
OPHTHALMOPLAGIC MIGRAINE

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MIGRAINE

Migraine is a periodic and paroxysmal protean disorder that affects nearly 10 % of the population. Many of its manifestations produce neuro-ophthalmologic symptoms & signs; which should be recognized by the clinician. The term migraine stems from Galen's usage of hemicrania to describe a periodic disorder comprising of paroxysmal blinding hemicranial pain, vomiting, photophobia, recurrence at regular intervals and relief by dark surroundings and sleep.

CLINICAL DEFINITION OF MIGRAINE

To make a diagnosis of Classic or Common migraine; the patient must have recurrent headaches accompanied with at least three of the following six criteria:

- Recurrent abdominal pain (with or without headache), nausea and vomiting (with headache).
- Hemicrania
- A throbbing, pulsatile quality to the pain.
- Complete or substantial relief after a brief period of rest.
- An aura which can either be visual, sensory or motor.
- A history of migraine headaches in one or more members of the immediate family.

THE WORKING DEFINITION FOR MIGRAINE

Benign recurrent headaches and or neurologic dysfunction usually attended by pain free interludes and almost always provoked by stereotyped stimuli. There is a hereditary predisposition towards attacks and the cranial circulatory phenomena that attend the attacks appear to be secondary to a primary C.N.S disorder.

Migraine gives rise to a number of well recognized syndromes', as well as a variety of "equivalents" less commonly considered as migraine. The symptom complexes or syndromes of migraine include Common migraine, Classic migraine, Complicated migraine, Cluster headaches and migraine equivalents.

COMPLICATED MIGRAINE

Migraine accompanied by prolonged visual or neurological deficits or by mental aberrations. Migraine in which the symptoms outlast the headache by more than 24 hours.

OPHTHALMOPLAGIC MIGRAINE

In 1860, Gubler first described a patient who had recurrent episodes of oculomotor nerve paresis during otherwise typical migraine attacks. This oculomotor nerve paresis persisted beyond the headache phase of the attack. Charcot in 1890 called this condition " Ophthalmoplegic Migraine".

DIAGNOSTIC CRITERIA

Walsh and O'Doherty outlined specific criteria essential for the diagnosis of Ophthalmoplegic Migraine.

History of typical migraine headache:

The headache is severe and throbbing in nature. Headache usually is unilateral, but occasionally bilateral or alternating.

Headache is typically of the crescendo type and may last several hours or days.

Ophthalmoplegia – including one or more nerves and may alternate sides with attacks.

Extraocular muscle paralysis may occur with the first attack or rarely precede it.

Exclusion of the other causes; by arteriography, surgical exploration or autopsy.

AGE OF ONSET

The majority of the patients experience their initial attack in the first decade of life, usually before five years of age. Ophthalmoplegic migraine most often occurs in infancy or early childhood, thus the first attack is usually incorrectly attributed to some other process such as trauma, aneurysm, infection or even recent immunization; only when the condition resolves and then recurs is the correct diagnosis made.

Rarely patients experience their first attack in adulthood, but such patients almost always have a history of typical migraine headaches with and without aura since childhood, a family history of migraine or both. Therefore never make a diagnosis of Ophthalmoplegic Migraine in an adult unless :

There is a strong family history of migraine.

The patient has a history of other types of migraine in the past.

Other causes of painful ophthalmoplegia have been excluded by appropriate laboratory and neuroimaging studies.

SEX

Ophthalmoplegic migraine does not occur more often in women than in men, unlike most other forms of migraine. It has a male predominance or occurs equally in both sexes, perhaps because it

almost always begins in childhood and the incidence of migraine is the same in both sexes until menarche.

FAMILY HISTORY

Rare; though not unknown, for ophthalmoplegic migraine to have occurred in other family members; but for other varieties of migraine like "Common" or "Classic"; a family history is invariably found.

PATTERN OF PRESENTATION

This condition follows remarkably constant pattern – the most important aspect of this syndrome. If the pattern is not followed then there is probably some cause other than migraine for the clinical symptomatology.

Pain, not oculomotor paresis, is the initial symptom, but because of the young patients in which it frequently occurs; the pain may not be clearly defined or localized. The older children describe pain in, around and above one eye, purely unilateral at first, but at the height of severity pain becomes less localizable. The

oculomotor nerve is most commonly affected. Ptosis begins at the time of severe pain and then followed by diplopia, which ceases when the ptosis becomes complete. The pupil is almost always affected, although in many cases it is only mildly dilated and reacts sluggishly to light and near stimulation. Nevertheless some patients with ophthalmoplegic migraine have a complete oculomotor nerve paresis except for total sparing of pupil.

The 3rd nerve of total oculomotor palsy takes about 10 – 18 days to resolve completely. Recovery is seen as improvement of ptosis and improved ocular motility or pupillary constriction to light or near stimulation.

It is the behavior of the subsequent attacks that makes a clinical diagnosis possible, after a complete symptom free interval of several months to 2 years, another identical attack will occur. Here however may come the most important diagnostic

feature, the symptoms and signs, still unilateral, affect solely the side opposite from the first occasion. This is most dramatic when the ophthalmoplegia is complete, for the alteration from side to side in different attacks could be mimicked by only the most unlikely coincidental, identical, bilateral structural lesions capable of behaving in an identical manner, hence is virtually diagnostic of ophthalmoplegic migraine.

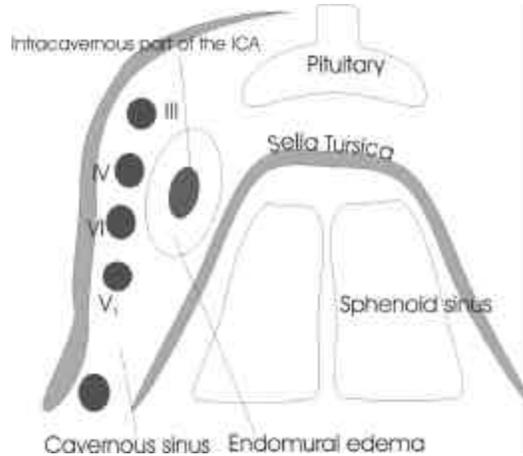
Gradually as the child gets older these attacks die out, often replaced by the more familiar varieties of migraine.

The Trochlear and Abducens nerve are affected less often in ophthalmoplegic migraine. More than one oculomotor nerves may be affected during an attack of ophthalmoplegic migraine. The Abducens

nerve is involved alone in 10% of the cases and occasionally oculomotor palsy is accompanied by trochlear, Trigeminal, facial or hypoglossal nerves.

PATHOGENESIS

The pathogenesis is unclear. The unilateral nature of the headache and the oculomotor nerve paresis that begins with ptosis and ends with complete paresis suggests a lesion in the peripheral portion of the oculomotor nerve. Two theories have



been suggested :

Compression theory

Ischemic theory

Some have ascribed ophthalmoplegic migraine to compression of one or more of the oculomotor nerves by a dilated or oedematous intracavernous portion of the Internal Carotid Artery. Angiographic studies have shown narrowing of the Intracavernous portion of the I.C.A.

A more plausible explanation for ophthalmoplegic migraine is that during an attack, a reduction in the blood flow through the I.C.A and perhaps through the posterior cerebral artery or Basilar artery, reduces flow to one or more of the oculomotor nerves, producing an ischemic oculomotor paresis. Since about 2/3 rd of the patients with ophthalmoplegic migraine characterized by oculomotor nerve paresis have a pupil that is completely spared or incompletely affected, an

ischemic process is more likely than a compressive lesion.

Other theories of the pathogenesis of ophthalmoplegic migraine are :

- Swelling of the posterior cerebral artery
- Pituitary swelling
- Unilateral brain swelling

SEQUELAE

As a rule oculomotor paresis recovers completely. Some children may have several attacks and may be left with permanent slight degree of ptosis, residual 3rd nerve paresis with slight pupillary dilatation. However diplopia is rare because of visual suppression. Some patients may develop secondary oculomotor synkinesis.

DIFFERENTIAL DIAGNOSIS AND INVESTIGATIONS

Features favouring the diagnosis of ophthalmoplegic migraine include concomitant or subsequent headaches that fulfil the criteria for the diagnosis of migraine and a family history of migraine, along with elimination of other possible causes, which invariably requires a CT scan. Included under the differential diagnosis of ophthalmoplegic migraine are :-

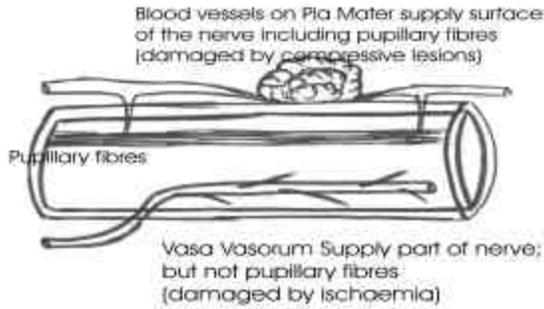
- Tolosa Hunt syndrome
- Oculomotor nerve schwannoma
- Aneurysms
- Tumors
- Diabetes
- Sphenoidal sinus mucoceles
- Myasthenia gravis
- Intermittent angle closure glaucoma with mydriasis

When the oculomotor nerve only is affected, the primary concern is an Intracranial aneurysm; even in a young child. The most common site of such an aneurysm is at the junction of the internal Carotid and the posterior communicating arteries or at the junction of the Basilar and the superior cerebellar arteries, at the tip of the basilar artery, within the cavernous sinus. CT scan & MRI permit identification of an aneurysm; and cerebral angiography is not necessary when either CT or MRI studies give normal results, particularly in children.

Myasthenia can be ruled out if the pupil is

involved and also to response to edrophonium (tensilon).

Increased intracranial tension can cause herniation of the hippocampal gyrus , producing an oculomotor paresis which maybe transient and recurrent and associated with severe headache.



Diabetic ophthalmoparesis – rarely occurs in children, beside the ophthalmoparesis that occurs in patients with diabetes mellitus, hypertension, giant cell arteritis , other systemic vasculopathies

persist longer than the ophthalmoplegia associated with migraine.

Mucocele of the sphenoidal sinus can cause painful ophthalmoplegia; as well as inflammatory lesions and tumors that invade the cavernous sinus.

In a few cases of ophthalmoplegic migraine the MRI showed enhancement and enlargement of the cisternal portion of the oculomotor nerve which spontaneously resolved after two and four years. Persistence of clinical recurrences was associated with long lasting presence of MRI findings.

TREATMENT

Very little evidence is available that there is any effective treatment for a particular attack, which in any case is self limiting. Ergotamine preparations are of no value. Attempts to treat with steroids to reduce the endomural oedema have raised equivocal results, but if one started treatment at the very onset of an attack, this might constitute a measure for ending the attack swiftly.

IS OPHTHALMOPLAGIC MIGRAINE MIGRAINOUS ?

The characteristics of headache and the frequent lack of associated symptoms during so called ophthalmoplegic migraine attacks indicate that the condition is not migrainous. The most likely possibility appears to be an orbital or retro- orbital inflammatory reaction – The Tolosa – Hunt Syndrome, which consists of recurrent attacks of orbital and periorbital pain and ophthalmoplegia.

When the clinical features of ophthalmoplegic migraine and Tolosa – Hunt Syndrome overlap, a positive MRI finding is one of the diagnostic criteria in the classification of Ophthalmoplegic Migraine and a trial of steroids is worthwhile in the

presence of enhancement of the oculomotor nerve, since ophthalmoplegic migraine may be non – infectious but inflammatory in etiology.

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