

Alternating Hemiplegia of Childhood

**Veena Kalra
Sanjay Rathi**

Alternating hemiplegia of childhood (AHC) is a rare symptom complex resulting from a variety of causes. The syndrome, first described in 1971(1), consists of recurrent episodes of hemiplegia lasting from few minutes to several days, typical onset before 18 months of age, paroxysmal occurrence of tonic/dystonic attacks, nystagmus, dyspnea and other autonomic disturbances. It often results in cognitive impairment and a choreoathetotic movement disorder. Seizures and progressive neurological deterioration are prominent feature (2). Sleep usually relieves both weakness and associated paroxysmal phenomena in a significant number of patients, only to reappear 10-12 minutes after awakening (3). AHC is unreported in the Indian literature. We report the clinical features and long term outcome in a 4-year-old girl.

Case Report

A 4-year-old girl born of a non-consanguineous marriage, at 36 weeks gestation following a cesarean section, initially reported to the Child Neurology Clinic in 1986. The pregnancy and neonatal period were uneventful. She achieved milestones normally during the first year of life and was able to walk

unsupported and speak 2-3 monosyllables before 18 months age. The first symptoms were noticed at 2 years of age when she developed sudden onset right-sided weakness from which she recovered completely in 2 days. There were no associated features like fever, seizures, loss of consciousness or headache preceding the episode. There was no family history of a similar disease, migraine-like symptoms or relation of weakness to diet.

Subsequently, she developed recurrence of similar episodes of transitory hemiplegia on either side without losing consciousness. The episodes lasted from few hours to few days. Such episodes recurred every 15-20 days without leaving any residual deficit. Quadriparesis occurred in a few of the episodes. The child used to move the paralytic limb in sleep and paralysis used to disappear after a good sleep to reappear within half-hour after awakening. The episodes of AHC were associated with nystagmus, upward and medial deviation of right eye without autonomic disturbances like sweating, vomiting, color change or respiratory disturbances. She was dysarthric when the episode was unilateral and mute when it was bilateral. The child became ataxic and disoriented during later stages of the illness. She also lagged developmentally behind her chronological age.

From the Department of Pediatrics, All India Institute of Medical Sciences, New Delhi 110 029.

Reprint requests: Dr. Veena Kalra, Additional Professor, Child Neurology Division, Department of Pediatrics, All India Institute of Medical Sciences, New Delhi 110 029.

Received for publication: November 9, 1994;

Accepted: March 10, 1995

Initial examination revealed a cooperative, conscious, alert child without any neurocutaneous stigmata or dysmorphic features. The blood pressure was normal and peripheral vessels equally palpable with no bruits. Muscle power was normal with brisk reflexes and flexor plantar response in between episodes and classic hemiparetic features during episodes. There was esotropia of right eye without affecting extraocular movements. Fundus examination and cranial nerves were normal. The EEG (both during the episodes and inter-hemiplegic period) was essentially normal except for a generalized slow rhythm. The brain stem evoked auditory responses, somatosensory evoked potentials, EMG and nerve conduction velocities were normal. A CT scan was within normal limits. Four vessel angiography was normal. The urine aminoacidogram, organic acid screen, blood levels of electrolytes, folate, vitamin B₁₂, lactate, lipids, immunoglobulins and alpha-tocopherol were within normal limits. Liver and renal function tests and creatine phosphokinase were normal. The random blood ammonia value was 63 $\mu\text{mol/l}$ and postprandial 46 $\mu\text{mol/l}$ (normal 18-80.4 $\mu\text{mol/l}$).

Considering the clinical profile and investigations a diagnosis of AHC was made. On follow-up at 12 years age her gait was diplegic. There were no cerebellar signs or nystagmus. Speech was dysarthric and intelligence subnormal, with a mental age of 5 years. The frequency of hemiplegic episodes was once every 3-4 weeks and duration of each episode was 6-8 h. The patient developed generalized seizures at 10 years of age which were treated with phenytoin.

In view of the paroxysmal nature of disease, the patient was initially treated with carbamazepine for one year without any success. She also received a calcium channel blocker, flunarazine in a dose of 7.5 ml/day at 3 years of age without decrease in frequency or duration of attacks.

Discussion

In our patient the first episode of paralysis appeared at 2 years of age. Nystagmus was present during some of the episodes. There were no associated or precipitating factors. She was symptom free in sleep even during the episodes and symptoms used to reappear soon after awakening. Evidence of developmental delay, mental retardation and neurological abnormalities including ataxia, dystonia were seen on follow up. The clinical profile suggested the diagnosis of AHC.

The criteria for diagnosis of AHC include: (i) typical onset before 10 months of age; (ii) repeated attacks of hemiplegia involving either side and generally relieved by sleep; (iii) presence of other paroxysmal phenomena such as tonic attacks, dystonic posturing, choreoathetoid movements, nystagmus or other oculomotor abnormalities, autonomic disturbances like sweating, color change and respiratory disturbances; and (iv) progressive mental and neurological handicaps resulting in severe disability as the disease advances(2,4). were seen on follow up. The clinical profile suggested the diagnosis of AHC.

The diagnosis is usually delayed despite typical manifestations. The exact cause of AHC is obscure(1,4-6). The differential diagnosis includes vascular disorders, aminoacidopathies and encephalo-myopathies with stroke like episodes(7).

The familial occurrence of AHC is reported(8). AHC complicating migraine often has a strong family history of classic migraine in at least one first degree relative(4). In our patient there was no such history. The typical onset of AHC is usually before 18 months of age(2), but this child had onset of the symptoms at two years. True seizures occur early in some patients with AHC(2) though our patient had delayed onset of seizures. The

use of carbamazepine in the first year may have suppressed an epileptic focus. Preservation of consciousness and pain during episodes was suggestive of AHC(3). The absence of hemicranial headache in our patient is in agreement with other reports(1). A normal serum lactate, amino acid and organic acid profile excluded a mitochondrial or neurometabolic disorder(7). Major vascular malformations were ruled out by a four vessel angiography(9).

Patients with AHC may show elevated levels of brain lactate during episodes(10). Cerebral hyperperfusion is reported on Tc-HMPAO scan and single photon emission computed tomographic (SPECT) imaging(9). Recent work on magnetic resonance spectroscopy of brain suggests that AHC is associated with an initial phase of relative focal cerebral hypoperfusion followed by prolonged hyperperfusion(4), akin to "luxury perfusion" of an acute stroke(11).

The lack of satisfactory response to drugs including calcium channel blockers(3,12,13) and anticonvulsants(1,3) is in agreement with the reported literature. However, treatment with flunarazine was continued in an attempt to limit the increase in severity of the condition.

REFERENCES

1. Verret S, Steele JC. Alternating hemiplegia in childhood. A report of eight patients with complicated migraine beginning in infancy. *Pediatrics* 1971, 47: 675- 680.
2. Aicardi J. Alternating hemiplegia of childhood. *Int Pediatr* 1987, 2:115-119.
3. Bourgeois M, Aicardi J, Goutieres F. Alternating hemiplegia of childhood. *J Pediatr* 1993,122: 673-679.
4. Arnold D, Silver K, Andermann F. Evidence for mitochondrial dysfunction in patients with alternating hemiplegia of childhood. *Ann Neurol* 1993, 33: 604-607.
5. Golden GS, French JH. Basilar artery migraine in young children. *Pediatrics* 1975, 56: 722-726.
6. Hosking GP, Cavanagh NPC, Wilson J. Alternating hemiplegia complicated migraine of infancy. *Arch Dis Child* 1978, 53: 656-659.
7. Pavlkis SG, Phillips PC, DiMauro S. Mitochondrial myopathy, encephalopathy, lactic acidosis and strokelike episodes. A distinctive clinical syndrome. *Ann Neurol* 1984, 16: 481-488.
8. Bradshaw P, Parson M. Hemiplegic migraine, a clinical study, *Quart J Med* 1965, 34: 65-85.
9. Aminian A, Strashun A, Rose A. Alternating hemiplegia of childhood. Studies of regional cerebral blood flow using ^{99m}Tc-hexamethylpropylene amine oxime single-photon emission computed tomography. *Ann Neurol* 1993, 33: 43-47.
10. Rust RS, Thomas A, Zupanc ML. High volume localized *in vivo* magnetic resonance spectroscopy and single-photon emission computed tomography in alternating hemiplegia. *Ann Neurol* 1992, 32: 452.
11. Lassen NA. The luxury perfusion syndrome and its possible relation to acute metabolic acidosis localized within the brain. *Lancet* 1966, 2:1113-1115.
12. Casaer P. Flunarizine in alternating hemiplegia in childhood. An international study of 12 children. *Neuropediatr* 1987,18: 191-195.
13. Casaer P, Azou M. Flunarizine in alternating hemiplegia in childhood. *Lancet* 1984, 2: 579.

