Recurrent headache in a five year old boy

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Abstract

Headache is infrequent in early childhood. Headache and neurological deficits associated with cerebrospinal fluid (CSF) lymphocytosis (HaNDL), a variant of migraine, is a rare disorder. A 5-year-old boy presented with recurrent episodes of headache for 6 months. Each episode lasted for a week and in the current episode, he was symptomatic for 3 days. All the episodes were associated with paresthesias and CSF lymplocytosis with normal protein and sugar. There was history of migraine in his family. His magnetic resonance imaging (MRI) brain with contrast with magnetic resonance (MR) angiography and venography were normal. Work-up for relevant causes of infection and vasculitis were negative. His symptoms subsided on oral antimigraine prophylaxis and he has been on remission for last 8 months. HaNDL should be considered in relevant clinical scenarios, as it prevents unnecessary investigations, therapy, and hospitalization.

Key Words

CSF lymphocytosis, HaNDL, headache, migraine, toddler

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Introduction

Headache is common in pediatric age-group and is increasingly reported in school-going years and adolesence. The common etiologies are migraine, sinusitis, refractive errors, intracranial space-occupying lesion, trauma, metabolic and vascular diseases. Recognition that pediatric headaches can result from primary and secondary causes is crucial for treatment.[1] One of the rare condition which can present as headache is headache and neurological deficits associated with cerebrospinal fluid (CSF) lymphocytosis (HaNDL), which is characterized by one or more episodes of severe headache, transient neurologic deficits, and CSF lymphocytic pleocytosis.[2] It is rarely reported in the pediatric age-group.[3]

A case of 5-year-old boy is being reported here who suffered from HaNDL. It is a self-limiting condition, prevents unnecessary investigations, prolonged antibiotic administration and hospitalization, and does not have any long-term sequelae.[4] Though etiology of this syndrome remains elusive till date, a possible association of migraine with HaNDL syndrome exists in current literature.[5]

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Case Report

A 5-year-old premorbidly normal boy presented to the current center with recurrent episodes of headache for last 6 months. First episode occurred 6 months back and the second episode 2 months prior to the current episode. Both the episodes lasted for around 1 week. In this episode, he was symptomatic for 3 days. All three times, including the current episode, his presenting complaints were continuous, generalized, throbbing, moderate to severe intensity headache associated with transient paresthesias (in the bilateral hands and thighs, which lasted for hours to days), getting partially relieved by medications and occasionally associated with non-projectile vomiting. These were not accompanied by any visual or auditory aura (photophobia or phonophobia). The previous two episodes were treated at a different center as meningitis based on CSF reports where he was hospitalized and received parenteral antibiotics, mannitol, and dexamethasone for 2-3 weeks. All three episodes were preceded by a viral prodrome 2-3 weeks before. There was no associated altered sensorium, seizures, cranial nerve palsies, flaccidity or tightness of limbs, abnormal involuntary movements, features suggestive of inco-ordination or bowel bladder disturbances. Fever was absent in all the episodes. There was a significant family history in the form of migraine like headache episodes in the grandmother and sister.

Table 1: Cerebrospinal fluid analysis report in all the three episodes

CSF parameters	1st episode	2nd episode	3rd episode
Cells (/μL)	78 (100% lymphocytes)	320 (100% lymphocytes)	60 (100% lymphocytes)
Protein (mg/dl)	17	30	18
Sugar (mg/dl) (CSF/blood)	56/83	133/162	56/92

CSF = Cerebrospinal fluid

His vital parameters, general physical and systemic examination were essentially normal. His fundus examination did not reveal any feature suggestive of papilledema.

His baseline hematological (hemogram with peripheral smear) and biochemical (serum electrolytes, blood sugar, liver and kidney function tests) investigations were normal. His CSF analysis revealed 60 lymphocytes with normal protein and sugar. Similar CSF findings were also seen in the first two episodes [Table 1]. Analysis of CSF for herpes simplex virus and tubercular polymerase chain reaction (PCR) were negative. Serological tests for Lymes disease and Brucellosis, human immunodeficiency virus (HIV), anti-nuclear and anti-neutrophilic cytoplasmic antibodies were negative. Magnetic resonance imaging (MRI) brain and spine with contrast, magnetic resonance (MR) angiography, and venography study were normal.

Based on the clinical database of recurrent episodes of headache, transient sensory symptoms in form of paresthesias, absence of fever, and normal neurological examination with characteristic CSF picture and normal neuroimaging, diagnosis of HaNDL was made.

Child was started on antimigraine prophylaxis with flunarizine (5 mg/day) in view of strong family history. He became asymptomatic over next 1 week and was discharged. He has not had any recurrence in the last 8 months.

Discussion

The syndrome of transient HaNDL was first described by Bartleson in 1981. It is a benign and self-limited disorder usually seen during the third and fourth decades of life.[2] The etiology remains elusive, the most plausible postulate being an inflammatory or infectious trigger precipitating a migraine like illness.[6,7] It is a diagnosis of exclusion and it is mandatory to rule out other conditions like viral encephalitis, meningitis, Lyme disease, neurobrucellosis, neurolupus, central nervous system vasculitis, HIV, and cerebral vascular diseases.[7] Absence of fever makes the possibility of infectious and inflammatory conditions highly unlikely as seen in the current case.

The most common transient neurological findings include sensory manifestations (78%), aphasia (60%), and motor symptoms (56%). Rarely migraine-like visual symptoms may be observed (12%).[8,9] Sensory signs frequently start with numbness in the hand and progress occasionally to arm, face, and tongue. The trunk and legs are rarely involved.[4] The current case also has sensory symptoms as a correlate for transient neurological deficits.

In this entity, the usual findings on lumbar puncture are raised opening pressure, lymphocytosis (10-760 cells/μL), normal to increased protein, and normal glucose. In the current case, CSF opening pressure was raised (subjectively as assessed by the flow through the lumbar puncture needle, not measured objectively by a manometer), with CSF lymphocytosis (78,320 and 60 cells/μL) and normal protein and sugar on all the three occasions. The CSF pleocytosis resolves with time, although exact duration has not yet been established.[8,9]

Normal brain imaging is a criterion for this syndrome, as observed in the current case, but patients with non specific MRI changes have been reported occasionally.[4] More recently, there have been some reports of cerebral hypoperfusion in HaNDL.[10] These, despite being a challenge for the established diagnostic criteria for the syndrome of HaNDL, suggest that early acute multimodal MRI might provide important additional clues to differentiate this entity from other conditions.[11]

Once the diagnosis of the syndrome of HaNDL is established, treatment should consist of symptomatic headache management. This syndrome has a relapsing remitting self-limited course; hence, education and reassurance of the patient and family is very important. HaNDL syndrome and its potential relationship to migraine have been evaluated in several studies.[12] Transcranial Doppler sonography and cerebral angiograms have been used to evaluate intracranial blood flow during and after episodes of HaNDL syndrome. Asymmetric velocities and pulsatilities were documented in bilateral middle cerebral arteries during an episode that normalized later simulating vasomotor features seen in migraine.[5] The current case has also shown favourable response to antimigraine prophylaxis.

Despite being a condition probably underdiagnosed and seldom reported in pediatric age-group, the syndrome of HaNDL should be considered in otherwise healthy children presenting with recurrent headache. It has a good prognosis and should be suspected at the earliest to avoid unnecessary investigations, therapies, and hospital stay.

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