Oculomotor Ophthalmoplegic Migraine in an Infant

R. Amit, M.D.* and D. Benezra, M.D.**

Departments of Pediatrics*, and Ophthalmology**, Hadassah University Hospital, Ein-Kerem, Jerusalem, Israel.

Reprint requests to: Rami Amit, M.D., Dept. of Pediatrics, Hadassah University Hospital, Ein-Kerem, P.O.B. 12000, Jerusalem, Israel.

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SYNOPSIS

Ophthalmoplegia is a rare presentation of migraine. Ophthalmoplegic migraine is frequently encountered in children, but has rarely been observed in infancy. We report a case of an infant with paroxysmal episodes of vomiting followed by a few weeks long periods of oculomotor nerve palsy. The first episode occurred at 14 months of age and the second at two years. In both episodes full recovery of the ophthalmoplegia occurred without treatment. The differential diagnosis of painful ophthalmoplegia and the variety of therapeutic regimens are discussed.

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INTRODUCTION

The occurrence of third nerve palsy in the pediatric age group is unusual, and the differential diagnosis of such events includes a variety of etiologic causes, such as aneurysm and transtentorial herniation, which bear serious implications.1 Ophthalmoplegia is a rare presentation of migraine. The occurrence of ophthalmoplegic migraine (OM) in infancy has been reported to date in only a few cases.2,3 We report an infant who presented with paroxysmal incomplete external oculomotor nerve palsy during the first attack, and involvement of the internal oculomotor during the second episode, six months later. The possible migrainous origin and the differential diagnosis of isolated painful ophthalmoplegia are discussed.

CASE REPORT

A 14-month-old female infant was admitted for investigation because of recurrent vomiting and an acute ophthalmoplegia of the left eye. She was the product of a normal full-term pregnancy, and was delivered by cesarian section because of fetal distress. The developmental milestones were appropriate for her age: she could sit at 5 months, stood up at 10 months, and was able to walk with assistance prior to her first admission at 14 months of age. She is the first child born to her unrelated, Ashkenazi Jewish, 36-year-old parents. The father is healthy and the mother suffers from migraine headaches.

The patient was initially hospitalized after recurrent episodes of vomiting of a week's duration. On the day of her admission, a left exotropia and hypertropia, with paresis of the medial and inferior recti, were noted. On examination she seemed comfortable and was afebrile. Weakness of adduction of the left eye was noted both on monocular and binocular movements. Convergence attempt failed to adduct the left eye. There was no ptosis. The pupils were equal and reacted to direct and consensual light stimuli. The fundi appeared normal. The rest of the physical and neurological examinations were normal. Laboratory workup that included erythrocytes sedimentation rate (ESR), complete blood count, urea, glucose and electrolytes, was normal. Computerized tomography of the head, with and without contrast was unrevealing. During the hospitalization she was treated only by partial occlusion of the sound right eye. The vomiting and the exotropia improved gradually. On follow up examination, three weeks later, the child was orthophoric with full eye movement, and resumed normal activities.

At the age of 2 years a second episode of vomiting, exotropia and hypertropia of the left eye occurred. The child was slightly apathetic and covered her left eye. It was difficult to assess whether the reason for covering the eye was ocular pain or diplopia. On examination, weakness of the left medial and inferior recti with dilated and hyporeactive pupil on the same side, were noted. ACT scan of the head with special views of the orbits, and a waking EEG, were normal. Because of the marked exotropia of 45 prism diopters and the non-alternating pattern of fixation, partial occlusion of the sound eye was again advised. Within 4 weeks, the exotropia again resolved completely with full eye movement and good binocular function.

DISCUSSION

The migrainous origin of the ophthalmoplegia in the case presented here is suggested by the following: the paroxysmal and temporary nature of the disturbance, the nausea and vomiting that preceded the ophthalmoplegia, the maternal history of migraine headaches, and above all the exclusion of...
other possible etiologies of third nerve palsy. Although the symptom of headache may be hard to evaluate in an infant, it is possible that during the second episode our patient suffered from pain around the left eye.

OM is encountered more frequently among young adults and children, and males are affected more than females.\textsuperscript{1,4,5} It has only rarely been reported in infancy,\textsuperscript{2,3} and the youngest patient to be reported to date was an 8-month-old female infant.\textsuperscript{2} In most of the cases the oculomotor nerve was affected both externally and internally.\textsuperscript{1} Less often the abducent nerve was involved.\textsuperscript{1,6} The case reported here is unusual due to the young age of the patient and incomplete involvement of the oculomotor nerve. In the first attack only the medial and inferior recti were involved. During the second episode, along with the medial and inferior recti, internal ophthalmoplegia of the sphincter pupilla of the left eye was observed. Recurrence of the oculomotor weakness on the same side, and persistence of the weakness for a few weeks, although rare, has been reported previously.\textsuperscript{1}

The differential diagnosis of painful ophthalmoplegia includes vascular malformations, inflammatory processes, trauma and neoplasia. Congenital aneurysm rarely becomes symptomatic during childhood. This possibility, as well as that of neoplasia, were excluded in our patient by the normal CT. Tolosa-Hunt syndrome, an inflammatory process involving the orbital fissures, is accompanied by elevated ESR, and responds favorably to steroids. The normal CT scan with special orbital views and the normal ESR excludes this possibility in our patient. Other causes of ophthalmoplegia, such as myasthenia and viral encephalitis, are unlikely in view of the paroxysmal and intermittent nature, and the clinical course described in our patient.

The cause of oculomotor nerve weakness in OM is believed to be due to compression of the nerve by different mechanisms, such as: transtentorial herniation of the temporal lobe,\textsuperscript{8} transient non-aneurysmal enlargement of the posterior cerebral artery,\textsuperscript{9} or of the intracavernous carotid artery.\textsuperscript{10} Treatment of OM with prednisone has been proposed by Smith et al.\textsuperscript{7} These authors report that an early treatment, within a few hours of the onset of the headache, resulted in decreased severity of the headache and shortened the time of diplopia. Prevention of recurrences may be important for the preservation of normal oculomotor function.\textsuperscript{3} Propranolol, which had been used for prophylaxis of migraine,\textsuperscript{5} has been used successfully in a 12-month-old infant as a preventive measure against recurrent episodes of migraine.\textsuperscript{3} Kuzemko el al reported a remarkable beneficial effect of ergotamin tartarate in the beginning of a migraine attack in an 11-year-old boy with OM.\textsuperscript{4} In our patient the symptoms in both attacks lasted for 3-4 weeks without treatment. Because of the severe exotropia that followed each attack and the possible suppression effect on visual input that might ensue, we believe that partial occlusion of the sound eye is mandatory in these cases in order to avoid amblyopia. The beneficial effects of prophylactic or symptomatic medical treatment of these patients have to be considered in different cases.

REFERENCES