

A CASE REPORT ON SYNDROME OF TRANSIENT HEADACHE AND NEUROLOGICAL DEFICITS WITH CEREBROSPINAL FLUID LYMPHOCYTOSIS (HANDL)

MAHMUD R¹, RASSEL MA², MONAYEM FB³, CHOWDHURY AH⁴, ISLAM KMN⁵

Abstract:

HaNDL syndrome (transient headache and neurological deficits with cerebrospinal fluid lymphocytosis) is a rare headache disorder previously known as Migraine with cerebrospinal fluid pleocytosis. The disease is characterized by one or more episodes of headache and transient neurological deficits associated with cerebrospinal fluid lymphocytosis. It is a benign disorder and a stroke mimicker. We describe a case of 50 years old lady presented with acute onset headache, right hemi sensory tingling and bilateral papilloedema. Her blood test, MRI of brain and MRV was normal. CSF study revealed lymphocytic pleocytosis. The patient was discharged with full recovery. HaNDL syndrome is a diagnosis of exclusion. High degree of suspicion and characteristic clinical and laboratory findings are important to recognize.

Key word: HaNDL, Pleocytosis, Migraine

DOI: <https://doi.org/10.3329/jdmc.v29i1.51177>
J Dhaka Med Coll. 2020; 29(1) : 89-91

Introduction:

The syndrome of transient headache and neurologic deficits with cerebrospinal fluid lymphocytosis (HaNDL syndrome) is a self-limited condition. It was previously known as Migraine with cerebrospinal pleocytosis; pseudo migraine with lymphocytic pleocytosis. It was first described in early 1980¹. It is a rare disorder with Migraine-like headache episodes (typically one to twelve) accompanied by neurological deficits including Hemi-paresis, hemiparesis and/or dysphasia, but positive visual symptoms only uncommonly, lasting several hours. There is lymphocytic pleocytosis. The disorder resolves spontaneously within 3 months². As an example of a rare disease. We described a case of 50 years old lady presented with acute onset headache, right hemi sensory tingling and bilateral papilloedema. Her blood test, MRI of brain and MRV was normal. CSF study revealed lymphocytic pleocytosis.

Case Description:

A 50 year old lady admitted in the inpatient of the Neurology Department of Dhaka Medical College on 15.02.20 with the history of severe throbbing head ache for 2 days associated with tingling sensation on the he right side of her body. The headache was global, throbbing in nature, severe in intensity and was unremitting. It was associated with several episodes of vomiting. There was associated hemi-paresis on her right side. There was no hemiparesis, confusion, visual disturbances, speech difficulty and gait disturbances. She did not have any fever, cough and respiratory distress. Neurologic examination revealed patient was fully conscious, there was no dysarthria or dysphasia. Cranial nerve examination revealed bilateral papilloedema, otherwise all cranial nerves were normal. Motor and sensory examinations revealed no focal deficit. There was no signs of meningeal irritation. Workup revealed unremarkable routine laboratory blood and urine examination.

1. Dr. Reaz Mahmud, Assistant Professor, Department of Neurology, Dhaka medical College.
 2. Dr. Mohammad Aftab Rassel, MD thesis student, Department of Neurology, Dhaka medical College.
 3. Dr. Farhana Binte Monayem, Medical officer, Sarkari karmachari Hospital.
 4. Dr. Ahmed Hossain Chowdhury, Associate Professor, Dept. of Neurology, Dhaka Medical College Hospital, Dhaka.
 5. Dr. KM Nazmul Islam, Assistant professor, Department of Neurology, Shaheed Suhrawardy Medical College, Dhaka
- Correspondence :** Dr. Reaz Mahmud, Assistant Professor, Department of Neurology, Dhaka Medical College. Phone - +88 01912270803. Mail- reazdmc22@yahoo.com

Received: 04-04-2020

Revision: 17-02-2020

Accepted: 21-03-2020

Initial CT scan of brain followed by MRI of brain with contrast and MRV were normal, except congenital absence of right transverse sinuses. CSF study revealed opening pressure 35 cm of water. Cell count revealed 170 cells per cubic millimeter with predominant lymphocyte (86%) and protein 175 mg/dl. CSF gram stain, AFB stain were normal, it was Negative for HSV serology. Blood for ANA and ENA profile were also normal. The patient was treated with Tab Topiramate 25 mg BD and Cap Nortriptyline 25 mg once daily and Tab Paracetamol as required. The head ache was subsided after 5 days of admission. She was discharged after 10 days. At her discharge CSF examination along with opening pressure was normal. Papilloedema was also subsided. Follow up after 2 month revealed no recurrence of headache with normal disc.

Discussion:

HaNDL (transient headache and neurological deficits with cerebrospinal fluid lymphocytosis) is a diagnosis of exclusion. The index case presented with migraine like headache with a transient neurological feature (hemiparaesthesia) persisted for > 4 hours, associated with CSF pleocytosis. Other possible etiologic studies were negative and the CSF pleocytosis improved with the improvement of the headache. So the case meets the diagnostic criteria² of HaNDL (transient headache and neurological deficits with cerebrospinal fluid lymphocytosis) given by international Headache society.

At presentation our provisional diagnosis was intracerebral haemorrhage (ICH) with raised intracranial pressure (ICP) though there was no confusion. The ICSOL with raised ICP, Venous stroke, subarachnoid hemorrhage and Meningitis were also our consideration. CT scan revealed no haemorrhage. MRI with contrast revealed no focal lesion and leptomeningeal enhancement. So we went for CSF study keeping in mind of benign intracranial hypertension (BIH) and viral meningitis though the patient had a focal Hemiparaesthesia. CSF pressure was raised but lymphocytic pleocytosis exclude the diagnosis of BIH. As there was no leptomeningeal enhancement and HSV serology was normal, viral etiology was also excluded.

So we made the diagnosis of transient headache and neurological deficits with cerebrospinal fluid lymphocytosis. Its important differential is Mollaret Meningitis where focal sign is absent, Mollaret cells might be found. The headache was subsided after 5 days. At 9th day we again performed the CSF study which was normal. This again goes in favor of our diagnosis, transient headache and neurological deficits with cerebrospinal fluid lymphocytosis.

In 1981 first reported cases of Bartleson¹ et al., patients of both sexes and 16-50 years of age was described where symptoms persisted for 1-12 weeks. Later Gomez- Aranda³ et al reported 50 cases with a wide variety of neurological deficits (such as sensory, motor, and language problems) and patient's experiencing 1-12 episodes. In their case series they found mean duration of headache was 19 hour and mean duration of neurologic deficit was 5 hour. In that cases CSF lymphocytes was found ranged from 10 to 760 per cubic millimeter and CSF protein was increased in 96% of patients. In this case reported in this article a lady of 50 years of age was diagnosed and the patient suffered for 1 episode lasting 5 days with no recurrence up to 2 months of follow-up. In 75% of cases it is found to be monophasic¹.

The etiology of this rare disorder is uncertain. Some researchers predict viral etiology⁴, some autoimmune etiology⁵ and some described it as a complicated migraine⁶. Kappler et al⁷. Found asymmetric velocities and pulsatilities in middle cerebral arteries using Transcranial Doppler sonography. Cerebral Angiograms that were performed subsequently were within normal limits. This findings provoked the authors to conclude that HaNDL syndrome could represent episodes of migraine due to the similarities of the vasomotor features. The studies conducted to find the association between HaNDL syndrome and acute ischemic stroke, found that despite the presence of neurologic deficits that were often prolonged, diffusion-weighted (DWI) MRI images were normal in all cases.

Conclusion:

This is probably the first reported case in Bangladesh. HaNDL syndrome is a diagnosis

of exclusion. High degree of suspicion and characteristic clinical and laboratory findings are important to recognize.

References:

1. Bartleson JD, Swanson JW, Whisnant JP. "A migrainous syndrome with cerebrospinal fluid pleocytosis." *Neurology* 1981; 31(10) :1257-1262.
2. The International Classification of Headache Disorders, 3rd edition (beta version) Cephalalgia 33(9), 720, International Headache Society 2013.
3. Gomez-Aranda F, Ca ´ nadillas F, Mart JF, ´ y-Masso . Pseudomigraine with temporary neurological symptoms and lymphocytic pleocytosis. A report of 50 cases," *Brain* 1997; 120, (7):1105-1113.
4. Devetag CF. Headache with temporary neurological symptoms and lymphocytis pleocytosis: a case report and etiologic hypothesis. *Neurological Sciences* 2002; 23(3):123-125, 2002.
5. Shikishima K, Kitahara K, Inoue K. Ophthalmologic manifestations in headache, neurologic deficits, and cerebrospinal fluid lymphocytosis (HaNDL) syndrome with nonspecific frontal lesions and hyperthyroidism. *Eye* 2006; 20(5):613-615.
6. Gekeler F, Holtmannspotter M, Straube A, Klopstock T, "Diffusion-weighted magnetic resonance imaging during the aura of pseudomigraine with temporary neurologic symptoms and lymphocytic pleocytosis," *Headache* 2002;42(4): 294- 296.
7. Kappler J, Mohr S, Steinmetz H, Cerebral vasomotor changes in the transient syndrome of headache with neurologic deficits and CSF lymphocytosis (HaNDL). *Headache* 1997; 37(8): 516-518.
8. Segura T, Hernandez-Fernandez F, Sanchez-Ayaso P, Lozano E, Abad L. Usefulness of multimodal MR imaging in the differential diagnosis of HaNDL and acute ischemic stroke. *BMC Neurology* 2010; 10(2):120.
9. Vallet A, Desestret V, Tahon F, Cho T, Nighoghossian N. Acute perfusion MR imaging in a HaNDL-like syndrome. *Cerebrovascular Diseases* 2009; 29(1):98-100.