

Claude's Syndrome associated with Neurosyphilis: Case Report

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Abstract

The alterations of the midbrain represent a great clinical challenge due to their anatomical location, it confers the passage of multiple tracts between the cerebral cortex and other subcortical structures such as cerebellum, pons and bulb, responsible for sensory-motor and autonomic control, which implies the difficult comprehension of clinical manifestations in the primary medical context, which delays the diagnosis and an adequate therapeutic approach. This article aims to make a clinical anatomical radiological correlation of the dorso-medial syndrome on the midbrain under the context of a patient diagnosed by meningovascular syphilis.

Keywords: Midbrain, neuroimaging, syndromes, brainstem, neurosyphilis

The midbrain is the most cephalic portion of the brainstem, extending from the pontomesencephalic junction to join the diencephalon. Its anterior limits are given by the crus cerebri and the interpeduncular fossa, while at a posterior level it is characterized by the presence of the superior and inferior colliculi (1). It contains important fibers that allow the transmission of information between the cerebral cortex, the cerebellum, the pons and the medulla, for which it participates in the processing of auditory and visual information, and movement control (2). Therefore, it is important that the clinician is familiar with the external and internal configuration of the midbrain to facilitate the location of structures that can be injured and identified in neuroimaging techniques.

At a clinical level, lesions of the midbrain, as in all structures of the brainstem, can be identified by the presence of cross-over syndromes, where ipsilateral cranial nerve alterations are evidenced that originate at this level associated with contralateral sensory or motor alterations (3). According to the literature reviewed, the main etiologies to be identified in brain stem syndromes are vascular, infectious, compressive, demyelinating and traumatic lesions, which give rise to syndromes such as Weber, Benedikt, Parinaud, Nothnagel and finally Claude, as the case may be. which will be described below (Semiological description of the midbrain syndromes - See table 1).

Case Report:

A 45-year-old man living on the street who consulted for a sudden onset of symptoms characterized by hemiparesis associated with dysidiadochokinesis and left hemiataxia with horizontal diplopia. Due to the limitation of adduction of the right eye, on admission with TA 100/60, fasting blood glucose 89 mg / dl, negative urine toxic tests, skull CT showing greater atrophy than expected for age, with no other detectable lesions, lipid profile within normality and a serology by RPR technique: POSITIVE (1: 256 DILS), FTA ABS: positive, HIV: negative.

A simple and contrasted brain MRI was performed (Figure. 1) showing an image that restricts diffusion in the dorsomedial region of the midbrain that, associated with the clinic, configures a Claude syndrome, in the context of a patient without associated cardiovascular risk factors, lumbar puncture (LP) was performed finding: leukocytes 8 / microL, proteinorrachia: 38 mg / dl, glucose index 0.7 mg / dl, india ink: negative, VDRL: reactive. Compromised CNS due to syphilis was confirmed, causing a vasculitic phenomenon in the territory of the perforating branches of the posterior cerebral artery (PCA). Treatment was started with crystalline penicillin 24 million units / day in continuous infusion to improve CNS penetration considering the pharmacodynamics and pharmacokinetics, obtaining a 90% improvement in symptoms at 6 months.

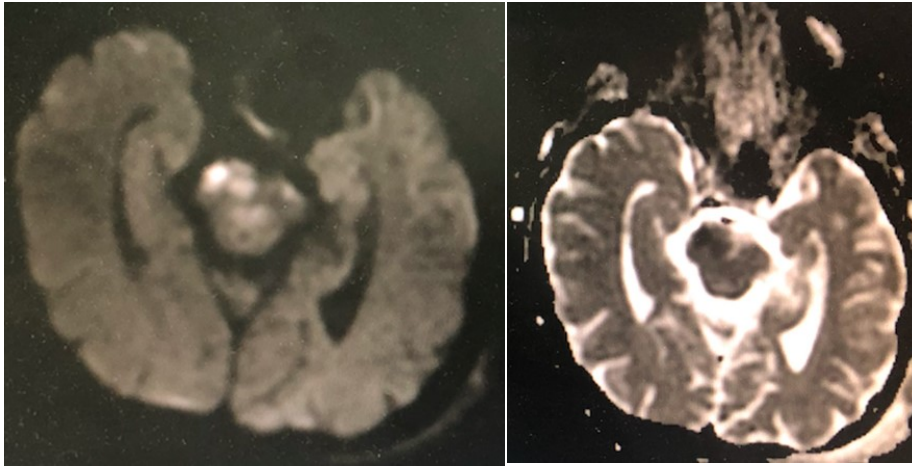


Figure 1. Image A (Diffusion DWI: hyperintense image that restricts diffusion in the right mesencephalic peduncle. Image B (ADC map shows hypointense image in the right mesencephalic peduncle).

Discussion:

The midbrain is one of the structures originated in the fourth week of gestation. It confers the passage of multiple structures and acts as a relay for sensory-motor information.

Alterations in this lead to the appearance of midbrain syndromes, which present a set of clinical manifestations that make their diagnosis difficult and despite its relatively low incidence maintains great clinical importance, considering that vascular causes are potentially interventible by means of new endovascular techniques.

Pathologic alterations in this level lead to the appearance of midbrain syndromes, which present a set of clinical manifestations that could make their diagnosis difficult and despite its relatively low incidence maintains great clinical importance, considering that vascular causes are the main etiology which can potentially be interventible by means of new endovascular techniques (1).

The clinical case presented has a very low frequency, since most of these syndromes are caused by primary vascular lesions, corresponding to 2.3% due to ischemia in nearby structures and 0.6% at the midbrain level (4). However, within the diagnostic approach of midbrain syndromes, traumatic, demyelinating, compressive and infectious lesions must be considered. According to the above, a clinical case of neurosyphilis related to a vasculitic phenomenon was presented, which compromised the dorsomedial territory of the midbrain.

Regarding syphilis, it is triggered by *Treponema pallidum* sub *pallidum*, which can remain dormant for years before becoming active in an adult. One of its complications corresponds to the involvement of the central nervous system, occurring in less than 10% of patients. During early stages, both the CSF and the meninges, as well as the vasculature, are affected, while the late forms involve the parenchyma of the spinal cord and the brain (5). The involvement of the blood vessels by the spirochete generates the appearance of vasculitis, associated with meningitis, which corresponds to one of the complications of greatest interest in clinical practice, reported in approximately 38.5% to 54.5% of all patients. cases of neurosyphilis (6). These vasculitic phenomena are more prevalent in the elderly, generally related to infarcts at the level of small, medium and large vessels, producing nonspecific clinical pictures, making diagnosis difficult (5).

On the other hand, despite the few records of neurosyphilis associated with vasculitis in young patients, this has become one of the main causes of early mortality in this age group (7). Therefore, it is recommended that all patients with stroke of unknown origin, get serology for syphilis.

The vasculitic phenomenon generated in this patient at the level of the perforating branches of the posterior cerebral artery generated involvement in the dorsomedial midbrain region, configuring the Claude syndrome, which is commonly confused with the Claude-Bernard-Horner syndrome. They differ in terms of the injured structures, where both the red nucleus and the nucleus of the oculomotor nerve ipsilateral to the injury are affected in the first, while in the second fascicles of the III and sympathetic nerve are affected together with structures adjacent to the site of the lesion, commonly without affecting red nuclei, producing additional clinical manifestations such as palpebral ptosis and anhidrosis (8).

Ultimately, it corresponds to a lesion on the dorsomedial aspect of the midbrain, typically caused by infarcts at the level of the perforating branches of the posterior brain, although it is equally associated with hemorrhagic and tumor causes (9). It manifests as ipsilateral paralysis of the oculomotor nerve with contralateral weakness of the upper rectum, sometimes pain, alterations in pupillary response (miosis), lack of coordination and cerebellar hemiataxia of the contralateral upper and lower extremities (2) (9). as evidenced in the clinical manifestations of our patient. However, there are some syndromes that can present incompletely.

Table 1. Midbrain syndromes description.

SYNDROME	LOCATION	STRUCTURES AFFECTED	CLINICAL MANIFESTATIONS
Weber	Ventromedial region	Cerebral peduncle and the ipsilateral tracts of the oculomotor nerve	Ipsilateral third nerve palsy, diplopia, ptosis, afferent pupillary defect, contralateral paralysis (peduncles), contralateral parkinsonian rigidity (substantia nigra)
Benedikt	Superior Colliculus	Red nucleus, superior cerebral peduncle, oculomotor tracts and inferior oculomotor nucleus	Ipsilateral cranial nerve III palsy, contralateral hemiataxia with intentional tremor, contralateral hemiparesis, and tendon reflexes hyperactive.
Parinaud	Superior Colliculus	Quadrigeminal tubercles	Classic triad: ascending paralysis, convergence retraction nystagmus, and pupillary hyporeflexia
Claude	Midbrain dorsomedial	Red nucleus and nucleus of the ipsilateral oculomotor nerve	Ipsilateral paralysis of the oculomotor nerve with contralateral weakness of the superior rectus muscle, cerebellar hemiataxia of contralateral upper and lower extremities.
Nothnagel	Quadrigeminal plate and superior cerebellar peduncle	Quadrigeminal tubercles and communicating fibers of the superior cerebellar peduncle	Unilateral or bilateral oculomotor nerve palsy and ipsilateral cerebellar ataxia.
Wernekinck	Wernekinck commissure	Communicating fibers of the superior cerebellar peduncle (dento-rubro-thalamic tracts such as dento-rubro-olivary tracts)	Bilateral cerebellar ataxia involving superior and inferior limbs, ocular alterations (nystagmus, diplopia) and palatal tremor.

Conclusion:

Despite their low incidence, syndromes at the midbrain level represent a great clinical challenge due to their varied clinical manifestations that can generate confusion at the time of clinical analysis in the context of primary medical care, for which it is important to take into account the anatomical correlation radiological, added to differential diagnoses that allow a timely diagnosis and an adequate therapeutic approach.

Within the diagnostic approach, the main etiologies such as vascular and infectious events must be recognized, the main cause being the primary vascular and syphilis within the infectious, since the risk of central nervous system affections is increased by up to 10% (10), which generates vasculitic phenomena that can trigger an ischemic event at the midbrain level.

Finally, it should be emphasized that those patients with meningovascular syphilis can cause infectious arteritis causing ischemia, which is why this should be one of the differential diagnoses in those patients with stroke without the presence of cardiovascular risk factors or those under 50 years of age.

Conflicts Of Interest:

The authors declare no conflict of interest.

References

1. Gregory A. Mihailoff., Duane E. Haines. *Fundamental Neuroscience for Basic and Clinical Applications E-Book*. 5th ed. Philadelphia: Elsevier; 2018.
2. Sciacca S, Lynch J, Davagnanam I, Barker R. Midbrain, Pons, and Medulla: Anatomy and Syndromes; Radio Graphics [Internet]. Pubs.rsna.org. 2020 [cited 4 September 2020].
3. Aminoff M, Daroff R. *Encyclopedia of the neurological sciences*. 2nd ed. San Diego: Elsevier; 2014.
4. Liu H, Qiao L, He Z, Wernekink commissure syndrome: a rare midbrain syndrome; Neurological Sciences [Internet]. Pubmed.ncbi.nlm.nih.gov. 2020 [cited 4 September 2020].
5. Feitoza L, Stucchi R, Reis F. Neurosyphilis vasculitis manifesting as ischemic stroke; Revista Sociedad Brasileira de Medicina Tropical [Internet]. Ncbi.nlm.nih.gov. 2020 [cited 4 September 2020].
6. Jiménez J, Ladino L, Uribe C, Guerra A, Ciro J, Hernández O, Ochoa J; Neurosífilis meningovascular con trombosis de la arteria basilar; Biomédica [Internet]. Scielo.org.co 2020 [cited 4 September 2020].
7. Shi M, Zhou Y, Li Y, Zhu Y, Yang B, Zhong L, Pan R, Yang D; Young male with syphilitic cerebral arteritis presents with signs of acute progressive stroke; Medicine (Baltimore) [Internet]. Ncbi.nlm.nih.gov 2020 [cited 4 September 2020].
8. Khan Z, Bollu P; Horner Syndrome; StatPearls [Internet]. Ncbi.nlm.nih.gov 2020 [cited 4 September 2020].
9. Tsuda H, Fujita T, Maruyama K, Ishihara M; Claude's Syndrome without Ptosis Caused by a Midbrain Infarction; Internal medicine (Tokyo, Japan) [Internet]. Pubmed.ncbi.nlm.nih.gov 2020 [cited 4 September 2020].
10. Jan K, Min R, Yen T, Singh S; Ischemic Stroke in an HIV Positive Patient: An Initial Presentation of Neurosyphilis; Neurological Medicine [Internet]. Ncbi.nlm.nih.gov 2020 [cited 4 September 2020].
11. Palacio, M., Núñez, T., Montiel, K., Ferrer, Y., Finol, F., & Parra, M. (2012). Síndrome de Weber hemorrágico: a propósito de un caso. *Revista Latinoamericana De Hipertensión* , (7), 45-47.
12. Burgueño-Montañés, C., Santalla-Castro, C., & Peña-Suárez, J. (2016). Síndrome de Parinaud «plus» en paciente con disgerminoma. *Archivos De La Sociedad Española De Oftalmología* , 91 (7), 341-345.

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