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"*HANDL SYNDROM* E": DIFFERENTIAL DIAGNOSIS OF HEADACHE IN THUNDER

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Case report: A 27-year-old female patient, previously healthy, was hospitalized with a thunderclap headache without associated acute focal deficit. He denies fever, infectious prodromes, recent vaccination or use of illicit drugs. On admission, cranial tomography and angiotomography were performed without alterations. The CSF had a xanthochromic aspect, 465 cells/mm ^{3} of lymphocytic predominance (93%), 155 mg/dL proteins, 60mg/dL glucose (97mg/dL capillary), in addition to a difference of 47 red blood cells between the first and second vials.. CSF cultures as well as the meningoencephalitis panel were negative. Referred to arteriography which did not show abnormalities. Serum infectious tests including HIV, VDRL, hepatitis and herpes virus negative. On the second day of hospitalization, the patient had hypoesthesia in the right hemiface, diplopia and global aphasia with a total duration of 12 hours and spontaneous improvement. In the context of acute focal deficit, cranial magnetic resonance imaging and intracranial arterial resonance angiography were performed without alterations. After 5 days, a second cerebrospinal fluid collection was performed and there was an improvement in the protein 26.6 mg/dL but maintenance of lymphocytic pleocytosis - 450 cells (98% lymphocytes). No specific treatment was administered. The patient had a favorable outcome without neurological sequelae, however, she maintained episodes of mild migraine headache pattern for another 40 days. There was no recurrence of new focal deficits.

DISCUSSION: *HaNDL syndrome (Headache with Neurologic Deficits and cerebrospinal fluid Lymphocytosis)* is a rare condition with a higher prevalence between the third and fourth decades of life, being an important differential diagnosis between secondary headaches. The syndrome is characterized by episodes of headache accompanied by transient neurological deficits lasting from 15 to 120 minutes, not exceeding a duration of three days. The thunderclap headache pattern is rare, with progressively worsening headache with migraine characteristic being more common. The CSF has a lymphocytic pleocytosis and proteinorrachia, in addition signs of intracranial hypertension such as visual symptoms, papilledema and abducens nerve paresis may be present. The etiology of the *HaNDL syndrome* is unknown, taking into account the inflammatory-appearing CSF, the monophasic course of the disease and the presence of a viral prodrome in one third of patients, an immune-mediated mechanism is suspected after inflammatory and infectious triggers. The association with herpes viruses type 1 and 2, herpes simplex type 6, *borrelia burgorferi* and less commonly cytomegalovirus is described in the literature. The spreading cortical depression is another pathophysiological factor of the disease not yet fully elucidated, it is believed that the viral disease triggers the release of inflammatory agents and, consequently, transient vasomotor changes. Abnormal neuroimaging findings are reversible, such as leptomeningeal uptake on cranial magnetic resonance imaging and hypoperfusion without diffusion restriction, not respecting vascular territory on functional neuroimaging - suggestive of oligoemia rather than ischemia. The electroencephalogram reflects the reversible period of cortical dysfunction and may show a pattern of focal or diffuse slowing. The *HaNDL syndrome* has the differential diagnosis of cerebrovascular accident, vasculitis of the central nervous system, migraine with aura, familial hemiplegic migraine and encephalitis, and its abrupt pattern with spontaneous resolution without specific treatment makes it different from

other pathologies. **Conclusion:** HaNDL *syndrome is a self- limiting* pathology with a favorable prognosis. In this case report, the patient meets the criteria for the diagnosis despite the absence of a viral prodrome. It is a diagnosis of exclusion that, although rare, must be suspected in cases of acute focal deficit and migraine headache, in young patients without cardiovascular risk factors.
Keywords: Thunderstorm headache; Lymphocytic pleocytosis; Pseudo migraine with lymphocytic pleocytosis.

REFERENCES

1. Morrison DG, Phuah HK, Reddy AT, *et al.* **ophthalmologic involvement in the syndrome of headache, neurologic deficits, and cerebrospinal fluid lymphocytosis.** *Ophthalmology.* 2003; 110:115.

2. Emond H, Schnorf H, Poloni C, *et al.* **Syndrome of transient headache and neurological deficits with CSF lymphocytosis (HaNDL) associated with recent human herpesvirus-6 infection.** *cephalalgia.* 2009; 29:487.

3. Vieira JP, Brito MJ, de Carvalho IL. **Borrelia lusitaniae infection Mimicking Headache, Neurologic Deficits, and Cerebrospinal Fluid Lymphocytosis.** *J Child Neurol.* 2019; 34:748.

4. Ferrante E & Prone V. **Possible clinical marker of first attack of HaNDL syndrome (stroke-like mimics) in emergency room : a case report.** *Neurological Science.* 2018;39(11):2003-2005.

5. Armstrong - Javors A, Krishnamoorthy K. **HaNDL Syndrome: Case Report and Literature Review.** J Child Neurol. 2019 Mar;34(3):161-167. doi : 10.1177/0883073818811546. Epub 2018 Dec 5. PMID: 30514135.