

CHILDHOOD HEADACHE SYNDROMES (PART I)

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Abstract

Objective

Headache is one of the most common reason that children are referred to the Pediatric Neurology Services. It is said that ten percent of children aged 5 to 15 years have migraine.

Subsequently, it is important for clinician to have a thorough, comprehensive and systematic approach to the evaluation and management of the child or adolescent who complain of headache.

This writing aims to explore the symptoms of headache, its epidemiology, classification, appropriate evaluation, differential diagnosis and management.

Headaches are divided into primary and secondary categories. Migraine and tension type headaches are prototype of primary headaches without underlying pathology. On the other hand, the type of headache which stems from organic diseases such as: brain tumor, increased intracranial pressure, systemic disease, drug toxicity or Ear Nose and Throat problems, are considered secondary.

On the whole, the majority of children with primary headache have two patterns of headache. One is a chronic low-grade and the other is an intermittent disabling headache. The cause of the former is either caffeine or analgesic abuse, and the latter is predominantly migraine.

Traditionally, if a child presents himself with chief complain of headache, care taker physician begins with history taking followed by thorough physical and neurological examinations.

In the majority of the cases, this initial process leads to a diagnosis or indicate the need for further testing.

Once the diagnosis is made, a program of management comes into being.

Keywords: Headaches, Children, Migraine

Introduction

Headache is a common problem in pediatric age group with up to 75% of children reporting a notable headache by the age of 15 years (1).

In one report, 59% of boys and 84% of girls between the age of 13 and 18 reported having experienced a headache within the past month (2). Also it is said that ten percent of children aged 5 to 15 years have migraine (3).

The effect of headache on the child's daily life including academic performance, school attendance, personality and social relationship is significant and its degree of influence depends on its etiology, frequency and severity.

As it is known, headache may be primary or manifestation of an underlying severe disorder like brain tumor. Therefore, having a child who suffers from recurrent,

progressive and severe headache, caretaker physician should feel careful evaluation of that child an obvious mandate.

As far as the primary headache is concerned, migraine is almost the only one that has the greatest effect on a child quality of life. Most migraine headaches are not severe and are easily managed by conservative measures without requiring medical attention.

Epidemiology

The modern era of the study of headache in children and adolescents was initiated by Bill in 1962 (4). He reported an extensive study of the epidemiology of pediatric migraine in 8933 children. Migraine was reported in 3.9% of children aged 7-15 years which increased to 5.3% in 15-year-old children.

Subsequent epidemiological studies have continued to show the high frequency of headache in children and adolescents with migraine being the most common type (5).

In a recent epidemiological study of 2669 children in Istanbul, Turkey, 46.2% of children aged 5-13 years reported having a debilitating headache, with 3.4% of these children, having migraine and a further 8.7% having probable migraine (6).

In a recent Italian study of 4386 adolescents aged 11, 13, and 15 years, 40% reported having at least one headache a week (7).

In 2351 individuals aged 15-19 years suffering from headache, Split and Newman, found that up to 28% of them had migraine, 19% having migraine without aura and 9% migraine with aura (8).

Of 10169 individuals aged 12-29 years, Stewart and co-authors found that the onset of migraine with or without aura was significantly earlier in male participant than in female with aura occurring in nearly a quarter of individuals (9).

Classification

Prior to 1998, the classification of headache was not uniform, and diagnostic criteria were not based on operational rules.

In 1998 The International Headache society (IHS) proposed a classification system for headache that the second edition of it was published in 2004.

Several pediatric authors have reviewed the initial IHS classification and have suggested that it is not applicable for children and adolescents.

Prior to and along with these (IHS) classifications, other clinical methods of classifying pediatric and adolescents headache have been proposed.

Winner and Rather suggested a clinical classification utilizing both the temporal pattern of a child's headache, plotted against its severity over time (10).

In this classification, five patterns were identified including:

Acute, Acute Recurrent, Chronic Progressive, Chronic non progressive and "Mixed "or Co-morbid headache (11).

Acute Headache

An acute headache is a single event with no history of a previous similar event. It may be generalized or localized and may be associated with neurological symptoms and signs or without them.

If an acute headache is noted in a critically ill child, a diagnosis needs to be made quickly and intervention may be lifesaving.

The differential diagnosis of acute headaches involves a wide variety of general medical as well as central nervous system etiologies.

Acute Recurrent Headaches

These types of headaches are separated by pain-free intervals. When an acute recurrent headache is associated with nausea, vomiting, photophobia and phonophobia, the headaches are usually migrainous in nature.

Chronic Progressive Headaches

These kinds of headaches get worse in frequency and severity over time. The progression may occur rapidly or slowly. The headaches may be accompanied by symptoms and signs of increased intracranial pressure or progressive neurologic disease.

The neurologic examination is frequently abnormal. An organic process is usually present and further testing is usually indicated.

Chronic Non- Progressive Headaches

Different names have been used to describe these types

of headaches, such as : Tension-type Headache, Muscle Contraction Headache, Chronic Daily Headache and Chronic nonprogressive headaches.

Chronic tension-type headache are usually not associated with symptoms of increased intracranial pressure or progressive neurologic disease. The neurologic examination is normal. Factors relating to school, stress, family dysfunction and medication overuse are frequently noted. Lipton have recently subdivided chronic daily headaches into chronic tension-type headaches, hemicranias continua, transformed migraine and new daily persistent headaches (12).

Mixed or Co-morbid Headaches

Some feel that these types of headaches also known as “the mixed headache syndrome” are the transformed migraine or chronic migraine which a number of years later evolve into chronic daily vascular headache.

Chronic daily Headache

Chronic daily headache are one of the most frequent types of headaches seen in the adolescent population. Some believe, chronic daily headache consists of combination of acute recurrent headaches and chronic non progressive one which is difficult to differentiate between it and co morbid headaches, transformed migraine and chronic migraine (11).

Pathophysiology of Headache

If indeed there are various distinct classes of headache, the pathophysiology of each type must be discussed separately. Both extracranial and intracranial structures are sensitive to pain. The sensitive extracranial structures include: the skin, subcutaneous tissues, muscles, mucous membranes, teeth and some of the larger vessels. Intracranial structures which are pain sensitive include: the vascular sinuses, larger veins and dura surrounding these structures and arteries at the base of the brain. The brain itself and most of the dura, ependyma and choroids plexus are insensitive to pain.

Any process causing inflammation, displacement, irritation, traction, dilation or physical invasion of any of these pain sensitive strictures will cause pain referred to either the face, top of the head, back of the head or neck (11).

Pathophysiology of Migraine

Knowing that pediatric migraine is the most frequent recurrent headache, it is wise to discuss the pathophysiology of this type of headache separately.

In 1938 Graham and wolf proposed the vascular theory of migraine (13).

This theory of migraine pathophysiology held that an attack has a prodromal phase marked by an aura characterized by vasospasm and second phase of intracranial vasodilatation responsible for pulsating headache.

It now has been shown that the aura is rarely accompanied by ischemia and the onset of headache occurs at a time when cortical blood flow is reduced and therefore not caused by vasodilatation (14). The vascular theory has been supplemented by a theory combines the vascular theory and the neuronal theory which is generally referred to as “trigeminal-vascular theory”.

This theory proposes that classical migraine (migraine preceded by an aura or other focal syndrome) is related to a paroxysmal depolarization of cortical neurons. During the initial phase of attack, a cortical spreading depression is elicited at the occipital pole of the brain.

The term “cortical spreading depression “ is used to describe a depression of spontaneous EEG and other cortical electrical activities spreading across the cerebral cortical surface. Propagation of the cortical spreading depression to the pain-sensitive meningeal trigeminal fibers that innervate the intracranial and dural blood vessels is believed to induce the headache.

Pathophysiology of Tension-type Headache

The term tension type headache implies that the basis of the pain seen in this disorder is related to muscle which is not really the case. Chronic tension type headache has many of the features of migraine but lacks the severe problem with nausea, photophobia and phonophobia. They also lack the usual triggering association such as menses, missing meals or altering sleep pattern. Possible mechanisms include genetic aspects, muscle mechanisms and central and/or peripheral sensitization. Stress also is identified as a trigger agent.

Neurophysiology of Cluster Headache

The clinical condition closely resembles paroxysmal

hemicrania or short lasting unilateral neurologiform headache with conjunctival injection and tearing.

There are three major aspects of the pathophysiology of this disorder: the trigeminal distribution of the pain, the autonomic features associated with the pain and the episodic pattern of the attacks (15).

The majority of patients affected are male and the neuroendocrine abnormalities of testosterone levels during cluster headaches have been known for over 3 decades (16).

Genetics

Even the familial transmission of migraine has been known for centuries, and an autosomal dominant pattern was implicated, current genetic data do not support this pattern. However, over the last few years, twin studies have supported a strong genetic component in the etiology of migraine, with a significant higher concordance rate among monozygotic twins as compared with dizygotic twins (17,18).

Clinical Manifestations

A child with acute Headache

The key to a correct diagnosis is a properly obtained history and thoroughly performed general physical and neurological examination. When it comes to a good understanding of the headache itself, the questions should be directed at the patient with the parent not participating until the entire history has been obtained from the child. Patients as young as 4 or 5 years of age may contribute significantly to a better understanding of their headache.

Specific questions as contained in the "Headache Data Base", help clinician to arrive at a specific headache diagnosis (10 & 11). Other questions that are related to the presence or absence of increased intracranial pressure, progressive neurologic disease, quality of life and impact upon daily activities follow. Important clues regarding potentially ominous headache include: the severity of headache, changes in a chronic headache pattern, consistently localized pain, pain that awakens the patient at night, pain associated with neurologic symptoms or signs and the patient declaring that "this is the worse pain I have ever had". The family history is quite important from both a genetic and environmental

perspective. Migraine is recognized as a familial disorder. Both migraine and non progressive headache often have a stress-related component and the latter occurs more frequently in dysfunctional families. The general physical examination may disclose abnormalities that are related to or causing the headache.

Examples of these cases include: elevated temperature or blood pressure, the presence of "café au lait" spots or other diagnostic skin abnormalities, short stature and tenderness over a specific localized area of scalp or skull. The neurologic examination should seek out signs of trauma, neck rigidity, head circumference, the presence of bruit, abnormalities of eye movement and/or the fundus. If the neurologic examination is abnormal, an underlying primary or secondary neurologic disorder should be suspected. The patient affect should be monitored throughout the examination and may be suggestive of stress or psychologically related problem. Once again in the majority of patients with migraine or stress related headaches, both the general physical and the neurologic examination are normal. After the initial history, physical and neurological examinations are completed, a differential diagnosis should be considered.

Combining the above with the clinical classification previously outlined, the tentative diagnosis is made. If a patient has intermittent headache with nausea and vomiting and no neurologic symptoms or signs, if these headache are indeed intermittent and separated by pain free intervals, and if there is a positive family history of migraine, the diagnosis of migraine should be suspected and no further laboratory interventions are needed.

On the other hand, if the patient has a relatively short history of a headache that is worsening quickly over time and is associated with neurologic symptoms and/or abnormalities on the neurologic examination, an organic disorder should be given a consideration. Further laboratory tests are generally indicated.

When a patient tells you that the headache is severe and at the same time appears to be in no distress, has no symptoms of increased intracranial pressure or progressive neurologic disease, and has a normal neurologic examination, then stress related chronic daily non progressive headache should be considered.

Laboratory Tests

Laboratory tests should be ordered based upon the history, character and temporal pattern of headache, physical and neurologic examination and differential diagnosis.

Most primary headaches can be diagnosed without diagnostic testing using a comprehensive history and neurologic and focused general examinations.

In some cases, however, diagnostic testing is necessary to distinguish primary from secondary causes that may share similar features (19).

The choice of which of the many laboratory tests should be ordered, rests upon this differential diagnosis.

Routine testing such as CBC, ESR, metabolic survey, thyroid function, and ANA profile are rarely indicated. On the other hand, if a patient is clinically ill, or history is suggestive of the presence of an underlying systemic or a neurologic disease, above mentioned tests plus further laboratory investigations are necessary.

These include: serum lead level, toxicology screen, neurologic testing for autoimmune disease, coagulation studies and titers for infectious diseases (11).

Neuroimaging

Guidelines for the evaluation of children with headaches have been developed through collaboration between the American Academy of Neurology, The Child Neurology Society and the American headache society (1 & 20).

The authors of these guidelines reviewed the available evidence for the evaluation of children with headaches and found that neurologic examination is the most crucial test for identifying potential serious complications, while also combining that occipital location warranted further evaluation.

In most cases of primary headaches that are long standing, recurrent and do not change with a normal neurologic examination, imaging studies are not necessary.

When further evaluation is needed, an MRI examination is the most sensitive test to identify structural abnormalities and should be the preferred neuroimaging test (21).

Even it is said the EEG is of limited value in the routine evaluation of headache in children (22). There is a report from this writer and my colleagues of a patient who presented with debilitating headache and severely abnormal EEG.

That child's headache was found to be a sole

manifestation of non convulsive status epilepticus. The patient's headache was relieved by antiepileptic drug and subsequently his electroencephalogram became clear (23).

CT scan is a useful diagnostic procedure under emergent circumstances and should be used to identify subarachnoid hemorrhage, ventricular enlargement, abscess, mass lesion and hemorrhage secondary to trauma (24).

However as mentioned above, if the diagnosis of a central nervous system lesion is suspected and the situation is not urgent, MRI at time coupled with Magnetic Resonance Angiography and/or Magnetic Resonance Venography (MRV) should be considered. It should be noted that as many as 40% of individuals, imaged for headache may have nonspecific abnormalities, including abnormalities of sinuses, nonspecific white matter abnormalities, arachnoid cysts, pineal cysts, venous angioma and chiaria malformation (25).

Lumbar puncture is useful when one suspects an infectious disorder or the presence of pseudotumor cerebri (11).

At the conclusion of the history, physical and neurologic examination and reviewing results of indicated paraclinical investigations, etiology of headache should be suspected. It is unjustified to start treatment unless a clear cut diagnosis has been made.

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