Secondary Headaches

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ABSTRACT

Purpose of Review: This article identifies the pertinent historical issues that lead to the identification of those headaches needing additional testing to exclude a serious underlying cause.

Recent Findings: Recurrences of giant cell arteritis, even after presumed successful treatment, are common. Postural orthostatic tachycardia syndrome is an often unrecognized cause of headache.

Summary: Patients with a primary headache disorder are more susceptible to the development of headache when a secondary cause occurs. Their headaches may be phenotypically similar to their primary headache disorder. Therefore, a secondary cause should be considered in patients with preexisting headache disorders who develop a significant increase in the number and severity of those attacks.

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In evaluating a patient with headache, the first task is to determine whether the headache is primary in origin or reflects underlying neurologic or systemic disease (ie, secondary headache). A high index of suspicion exists for secondary headache when the headache reaches full intensity rapidly, is associated with abnormal neurologic signs, is progressive in intensity and duration over time, is increased by Valsalva maneuver, is worse upon standing, or initially develops after the age of 50. Known coexisting systemic and neurologic disorders also increase the likelihood that the headache is secondary. Headaches that worsen upon leaning forward, classically described as a symptom of sinusitis, are generally not of diagnostic value. Secondary headaches occur more often and may be relatively more prominent in patients with a preexisting primary headache disorder. Accordingly, a clear progression in the intensity and frequency of the primary headache should prompt a reevaluation.

The International Classification of Headache Disorders, Second Edition (ICHD-II) defines secondary headaches as follows (only some terms will be discussed in this article):

- Headache attributed to head and neck trauma
- Headache attributed to cranial or cervical vascular disorder
- Headache attributed to nonvascular intracranial disorder
- Headache attributed to a substance or its withdrawal
- Headache attributed to infection
- Headache attributed to disturbance of homeostasis
- Headache or facial pain attributed to disorder of cranium, neck, eyes, ears, nose, sinus, teeth, mouth, or other facial or cranial structures
- Headache attributed to psychiatric disorder

BRAIN TUMOR HEADACHE

As with many other medical conditions, headaches are more common in patients with brain tumors if they have a preexisting primary headache disorder. In 30% of cases involving brain tumor,

KEY POINTS

- Individuals with a preexisting primary headache disorder who develop a secondary cause commonly experience a change in their preexisting headache type rather than develop a new headache type.
- Headaches associated with brain tumors commonly wake patients from sleep, but this also occurs frequently with migraines, cluster headaches, and hypnic headaches.

headache is a major concern, but only 1% of these individuals have headache as the sole clinical manifestation of the tumor. In one study, 5% of those presenting with a brain tumor or another structural neurosurgical disorder presented with headache, and none had headache as the sole concern.[1] Individuals with primary headache syndromes commonly experience a change in the preexisting headache, with an increase in frequency, severity, and duration of symptoms. Headaches associated with a brain tumor usually increase upon Valsalva maneuver and exertion (although this also occurs frequently with migraine). Headaches associated with brain tumors may awaken the individual from sleep, but this is also common with cluster headaches and migraines.

HEADACHES OF OTHER MASS LESIONS

Brain Abscess

Brain abscesses evolve from a localized region of cerebritis to become a walled capsule, which is a mass lesion. As such, the associated headaches are the same as with a tumor. The presence of a headache accompanying a mass lesion with fever is strongly suggestive of a brain abscess, but fever is seen in less than half of these cases.[2] An impaired sensorium, focal neurologic signs and symptoms, and seizures are common. In the past, a high percentage of brain abscesses were seen in young children and were typically a complication of otitis media, but this is no longer the case. In areas where AIDS is prevalent, toxoplasmosis is the most common cause of brain abscess. The headache of brain abscess is indistinguishable from that of a tumor, but the associated clinical course can be influenced by the virulence of the infecting organism and any underlying disorder that predisposed to the development of the abscess.

Subdural Hematomas

Particularly in older adults, chronic subdural hematomas can present as headache. Eighty percent of patients with chronic subdural hematomas report headache, and a change in sensorium is common. Because these are mass lesions, the headache's characteristics are similar to those of headaches associated with brain tumors. The patient may not be able to describe a history of trauma or it may be described as remote from the development of the headache.

IDIOPATHIC INTRACRANIAL HYPERTENSION

The term idiopathic intracranial hypertension (IIH) is often used synonymously with pseudotumor cerebri, but the former is a specific diagnosis, whereas the latter is a syndrome with many potential causes, of which IIH is the most common. Because it involves diffuse swelling of the brain and increased intracranial pressure (ICP), the headache is the same as that associated with a brain tumor but without the clinical consequences of a tumor compressing adjacent structures. Similar to individuals with brain tumors, patients with IIH and a primary headache disorder often experience a symptomatic worsening of the preexisting disorder. In a review of 82 patients with IIH, 30% had symptoms of episodic tension-type headaches, and 20% had symptoms consistent with migraine without aura.

Most patients with IIH are women who are obese and of childbearing age, but this disorder can occur at any age regardless of sex or weight. A recent weight gain, even in non-obese individuals, increases the risk for the development of IIH. Pulsatile tinnitus or other intracranial noises are commonly experienced. Diplopia, commonly from a sixth nerve palsy, and transient visual obscurations may occur. Papilledema is typical but not invariable, although

patients with IIH without papilledema tend to have lower opening pressure.[3] Photopsias and retrobulbar pain are common. Radicular pain can be seen as the elevated pressure is transmitted down to all root sleeves. Cessation of previously identified spontaneous venous pulsations seen with ophthalmoscopy strongly suggests raised ICP.

The diagnosis is confirmed by normal imaging studies. Careful evaluation of MRI scans can reveal changes: an empty sella, flattening of posterior globes, protruding of optic nerve heads, and vertical tortuosity of optic nerves.[4] Lumbar punctures reveal normal fluid composition under elevated pressure. Magnetic resonance venography can exclude venous sinus obstruction, although this study is often difficult to interpret because a high degree of natural variability occurs in the anatomy of this structure. Patients with venous sinus thrombosis, however, may exhibit all of the classic signs and symptoms of IIH.

The treatment of IIH generally involves the use of a carbonic anhydrase inhibitor such as acetazolamide or topiramate. Loop diuretics may also be of value. Corticosteroids, which can significantly increase weight, are not generally appropriate. Octreotide is a promising therapy.[5] For obese patients, weight reduction, including bariatric surgery in patients with morbid obesity, can be helpful, but it is not a short-term treatment. Serial lumbar punctures as a therapy have not been sufficiently studied, are very uncomfortable, and are not recommended. For cases refractory to medical therapy, a lumbar peritoneal shunt, ventriculoperitoneal shunt, or optic nerve sheath fenestration may be necessary to preserve vision and often can improve headache. Transverse sinus stenting has not been sufficiently studied as a treatment for IIH.

The headache of IIH and the visual signs and symptoms do not often parallel each other, and the treatment of one does not necessarily help the other. Accordingly, a careful ocular evaluation, including periodic perimetry, and an evaluation for headache management need to be performed. If the head pain does not resolve with the agents used to reduce ICP, the headaches are usually treated based on their phenotype, often involving the use of preventive antimigraine drugs.

This condition is not benign, and blindness can ensue. This complication is more common in African Americans than whites.

KEY POINT

- Changes on MRI scans such as empty sellas, flattening of the posterior globes, protruding optic nerve heads, and vertical tortuosity of optic nerves can strongly suggest idiopathic intracranial hypertension.

ORTHOSTATIC HEADACHES

Most orthostatic headaches are caused by intracranial hypotension, which commonly occurs after a lumbar puncture. Orthostatic headaches often begin within 48 hours of lumbar puncture but can be delayed up to 14 days. These headaches occur in 30% of lumbar punctures, less often with the use of atraumatic needles and more often in patients with low body mass indexes. Prolonged bed rest following a lumbar puncture is ineffective in preventing the headache.

Causes of orthostatic headaches are shown in **Table 3-1**.

Typically the pain begins rapidly upon arising and is relieved by reclining, abdominal compression, Valsalva maneuver, or exertion. It tends to be absent or not severe in the early hours of the day, appearing or worsening as the day progresses. The pain of a low-pressure headache is typically dull but can be pulsatile and is typically felt bilaterally in the occipital and frontal regions. In 15% of patients, the headache has a thunderclap onset. Stiff neck and nausea are common and are often accompanied by blurred vision, pulsatile tinnitus, and radicular upper extremity symptoms. Cases of low-pressure headache presenting as a cough headache have been reported.

TABLE 3-1 Orthostatic Headaches

- **With Cranial and Spinal Leaks**
  - Lumbar puncture
  - Neurosurgical procedure
  - Trauma
  - Meningeal diverticula/Marfan syndrome, Ehlers-Danlos syndrome, and other connective tissue abnormalities of spinal dura
  - Absent nerve root sleeve
  - Tumor invasion of nerve roots
- **Unassociated With CSF Leak**
  - Postural orthostatic tachycardia syndrome
  - Overshunting
  - Dehydration, uremia
  - Carbonic anhydrase inhibitors

Symptoms typically resolve a few days after the lumbar puncture, but persistent low-pressure headaches can occur. Some individuals with new daily persistent headache have spontaneous intracranial hypotension.

Spontaneous intracranial hypotension has other causes that may not always be evident and may require a more detailed evaluation. True hypovolemia can be responsible for the CSF hypovolemia. Overshunting can result in this disorder. This can be the consequence of the administration of a carbonic anhydrase inhibitor, such as topiramate, in a patient with a previously stable stunt.

Other causes of leaks include meningeal diverticula, dural root sleeve tears, excessive coughing, erosion of the dura from adjacent lesions, and head trauma. The characteristic meningeal enhancement can be confused with carcinomatosis, lymphoma, sarcoidosis, and rheumatoid arthritis.

MRI may reveal pachymeningeal enhancement and thickening, and downward displacement of the cerebellar tonsils and even subdural effusions may be present. The tonsillar descent may be confused with a Chiari type 1 malformation. The ventricles may be small. Pituitary enlargement is common, and spinal epidural venous plexus may be prominent.

If CSF rhinorrhea is suspected, the fluid can be assayed for beta-2 transferrin, which is found only in CSF, perilymph, and aqueous humor. This test has a sensitivity of 94% to 100% and a specificity of 98% to 100%.[6] If the source of a presumed leak is not identified clinically, further radiologic evaluation is necessary and may include CT myelography and indium cisternography. Both procedures involve a lumbar puncture, at which time the CSF pressure is commonly unobtainable or low, although normal pressures have been reported. The CSF protein level can be normal or significantly elevated. A lymphocytic pleocytosis of up to 50 cells/mm^3 is common. A radioisotope cisternogram can detect smaller volumes of leak, even though the resolution is poor. It is more common for little or no radioactivity to appear over the cerebral convexities at 24 to 48 hours. Indirectly, early radioactivity to appear in the kidneys and bladder suggests low CSF pressure. A CT myelogram has better spatial resolution than cisternography but is less sensitive to low-volume leaks. Both can be performed with a single lumbar puncture. Nasal pledgets

can be scanned for radioactivity. An alternative to indium cisternography is magnetic resonance myelography and cisternography performed several hours after the CT myelography.

In the case of a post–lumbar puncture headache, IV caffeine 500 mg can terminate the attacks but is expensive and often has a short-lasting effect. An epidural blood patch can be immediately and dramatically effective. The mechanism of action of an epidural blood patch is not known but may be due to displacement of an engorged epidural venous plexus, which secondarily increases ICP. A direct repair of the source of the leak, if identified, may be necessary. In cases in which no source of leak has been identified, an epidural blood patch may still be of value.[7] The difficulty in treating these individuals is illustrated in **Case 3-1**.

Postural Orthostatic Tachycardia Syndrome

Postural orthostatic tachycardia syndrome (POTS) is an important cause of orthostatic headache, particularly in young, postpubertal women. The headaches also may be non-orthostatic, often coexisting with the orthostatic variety. Many patients have comorbid migraine. A tachycardia with an increase of at least 30 beats/min after standing for 5 to 30 minutes is diagnostic. The disorder often follows an infectious disease. In addition to headache, fatigue, decreased concentration, exercise intolerance, and presyncope/syncope may occur.[8] Physical activity may worsen nonpostural headaches in these individuals, but their headaches resolve when supine. Because chronic fatigue, palpitations, and anxiety are frequently

Case 3-1

A 36-year-old woman presented with a 2-year history of headaches. The pain was described as generalized. She would awaken feeling well, but within 30 minutes of arising the pain would begin and progress as the day advanced. By the end of the day, she noted that the pain was in her cervical region as well as her head. She would experience relief when she was supine. She reported no history of a lumbar puncture, any other trauma, or any nasal discharge. No radicular symptoms were present.

She underwent a CT myelogram, which was entirely normal. Following this study, an epidural blood patch was performed. She described immediate and dramatic improvement. During this time, she noted a strong salty taste in her mouth that she could not explain. Three days later the pain resumed, coincident with a disappearance of the salty taste. She was unable to collect a sufficient volume of nasal secretions to assay for beta-2 transferrin.

An indium 111 cisternogram was normal, except excessive radioactivity was noted in the nasal pledgets.

A high-resolution sinus CT (**Figure 3-1**) revealed a small air bubble along the posterior left margin of the crista galli with poor ossification of the cribriform plate on the left side.

Following an endoscopic repair of the leak, her symptoms immediately resolved.

Comment. Determining the site of the leak in a low-pressure headache due to CSF oligemia is often difficult. Repeated CT myelography or indium cisternograms may be necessary.

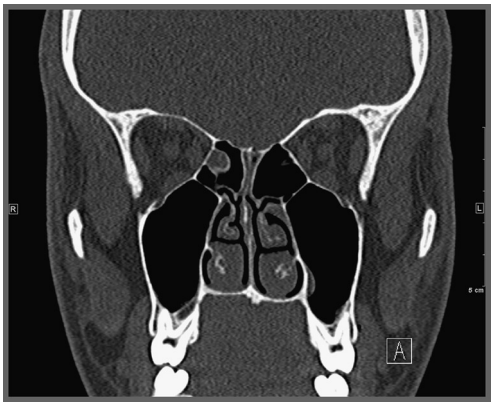

FIGURE 3-1 High-resolution sinus CT reveals a small air bubble along the posterior left margin of the crista galli with poor ossification of the cribriform plate on the left side.

KEY POINT

■ Because primary (benign) cough headache is rare, all patients with cough headache should be thoroughly evaluated for structural causes.

present, POTS is often diagnosed as a psychiatric disorder.[9] Treatment of POTS may improve the associated headache; such treatment includes increased hydration and salt intake, exercise, and the use of elastic stockings. Beta-blockers, indomethacin, midodrine, and fludrocortisone may be added in more refractory cases.

EXERTIONAL HEADACHES

Primary cough headache, primary exertional headache, and primary headache associated with sexual activity are classified as primary headaches, but many headaches identical to these have a secondary cause.

Cough Headache

Primary cough headache and its variants are seen predominantly in men and are characterized by the abrupt onset of severe head pain triggered by coughing, bending, stooping, or sneezing. The typical duration of symptoms is seconds to minutes (unlike the thunderclap headache of subarachnoid hemorrhage [SAH], which tends to last much longer). Cough headache is commonly triggered by a respiratory infection and is seen in only 1% of the population, most commonly in the fourth decade of life.[10] Because the primary disorder is rare, neurologic evaluation must include neuroimaging; 25% of patients presenting with apparent cough headache have a symptomatic structural abnormality, the most common being a Chiari malformation. Primary cough headache is benign and self-limiting. Indomethacin can be a highly effective treatment, and a lumbar puncture with a high volume of drainage can be effective for about 50% of individuals. Even though cough-triggered headache can occur as a result of increased ICP, it also occurs as a symptom of low ICP.

The preferred treatment for primary cough headache is prophylactic indomethacin 25 mg to 50 mg 3 times a day. Acetazolamide also may be effective. While their mechanism of action in this setting is unknown, both of these agents reduce ICP. Small studies with topiramate, a carbonic anhydrase inhibitor like acetazolamide, have suggested benefit in some patients.

Coital and Other Exertional Headaches

Primary exertional headaches are defined as being pulsatile, lasting 5 minutes to 2 days, and occurring during or after physical exertion. Unlike cough headache, which is triggered by Valsalva maneuver, primary exertional headaches are triggered by strenuous physical activity.

As with cough headache, more men than women experience coital headache. Most coital headaches occur close to orgasm and have an apoplectic onset. A diagnosis of SAH must be entertained on the headache's first occurrence. If attacks have been present for weeks to months, however, an aneurysm or recurrent SAH from any source is quite unlikely as the cause. In all cases of headache precipitated by Valsalva maneuver (ie, exertion against a closed glottis), an MRI scan is appropriate to exclude an intracerebral lesion.

Exertional headaches can occur with other sustained physical activity (eg, running and swimming), but in these cases the headache has a subacute onset. Sneezing, laughing, and crying can be triggers of exertional headaches. As-needed anticipatory pretreatment with indomethacin, triptans, or ergotamine may prevent attacks. Scheduled prophylactic treatment with propranolol or nadolol can be effective.

Cardiac Cephalalgia

Headaches that occur with exertion and typically subside with rest are occasionally

caused by cardiac disease, which can be considered an ''anginal equivalent.'' In some cases, the headache occurs when the patient is at rest.[11] Headaches associated with cardiac cephalalgia commonly involve the forehead, occiput, or jaw but can affect any structure above the umbilicus. Cases of such headaches responding to nitroglycerin have been described, and administration of that drug might be useful if a cardiac etiology for headache is suspected. The headaches of cardiac cephalalgia resemble either migraine or tension-type headache and vary in location. In 27% of cases of cardiac cephalalgia, headache is the sole manifestation.[12] An ECG performed during the symptomatic period may reveal ischemic changes, although coronary angiography may be necessary to confirm the diagnosis.

In contrast to the previously described exertional headaches, the onset of cardiac cephalalgia is gradual with exertion and resolves with rest. The pain of primary cough headache and headache with orgasm (benign sexual headache/explosive type) typically has an abrupt onset. Headaches associated with cardiac cephalalgia typically do not respond to analgesics and triptans; in fact, triptans and ergots are contraindicated in patients with known coronary artery disease.

HEADACHE AS A SYMPTOM OF CEREBROVASCULAR DISEASE

Headache is a common symptom of stroke. In a study by Tentschert and colleagues,[13] 27% of stroke patients had headache at the onset of their stroke before other symptoms occurred. Headache at stroke onset is correlated with a preexisting history of migraine, younger age, female sex, cerebellar stroke, and blood pressures less than 120 mm Hg systolic and less than 70 mm Hg diastolic. Larger strokes are more likely than small ones to be associated with head pain. Headaches are more common in strokes involving the posterior circulation than those involving the anterior circulation.[14] In another study,[15] most headaches were described as pressurelike, bilateral and anterior, increasing with cough, and lasting a mean of 3.8 days. When ICHD-II criteria were applied, the most common description was that of a tension-type headache or migraine.[16] As with individuals with many other secondary headaches, patients with a preexisting primary headache disorder were more likely to present with headache as a symptom of stroke, and in this study the stroke-related headache resembled that of the preexisting headache type in about half of the patients. Nausea and vomiting were present on day 1 in 40% of cases, which could make the differentiation from migraine problematic. Additionally, individuals with migraine with aura are at an increased risk for stroke, particularly if they also smoke and use an estrogen-containing oral contraceptive. Stroke from migraine is quite rare, however, especially when compared to the prevalence of a migrainelike headache accompanying acute stroke from another cause.

Subarachnoid Hemorrhage

The usual description of an SAH is the sudden onset of ''the worst headache of my life,'' often developing with exertion. Such thunderclap headaches can be primary or secondary, and the features of primary thunderclap headache are summarized in **Table 3-2**.[16] So-called sentinel headaches occur in 20% to 50% of cases with subsequent aneurysmal SAH, and the recognition of the significance of these antecedent headaches can be lifesaving. Sentinel headaches have a thunderclap onset and may last from hours to days. Meningismus tends to be absent.

KEY POINT

- In an individual with atherosclerotic risk factors, consider a cardiac cause of head pain when it is triggered by exertion and subsides with rest.

KEY POINT

■ Nearly half of headaches from subarachnoid hemorrhage deviate from the classic description.

TABLE 3-2 Primary Thunderclap Headache[a]

A. Severe head pain fulfilling criteria B and C
B. Both of the following characteristics:
   1. Sudden onset, reaching maximum intensity in <1 minute
   2. Lasting from 1 hour to 10 days
C. Does not recur regularly over subsequent weeks or months
D. Not attributed to another disorder

[a] Adapted from Headache Classification Committee of the International Headache Society, Cephalalgia.[16] © 2004, with permission of SAGE. *cep.sagepub.com/content/24/1_suppl/9.long*.

Nearly half of patients with SAH have headaches that deviate from the classic description. In a large series, 34% of headaches occurred during nonstrenuous activity, and 12% developed during sleep.[17] No characteristic location has been determined, and the headache may be self-limited or relieved by analgesics or triptans. The development of neck pain is common, but neck pain is also common in migraine (potentially leading to incorrect diagnosis). The abrupt onset of headache is always of concern, but in one study this occurred only half of the time in patients with SAH and more often (two-thirds of the time) in patients with benign causes of thunderclap headaches.[18] In this study, thunderclap headache, female sex, seizures, vomiting, loss of consciousness, and focal symptoms increased the likelihood of SAH being the cause of the headache.

Other causes of thunderclap headache and their distinctions from primary thunderclap headache are listed in **Table 3-3**.

Other Vascular Headaches

Reversible cerebral vasoconstriction syndromes, a heterogenous group of conditions, lead to multifocal narrowing in intracerebral arteries. Patients with one of the reversible cerebral vasoconstriction syndromes may present with thunderclap headache and exhibit focal neurologic abnormalities. Individual headaches may last minutes to hours, and the headache disorder generally resolves over days to weeks.

Cerebral sinus thrombosis is accompanied by headache in 75% to 90% of cases. The headache is insidious in onset or, less commonly, thunderclap.[19] As with other headaches associated with

TABLE 3-3 Secondary Causes of Thunderclap Headache

- ▶ Intracerebral or subarachnoid hemorrhage (ruptured and unruptured aneurysms are the most common cause of subarachnoid bleeds)
- ▶ Cerebral venous sinus thrombosis
- ▶ Vertebral dissections
- ▶ Acute hypertension
- ▶ Acute low CSF pressure (including CSF oligemia)
- ▶ Cough, micturition (with or without bladder pheochromocytoma), exercise, and sexual activity
- ▶ Colloid cyst of the third ventricle
- ▶ Reversible cerebral vasoconstrictive syndrome (all of its associated syndromes)
- ▶ Psychiatric conditions, including panic disorder

increased ICP, they tend to intensify with Valsalva maneuver. An alteration in level of consciousness, seizures, and focal neurologic signs may accompany the headache, but in 15% to 30% of patients with cerebral sinus thrombosis headache is the sole symptom.[20]

Cervical artery dissections occur more often than is commonly recognized. They probably arise from an intimal tear and the subsequent development of an intramural hematoma. Headache is the most common presenting symptom, seen in 60% to 95% of patients with carotid dissections and in 70% of patients with vertebral dissections.[21] In carotid dissection, pain typically occurs in the ipsilateral face and neck along with an ipsilateral headache. An ipsilateral partial Horner syndrome is present in slightly less than half of cases, and signs and symptoms of cerebral or retinal ischemia may occur up to a month following anatomic dissection.[22] In vertebral artery dissection, cervical pain and occipital headache are common, and the headache is often throbbing and steady in character. Symptoms of stroke may occur at the time of dissection or up to 2 weeks later.

Carotid or vertebral artery dissection may complicate trivial or major neck/head trauma, and in a substantial proportion of cases no antecedent history of trauma whatsoever is reported.

INFECTION

According to the ICHD-II, headaches attributed to infection can be divided into four categories:

(1) Headache attributed to intracranial infection
(2) Headache attributed to systemic infection
(3) Headache attributed to HIV/AIDS
(4) Chronic postinfection headache

Headaches attributed to intracranial infections include those associated with meningitis, encephalitis, brain abscess, or subdural empyema.

Headaches attributed to systemic infections include those associated with viral and bacterial systemic infections.

HIV can cross the blood-brain barrier, so neurologic complications of HIV/AIDS are common. These include encephalopathy, cryptococcal meningitis, herpes and cytomegalovirus encephalitis, progressive multifocal leukoencephalopathy, cerebral toxoplasmosis, and primary CNS lymphoma. Headache occurs at some time in the course of HIV infection in 50% of patients, and when a person with HIV/AIDS reports a significant headache (especially of new onset), a thorough evaluation is essential.[23] In one series, 82% of HIV-positive patients with a report of headache had a significant and potentially treatable secondary cause (primarily toxoplasmosis abscess or cryptococcal meningitis).[24] In another study, headaches occurred in 38% of HIV-positive patients, with 66% having primary headaches and 34% having secondary causes.[25] Given the age of these studies and the fact that current treatment has improved the outcome associated with the primary infection, these numbers are likely to be lower now. Regardless, CD4 counts below 200 cells/mm^3 strongly suggest a secondary cause for the headache, and in such circumstances MRI and lumbar puncture typically are indicated.[26] A change in preexistent secondary headache has been associated with stages of the infection and mild encephalopathy but is not correlated with the CD4 count or antiretroviral therapy.[27]

Chronic postinfection headache can be diagnosed only when the actual intracranial infection resolves but the headache persists for 3 or more months. The ICHD-II states that a persistent headache represents a continuation of the infection in bacterial meningitis,

KEY POINT

■ Most patients with AIDS who present with new-onset headache have a secondary cause.

KEY POINT

- Inflammatory markers in giant cell arteritis are not always predictive of disease activity.

but in other forms of postinfectious headache the symptoms occur after the infection has resolved. The headaches are probably immune-mediated or inflammatory in origin. It has been theorized that new daily persistent headache is a form of postinfectious headache, but that remains speculative.[28] It is also not proven that early treatment of infection reduces the risk for the development of postinfectious headache. Although controlled studies are small, the use of immunosuppressant agents, including corticosteroids, may be valuable in the management of individuals with postinfectious headache.[29]

OTHER SECONDARY HEADACHE TYPES

Headaches, Neurologic Deficit, and Lymphocytosis Syndrome

Often referred to as pseudomigraine, headaches, neurologic deficit, and lymphocytosis (HaNDL) syndrome causes headache with neurologic deficits lasting from hours to 3 days. Although not known to be of infectious origin, it can be associated with fever. The CSF demonstrates an elevated protein level and pleocytosis.[30] The cause is elusive. Although transient, recurrences are the norm, generally occurring within 3 months of the original event. No specific treatment is available, and the condition itself is benign and self-limited.

Giant Cell Arteritis

Giant cell arteritis is a serious cause of headaches that is generally seen in individuals older than 60. Mostly whites are affected. Fifty percent of individuals with giant cell arteritis have polymyalgia rheumatica, and 15% of those with polymyalgia rheumatica ultimately develop giant cell arteritis. The mean age at onset of symptoms for both giant cell arteritis and polymyalgia rheumatica is 70.[31] Signs and symptoms include fever; malaise; scalp tenderness; anemia; jaw or tongue claudication; painful dysphagia; hoarseness; myalgias and muscle stiffness of neck, shoulders, and pelvic girdle; and occasionally visual loss. Symptoms of the disease frequently change over time, which can confound diagnosis.

Headache is the most common symptom of giant cell arteritis, occurring in 90% of patients. It is frequently temporal but can be occipital and is generally bilateral. Scalp discomfort is common and can be confused with the cutaneous allodynia often accompanying prolonged or chronic migraine. Jaw claudication occurs in one-third to one-half of these cases due to masseter ischemia consequent to stenosis of the maxillary artery.[32] This can be confused with temporomandibular dysfunction, which occurs with any jaw movement, whereas jaw claudication occurs after a few minutes of mastication and resolves rapidly with rest. Painful oral symptoms can occur with giant cell arteritis (eg, throat pain, trismus, glossitis, and dysphagia).[33]

Serologic inflammatory markers are not always predictive of the disease's presence or its activity. Relapses of polymyalgia rheumatica and giant cell arteritis are common, and in 50% of these relapses the C-reactive protein and erythrocyte sedimentation rate are not elevated.[34] Recurrences after therapy is completed are also frequent and commonly present with recurrent headache.[35]

Posttraumatic Headaches

Postconcussive headache encompasses a heterogeneous group of headaches that follow a head injury. A loss of consciousness is not a prerequisite for these headaches. An inverse relationship between the severity of head injury and the severity of headache has been described. In general, posttraumatic headaches resolve within 4 weeks of the injury.

As with many causes of secondary headaches, individuals with a preexisting headache disorder are more likely to develop headaches following head trauma. Because head trauma can be associated with injury to many pain-sensitive structures, the headaches are best managed according to their phenotype.

Headaches and the Sinuses

Acute sinusitis can be associated with headache, and its diagnosis requires the presence of purulent nasal drainage, pathologic sinus findings on CT or MRI, x-ray, or transillumination, the simultaneous onset of headache with sinusitis, and the localization of head pain to specific facial and cranial sites near the sinuses.[16]

The following are ICHD-II criteria for rhinosinusitis headache:

(1) Diagnostic criteria: pain in one or more regions of the head, face, ears, or teeth
(2) Clinical criteria: laboratory or imaging evidence of acute rhinosinusitis (eg, purulent nasal drainage; nasal obstruction; fever hyposmia or anosmia; CT, MRI, or fiberoptic nasal endoscopic findings)
(3) Simultaneous onset of headache and rhinosinusitis
(4) Headache lasts more than 7 days after remission or successful treatment of acute rhinosinusitis

With the exception of sphenoid and frontal sinusitis, acute sinusitis is more common in children than adults.

The ICHD-II states, ''Chronic sinusitis is not validated as a cause of headache or facial pain unless relapsing into an acute stage.''

Schreiber and colleagues found that 88% of physician-diagnosed or self-diagnosed cases of sinus headache met the ICHD-II criteria for migraine or migrainous headache.[36] One reason for the high rate of misdiagnosing migraine as sinus headache is the high prevalence of lacrimation and nasal congestion seen in migraine.[37]

Isolated sphenoid sinusitis, although relatively rare, is a serious disorder that can resemble chronic tension-type headache or chronic migraine. CT scanning is the preferred diagnostic test, and this disorder is often entirely missed or underestimated by MRI.

Mucosal contact points have been considered to cause or contribute to headache or facial pain. However, a large study[38] suggested that their coexistence is likely to be coincidental. Even so, if the pain is reliably terminated by endoscopically placed lidocaine on the contact point, surgical management may be helpful.[39]

Hypertension

Modest hypertension is generally not associated with headache, but 20% of patients presenting with an acute hypertensive crisis do have headache, and occasionally these headaches have a thunderclap onset.[40] Such headaches typically affect the occiput and upper cervical region and are often associated with chest pain, dizziness, and focal neurologic signs.

Paradoxically, elevated blood pressures can be associated with a lowered risk of headache. One study showed that systolic hypertension above 150 mm Hg and diastolic elevations were associated with a lowered risk of nonmigrainous headache.[41]

KEY POINT

- As with other causes of secondary headaches, individuals with primary headache disorders are more likely to develop a secondary headache after head injury, and treatment is based on the phenotype of that headache.

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