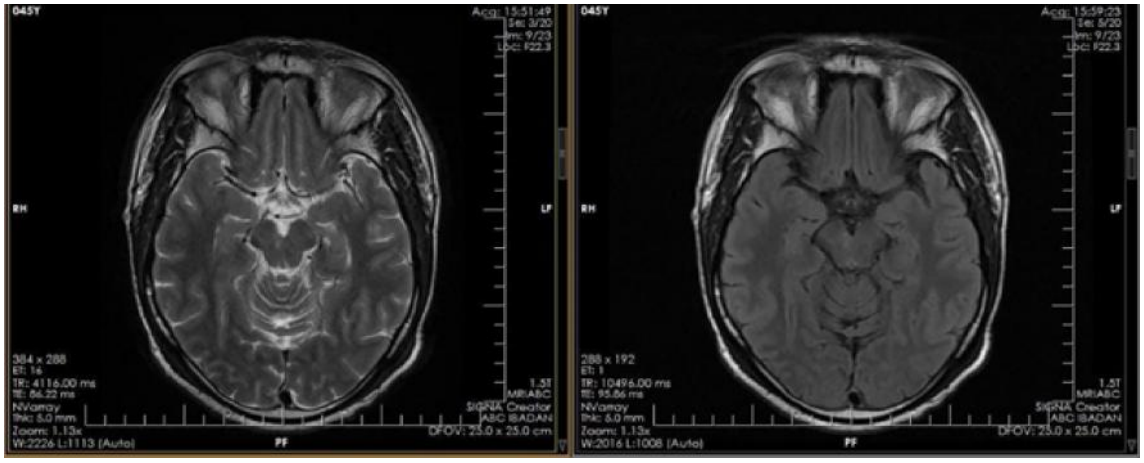

Figure 2: Axial brain MRI T2 weighted and FLAIR Image with the blue arrows showing a high signal intensity at the right ventromedial midbrain.

over this time. Discharge prescriptions included aspirin 300mg for an additional 1week, lifelong clopidogrel at 75mg and atorvastatin at 40mg daily according to nice guideline for secondary prevention. The patient presented for outpatient clinic 2 weeks following discharge and was found to have made a sustained and remarkable improvement

DISCUSSION

The midbrain, a structure that originates in the fourth week of gestation, confers the passage of multiple structures and consequently acts as a relay for sensory-motor information. Its anterior boundary is given by the crus cerebri and the interpeduncular fossa, while the posterior limit is characterized by the presence of the superior and inferior colliculi.[9] Alterations in these structures lead to the appearance of midbrain syndromes, which manifests as a subset clinical symptoms and signs that make their diagnosis unique and maintains great clinical importance as a result of potential intervention by new endovascular techniques considering that vascular causes are potentially intervenable.

A syndrome of such clinical importance, was described by a French psychiatrist and neurologist Henri Claude (1869-1945) who described a case of unilateral oculomotor palsy of midbrain origin with contralateral ataxia in a house painter. In his original report, there was pupillary involvement in the form of a dilated and fixed pupil and impaired convergence of both eyes. His patient had a paramedian mesencephalic infarction with involvement of superior cerebellar peduncle, medial half of red nucleus, and medial longitudinal fasciculus and oculomotor nerve fascicles.[10] After several years of this description, Claude syndrome has been a subject of rare case reports, with some disagreement over the precise localization of the lesion.

Our case clearly reveals a correlation between ischemia in the ventromedial midbrain infarction confirmed by MRI of the brain and the clinical presentation of Claude syndrome. More interestingly, our case is seen in an individual with a low vascular risk factor (ASCVD 1.3%). Most cases of this cerebrovascular event rate were 3 times higher among patients with a history of ASCVD who met the definition of very high risk in the 2018 AHA/ACC blood cholesterol guideline than among their counterparts who did not meet this definition [11] this is in contrast with our index patients. The patient presented with right fascicular 3rd nerve palsy with a fixed, dilated pupil with impaired convergence and complete ptosis. There was also the presence of contralateral ataxia. Magnetic resonance imaging revealed a paramedian infarction of the ventral midbrain at the level of the superior colliculus, which confirms the clinical symptoms and signs as affectation of the 3rd nerve fascicle and the red nucleus (cerebellorubral tract) would explain the ophthalmoplegia, papillary abnormalities and the contralateral ataxia respectively. As the contralateral hemiataxia was originally explained to be from the involvement of the red nucleus, other studies suggested that involvement of the superior cerebellar peduncle gives rise to the contralateral ataxia seen in this syndrome.[12]

CONCLUSION

It is also worthy of note that this case classically fits into the earliest case report by Henri Claude in 1912 and is unique in that it is the first case description of Claude syndrome in a patient with low ASCVD risk score (1.3%).

REFERENCES

1. **Sciacca S,** Lynch J, Davagnanam I, Barker R. Midbrain, Pons, and Medulla: Anatomy and Syndromes. RadioGraphics. 2019 Jul;39(4):1110–1125.
2. Atherosclerotic cardiovascular disease risk assessment: An American Society for Preventive Cardiology clinical practice statement - PMC [Internet]. [cited 2023 Jul 14].
3. **Aoki Y,** Hashimoto T. Paramedian midbrain infarction presenting with bilateral ptosis and unilateral median longitudinal fasciculus syndrome: A peculiar midbrain syndrome. Case Rep Neurol. 2022;14:197–201.
4. **Pj M,** Hm C, Lr C. Midbrain infarction: associations and aetiologies in the New England Medical Center Posterior Circulation Registry. J Neurol Neurosurg Psychiatry [Internet]. 1998 Mar [cited 2022 Nov 28];64(3).
5. **Adachi T,** Takagi Y. [A case of left third nerve palsy and contralateral vertical gaze palsy with medial midbrain infarction]. Rinsho Shinkeigaku. 2000 Aug;40(8):807–810.
6. **Gowda SN,** De Jesus O. Brainstem Infarction. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2022 [cited 2022 Nov 29]. Available from: http://www.ncbi.nlm.nih.gov/books/NBK560896/
7. **Ruchalski K,** Hathout GM. A Medley of Midbrain Maladies: A Brief Review of Midbrain Anatomy and Syndromology for Radiologists. Radiol Res Pract. 2012;2012:1–11.
8. **Feroze KB,** Patel BC. Parinaud Syndrome. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2023 [cited 2023 Jul 14].

9. **Gregory A.** Mihailoff., Duane E. Haines. Fundamental Neuroscience for Basic and Clinical Applications E-Book. 5th ed. Philadelphia: Elsevier; 2018. - Google Search [Internet]. [cited 2022 Nov 29].
10. **Sheetal.** Claude syndrome: A report of two cases and review of literature [Internet]. [cited 2023 Jan 3]. Available from: https://www.mdcan-uath.org/ 2018;7;(11); 29-31.
11. **Colantonio LD,** Shannon ED, Orroth KK, *et al.* Ischemic Event Rates in Very-High-Risk Adults. J Am Coll Cardiol. 2019 Nov 19;74(20):2496–507.
12. **Song TJ,** Suh SH, Cho H, Lee KY. Claude's Syndrome Associated with Neurocysticercosis. Yonsei Med J. 2010 Nov 1;51(6):978–979.