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Case Report

Rare mimicker of transient ischaemic attacks: the syndrome of transient headache and neurological deficits with cerebrospinal fluid lymphocytosis

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ABSTRACT

The syndrome of transient headache and neurologic deficits associated with cerebrospinal fluid lymphocytosis (HaNDL) is a benign and self limiting disorder characterized by 1 or more episodes of severe headache and transient neurologic deficits with lymphocytic pleocytosis in the cerebrospinal fluid. We report a case of a 30-year-old male who presented with four episodes of headache followed by hemi paresis and hemisensory loss in a week the patient was completely asymptomatic in between each episode of headache with neurological deficits. Persistent serial imaging to visualize the brain were normal with evidence of cerebrospinal fluid lymphocytosis, which lead to the diagnosis of HaNDL. We need to advocate a high degree of suspicion for HaNDL in the background of strong clinical history and findings, when imaging is normal.

Keywords: HaNDL, TIA

INTRODUCTION

The syndrome of transient headache and neurologic deficits associated with cerebrospinal fluid lymphocytosis (HaNDL) is a benign and self limiting disorder characterized by 1 or more episodes of severe headache and transient neurologic deficits like hemisensory loss, hemi paresis, language disturbances and occasionally visual symptoms with lymphocytic pleocytosis in the cerebrospinal fluid. It occurs usually in young to middle aged men.[1] HaNDL should be considered among the differential diagnosis in patients presenting with recurrent headache and acute neurologic deficits. The episodes lasts from a few hours to 3 days and can have about 1-12 episodes during this monophasic illness. The exact Pathogenesis is not known, it could be immune mediated, the positive serum viral antibody titres and auto antibodies to the subunit of T-type voltage-gated calcium channel (CACNA1H), mitogen activated protein kinase-4 (MAPK-4), and DNA-dependent protein kinase catalytic subunit (DPKCU).[2] Management is conservative, but drugs reported be useful are corticosteroids, calcium channel blockers, valproic acid, and acetazolamide. The prognosis is usually good.

CASE REPORT

A 30-year-old male presented to the hospital with sudden onset benumbed sensation of the left half of the body which lasted for 20 minutes. After 10 minutes he developed a diffuse dull aching headache which persisted for 8 hours. The intensity of the headache was slowly progressive. Fifteen minutes from the onset of headache he developed left hemiparesis and facial lag, which lasted for 1 hour and resolved spontaneously. On presentation to the hospital his neurological and systemic examination was normal. Patient underwent neuroimaging studies, Mri BRAIN with MR angiogram and CT angiogram of neck and intracranial vessels done were normal. The patient

was diagnosed to have a transient ischemic attack and was started on antiplatelets. Four days after the first episode patient again developed numbness of left arm lasting for 10 min. Ten minutes from the onset of numbness he a developed headache followed by slurring of speech and deviation of angle of mouth lasting for 2 hrs and the headache persisted for 4 hrs. The headache was associated with photophobia and vomiting. The MRI brain was repeated again and was found to be normal. He was asymptomatic for two days, and later in the night he had a severe throbbing occipital headache with slurring of speech, the headache persisted for 4 hours and the dyarthria recovered over 45minutes. During all his episodes, blood pressure was documented to be normal. When asked retrospectively patient said that he had history of headaches prior to this presentation, but he has never had hemiparesis or dysarthria. The differentials we considered at this point were sporadic hemiplegic migraine, meningitis, reversible cerebral vasoconstriction syndrome. Electroencephalogram showed frontocentral slowing more on the right side than the left. To rule out reversible cerebral vasoconstriction syndrome patient was proceeded with a DSA of the cerebral vessels which was normal and didn't show any signs of vasospasm. After DSA he again had sudden onset headache which continued for 8 hrs, 15 minutes from the onset of headache he developed deviation and slurring of speech which lasted for 1 hour and resolved spontaneously. The intensity of the headache he developed after the DSA was the worst compared to the previous episodes.

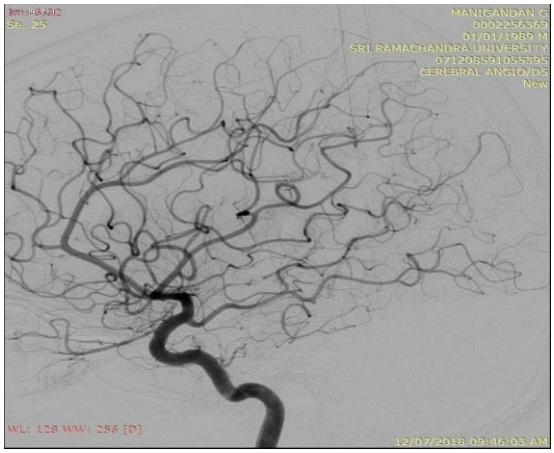

Figure 1: Digital substraction angiography of cerebral vessels.

His routine blood parameters and the cardiac evaluation with an echocardiogram and Holter were normal. To rule out CNS infection patient underwent cerebrospinal fluid analysis which showed an opening pressure of- 13CM OF H2O, protein-203 sugar -57 chloride -125 WBC- 95 with polymorphs -30% and lymphocyte-70%, Adenosine deaminase- 7.2 with no xanthochromia with CSF TB PCR and gene expert being normal. His CSF viral encephalitis, autoimmune encephalitis and lyme antibodies were negative. The CSF findings were suggestive of a CNS infection but the patient being completely asymptomatic between the episodes and the dramatic change in the clinical picture between the normal phase and headache phase made us suspect HaNDL. The patient was treated with analgesics and calcium channel blockers and his intensity and episodes of headache didn't recur. After three months of these episodes and starting treatment the patient underwent repeat cerebrospinal fluid (CSF) analysis which was found to be normal (CSF cell count-absent, sugar-70 and protein- 42) this confirmed our diagnosis of HaNDL.

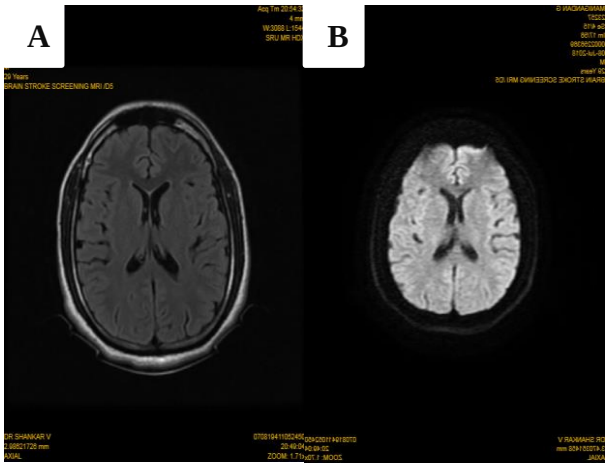

Figure 2: MRI. A) MRI Brain, B) DWI sequence.

DISCUSSION

HaNDL is a self-limiting syndrome that presents as a sudden onset of headache with temporary neurologic deficit and CSF lymphocytosis. The common presenting symptoms in adults are headache with sensory disturbances and in children its speech impairment.[3] It lasts an average of 19 hours, and it may be accompanied by nausea, vomiting, photophobia, or phonophobia, but most patients don't have a prior history of migraine headaches. Sensory symptoms, language disturbances, and hemiparesis lasting upto 5 hours are the frequent focal deficits. Cortical spreading depression (CSD) triggered by a preceding or concurrent viral illness leading to transient vasomotor changes has been suggested as a pathogenic mechanism.[4] Leptomeningeal enhancement has also been noted in MRI imaging in some patients, this suggests that impaired cerebrospinal fluid resorption might be the reason for elevated intracranial pressures noted in HaNDL syndrome.

The defining features of HaNDL is a cerebrospinal fluid (CSF) lymphocytic pleocytosis with elevated CSF opening pressure and increased CSF protein levels, with a mean protein level of 94 mg/dL.[5] HaNDL is mostly a diagnosis of exclusion after investigating all the common diseases that presents as headache and transient neurologic deficits. The presentation of HaNDL can mimic a stroke and patients might be treated with thrombolysis. EEG can show focal or bilateral slowing with delta or theta frequencies and SPECT scans can

show hypoperfusion. In HaNDL, cerebral angiography can trigger an episode of neurological dysfunction, and this should be borne in mind when investigating headaches associated with focal neurological symptoms.[6] Investigating the patient repeatedly with invasive and expensive laboratory and imaging investigations can be avoided when the diagnosis of the syndrome HaNDL is certain.

CONCLUSION

HaNDL is regarded as a rare disease as its mostly underreported or misdiagnosed. The patients who present with another characteristic attack within three months of the initial attack need not require extensive investigation for the subsequent attacks. A repeat head CT or MRI for each attack and a lumbar puncture for the second attack would suffice.

These investigations are required for reassuring the diagnosis as HaNDL and not a coincidental secondary etiology such as infarction or central nervous system infection. However, more extensive repeat testing may be needed when the diagnosis of HaNDL is not well established. The clinical presentation of the disease can be quite dramatic, but the prognosis is good. Increased recognition of this syndrome helps in preventing unnecessary tests and treatments.

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