Alternating Hemiplegia of Childhood Associated With Bilateral Ptosis in A 3-Year-Old Girl

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ABSTRACT

Alternating hemiplegia of childhood is a rare and poorly understood clinical syndrome that has hemiplegia as the major feature. The syndrome has been linked to migraine, epilepsy and familial paroxysmal choreoathetosis. Cranial nerve dysfunctions are very rare in Alternating hemiplegia of childhood. We present a child with Alternating hemiplegia of childhood who had two attacks associated with ptosis. Flunarizine therapy was started to prevent a new attack of Alternating hemiplegia of childhood and she has remained free of symptoms with no residual signs during the 18-month follow-up period. As in our case, different forms of Alternating hemiplegia of childhood may be seen and an attack including hemiplegia-p toesis may become an Alternating hemiplegia of childhood episode.

Key words: Alternating hemiplegia of childhood, ptosis, cranial nerve dysfunction.

Alternating hemiplegia described by Verret & Steele in 1971 is a rare and poorly understood clinical syndrome that has hemiplegia as the major feature. The syndrome has been linked to migraine, epilepsy and familial paroxysmal choreoathetosis. Alternating hemiplegia of childhood develops in infants and young children between birth and in the first six year of life. The duration of motor attack (hemiplegia & dystonia) and nystagmus varies from minutes to days and the intensity waxes and waves during a single episode. Low intellectual status and progressive neurological deterioration is recognized with time.

Some children with migraine develop the syndrome of alternating hemiplegia, which has its onset during infancy. Acute hemiplegia may be initial manifestation of migraine and may recur, affecting on side and then the other. Frequent episodes of vasoconstriction associated with ischemia may result in irreversible cerebral injury leading to mental retardation and an episode in this subgroup of children.

Although a few cases of Alternating hemiplegia of childhood associated with facial palsy are reported, as fas as we know, no case of Alternating hemiplegia of childhood associated with ptosis has been reported.

CASE REPORT

A 3-year-old girl was admitted to our clinic because of left hemiplegia and choreoathetosis associated with bilateral ptosis. Her medical history showed that she has had a non-specific upper respiratory tract infection a week ago previously and she had a suffered from left ptosis lasting a few minutes three months before admission. Her neurological examination revealed left hemiplegia, choreoathetosis and bilateral ptosis. Her electroencephalography (EEG) displayed sharp wave activity in the right parietal region. Bilateral ptosis did not improve with intramuscular neostigmine. The results of metabolic, collagen-vascular and routine cerebrospinal fluid analysis, bacterial cultures and serologic test in CSF and / or sera, purified protein derivative skin test were all normal. The patient has followed in our clinic where she showed a marked improvement and she was discharged from the hospital after a week with no residual signs. Two months following this attack, right central facial palsy developed and this last attack resolved completely in five days. Ruling out acute bacterial infection, collagen-vascular diseases as well as the remaining causes of the remainder of childhood hemiplegia, and based on the recurrent neurological attacks and full clinical recovery of each attack, a diagnosis of Alternating hemiplegia of childhood was made. Flunarizine therapy was started to prevent a new attack.
of Alternating hemiplegia of childhood, and she has remained free of symptoms with no residual signs including normal IQ level during the 18-month follow-up period.

**DISCUSSION**

On the basis of clinical episodes with spontaneous clinical recoveries, normal cranial MRI and ruling out acute infection, our patient was diagnosed as Alternating hemiplegia of childhood. Although Alternating hemiplegia of childhood is clinically characterized with hemiplegia, dystonia, migraine and abnormal eye movements have also been reported in different cases (7, 8), but to the best of our knowledge, no case of Alternating hemiplegia of childhood associated with ptosis has yet been described yet. Alternating hemiplegia of childhood is occasionally associated with migraine, but in most cases the cause is unknown. Alternating hemiplegia of childhood develops in infants and young children between birth and in the first six year of life (2, 3). Hemisyndromes are more common in children and may be characterized by numbness of the face arm and leg; unilateral weakness; and aphasia. Rarely, both sides area involved during an attack. Choreathetosis and dystonic movements are commonly observed of the hemiparetic extremity. These hemisyndromes are more than one attack is uncommon in the pediatric age group. There are two types; benign and classic Alternating hemiplegia of childhood. Frequent episodes o vasoconstriction associated with ischemia may result in irreversible cerebral injury leading to mental retardation and epilepsy in this subgroup of children.(5,6,8-10).

It might be said that the patient had two Alternating hemiplegia of childhood attacks with ptosis, if it is considered that the first attack occurred at home. It has been reported that the attacks of Alternating hemiplegia of childhood may result from cytotoxic edema of the related parts of the brain (2) and therefore acetzolamide has been useful in Alternating hemiplegia of childhood (11). In our case other ocularmotor findings were not coexistent with bilateral ptosis. This suggests that Alternating hemiplegia of childhood can involve local neural fiber and be disordered. Facial palsy was also seen rarely in Alternating hemiplegia of childhood. We did not find any literature except for that of Houriuchi, who reported supranuclear facial paralysis (12). The patient is third attack went occurred with central facial paralysis; this showed that the number might change according to the indolent place.

The fact that the attacks of our patient ended up in full recovery suggests that the prognosis of Alternating hemiplegia of childhood is excellent, but low intellectual status and progressive neurological deterioration may develop in time (2, 3). Alternating hemiplegia of childhood can also be provoked by some other factors such as excitement, cold weather, hot weather, illness and emotional stress (2, 4). Therefore, in our case, a non-specific upper respiratory tract infection was meaningful. Provocative factors for Alternating hemiplegia of childhood may be prevented and these measures might decrease the number of attacks.

Anticonvulsant and antimigraine medications have been used to prevent attacks or progression. As far as calcium channel-blocking agents including flunarizine have been useful (4). Thereafter, other medications such as IVIG, amantadin, carnitine, acetazolamide and corticosteroids are recommended to reduce the frequency of attacks (4,9,11-13). In our patient, flunarizine therapy was started to prevent a new attack of Alternating hemiplegia of childhood and she has remained free of symptoms with no residual signs during the 18-month follow-up period.

In conclusion, as in our case, different forms of Alternating hemiplegia of childhood may be seen and an attack including hemiplegia-ptosis may become an Alternating hemiplegia of childhood episode.

**REFERENCES**


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