Paparella: Volume I: Basic Sciences and Related Principles

Section 9: Otolaryngologic Manifestations of Systemic Diseases and Pain

Chapter 42: Headache

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Headache is probably the most frequent complaint in medicine. This familiar problem afflicts 80 per cent of the population in any given year, and 50 per cent of sufferers have severe and disabling headaches. Mathew has claimed that as many as 20 per cent of all women suffer from migraine. Each year between 10 and 20 per cent of the population consult physicians with headache as their primary symptom. Huge amounts of nonprescription analgesics are consumed annually in the United States - 16.000 tons of aspirin or a half-tablet per person - mainly for relief of pain around the head.

Most headache sufferers have neither a severe physical nor emotional problem. Patients and physicians are keenly aware, however, that some headaches can be symptomatic of serious disease.

Most headaches are benign and secondary to some relatively benign illness, which is usually viral and febrile, or they can be secondary to various disorders affecting the nervous system or structures about the head. Headaches can also be *primary;* such as tension headaches and vascular or migraine headaches.

The evaluation of the patient with headache by appropriate history, examination, and laboratory studies usually enables the physician to make the correct diagnosis and to treat the patient successfully.

Pain-Sensitive Structures About The Head

Although the brain parenchyma itself is probably insensitive to pain, this is not true for its integral and surrounding structures. The venous sinuses, parts of the dura mater, especially at the base, the arteries of the dura, and the cerebral arteries of the base of the brain are all pain-sensitive. The cranium, the actual brain substance, most of the dura and arachnoid, the ependymal lining of the ventricles, and the choroid plexuses are insensitive to pain.

Headache is thought to result from four basic mechanisms: traction, distention, inflammation, or direct pressure. Traction on the veins that pass to the deep venous sinuses, on the middle meningeal arteries, or on large arteries at the base of the brain and their main branches produces head pain. Distention and dilatation of intracranial arteries can also cause pain. Inflammation in or about any of the pain-sensitive structures of the head causes pain. Direct pressure on the cranial and cervical nerves containing pain fibers produces pain. In turn, by any

of these mechanisms, the pain can be localized or generalized.

The search for the substance(s) that produce the pain of migraine remains a major challenge, despite many studies in the last 50 years. Vasoactive substances and related agents have been tested, but most have not yielded consistent results. These substances include epinephrine, serotonin, bradykinin, histamine, tyramine, prostaglandins, endorphins, enkephalins, and substance P. Dyslipoproteinemic states, platelet abnormalities, and disturbance of brain oxygenation have also been implicated. Probably, several mechanisms contribute to migraine pathophysiology.

Clinical Classification

Head pain can be classified in several ways (see above), including primary and secondary headache. Primary headache is not caused by general medical diseases, diseases affecting the nervous system, or diseases of structures of the head and neck. Thus, primary headache disorders are comprised of two basic syndromes that can occur distinctly or that can occasionally merge: tension headache and vascular or migrainous headaches. Secondary headache is a result of general medical diseases, diseases affecting the central nervous system, or diseases of structures of the head.

Clinically, we recommend the headache classification scheme proposed by Rothner and others that uses a temporal and graphic method, rather than methods based on the presumed location of the abnormality, the symptom complex, or the pathophysiology.

This useful classification divides headaches as follows:

1. Acute: a single headache event with no previous history of a similar event.
2. Acute recurrent: these can be subclassified into localized or generalized.
3. Chronic and progressive.
4. Chronic and nonprogressive.

Acute Headache

The differential diagnosis of the first-time acute headache encompasses a wide spectrum and can even include life-threatening conditions, especially if accompanied by other neurologic symptoms and signs. Acute headache can accompany the following: systemic infection, especially febrile illnesses; central nervous system infection, especially meningitis and encephalitis; hemorrhage and/or thrombosis within the central nervous system (CNS); trauma; sudden elevations of intracranial pressure; and acute sinusitis.

Acute headache can also be seen if toxins are ingested or absorbed (eg carbon monoxide, lead) or with anemia, hypoxia, electrolyte imbalance, hunger, hangover, hypoglycemia, exertion, or lumbar puncture, following epileptic seizures, or in association with collagen-vascular disorders, neoplastic diseases (eg leukemia), temporal arteritis, and trigeminal neuralgia.

Generalized and Localized Headache

Thus, an acute headache occurring for the first time can be difficult to diagnose. Its classification can be usefully separated further into those suffering from generalized pain and those in whom the pain is localized. Dhopesh and colleagues studied acute generalized headache in an emergency room setting and found the following breakdown of causes; non-CNS infections, 10 per cent; tension, 20 per cent; trauma, 10 per cent; migraine, 5 per cent; and hypertension, 5 per cent. The other 15 per cent comprise equally serious or life-threatening conditions, such as increased intracranial pressure, subarachnoid hemorrhage, central nervous system leukemia, and the exercise-induced syndrome.

Wald noted that acute localized headache is usually caused by one of the following: sinusitis, otitis, orbital cellulitis, optic or retrobulbar neuritis, orbital pseudotumor, dental problems, temporomandibular joint problems, trauma, or occipital neuralgia.

Migraine

Acute recurrent headaches occur periodically in patients who are not ailing between episodes. Most cases of recurrent severe headaches occurring in patients who are asymptomatic in between the attacks are caused by migraine in one of its forms.

Migraine can be defined as a group of constitutional disorders characterized by paroxysmal disturbances of cephalic neurovascular function and presenting mainly as episodic headaches associated with autonomic, visual, somatosensory, and/or psychological symptoms of variable prominence. Migraine is commonly subdivided into classic (with aura), common (without any aura), complicated, and cluster headaches.

Common Migraine. In common migraine the headache is the conspicuous feature and there is little neural accompaniment. It is the most frequent type, occurring in up to 15 per cent of the population, with a 3:1 female-to-male predominance.

In children of grad-school age, 5 to 10 per cent have migraine, and 50 per cent of patients have their first attack before the age of 20. The family history is often strongly positive, although migraine can skip a generation. Preadolescent boys are more frequently affected than girls but, after puberty, girls are more commonly affected. The attacks can occur for no obvious reason but many precipitating factors are recognized, including stress, hunger, fatigue, menses, illness, exercise, minor head trauma, and environmental changes.

The prodrome in common migraine can come on hours before the headache. It includes behavioral changes and irritability, gastrointestinal disturbances, and autonomic symptoms, such as pallor.

The headache itself can be one-sided (in two-thirds of adults) or bilateral. The pain is pulsatile, throbbing, and disabling. The patient frequently also experiences anorexia, nausea and

vomiting, and abdominal pain.

Fatigue, photophobia, and phonophobia accompany the attack. Thus, the patient often voluntarily goes to a quiet, darkened place and tries to go to sleep.

The attack lasts from 2 to 6 hours or longer. The frequency is usually once or twice per month, sometimes weekly but seldom more often.

Classic Migraine. In classic migraine there is headache with conspicuous neural accompaniment, usually in the form of visual symptoms. It occurs in less than 30 per cent of migraineurs.

Here the prodrome can involve sharply defined visual, sensory, or motor symptoms that occur relatively shortly before the onset of the headache and last usually from 10 to 20 minutes. A visual aura is the most common manifestation, and can consist of a bright-light phenomenon - "like you looked at the sun". Halos, zigzags, distortions of size and perspective (an "Alice in Wonderland" phenomenon), or scintillations can occur. These visual symptoms might follow neurophysiologic and anatomic rules and be truly hemianopic, but are often difficult to explain neurologically. It should be remembered, however, that they are no less real for being difficult to explain.

The headache stage in classic migraine is similar to that in common migraine.

Complicated Migraine. Complicated migraine includes hemiplegic, basilar, ophthalmoplegic, and ophthalmic migraine, and late-life migraine accompaniments.

Hemiplegic migraine affects children and adolescents and only rarely continues beyond the 20s. Typically, the first symptom of an attack is weakness of one side of the body. The face and arm are more involved than the leg and paralysis is usually not total. It is not unusual for paresthesias to accompany the weakness; tingling can affect the face and tongue, which is strongly suggestive of a migrainous etiology. If the right side of the body is affected dysphasia might be present. The headache can begin simultaneously with weakness or can appear 10 to 30 minutes later. The pain is usually hemicranial and involves the side of the head opposite the weakness. The pain is typically vascular, throbbing, often severe, and exacerbated by movement. Unlike classic and common migraine, nausea and vomiting are unusual in those with hemiplegic migraine.

Basilar migraine also affects young patients, more often girls. The initial symptoms are usually visual and consist of blurred vision or frank visual field defects. Bilateral sensory disturbances and vertigo, ataxia, and dysarthria and obtundation can occur, as well as diplopia and quadriparesis. After these symptoms have been present for 10 to 60 minutes the headache usually begins. It is posterior in location, and is severe and throbbing, and lasts several hours. Nausea and vomiting are more common than in hemiplegic migraine.

Ophthalmoplegic migraine usually affects children who are already known migraineurs. Unlike hemiplegic or basilar migraine, headache usually comes first. The pain is severe and located in or around one eye. After some hours of pain the patient can develop double vision, ptosis, strabismus, or sometimes dilatation of the pupil. The headache clears within hours but the ophthalmoplegia may persist.

Cluster Headache. Cluster headaches are characterized by attacks that occur approximately one to three times per day, last about 45 minutes in duration, and are unilateral and oculotemporal or oculofrontal in location, excruciating in severity, and boring and non-throbbing in character. Associated symptoms are also unilateral and consist of lacrimation, rhinorrhea and nasal stuffiness, and a partial Horner's syndrome, including ptosis and myosis. Certain associated features are believed to be almost pathognomonic of the disorder; these include pacing, walking, sitting and rocking.

The episodic type of cluster headache is the most common (80 per cent of cases). This type is defined by periods of susceptibility to headache known as cluster periods, which alternate with remissions. It is predominantly a male disorder with a wide age range, but most of the sufferers are middle-aged.

The differential diagnosis for cluster headache includes migraine, trigeminal neuralgia, temporal arteritis, pheochromocytoma, and Raeder's paratrigeminal syndrome.

Other causes of recurring and disabling headaches include intermittent increases in intracranial pressure, tension headaches (occasionally), and, rarely, vascular malformations.

Chronic Headache

Chronic headaches can be progressive or nonprogressive.

Chronic Progressive Headaches

Chronic progressive headaches increase in severity and frequency over time. They almost always have an organic cause and are usually secondary to increased intracranial pressure.

These headaches are usually accompanied by other neurologic symptoms and signs and are generally present for days or weeks. The differential diagnosis includes space-occupying lesions (leading to increased intracranial pressure secondary to cerebral edema) and/or obstructive hydrocephalus. Occasionally, such chronic progressive headaches can be caused by a non-space-occupying lesion, such as in certain anemic states, in hypertension, and following trauma.

The major differential diagnoses for headaches that are progressive in severity and frequency include: brain tumors, pseudotumor cerebri, hydrocephalus, brain abscess, and subdural hematoma.

Chronic Nonprogressive Headache

Chronic nonprogressive headaches can occur constantly or several times daily or weekly without much change in severity. They are not usually associated with neurologic symptoms or signs and generally have a psychological basis.

The muscle contraction headache is defined as "an ache or sensation of tightness, pressure or constriction widely varied in intensity, frequency and duration, sometimes long-lasting and commonly sub-occipital. It is associated with sustained contraction of skeletal muscles in the absence of permanent structural change, usually as part of the individual's reaction to life stress".

In the typical case, the patient complains of headache that begins in midmorning or early afternoon and reaches a maximum toward evening. The headache might be present every day and the pain is predominantly occipital. Patients have described a vicelike band of pressure traveling across the head in a circular manner. There can be associated tenderness of the scalp, particularly at the position of insertion of the muscles around the head, at the mastoid line (the superior temporal insertion of the temporalis), and just above the eyebrows. The pain is often described as a steady aching pain. Sometimes the patient actually ascribes a "toothache" quality to the pain. The pain is not uncommonly greater on one side of the head than the other.

Sometimes localized problems about the head can produce this type of clinical headache picture, including disorders of the temporomandibular joint, refractive errors, hypertension, and anemia.

Patient Evaluation

History

Bearing in mind that headache has no single cause, that some headaches signal a serious and urgent condition, and that the patient is often concerned for the worse, the correct diagnosis can usually be established from a properly obtained history. It is recommended that information be obtained about the following: duration of symptoms and of headache; frequency; site; whether more than one type of headache is involved; whether the problem is getting better or worse, or is static; precipitating factors; the quality of the pain; mode of onset, and offset; associated symptoms or signs; family history; and what provides relief.

Headaches sufferers, particularly migraineurs, are often eloquent historians and should be encouraged to describe their typical headache. A description of stereotyped, paroxysmal, disabling, and relatively short attacks is highly suggestive of migraine. A history of early morning vomiting, personality change, and disturbance of movement suggests increased intracranial pressure. The headache that is present from morning until night, is tolerated, and is located like a band over the top or around the head is typical of a tension headache. Localized pain suggests localized disease. Meningeal irritation is characterized by a relatively rapid change, fever, malaise, and hypoactivity.

Examination

The clinical examination should include assessment of the patient's mood, affect, and speech. The head should be examined and, in younger patients, the head circumference measured and plotted. Palpation and percussion of the head should be performed and bruits should be listened for over the head, eyes, and neck. The patient should be asked to flex and extend the neck, laterally rotate the neck, and laterally flex the neck in opposition to the examiner. Cranial nerve examination must be performed, including the visual fields and optic fundi. The motor, reflexes, sensory, and cerebellar systems should be examined and attention paid to inspection and palpation of the spine. The skin should also be checked, particularly for any midline skin defects or neurocutaneous lesions.

The general physical examination should include measurement of the height, weight, and blood pressure and a search for other appropriate factors, such as evidence of vasculitis or abdominal tumors.

Significant deviations in stature can indicate a hypothalamic or pituitary disorder. Head enlargement can be familial or raise the possibility of hydrocephalus or occult tumor. Vascular anomalies can produce asymmetric bruits. Localized tenderness about the scalp, face, or neck can indicate trauma or local inflammation. Papilledema, retinal hemorrhage, or optic disc pallor can be clues to serious CNS disease. Neck stiffness suggests meningeal inflammation and sometimes early cerebellar tonsillar herniation. Focal neurologic signs can lead to localized CNS pathology. Cerebellar ataxia can be caused by posterior fossa disease or hydrocephalus. Skin examination can reveal hyper- or hypopigmented areas, trauma, adenoma sebaceum, or midline lesions.

Thus, a perfectly normal and thorough examination frequently excludes serious causes of headache. The "laying on of hands" holds much weight in this era of excess faith and dependence on resource-consuming laboratory investigation.

Laboratory Tests

In most cases of headache, after a proper history and physical examination, it is possible to arrive at the most likely, if broad, clinical diagnosis.

Then what? We do not believe that it is necessary for laboratory studies to be done in all cases of headache. Bearing in mind the wide variety of etiologies for headache, it is necessary to be selective. For instance, radiographs of the skull and sinus are relatively safe and inexpensive but of low diagnostic yield; this is also true of electroencephalography.

Computed tomography of the head is the test of choice for excluding most structural or space-occupying lesions. It is safe, accurate, and rapid (although expensive), but has a low yield in the presence of a benign history and examination. Vascular headache-producing lesions cannot be entirely excluded by this technique and it often has to be supplemented by other tests. Joseph has estimated that the necessity for computed tomography to be done can be reliably determined

by the history and examination; probably only about 1 in 300 patients seen in a headache clinic setting requires this test.

Nuclear magnetic resonance imaging has the advantage over computed tomography in its ability to provide better images of the deeper structures of the brain, especially in the hypothalamic area, brain stem, posterior fossa, and spine.

Positron emission tomography holds promise in the investigation and understanding of headache, but is largely experimental at this time.

Simple laboratory tests such as the complete blood count to check for anemia and leukemia, measurement of the erythrocyte sedimentation rate, electrolytes, and blood urea nitrogen and blood glucose levels, and urinalysis are useful in some cases. Antibody tests for autoimmune diseases might be considered. In patients with recurrent severe migraine, lipid determination has been recommended.

Lumbar puncture with spinal fluid analysis, including measurement of the pressure, is the test of choice in determining the presence of CNS infection. This test is contraindicated in patients in whom intracranial mass lesions are suspected, because of the risk of herniation.

In some individuals with tension or psychologically induced headaches, and in those with stress-triggered migraine, psychological testing is often useful.

Therapy

The emphasis here is on the treatment of the primary headache disorders: tension and migraine headaches. The objectives of treatment should be the symptomatic relief of the individual headache and prophylactic treatment to decrease the frequency of attacks. If prophylactic treatment is successful, the severity and duration of the attacks are also decreased.

Often the headache disorder is severe enough to require both pharmacologic and psychotherapeutic approaches. Treatment should be guided by its effectiveness, versatility, and relative safety, and should be free of the danger of addiction or abuse.

These treatment strategies are generally appropriate for healthy children and adults. The choice of drug and dosage must also consider any complicating medical illness in those of all ages.

Given that most patients with headaches have neither major psychological, structural, or emotional disorders, simple measures such as reassurance, education, and counseling are helpful.

Tension Headache

If headaches have been very frequent or have continued for more than 6 weeks in the absence of neurologic symptoms and signs, they are usually functional. This is especially true when accompanied by prolonged absence from work or school.

If the diagnosis is apparent early it should be discussed. A purely pharmacologic approach is rarely successful, and the psychodynamics of the situation require investigation.

Treatment is most successful if it includes nonpharmacologic approaches (eg counseling, psychoanalysis, psychotherapy, biofeedback, relaxation exercises) in conjunction with non-narcotic analgesics alone or in combination with caffeine and a mild sedative (Table 1). If depression is a major component of the tension headache syndrome, antidepressants and counseling are effective.

Table 1. Treatment of Tension Headache

Nonpharmacologic Approach	Pharmacologic Approach	
	Drug Class	**Popular Class**
Reassurance, counselling, psychotherapy, biofeedback	Analgesics (non-narcotic)	Aspirin, propoxyphene, ibuprofen
	Caffeine	Fiorinal, Empirin Aspirin, Midrin various combinations
	Sedative	Diazepam
	Antidepressant	Amitriptyline (Elavil).

Migraine

Educating the patient about the nature of the disease and the attack, providing reassurance that the condition is a major nuisance rather than a major handicap, and sharing a confident outlook that the condition is highly treatable are all important components of migraine therapy. The literature contains many suggestions on how to treat migraine, but finding a successful mode of therapy for the individual patient often requires a trial-and-error approach. A logical therapeutic plan is advised, bearing in mind that many approaches are available. Because there is so much variability in the nature of migraine itself, both among patients and in the pharmacologic actions of any drugs used, therapy must be flexible, confident, optimistic, and positive.

Women migraine sufferers who are taking oral contraceptive pills should be advised to stop.

Nonpharmacologic Approaches. These include alteration in lifestyle, which is seldom easy but designed to avoid triggering episodes. These changes include the learning of relaxation techniques and occasionally dietary modifications (Table 2).

Table 2. Treatment of Migraine

Nonpharmacologic Approach	Pharmacologic Approach	
	Symptomatic	**Prophylactic**
Reassurance, education, positive outlook, dietary and life	Ergotamine tartrate Chlorpromazine (?) Hypnotic	Ergotamines, "mixtures" Beta blockers Naproxen; calcium channel blockers; others - eg consider cyproheptadine, methysergide, tranquilizer.

Pharmacologic Measures. Medications effective in the treatment of migraine include use of the following (Table 3): analgesics; extracranial vasoconstrictors (eg ergot alkaloids, isometheptene mucate); drugs diminishing response to vasodilatation (eg beta-adrenergic blockers, omega fatty acids - ?); vasoactive agents (calcium channel blockers - flunarizine, verapamil hydrochloride); serotonin or a 5-HT antagonist (eg methysergide, cyproheptadine); nonsteroidal anti-inflammatory drugs, NSAIDs (prostaglandin inhibitors - indomethacin, naproxen, tolmetin sodium, zomepirac sodium); antiplatelet-aggregation agents (eg aspirin, dipyridamole (Persantin), omega fatty acids - ?); drugs that block central motor reflexes and diminish vascular reactivity (eg clonidine); drugs that influence monoamine metabolism and reuptake (tricyclic and tetracyclic antidepressants, monoamine oxidase inhibitors); tranquilizers (eg benzodiazepine, chlorpromazine); steroids; mood-altering drugs (eg lithium); and combinations of these.

The usual therapeutic approach to the migraine sufferer includes (see Table 2): ergotamines (used acutely or prophylactically), beta-adrenergic blockers, naproxen, calcium channel blockers, and other selected approaches.

The recommended treatment of the patient with an acute, severe, and established attack of migraine is a combination of an analgesic and hypnotic (avoiding narcotic analgesics), chlorpromazine, hypnotic alone, or consideration of a steroid.

The therapeutic approach to cluster headache remains difficult and controversial. Remedies include histamine desensitization, lithium carbonate, verapamil, a local anesthetic agent, and oxygen inhalation.

Table 3. Drugs Used for the Treatment of Migraine

Class	Drug	Trade Name
Analgesic	Aspirin	
	Propoxyphene	Darvon
	Ibuprofen	Motrin
Ergot alkaloid	Ergotamine tartrate	Cafergot; Gynergen; Medihaler Ergotamine (aerosol)
Antivasodilators	Propranolol	Inderal
	Fish oil (?)	MaxEPA
Vasoactive agent (Ca channel blocker)	Flunarizine, verapamil HCL	
Serotonin or 5-HT antagonist	Methysergide	Sansert
	Cyproheptadine	Periactin
NSAID	Indomethacin	Indocin
	Naproxen	Naprosyn; Anaprox
	Tolmetin sodium	Tolectin
	Zomepirac sodium	Zomax
Antiplatelet	Aspirin, dipyridamole	Persantine
Vascular "calmer"	Clonidine	Catapres
Monoamine "alterer"	Tricyclic antidepressants:	
	Amitriptyline	Elavil
	Monoamine oxidase inhibitors	Nardil; Parnate
Tranquilizer	Benzodiazepines:	
	Diazepam	Valium
	Chlorpromazine	Thorazine
Steroid	Prednisone	
Mood alterer	Lithium carbonate	
Combination	Caffeine, aspirin, butalbital, others	Fiorinal, Bellergal-S; Midrin.