careful examination of distal blind end is important in making the diagnosis.

Contributors: SD and RB collected patients' records, drafted the manuscript and performed the review of the literature. AN was involved in patient management and reviewed the manuscript. All authors approved the final manuscript.
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Sporadic Hemiplegic Migraine

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We report a 4-year old boy with probable sporadic hemiplegic migraine. The present case did not fulfill the International Classification of Headache Disorders diagnostic criteria for the disease completely, as it is unclear whether the child had any headache or not. The differential diagnoses are discussed. The case is reported for its rarity and to increase awareness.

Key words : Child, Headache, India, Sporadic hemiplegia migraine.

The phenotype of Sporadic hemiplegic migraine (SHM) attacks may include fever, lethargy, dysphasias, confusion, hemiparesis, hemisensory symptoms, hemianopia and scintillating scotoma. The symptomatology may resemble a stroke. Diagnosis of SHM in most cases is essentially one of exclusion as there is no diagnostic marker, genetic testing is not widely available, and not all genes associated with SHM are yet known [1]. Differential diagnoses include stroke, Todd's palsy, the syndrome of headache, transient neurologic deficit and CSF lymphocytosis (HaNDL), mitochontrial encephalopathy with lactic acidosis and stroke-like episode (MELAS), alternating hemiplegia in childhood, Takayasu disease, and sickle cell anemia [3-4]. Only two reports have previously been published from India [5,6].

CASE REPORT

A four-year old child presented with a transient weakness of the right half of face lasting for 10 minutes. There had been no fever, seizure, headache, visual phenomenon or alteration of sensorium either preceding or during this spell. Speech was normal. Forty eight hours later, he lost his motor speech (comprehension intact) without any hemiparesis and recovered fully in 15 minutes. On the same evening, he again lost his speech, with right sided weakness and facial deviation to the left. This episode was also unassociated with any headache or seizures. He was admitted at a nearby hospital where he recovered in about 3 days time. A CT scan of brain (plain) and a CSF study were normal. He had no family history of vascular disease but his mother suffered from migraine without aura.

When seen about a week later, the child was well and had normal blood pressure and had no neurologic signs or symptoms. All his peripheral pulses were normal and equal. The child was born of non-consanguineous parents and had a normal birth history and motor development. Speech was delayed, and at 4 years, he could only speak monosyllables, though his hearing was normal and comprehension for verbal speech was intact. Routine hematological and biochemical investigations were

normal including coagulation and serum lipid profiles. Sickling test was negative; blood lactate, pyruvate, and homocystine levels were also normal. Contrast enhanced MR Scan of brain, MR cerebral angiography and Venography yielded normal results. ECG and echocardiography were also normal. Doppler study of neck arteries did not reveal any stenosis. A six hour video EEG record was normal. Molecular genetic study could not be performed due to lack of facilities. On the basis of the clinical diagnosis of probable SHM, the child was started on migraine prophylactic regime with cyproheptidine. He was kept under follow up for three months, during which no similar episode occurred. Thereafter, he was lost to follow-up.

DISCUSSION

The diagnostic criteria for sporadic hemiplegic migraine was published by the International Headache Society (HIS) in 2004 [2]. Diagnosis of SHM in the present case had been one of exclusion. Vascular disease was excluded as the child had normal brain imaging, angiography, cardiac and biochemical investigations including lactate / pyruvate measurements. Mitochondrial myopathy with ragged red fibers was not considered as the child had only stroke like episodes, no clinical evidence of any muscle weakness in between attacks and normal serum creatine phosphokinase level.

The problem was the absence of any headache associated with the episodes. However, one needs to consider that the child's speech development had been poor and hence he probably could not express his symptom appropriately. On the other hand, the positive family history of migraine had been supportive. It may also be noted that ICHD-2 (which classifies both forms of hemiplegic migraine under the common rubric of migraine with aura) [2] also mentions of one entity as typical aura (visual, sensory or speech) with non-migraineous (i.e. not fulfilling diagnostic criteria of migraine) headache. And also the childhood periodic syndromes (like cyclical vomiting, abdominal migraine and benign paroxysmal vertigo of childhood) without any headache, are recognized by ICHD-2 as precursors of migraine. The present case can certainly be an unusual form of such a periodic syndrome.

Two major differential diagnoses were considered. HaNDL, first clearly delineated by Bartleson, *et al.* [7] and included in ICHD -2 under the rubric of headache due to nonvascular intracranial disorders, seemed unlikely in view of the absence of CSF pleocytosis and normal protein content. Alternating hemiplegia in childhood (AHC) may be a closer mimic. Early workers considered AHC to be a rare form of complicated migraine [8]. The current status remains undermined in ICHD-2. However, that AHC is often associated with neurodegenerative diseases and that onset is always below 18 months, would be points against considering this diagnosis in the present case. No evidence of a neurodegenerative disease was found in this child.

Jen, *et al*. [9] suggested that hemiplegic migraine should be considered in every child with prolonged hemiplegic symptoms, as well as in adults. SHM in children may be complicated with persistent aura without cerebral infarction and rather infrequently with migraneous infarction. The treatment of acute SHM attacks, as well as prevention in children remains an unresolved issue due to rarity of the disease and lack of randomized controlled trials.

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