ISSN- O: 2458 - 868X, ISSN–P: 2458 – 8687
Index Copernicus Value: 68 . 16
PubMed - National Library of Medicine - ID: 101731606

International Journal of Medical Science and Innovative Research (IJMSIR)
IJMSIR : A Medical Publication Hub
Available Online at: www.ijmsir.com
Volume – 5, Issue –2, April - 2020, Page No. : 47 - 50

A case report of transient headache and facial palsy with cerebrospinal fluid leucocytosis

[1]Dr. Neetu Mariam Alex, Department of Internal Medicine, Apollo Main Hospitals, Greams road, Chennai, Tamil Nadu.
[2]Dr. M. Phani Krishna, Department of Internal Medicine, Apollo Main Hospitals, Greams road, Chennai, Tamil Nadu.
[3]Dr. J R Subramaniam, Department of Internal Medicine, Apollo Main Hospitals, Greams road, Chennai, Tamil Nadu.
Corresponding Author: Dr. Neetu Mariam Alex, Department of Internal Medicine, Apollo Main Hospitals, Greams road, Chennai, Tamil Nadu.

Citation this Article: Dr. Neetu Mariam Alex, Dr. M. Phani Krishna, Dr. J R Subramaniam,"A case report of transient headache and facial palsy with cerebrospinal fluid leucocytosis ", IJMSIR- April - 2020, Vol – 5, Issue -2, P. No. 47 – 50.
Type of Publication: Case Report
Conflicts of Interest: Nil

Abstract

The syndrome of transient headache and neurological deficits with cerebrospinal fluid lymphocytosis (HaNDL) is a self limited, benign entity which was first described in the year 1980 by Swanson, Bartleson and Whisnant. It is characterised by varied but transient neurological deficits, headache and cerebrospinal fluid (CSF) pleocytosis. Although it is a self limited condition an extensive work up is warranted to rule out other serious conditions. In this case report we describe a 49-year-old female with past medical history of hypothyroidism presented with history of fever for 4 days, headache and left sided upper motor neuron type facial palsy. Extensive infective and autoimmune workup done in our patient turned out to be negative. This made us to consider this rare syndrome as the most possible diagnosis. She was managed symptomatically with NSAIDs and Acetazolamide and recovery was complete within a week. This case is presented in view of its rarity and clinical mimicry with much more serious conditions and complete recovery from neurological deficit, knowledge of which helps in avoiding unnecessary workup and therapies, as treatment for this condition is largely symptomatic.

Keywords: HaNDL syndrome, Transient headache, CSF pleocytosis, migranous headache

Introduction

The syndrome of transient headache and neurological deficits with cerebrospinal fluid lymphocytosis (HaNDL) is a self limited, benign entity that is characterised by one or more episodes of severe headache, transient neurologic deficits, and lymphocytic pleocytosis in CSF. Neurologic deficits typically last 15–120 minutes, although a range of 5 minutes to 3 days has been reported [(1)]. Its aetiology is not fully understood and it can mimic a variety of conditions including aseptic meningitis, meningoencephalitis, migraine with aura and stroke [(2)]. Although it is a self limited condition an extensive work up is warranted to rule out other serious conditions. Once a diagnosis of HaNDL is established, treatment is symptomatic management of headache. Knowledge about this rare neurological disorder is important for clinicians in view of its self-limited nature and favourable outcome, besides symptomatic

treatment, other therapeutic interventions are largely unnecessary.

Case Presentation

Our case was a 49-year-old female with past medical history of hypothyroidism presented with history of fever for 4 days 1 week ago and complaints of severe headache and nausea for 1 week. She had complaints of reduced appetite, disturbed sleep and constipation for past 1 week. She also had complaints of numbness over the left side of the face for the past 2 days.

She was initially evaluated as outpatient and Magnetic resonance image (MRI) Brain with MR venogram done revealed partially empty sella, prominent bilateral perioptic CSF spaces, and vertical tortuosity of right optic nerve. FLAIR signal hyper intensity in cortical region of medial bi- frontal lobe involving cingulate gyrus, medial aspect of left fronto-parietal lobe, suggestive of benign intracranial hypertension **(Figure - 1, 2, 3, 4).**

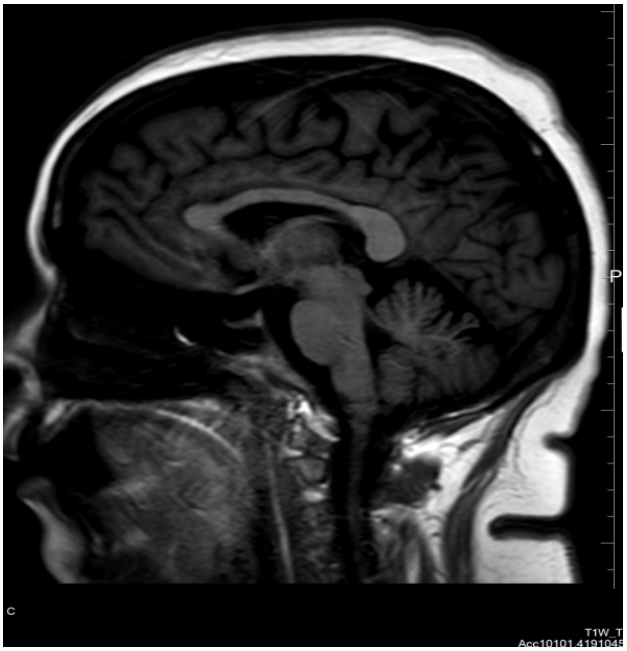

Figure 1: MRI brain showing partially Empty sella

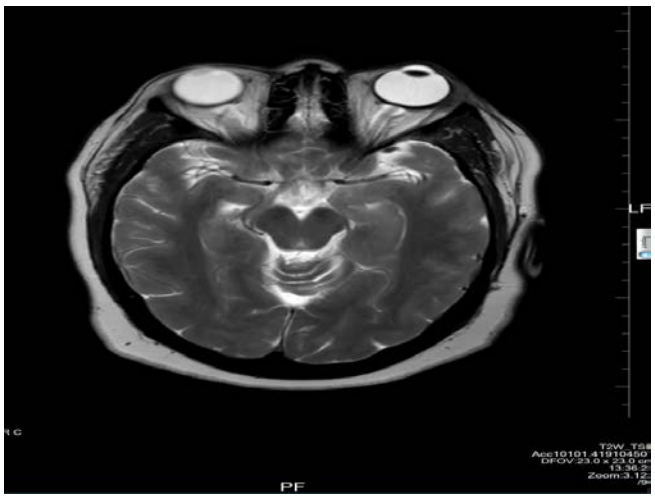

Figure 2: MRI Brain showing prominent bilateral perioptic CSF spaces

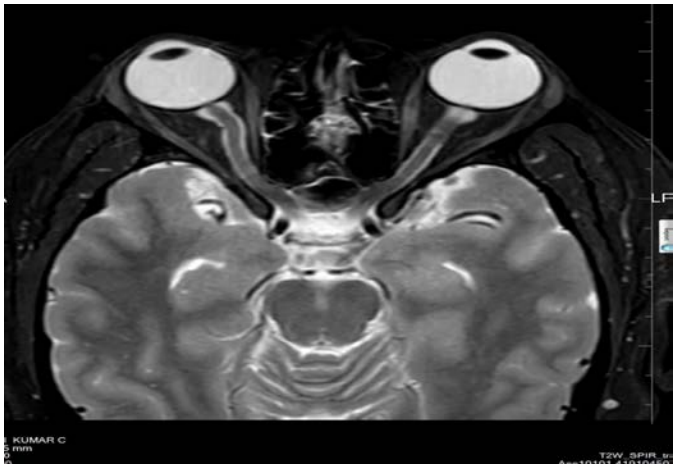

Figure 3: MRI Brain showing vertical tortuosity of right optic nerve

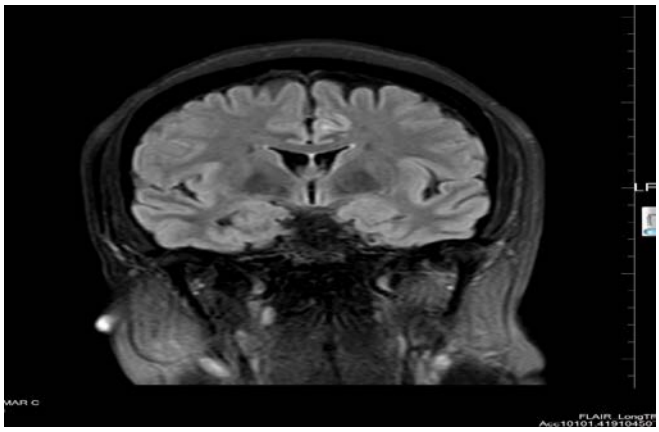

Figure 4: MRI Brain showing FAIR Hyperintensities in cortical region involving cingulate gyrus.

In view of persistent headache and numbness over face she presented to the emergency department. At the time of admission she was conscious, oriented and afebrile with stable haemodynamics. Nervous system examination was notable for left sided upper motor neuron facial palsy and horizontal nystagmus to the right side. Rest of the systemic examination did not reveal any significant abnormality. Baseline investigations were within normal limits except for elevated TSH and low Free T3 levels. Visually evoked potential (VEP) was suggestive of bilateral anterior visual pathway dysfunction and papilledema. She was started on IV Ceftriaxone 2gm OD and NSAIDs. Guarded lumbar puncture was done after clearance from ophthalmologist in view of raised ICT. Opening CSF pressure was 32cms of CSF. CSF analysis reports were as follows:

Table-1: CSF Analysis of patient

CSF Analysis	
Protein	222.3mg/dl
Glucose	32mg/dl
Chloride	116mEq/L
LDH	45
Appearance	Slightly turbid
Total WBCs	1000 (Neutrophils-1%, lymphocytes-99%)
Gram stain	Moderate pus cells and no bacteria seen
Fungal stain	No fungal elements including Cryptococcus seen
AFB smear	No AFB seen
X-PERT MTB/RIF	Not detected

Auto immune workup was negative (ANA, dsDNA, ANCA). HIV and TPHA (Treponema pallidum hemagglutination assay) were negative. Serum calcium and Angiotensin converting enzyme (ACE) levels were within normal limits. Blood cultures and urine culture did not show any significant growth. Meanwhile she was managed symptomatically with NSAIDs , Carbonic Anhydrase inhibitors and empirical antibiotics. In view of extensive infective and autoimmune workup being negative, a diagnosis of syndrome of transient headache with neurological deficits and CSF lymphocytosis (HaNDL) was considered, with partially treated bacterial meningitis being considered as other possibility. After a detailed discussion with Infectious diseases and Neurology specialists, based on available lab and imaging reports and in view of complete and rapid recovery, HaNDL was considered to be more likely diagnosis in this patient.

Discussion

HaNDL syndrome was first described in 1980 by Swanson, Bartleson and Whisnant, who described seven patients with migrainous headache with pleocytosis in the CSF at the American Academy of Neurology [(3).] It is more commonly reported in 3rd and 4th decades of life. A minority of cases may have family history of migraine and a few cases may have a history of preceding viral illness. This syndrome is characterised by a classical triad of headache, neurological deficit and CSF pleocytosis, all of which were present in our case. Headache is throbbing type, and usually moderate to severe. Most frequent neurologic deficits we see in this are hemiparesis, hemisensory disturbances and aphasia. In our patient neurological deficit was in the form of facial palsy and papilledma, from which she recovered completely in concordance with benign and self limiting nature of this entity. Although focal deficits are more common, there

have been case reports with more diffuse deficits like acute confusional states.

Intracranial vasomotor changes and perfusion disparity plays a role in the pathophysiology of HaNDL syndrome. High titres of antibody against CACNA1H protein which is one of the subunits of T-type calcium channel were found in 2 patients with HaNDL syndrome, supporting the view that ion channel autoimmunity may partially contribute to the pathogenesis of the syndrome [(4).] The differential diagnosis of a patient presenting with focal neurological deficits with headache include stroke; structural brain lesions; epileptic seizures; neuroinfectious diseases, Mollaret's meningitis, encephalitis , neurolupus, familial hemiplegic migraine type 1 (FHM1); vasculitis of the central nervous system and Hashimoto's encephalopathy [(3)]. Extensive infective and autoimmune workup done in our patient turned out to be negative. This made us to consider this rare syndrome as the most possible diagnosis. She was managed symptomatically with NSAIDs and Acetazolamide and recovery was complete within a week.

It is very important to have knowledge about this rare neurological disorder in view of its benign self-limited nature and to differentiate it from other close differentials thereby avoiding unnecessary therapies like long term antibiotics and thrombolysis. In a patient who has headache accompanied by recurrent neurological disorder episodes, CSF pleocytosis and normal imaging tests, HaNDL syndrome should be absolutely considered [(4)]. Once the diagnosis of HaNDL syndrome is established treatment is mainly symptomatic. However, corticosteroids, calcium channel blockers and acetazolamide have been previously used in treatment [(5)].

Conclusion

Through this case report we would like to bring to the notice of medical fraternity about HaNDL syndrome, which is a rare disease characterised by headache, wide range of transient neurological deficits and CSF pleocytosis. It is mainly a diagnosis of exclusion after ruling out more serious and life threatening conditions. Knowledge about this condition helps in avoiding unnecessary workup and therapies, as treatment for this condition is largely symptomatic.

References

1. Esteban-Zubero et al. "Transient headache and neurological deficits with cerebrospinal fluid lymphocytosis (HaNDL Syndrome): A case report". Health Prim Car, 2017, Volume 1(1): 1-3.
2. Filipa Silva et al. "How to HaNDL This Case?" Journal of Medical Cases, Vol. 7, No. 3, Mar 2016.
3. Rivero-Sanz E, et al. "HaNDL syndrome in a 14-year-old girl" BMJ Case Rep 2016. Doi: 10.1136/bcr-2015-213018.
4. Çoban et al. "HaNDL Syndrome" Archives of Neuropsychiatry 2013; 50 Supplement 1: S52-S55.
5. Al Hadidi et al. "Syndrome of transient headache and neurologic deficits with cerebrospinal fluid lymphocytosis masquerading as meningitis in HIV patient" Ann Transl Med 2017; 5(6):138.