CHAPTER 101 Headache

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Headache is a common complaint, more frequent than the common cold, and accounts for approximately 3 million visits to the emergency department (ED) per year in the United States.[1] In addition, many more patients present with headache as part of a constitutional illness, making the symptom of headache one of the most frequent complaints in the ED.[2]

Headache commonly is divided into *primary* and *secondary* disorders. The primary headache disorders include migraine, cluster, and tension-type headaches, which represent greater than 90% of headaches seen in clinical practice.[3] Secondary headache disorders include a variety of organic illnesses in which head pain is a symptom of an identifiable, distinct pathologic process. To facilitate a standardized approach to headache management, the International Headache Society (IHS) published classification and diagnostic criteria for "headache disorders, cranial neuralgias, and facial pain" in 2004.[4] This comprehensive and widely accepted system includes 14 categories of headache disorders and uses specific operational diagnostic criteria to define each headache type (Box 101-1). This revised classification system offers better separation of primary and secondary headaches and more standardized criteria for diagnosing secondary headaches.[5]

The vast majority of patients presenting with headache have a benign primary headache disorder requiring only symptomatic treatment and referral. The challenge for the emergency physician is to identify the very small subset of patients who have headache as a symptom of a serious or potentially life-threatening disease (see Chapter 16).

PRIMARY HEADACHE DISORDERS

Migraine Headache

Principles of Disease. Migraine is a common, chronic, sometimes incapacitating neurovascular disorder, characterized by attacks of severe headache, autonomic nervous system dysfunction, and, in some patients, an aura involving neurologic symptoms.[6] It is a primary headache disorder believed to have a genetic basis.[7]

Migraine headaches account for approximately 1 million visits to the ED per year.[8] They typically begin in the second decade of life, peaking in early to midadolescence, and are more prevalent among women (17%) than among men (6%).[7,9,10] During childhood, however, there is no gender difference in the prevalence of migraine.[3] After menarche a correlation between migraine headache and menses is found in approximately 15% of female migraine sufferers, possibly related to fluctuating estrogen and progesterone levels. After menopause, women also tend to experience fewer migraine headaches. The lifetime prevalence of migraine is at least 18%.[6] As many as 60% of women with migraines report an association between migraine and menstruation.[11]

Historically, migraine headaches have been considered to be vascular in origin. According to this hypothesis, an initial phase of cerebral vasoconstriction resulting in neurologic symptoms (migraine with aura) is followed by a vasodilatory phase, manifested by the typical pounding headache of migraine. Appropriate changes in blood flow have been demonstrated for the classic migraine attack, and pain relief provided by vasoconstriction has been cited as further support for this hypothesis.[12] However, this mechanism does not fully explain the entire spectrum of migraine attacks, and migraine is no longer thought to be caused by a primary vascular event.[6] It is now believed that the pathophysiologic cause of migraine may actually originate in the brainstem within its descending and ascending circuitry, including the ascending pain-modulating projections from the midbrain raphe nuclei.[13] Evidence suggests a perturbation of neural activity within this serotoninergic system as an important precursor to migraine.[14] Changes in serotoninergic activity can alter the cranial circulation, triggering a "vascular phase." In addition to constriction and dilatation of intracranial and extracranial arteries, this neurovascular reaction activates the nociceptive trigeminal vascular system.[15,16] Neural connections between cerebral blood vessels and the trigeminal nerve release neuropeptides that can induce a painful neurogenic or sterile inflammation.[17]

Agonists of the 5-hydroxytryptamine (5-HT) 1B/1D receptor, such as sumatriptan or dihydroergotamine, block the inflammatory process. Effective prophylactic agents are believed to act as antagonists of the 5-HT_2 receptor site.[18]

Migraine is further divided into two major categories. Migraine without aura, or "common migraine," is the most frequent form of migraine and accounts for approximately 80% of all cases (Box 101-2). "Classic migraine," or migraine with aura, has specific reversible neurologic symptoms that precede the actual headache (Box 101-3) and is seen less frequently.

Clinical Features. Migraine headaches tend to be chronic and recurrent. The headache often is unilateral, pulsating in quality, moderate to severe in intensity, and exacerbated by routine activities. The side of the headache can vary with individual attacks, and the headache may be bilateral in 40% of patients. The onset usually is gradual, and the attacks typi-

BOX 101-1 International Headache Society Classification of Headache

1. Migraine
2. Tension-type headache
3. Cluster headache and trigeminal autonomic cephalgias
4. Other primary headaches
5. Headache attributed to head and/or neck trauma
6. Headache attributed to nonvascular intracranial disorder
7. Headache associated with nonvascular intracranial disorder
8. Headache attributed to a substance or its withdrawal
9. Headache associated with noncephalic infection
10. Headache attributed to disorder of homeostasis
11. Headache or facial pain attributed to disorder of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth, or other facial or cranial structures
12. Headache attributed to psychiatric disorder
13. Cranial neuralgias and central causes of facial pain
14. Other headache, cranial neuralgia, central or primary facial pain

Available at http://ihs-classification.org/en/.

BOX 101-2 Migraine without Aura (Common Migraine): International Headache Society Criteria

A. At least five attacks fulfilling criteria in B, C, D, and E
B. Attack lasts 4 to 72 hours (untreated or unsuccessfully treated)
C. Headache has at least two of the following characteristics:
   1. Unilateral location
   2. Pulsating quality
   3. Moderate to severe pain intensity
   4. Aggravation by or causing avoidance of routine physical activity (e.g., walking or climbing stairs)
D. During headache, at least one of the following:
   1. Nausea or vomiting (or both)
   2. Photophobia and phonophobia
E. Not attributable to another disorder

Available at http://ihs-classification.org/en/.

BOX 101-3 Migraine with Aura (Classic Migraine): International Headache Society Criteria

A. At least two attacks that fulfill criterion B
B. Presence of at least three of the following four characteristics for a diagnosis of classic migraine:
   1. One or more fully reversible aura symptoms indicating focal cerebral cortical or brainstem dysfunction (or both)
   2. At least one aura symptom developing gradually over more than 4 minutes, or two or more symptoms occurring in succession
   3. No single aura symptom lasting longer than 60 minutes
   4. Headache beginning *during* aura or *afterward*, with a symptom-free interval of less than 60 minutes (also may begin *before* aura)
C. Exclusion of related organic diseases by means of an appropriate history, physical examination, and neurologic examination with appropriate diagnostic tests

Available at http://ihs-classification.org/en/.

cally last from 4 to 72 hours. Headache frequency is quite variable, and some patients experience several episodes per month. Associated symptoms and signs include nausea, vomiting, anorexia, photophobia, phonophobia, osmophobia (aversion to odors), blurred vision, lightheadedness, and nasal congestion. Some patients experience cognitive impairment producing forgetfulness, irritability, and depression, whereas others may be manic, with outbursts of anger that can be disruptive in the ED setting. Many patients have dramatic light and sound sensitivity and seek a cool, dark, and quiet room.

The aura of classic migraine consists of focal neurologic symptoms that precede and herald the migraine attack. By definition, the aura is fully reversible and typically lasts 10 to 20 minutes, although it may continue for as long as 1 hour. The most common aura is visual; features may include scintillating scotomas (bright rim around an area of visual loss), teichopsias (subjective visual image perceived with eyes open or closed), fortification spectrums (zigzagged wall of fortress slowly drifting across visual field), photopsias (poorly formed brief flashes or sparks of light), and blurred vision. Less common auras include somatosensory phenomena such as tingling or numbness, motor disturbances, and cognitive or language disorders.[19]

Ophthalmoplegic migraine is a rare syndrome associated with paresis of one or more ocular nerves, most commonly the third cranial nerve. Patients typically present with ipsilateral headache associated with extraocular muscle paresis and occasionally pupillary changes. The ophthalmoplegia or pupillary changes may last for days to weeks and, rarely, may become permanent.[20] Because of the neurologic abnormalities, secondary causes including intracranial aneurysm and mass lesion must be ruled out.

Hemiplegic migraine is characterized by episodic hemiparesis or hemiplegia as an aura to the migraine attack. The progression of the motor deficit is slow or marching in quality and in most cases is accompanied by a sensory disturbance as well. The neurologic symptoms last 30 to 60 minutes, followed by a severe pulsating headache. Rarely, the motor deficit is persistent, resulting from a true migraine-induced stroke.

Basilar-type migraines arise with an aura referable to the brainstem and are associated with multiple neurologic findings, including visual symptoms (often total blindness), dysarthria, tinnitus, vertigo, bilateral paresthesias, paresis, and altered level of conciousness.[21] The symptoms are stereotypic and resolve spontaneously.

Status migranosus is a severe migraine headache that persists longer than 72 hours. Associated symptoms are debilitating, and patients often require hospitalization for pain management and supportive care.

Many factors can trigger migraine headaches in predisposed persons. Common precipitants include sleep deprivation, stress, hunger, hormonal changes including menstruation, and the use of certain drugs including oral contraceptives and nitroglycerin.[8] In addition, some patients report specific food sensitivities including chocolate, caffeine, and foods rich in tyramine, monosodium glutamate, and nitrates.[22,23] Alcohol, specifically red or port wine, has also been implicated. In others, certain sensory stimuli such as a strong glare or strong odors, loud noises, or weather changes can trigger an attack.[24]

Differential Diagnosis. Because of the complexity of the symptomatology, migraine headaches may be difficult to distinguish from other, secondary causes of headache. Other disorders that mimic migraine include ruptured berry aneurysm, arteriove-

nous malformation, intracranial mass lesions, giant cell arteritis, and cerebrovascular disease.

Diagnostic Evaluation. Routine neuroimaging is not necessary for patients with typical recurrent migraine headaches. However, neuroimaging must be considered for patients with new-onset headaches, headaches with a progressive course or change in pattern, headaches that never alternate sides, and headaches associated with any neurologic abnormalities or seizures. Such patients have a substantially higher likelihood of having a secondary cause such as tumor, arteriovenous malformation, or structural lesion.[25] In addition, patients who present with a severe headache or the "worst headache of [their] life" require a lumbar puncture to rule out subarachnoid hemorrhage if findings on a computed tomography (CT) scan are negative.

Treatment. The pharmacologic treatment of migraine is divided into *abortive* therapies, which attempt to limit the intensity and duration of a given episode, and *prophylactic* therapies, which are intended to decrease the frequency and intensity of attacks.[26] The goals of acute migraine therapy include treating attacks rapidly and consistently to avoid headache recurrence, restoring the patient's ability to function, and minimizing the use of backup and rescue medications.[24]

Patients who are unable to control their headaches at home often present to the ED for better pain control or supportive therapy. There are several approaches to treating the acute headache episode, depending on the severity of the attack (Table 101-1). In addition, patients with a history of migraine may relate specific interventions that have been successful. The choice of agents depends on several factors, including the patient's previous response to specific therapies, the existence of comorbid conditions, and the presence or absence of nausea or vomiting. Gastric stasis is common during acute migraine attacks and may limit the effectiveness of oral agents.

For mild to moderate attacks, the IHS recommends simple analgesics such as acetaminophen or nonsteroidal anti-inflammatory drugs (NSAIDs). In the presence of nausea or vomiting, adding an agent such as metoclopramide enhances the absorption and effectiveness of these medications. Appropriate doses and possible side effects are listed in Table 101-1.

For moderate to severe attacks, several classes of medications are available to treat the pain in addition to the nausea and vomiting that frequently accompany the headache. Specific agents available for treating severe migraine include DHE and the triptans. DHE should be given intravenously (IV) in a dose of 1.0 mg over 2 minutes; this can be repeated in 1 hour if pain control has not been achieved. Because DHE can cause nausea and vomiting, patients should be pretreated with an antiemetic such as metoclopramide 10 mg IV or prochlorperazine 5 mg IV. Repeated administration of the intravenous form of DHE has been shown to be very effective in patients with intractable migraine and status migranosus (i.e., a migraine attack lasting more than 72 hours). Contraindications to using DHE include pregnancy, breast-feeding, poorly controlled hypertension, coronary artery disease, and peripheral vascular disease. DHE should not be used if the patient has already taken any drug in the triptan class.

Sumatriptan, the first-approved medication of the triptan class, is a selective 5-HT (1B/1D) receptor agonist. Other triptans that are available include zolmitriptan, naratriptan, and rizatriptan, but only sumatriptan is available for subcutaneous administration, and it is the most common preparation used in the ED setting. The initial dose is 6 mg given subcutaneously, which may be repeated once in 1 hour if the patient has a partial response to the first dose. Common side effects include tingling, flushing, warm or hot sensations, and heaviness in the

Table 101-1 Selected Medications for Acute Migraine Attacks

MEDICATION	DOSE AND ROUTE ADMINISTERED	COMMENTS
Mild to Moderate		
Acetaminophen	500–1000 mg PO	Gastrointestinal upset
Aspirin	650–1000 mg PO	Gastrointestinal upset
Ibuprofen	600–800 mg PO	Gastrointestinal upset
Naproxen sodium	275–550 mg PO	Gastrointestinal upset
Tolfenamic acid	200–600 mg PO	Gastrointestinal upset
Moderate to Severe		
Dihydroergotamine	1 mg IV or IM; may be repeated in 1 hour	Gastrointestinal upset (pretreat with antiemetic) Chest pain, throat tightness, flushing
Triptans		
Sumatriptan	6 mg SC; may be repeated once in 1 hr if partial response	Contraindicated with hypertension, coronary artery disease, peripheral vascular disease, and pregnancy
Sumatriptan	25–100 mg PO	Cannot be used within 24 hours of ergot usage
Rizatriptan	5–10 mg PO	
Zolmitriptan	2.5–5 mg PO	
Naratriptan	1–2.5 mg PO	
Prochlorperazine	10 mg IV or IM; may be repeated in 30 to 60 minutes	Sedation and dystonic reaction
Metoclopramide	10 mg IV	Dystonic reaction
Ketorolac	30 mg IV or 30–60 mg IM	Gastrointestinal upset; avoid this medication in elderly and in patients with renal insufficiency
Morphine	2–4 mg IM or IV	Opioids less efficacious than other treatment modalities
Refractory Attack, Status Migranosus		
Dihydroergotamine	1 mg IV q8h	Use in conjunction with antiemetic (e.g., metoclopramide, prochlorperazine)
Steroids	Various regimens	Gastrointestinal bleeding, infection, cataracts, aseptic necrosis, memory disturbances

chest. Sumatriptan has contraindications similar to those for DHE and should not be used within 24 hours of administration of an ergotamine-containing medication or DHE.[27] In addition to the subcutaneous preparation, sumatriptan and the other triptan agents are available in oral formulation for the treatment of acute migraine attacks.

Neuroleptics also have been shown to be effective in treating acute migraine attacks. Prochlorperazine can be administered as a slow 10-mg intravenous bolus, which can be repeated once in 30 to 60 minutes.[16,28] The most common side effects after parenteral administration include sedation, postural hypotension, and extrapyramidal symptoms including acute dystonic reactions.

Narcotic analgesics such as morphine should be reserved for patients who do not respond or have contraindications to standard migraine therapies. Although frequently used, narcotics have been shown to be less efficacious than other agents and are associated with a risk of addiction; however, some patients obtain relief with this class of medications.

The use of steroids for the treatment of migraine remains controversial. Anecdotal evidence suggests that they may be effective for prolonged migraine attacks that are refractory to standard therapies and for treating status migranosus.[13,29]

Occasionally, patients do not respond to initial therapy in the ED and require hospitalization for continued pain control and supportive therapy.

Prophylactic Therapy. Prophylactic therapy is indicated for patients who have frequent attacks (more than two or three episodes per month), prolonged attacks lasting more than 48 hours, or attacks that are severe and debilitating. Of note, prophylactic medications are seldom more than 55 to 65% effective.[30]

Several classes of medications are used for the prophylaxis of migraine. Many of these medications have significant side effects, especially among women of childbearing age; therefore, after headaches have decreased, attempts should be made to taper and discontinue treatment when possible.

Beta-adrenergic blocking agents reduce both the frequency and severity of migraine headache and are the drugs most widely used for prophylaxis in patients with recurrent migraine.[8] Propranolol has been the most extensively studied medication. Patients who do not respond to propranolol may respond to another drug in this class, which includes atenolol, metoprolol, timolol, and nadolol.[31] Contraindications to beta-blockers include pregnancy, asthma, heart failure, Raynaud's phenomenon, and diabetes mellitus.[13]

Other medications used for migraine prophylaxis include calcium channel blockers, tricyclic antidepressants, anticonvulsants including divalproex sodium and sodium valproate, and monoamine oxidase inhibitors.[13,31]

Methysergide, a semisynthetic ergot preparation, also has been widely used for prophylaxis. It is a potent peripheral serotonin antagonist with a presumed mechanism of action similar to that for other ergot drugs. Its use is contraindicated in patients with coronary artery or peripheral vascular disease. Prolonged use has been associated with retroperitoneal, pulmonary, and endocardial fibrosis.[32]

Cluster Headache

Perspective. Cluster headache is the only headache syndrome that is more common in men than in women. It typically occurs in young to middle-aged adults who smoke, with a peak incidence in the late 20s.[33] The headaches tend to occur repeatedly over a defined time interval—hence the term "cluster." Several attacks can occur in 1 day, and a typical cluster period may last 6 to 8 weeks. Several precipitating factors have been implicated, most notably the ingestion of alcohol. Stress and climatic changes may also play a role in susceptible persons.

Clinical Features. Cluster headaches occur suddenly with little warning, and several episodes can occur within a 24-hour period. Each headache lasts from a few minutes up to 2 hours. The headache typically begins with a unilateral sharp, stabbing pain in the eye, which may awaken the patient from sleep. The attacks tend to occur exclusively in the territory of the trigeminal nerve.[34] Unlike the migraineur, the patient with cluster headache presents in a predictable fashion (i.e., holding a hand to the affected eye, rocking, rubbing the head, and pacing). The attack subsides rapidly, often leaving the patient exhausted.

Up to 30% of patients have a partial Horner's syndrome with ptosis and miosis.[35] The eye often is injected and tearing, and many patients have unilateral nasal congestion.[12]

Differential Diagnosis. Other headache disorders that mimic cluster headache include migraine, trigeminal neuralgia, and chronic paroxysmal hemicrania (CPH). With migraine, the clinical presentation, gender, and age distribution usually are different. With trigeminal neuralgia, the pain peaks within seconds, lasts only a couple of minutes, and can be provoked by specific trigger points on the face or oral mucosa. CPH is manifested by a brief unilateral headache that recurs at least 15 times a day, often induced by rotation or turning of the head or by pressure on the cervical spine.[36]

Treatment. Because cluster headaches are abrupt in onset, treatment must be initiated rapidly to be effective. At present, subcutaneous sumatriptan 6 mg is the preferred abortive therapy in most cases if it can be given very early after the onset of the attack[37]; however, by the time the patient presents to the ED, a full-blown headache usually has developed, and symptomatic treatment is indicated. High-flow oxygen at a rate of 7 to 10 L/minute has been shown to abort the headache within several minutes.[38] DHE 1 mg given IV or intramuscularly (IM) also has been shown to be effective, but it is less practical than oxygen administration and has more side effects. For patients who do not respond to these measures, intranasal application of cocaine[30] or lidocaine to produce anesthesia of the sphenopalatine region has been advocated by some clinicians but has not gained widespread acceptance.[39]

In addition to acute therapies, several medications have been shown to be effective for the prophylactic treatment of cluster headaches. A short course of oral prednisone may effectively abort a cluster attack in some patients. A recommended regimen is 60 mg of prednisone daily for 10 days, followed by a 1-week taper.[39] To prevent breakthrough headaches after the steroid taper, patients may require the concurrent administration of another prophylactic agent (e.g., verapamil, lithium carbonate, methysergide).

Tension Headache

Perspective. Tension headache is the most common recurrent pain syndrome, affecting more than 78% of the population.[40,41] Women are affected more frequently (80% in all women) than men (66%), and most patients are middle-aged.[7] The headaches typically do not cause significant disability, and patients are able to continue with their normal daily activities.[42] The median frequency of headaches is six per month, and stress and lack of sleep are implicated as triggering factors.[43,44] The average duration of the headache is 4 to 13 hours, with a maximum of 72 hours.[44]

Little is known about the pathophysiology of tension headache.[40] There is no clear evidence that increased muscle activity is present, and the physical examination will reveal tender areas of the scalp and neck with both tension and

migraine headaches. Evidence suggests that tension and migraine headaches may be part of a continuum with similar pathophysiology.

Clinical Features. Patients typically complain of a tight, bandlike discomfort around the head that is nonpulsating and dull. They also may experience tightening of the neck muscles. A majority do not seek medical assistance because the headache usually is mild in intensity and of relatively short duration. Occasionally, the discomfort can build up slowly and fluctuate in severity over several days. Unlike in migraine, the headache does not worsen with physical activity, and accompanying symptoms such as nausea, vomiting, phonophobia, or photophobia are unusual. Anxiety and depression may coexist with chronic tension headache, which by definition occurs more than 15 days a month and can be daily and unremitting.[33]

Differential Diagnosis. Tension headache is the least distinct of all of the primary headache disorders, and its diagnosis is based mainly on the absence of features that would suggest another cause for the headaches. Because of this lack of specificity, the clinician often may hesitate to make this essentially benign diagnosis without other investigations to exclude organic disease.[45] The most common disorders mimicking tension headache include idiopathic intracranial hypertension, oromandibular dysfunction, cervical spondylosis, sinus or eye disease, and intracranial masses.

Treatment. For a majority of patients with tension headaches, simple analgesics such as acetaminophen or an NSAID are adequate for pain control. Because tension-type headache is more common in sedentary persons, a regular exercise program may help.[33] Patients with chronic symptoms may exhibit signs of depression or anxiety, and these patients often respond to medications and nonpharmacologic regimens that treat these conditions. Some nonpharmacologic regimens are meditation, massage, and biofeedback. For long-term management, psychotherapy may be of value in teaching patients to deal with tension effectively.

SECONDARY HEADACHE DISORDERS

Subarachnoid Hemorrhage

Principles of Disease. Subarachnoid hemorrhage (SAH) refers to extravasated blood in the subarachnoid space. Presence of the blood activates meningeal nociceptors, leading to diffuse occipital pain along with signs of meningismus. SAH accounts for up to 10% of all strokes and is the most common cause of sudden death from a stroke.[46]

Approximately 80% of patients with nontraumatic SAH have ruptured saccular aneurysms.[47] Other causes include arteriovenous malformations, cavernous angiomas, mycotic aneurysms, neoplasms, and blood dyscrasias. SAH may be caused secondarily by an intraparenchymal hematoma that dissects its way into the subarachnoid space.

The risk for aneurysmal SAH increases with age, with most cases occurring between the ages of 40 and 60 years.[48] In children and adolescents, aneurysms are uncommon, and when SAH occurs it usually is secondary to an arteriovenous malformation.[49] It is estimated that 5% of the general population harbor a berry aneurysm, and the risk of rupture may increase with aneurysmal size. Other risk factors associated with SAH include hypertension, smoking, excessive alcohol consumption, and sympathomimetic drugs.[50,51] Increased systolic blood pressure values and long-term hypertension before aneurysm rupture seem to predict fatal SAH independently of aneurysm size or the patient's age at the time of rupture; patient gender also does not influence mortality.[52] A familial association of cerebral aneurysms with several diseases, including autosomal dominant polycystic kidney disease, coarctation of the aorta, Marfan's syndrome, and Ehlers-Danlos syndrome type IV, has been described.

Of all patients presenting to the ED with headache, 1 to 4% have SAH. Many patients with SAH die before reaching the hospital, with preadmission mortality rates ranging from 3% to 26%.[47] Because of the significant morbidity and mortality associated with this condition (with reported rates of up to 50%) and the high likelihood of clinical deterioration in patients who initially are misdiagnosed, SAH should be a primary consideration in the initial ED evaluation. Accordingly, familiarity with its presentation is essential.[47]

Clinical Features. A majority of patients with SAH present with a sudden, cataclysmic "thunderclap" headache, which often is described as "the worst headache of [their] life." The onset of headache may be associated with exertional activities such as exercise, the Valsalva maneuver, or sexual intercourse in up to 20% of patients.[46] One study demonstrated that moderate to extreme physical exertion in the previous 2-hour period was associated with a tripling of the risk for SAH.[53] Associated signs and symptoms include nausea and vomiting in approximately 75% of patients, neck stiffness in 25%, and seizures in 17%.[48] Some patients experience a headache within the previous 6 to 8 weeks, indicating a warning leak or sentinel hemorrhage. Physical findings depend on the extent of the SAH. Meningismus is present in more than 50% of patients,[49] and up to 20% have focal abnormalities.[50] Funduscopic examination may reveal retinal or subhyaloid hemorrhages, and patients also may have an isolated third or sixth nerve palsy. Oculomotor (third) nerve compression secondary to an expanding aneurysm leads to pupillary dilation. Approximately 50% of patients with a ruptured aneurysm are restless or have an altered level of consciousness. Although a majority do not have focal neurologic signs, such signs when present may indicate the site of the aneurysm.[54]

The patient's prognosis is related to neurologic status at hospital admission. The Hunt and Hess scale stratifies patients according to their clinical signs and symptoms at the time of presentation and is predictive of outcome[46,55] (Table 101-2). Patients who present with a grade I or II hemorrhage tend to have a good prognosis, and patients in grades IV and V tend to do poorly. These latter patients have an altered mental status, ranging from stupor to deep coma, together with focal neurologic signs and symptoms. Patients with grade III hemorrhage present with drowsiness or confusion and are at risk for rapid clinical deterioration.

Diagnostic Studies. When the diagnosis of SAH is considered, a CT scan should be ordered emergently. Figure 101-1 shows an example of SAH on a CT scan. For acute hemorrhage less

Table 101-2 Hunt and Hess Clinical Grading Scale for Cerebral Aneurysms and Subarachnoid Hemorrhage

GRADE	CONDITION
0	Unruptured aneurysm
1	Asymptomatic or minimal headache and slight nuchal rigidity
2	Moderate or severe headache, nuchal rigidity, no neurologic deficit other than cranial nerve palsy
3	Drowsiness, confusion, or mild focal deficit
4	Stupor, moderate to severe hemiparesis
5	Deep coma, decerebrate posturing, moribund appearance

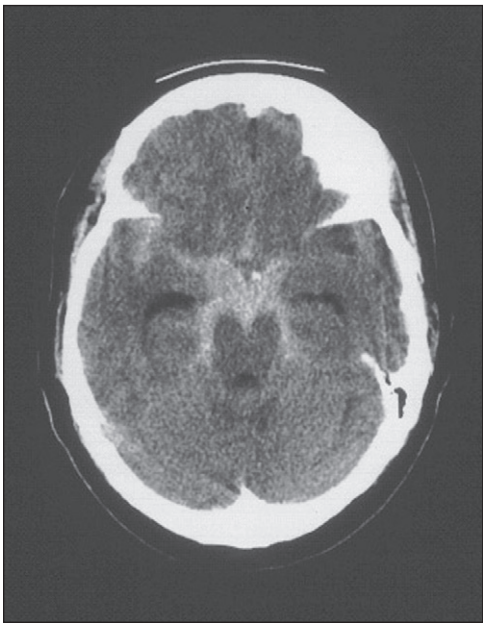

Figure 101-1. Cerebral aneurysm. Shown is a computed tomography scan of an aneurysmal subarachnoid hemorrhage in a 55-year-old woman. Subarachnoid blood can be seen within the interpeduncular and ambient cisterns and the right sylvian fissure, from a ruptured aneurysm at the junction of the right carotid artery and the posterior communicating artery. (From Soliman E, Kader A, Perez N: Cerebral aneurysm. Online article at eMedicine.com. Available at http://www.emedicine.com/med/topic3468.htm, picture 8.)

than 24 hours old, the sensitivity of CT in identifying hemorrhage is greater than 90%; however, it decreases to approximately 50% by the end of the first week.[47] When the CT scan is negative, a lumbar puncture should be performed. Using lumbar puncture as a first strategy, postulated to be cost-effective in carefully selected patients who have completely normal physical examinations, may be safe but has not been studied clinically.[56] To differentiate a traumatic lumbar puncture from SAH, the patient's cerebrospinal fluid (CSF) should be spun and the supernatant observed for xanthochromia. The yellowish pigmentation is secondary to the metabolism of hemoglobin to pigmented molecules of oxyhemoglobin and bilirubin, a process that takes approximately 12 hours to occur.[57,58] The method of comparing the red blood cell count in the first and the last tubes of CSF has been shown to be unreliable.[47] CSF xanthochromia in association with negative findings on the CT scan is diagnostic of SAH. After the diagnosis is established, angiography should be performed to study the vascular anatomy and identify the source of hemorrhage in patients who are candidates for surgical intervention.

Most authorities agree that the presence of xanthochromia as measured by spectrophotometry, which is much more sensitive than visual inspection,[47] is the primary criterion for a diagnosis of SAH. However, because xanthochromia may require up to 12 hours to be present after the initial bleeding, patients with persistently bloody CSF without xanthochromia should undergo vascular imaging when the level of clinical suspicion of SAH is high.[47]

Up to 90% of patients with SAH have cardiac arrhythmias or electrocardiographic abnormalities suggestive of acute cardiac ischemia, which may lead to an erroneous primary cardiac diagnosis.[47] Typical electrocardiographic findings include ST-T wave changes, U waves, and QT prolongation.[59]

Treatment. The management of SAH is complex and includes initial resuscitation, stabilization, and emergent neurosurgical consultation. The goals of management are to treat the acute medical and neurologic complications, prevent recurrent hemorrhage, and forestall the ischemic complications of vasospasm.[60] Because of an altered level of consciousness, patients with SAH of grade III or higher are at risk for respiratory depression and hypercapnia, which can lead to further increases in intracranial pressure (ICP); therefore, these patients may require early endotracheal intubation. Blood pressure also must be closely monitored because of the risk of continued bleeding or recurrent hemorrhage. Nimodipine, a calcium channel blocker, should be started soon after a diagnosis of aneurysmal SAH is made, to lessen the likelihood of ischemic stroke. Because nimodipine may cause transient hypotension in some patients, hemodynamic monitoring is required during its administration. The recommended dose is 60 mg by mouth or nasogastric tube every 4 hours. Calcium antagonists reduce the risk of poor outcome and secondary ischemia after aneurysmal SAH.[61] A Cochrane review concluded that treatment with antifibrinolytics (e.g., aminocaproic acid) does not improve overall outcome because the reduction in the rate of rebleeding is offset by an increase in poor outcome caused by cerebral ischemia.[62]

Blood pressure management should be determined by the patient's clinical status with involvement of the treating neurosurgeon. A typical goal would be a systolic blood pressure less than 160 mm Hg or a mean arterial pressure less than 130 mm Hg unless vasospasm is present.[63]

Analgesics, including opioids, should be used for persistent headache. In patients who are nauseated or at risk for vomiting, antiemetics also must be administered. Agitated patients require sedation, and all patients should be placed at bedrest in a quiet and dark environment. Clinically evident seizures should be treated with anticonvulsants, but the prophylactic use of these drugs is controversial.[59] A majority of these patients require hemodynamic and ICP monitoring in an intensive care setting. The role of surgery (e.g., aneurysmal clipping) versus endovascular coil embolization is not yet fully defined.[64]

Brain Tumor

Principles of Disease. Headache is the most common presenting complaint with brain tumor, being reported by approximately 50% of the patients.[65] A majority of these patients are elderly and have a cerebral metastasis as a cause of their headache.[66] The most common causes of metastasis are lung and breast carcinoma, followed by malignant melanoma and carcinomas of the kidney and gastrointestinal tract.[54] Primary brain tumors are much less common and typically occur in adults younger than 50 years.

The headache can be caused by several mechanisms, including direct involvement and traction on pain-sensitive structures such as meninges or larger cerebral vessels, or may be a symptom of increased ICP. The pain patterns produced are highly variable, depending on the location of the mass and the structures involved.[54] Headaches often but not always are on the same side as the tumor. With increased ICP, the pain often is bifrontal or bioccipital and may be accompanied by vomiting. Brain tumors also may disrupt sleep, awakening the patient during the night. This effect may be related to increases in cerebral pressure that occur with recumbency and sleep-related carbon dioxide retention.[9] Rapidly growing tumors are more likely to be associated with headache.[67]

Clinical Presentation. The typical patient presents with complaints of a worsening headache that has been present for weeks to months. The headache may have been present initially only on awakening, gradually becoming continuous. The classic triad of brain tumor headache—sleep disturbances,

severe pain, and nausea and vomiting—is seen in only one third of patients.[68] Vomiting, when present, may be projectile and not preceded by nausea. If increased ICP is present, the headache often is bilateral and worsened by coughing, sneezing, bending, defecation, and sexual intercourse.[69] Although patients may not complain of focal neurologic deficits, abnormal findings are often found with neurologic testing.[70] Other presentations include seizures, personality changes, and cognitive difficulties.

Diagnostic Evaluation. The diagnosis of brain tumor is often suspected from the history and neurologic examination. Neuroimaging with CT or magnetic resonance imaging (MRI) is the most efficient way to confirm the diagnosis. Contrast enhancement on CT often improves the identification of the underlying mass lesion and helps differentiate it from other causes, including abscess, hematoma, and vascular malformation.[70]

Treatment. Management consists of urgent referral to neurosurgery and treatment of any acute complications, including increased ICP and seizures. For patients who present with symptoms suggestive of increased ICP (e.g., headache, nausea, vomiting, confusion, weakness), treatment with steroids has been shown to be beneficial. Dexamethasone is the high-potency steroid used most often to treat edema associated with brain tumors. It has several advantages over other glucocorticoids, including a longer half-life, reduced mineralocorticoid effect, and a lower associated incidence of cognitive and behavioral complications.[70] The exact dose of steroids necessary for each patient varies in accordance with histology, size, and location of the tumor and the amount of edema present. In general, most patients require between 8 and 16 mg of dexamethasone per day. An appropriate starting dose in the ED is 10 mg IV, followed by 4 mg every 6 hours.

Patients with a seizure (generalized or partial) should receive anticonvulsant therapy. Appropriate first-line agents include phenytoin, carbamazepine, and valproic acid. Empirical or prophylactic treatment does not appear to delay or prevent the onset of seizure activity and may expose the patient to unnecessary complications and toxicity.[70]

Giant Cell Arteritis

Principles of Disease. Giant cell arteritis, or temporal arteritis, is a systemic inflammatory process of the small and medium-sized arteries. Extracranial branches of the aortic arch and the ophthalmic vessels most commonly are involved, but the process may affect any artery in the body.[71] The mean age at onset is 71 years, and it is rare before age 50. Females are affected more commonly than males.

Clinical Presentation. Headache is the most common initial manifestation of giant cell arteritis and occurs in more than 70% of patients with this disorder.[72] The headache often is of 2 to 3 months' duration and can be continuous or intermittent and often worsens at night or on exposure to cold. The pain may be described as sharp, throbbing, boring, or aching and usually is localized to the temporal region but may occur anywhere in the head. The physical examination may reveal tenderness over the scalp in the area of the temporal artery, with exacerbation of the pain by wearing a hat or resting the head on a pillow. Patients also may experience jaw claudication secondary to vascular insufficiency of the masseter and temporalis muscles. Systemic signs and symptoms including fever, anorexia, and weight loss often are present. Approximately 40% of patients complain of pain in their large proximal joints, with symptoms referable to the neck, torso, and lower back. Typically, pain and stiffness are worse in the morning and lessen as the day goes on.[72] This condition, known as *polymyalgia rheumatica*, can occur in the absence of giant cell arteritis.

The most serious complication of giant cell arteritis is permanent visual loss, which eventually occurs in 36% of untreated cases.[73] Amaurosis fugax also can occur before permanent visual loss. Other complications include peripheral neuropathies, transient ischemic attacks, and stroke.

Diagnostic Evaluation. The physical examination may reveal abnormalities of the temporal arteries, including tenderness, reduced or absent pulsations, erythema, and nodularity or swelling, best detected by light palpation just anterior and slightly superior to the tragus of the ear.[74] Visual acuity and visual field testing and a thorough funduscopic examination also should be performed.

A majority of patients have a significant elevation of the erythrocyte sedimentation rate (ESR), usually to more than 50 mm/hour and often more than 100 mm/hour, although an elevated ESR is not specific for the disorder and a normal value does not rule out the diagnosis. Other abnormalities on laboratory studies include mild to moderate anemia, elevated C-reactive protein level, and liver function abnormalities.[54] An elevated platelet count (greater than 400,000/μL) may be a risk factor for permanent visual loss.[75] The diagnosis is confirmed by temporal artery biopsy. Because this is a patchy disease, multiple biopsy specimens of a long segment of the artery may need to be examined.

Treatment. Because of the risk of visual loss, giant cell arteritis constitutes a medical emergency, and treatment should be initiated promptly when the diagnosis is suspected. Steroids are the mainstay of therapy; the recommended initial dose of prednisone ranges from 60 to 120 mg/day. Symptomatic response usually occurs rapidly over days, although therapy must be continued for months, with close ESR monitoring.

Carotid and Vertebral Dissection

Principles of Disease. Carotid and vertebral dissections are more common than previously realized. They are the most frequent cause of stroke in persons younger than 45 years, accounting for approximately 20% of all cases in this age group.[76] Although dissections may occur spontaneously, careful history taking frequently identifies an association with sudden neck movement or trauma preceding the event.[76,77] Reported mechanisms include neck torsion, chiropractic manipulation, coughing, minor falls, and motor vehicle accidents. Early symptoms and signs are often subtle, and in the absence of neurologic findings delays in diagnosis are common. The median delay from symptom onset to diagnosis was seven days in one report.[77]

The pathologic lesion is intramural hemorrhage within the media of the arterial wall. The hematoma can be localized or extend circumferentially along the length of the vessel, resulting in partial or complete occlusion. Platelet aggregation and thrombus formation also occur, further compromising vessel patency or causing distal embolization. The timing of these events is variable, and a patient may experience symptoms of cerebral ischemia days to years after dissection.[78,79]

Clinical Presentation. The typical presentation of the patient with carotid or vertebral dissection is the abrupt onset of pain in the neck or face. Neurologic findings usually occur within the first few hours, but autopsy studies have shown that strokes may occur months later.[76]

Carotid Dissection. The classic triad of symptoms for carotid dissection includes unilateral headache, ipsilateral Horner's syndrome, and contralateral hemispheric findings that may include aphasia, neglect, visual disturbances, or hemiparesis. The headache is often severe and throbbing but may be subacute and similar to previous headaches. Acute severe retro-orbital pain in a previously healthy person with no history of cluster headaches is particularly suggestive of carotid dissection.[12]

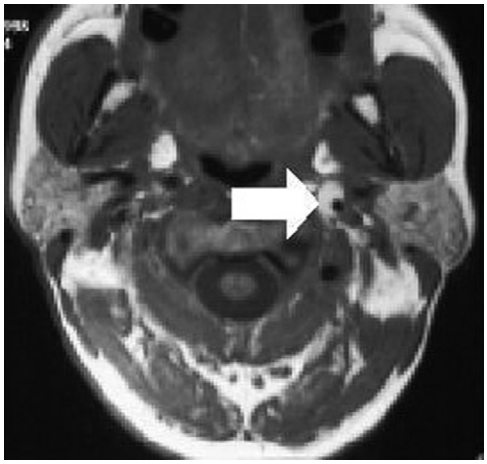

Figure 101-2. Axial T_1-weighted magnetic resonance image demonstrating a crescent sign *(arrow)* in a patient with a left internal carotid artery dissection. (From Kidwell C: Dissection syndromes. Online article at eMedicine.com. Available at http://www.emedicine.com/neuro/topic99.htm, picture 2.)

Most patients eventually develop signs of cerebral ischemia. Warning symptoms include transient ischemic attacks, amaurosis fugax, episodic light-headedness, and syncope. Spontaneous dissection of the carotid artery has a favorable prognosis and recurrence is uncommon.[79] Factors associated with a worse prognosis include old age, occlusive disease on angiography, or stroke as the initial presenting symptom.[80]

Vertebral Dissection. Vertebral artery dissections are less common than carotid dissections. The classic presentation is that of a relatively young person with severe, unilateral posterior headache and neurologic findings.[81] The majority of patients develop a rapidly progressive neurologic deficit with symptoms of brainstem and cerebellar ischemia. Common findings include vertigo, severe vomiting, ataxia, diplopia, hemiparesis, unilateral facial weakness, and tinnitus.[82] Spontaneous vertebral artery dissection appears to be relatively rare. Approximately 10% of patients who develop a vertebral dissection die during the acute phase, secondary to massive stroke. For patients who survive, the prognosis is usually good.[77]

Diagnosis and Treatment. The diagnosis of dissection may prove to be difficult. A CT scan should be obtained first but is often normal in uncomplicated dissection. Further imaging studies, including MRI, magnetic resonance angiography, or catheter angiography are required to confirm the diagnosis.[76] Figure 101-2 shows an example of carotid artery dissection on MRI. Duplex imaging is of limited value.[76] Treatment is aimed at stroke prevention and usually includes early anticoagulation followed by antiplatelet therapy.

Identifying patients with dissection is challenging. More than 50% of patients see their physician for symptoms before admission. The emergency physician must consider the diagnosis in any young patient who presents with head or neck pain with focal neurologic findings.

Cerebral Venous Sinus Thrombosis

Principles of Disease. Thrombosis of the intracranial veins and sinuses is an uncommon cause of stroke, in contrast with arterial causes. Because of the significant associated morbidity, however, cerebral venous sinus thrombosis (CVST) is an important consideration in the differential diagnosis for headache in patients with suggestive signs and symptoms.

Numerous factors have been associated with the development of CVST, including genetic and hypercoagulable disorders, pregnancy and the puerperium, inflammatory systemic disorders including vasculitis and connective tissue disorders, head trauma, CNS infections, medications (e.g., oral contraceptives, steroids), and neurosurgical procedures (e.g., dural puncture, internal jugular vein infusions).

Clinical Presentation. The clinical presentation can be quite variable, depending on the location of the thrombosis. Common symptoms and signs include headache, nausea and vomiting, seizures, decreased level of consciousness that may progress to coma, and focal neurologic deficits. Papilledema is frequent with chronic cases but is less common with acute presentations. With cavernous sinus thrombosis, the clinical picture is dominated by ocular findings including orbital pain, proptosis, and paralysis of extraocular movements.

Diagnostic Evaluation. An elevated D-dimer level is present in approximately 90% of cases of CVST and, along with the clinical findings, can be used to determine the need for further diagnostic testing in individual patients.[83,84] The diagnosis of CVST is based on neuroimaging of the area of thrombosis. CT by itself may reveal nonspecific lesions such as an infarct, hemorrhage, or edema. The key to diagnosis is to image the venous system itself, and this is best accomplished by a combination of MRI to visualize the thrombosed vessel and magnetic resonance venography (MRV) to detect nonvisualization of the same vessel.[85] CT angiography/venography has also been used to visualize the cerebral venous system.

Treatment. Specific treatment of CVST includes anticoagulation to prevent propagation of the thrombosis as well as complications (e.g., pulmonary embolism). In patients whose clinical condition worsens despite anticoagulation, thrombolysis or thrombectomy may be considered in centers with expertise in interventional procedures. Seizures should be treated with anticonvulsants.

The prognosis with CVST is based on the underlying etiology and the development of complications. During the acute phase, the overall death and dependency rate is approximately 15%. Approximately two thirds of patients will recover without sequelae, however—a rate far superior to that among patients who suffer an arterial stroke.

Idiopathic Intracranial Hypertension

Principles of Disease. Idiopathic intracranial hypertension (IIH) also is known as "pseudotumor cerebri" or "benign intracranial hypertension." The term *idiopathic intracranial hypertension*, however, is preferred, because this disorder is not always benign and may have significant neurologic sequelae in affected persons.

IIH is a relatively common neurologic disease seen primarily in young obese women of childbearing age. Several predisposing factors have been identified, including the use of oral contraceptives, anabolic steroids, tetracyclines, and vitamin A.[86,87]

Pathophysiology and Clinical Features. The pathophysiology of this disease remains controversial, with increased brain water content and decreased CSF outflow considered the two major causative factors.[88] The most prominent symptom is generalized headache, which often is gradual in onset and of moderate intensity. No specific localizing pattern has been documented, although in some patients the headache is worsened by eye movement. It may awaken the patient from sleep and is exacerbated by bending forward and the Valsalva maneuver, both of which impede cerebral venous return.

Visual complaints are common, and patients may experience transient visual obscuration several times a day secondary to

BOX 101-4 ICHD-II CRITERIA FOR DIAGNOSIS OF IDIOPATHIC INTRACRANIAL HYPERTENSION

1. Alert patient with neurologic examination that either yields normal findings or demonstrates any of the following abnormalities:
   A. Papilledema
   B. Enlarged blind spot
   C. Visual field defect (progressive if untreated)
2. Increased CSF pressure (>200 mm H_2O in nonobese patients, >250 mm H_2O in obese patients) measured by lumbar puncture in the recumbent position or by epidural or intraventricular pressure monitoring
3. Normal CSF chemistry (low CSF protein acceptable) and cellularity
4. Intracranial disease (including venous sinus thrombosis) ruled out by appropriate investigations
5. No metabolic, toxic, or hormonal cause of intracranial hypertension

CSF, cerebrospinal fluid; ICHD-II, International Classification of Headache Disorders, 2nd ed.
Available at http://ihs-classification.org/en/.

ischemia of the visual pathways. These episodes can be followed by prolonged periods of visual loss, which can become permanent in up to 10% of patients.[89] Patients also may complain of nausea, vomiting, and dizziness. The physical examination will reveal papilledema and visual field defects, including an enlarged blind spot initially, followed by loss of peripheral vision. Occasionally, a sixth nerve palsy is noted.

Diagnosis. The diagnosis of IIH should not be made without neuroimaging and measurement of ICP. The diagnostic criteria are listed in Box 101-4.

Treatment. Predisposing factors (e.g., discontinuation of implicated medications) should be corrected. Symptomatic treatment often includes lowering ICP and managing the headache. Acetazolamide (a carbonic anhydrase inhibitor) can be used to decrease CSF production alone or with a loop diuretic such as furosemide. Steroids also have been used, although their mechanism of action is unclear. Prolonged therapy is problematic, and rebound IIH often occurs when doses are tapered. Repeated lumbar punctures can be attempted, but most patients find this approach objectionable. In patients with impending visual loss or incapacitating symptoms, placement of a ventricular shunt or optic nerve sheath fenestration may be indicated.

Post-traumatic Headache

Headache is the most common symptom after minor head injury. It often is part of a complex syndrome that can include dizziness, fatigue, insomnia, irritability, memory loss, and difficulty with concentration. The prevalence of headache with post-traumatic syndrome is not known, because most patients are not admitted for this condition. There are approximately 2 million closed head injuries per year, and post-traumatic headache (PTHA) occurs in an estimated 30 to 50% of patients with these injuries.[90] Acute PTHA develops hours to days after the injury and resolves within 3 to 6 months.[91] Chronic PTHA may last from several months to years and may mimic other forms of headache, including tension and migraine headaches. The presence of headache, dizziness, or nausea on initial presentation is strongly associated with the development of chronic PTHA.[92]

Patients in whom PTHA develops after minor head injuries have normal findings on neurologic examination and neuroimaging studies. The pathophysiologic mechanism for their symptoms is unclear and may have both anatomic and functional components. Most patients are more concerned about the cause of the headache rather than the headache itself.

Treatment is symptomatic. For acute PTHA, analgesics such as acetaminophen or NSAIDs are adequate for pain control. For chronic PTHA, treatment must be individualized depending on the type of headache and associated symptoms the patient is experiencing. Novel therapies, such as antidepressants and beta-blockers, may be effective in selected patients.

Acute Glaucoma

Patients with acute angle closure glaucoma present with sudden onset of severe pain localized to the affected eye that may radiate to the ear, sinuses, teeth, or forehead.[69] Visual symptoms, including blurriness, halos around lights, and scotomas, typically are present, and many patients also experience nausea and vomiting. The underlying pathophysiologic mechanism is congenital narrowing of the anterior chamber angle that, under certain conditions, closes, resulting in a significant rise in intraocular pressure (IOP). Episodes can be precipitated by entering a low-light environment such as a movie theater, with resultant pupillary dilatation, or by the use of medications such as mydriatics (e.g., for dilated ocular examination), sympathomimetics (e.g., pseudoephedrine), or agents with anticholinergic properties (e.g., antiemetics, antihistamines, antipsychotics, antidepressants).[86,93]

Physical examination reveals a red eye with a fixed, mid-dilated pupil, corneal clouding, and shallow anterior chamber. The diagnosis is confirmed by demonstrating markedly elevated IOP in the range of 60 to 90 mm Hg (normal is less than 21 mm Hg).

Treatment includes topical miotics, topical beta-blockers, oral carbonic anhydrase inhibitors (e.g., acetazolamide, 250 mg four times daily), intravenous osmotic agents (e.g., mannitol), and prompt referral to an ophthalmologist. The potential for diagnostic confusion between acute glaucoma, iritis, and cluster headache must be recognized. Although cluster headache may arise with pain, nausea, and a red eye, vision is not affected and the pupil generally is small and the eyelid is ptotic (from an oculosympathetic paresis).[94] Acute iritis also arises with a painful red eye, but only acute angle closure glaucoma is associated with markedly elevated IOP.

Post–Dural Puncture Headache

Principles of Disease and Pathophysiology. Headache is the most common complication of lumbar puncture, occurring in up to 40% of patients.[95] The incidence is highest in the 18- to 30-year age group, but this complication is uncommon in young children and in adults older than 60. Although the onset often is immediate, patients may not report symptoms for several days. In a majority of affected persons, the duration of headache is less than 5 days.[95,96]

The cause of post–dural puncture headache (PDPH) is not entirely clear. The most likely explanation is a persistent CSF leak that exceeds CSF production, resulting in CSF hypotension. If sufficient CSF is lost, the brain descends in the cranial vault when the patient assumes the upright position, leading to increased traction on the pain fibers.[97] Thus, the headache is characteristically positional and increases with the upright position and decreases with recumbency. The amount of time a patient remains recumbent after lumbar puncture does not appear to affect the incidence of headache.[12]

Certain factors have been implicated as causes of PDPH, including the size or diameter of the spinal needle, the orientation of the bevel during the procedure, and the amount of fluid withdrawn. Smaller-diameter needles cause less leakage, and it is postulated that inserting the needle with the bevel up (i.e., bevel pointing up when the patient is in the lateral position) minimizes damage to the dural fibers. Using atraumatic needles or pencil-point needles (e.g., Whitaker[98] or Sprotte[99]) also has been shown to reduce significantly the incidence of PDPH.[100,101]

Clinical Features. PDPH typically is bilateral, throbbing, and exacerbated by the upright position. Associated signs and symptoms include neck stiffness; nausea; vomiting; auditory disturbances, including tinnitus and hearing loss (hypoacusis); and ocular symptoms, including blurred vision and diplopia.[97]

Treatment. Most PDPHs resolve spontaneously within a few days with bedrest, adequate hydration, and mild analgesics. For persistent headaches, methylxanthine agents have been found to help some patients. Oral caffeine (300 mg every 4 to 6 hours), caffeine sodium benzoate (500 mg in 1 L of fluid), or theophylline (300 mg PO every 8 hours) may be effective.[95] For severe headaches lasting longer than 24 hours, an epidural blood patch (autologous blood clot) relieves the headache in the majority of patients.[101]

Intracranial Infection

Headache is common among patients with intracranial infections, including meningitis, brain abscess, encephalitis, and acquired immunodeficiency syndrome. The severity and type of headache vary depending on the specific infection.

With acute bacterial meningitis, the patient often has a severe bursting headache that rapidly increases in severity over a short period.[102] These patients typically have significant meningismus, with both Kernig's and Brudzinski's signs. With viral meningitis, patients also may complain of severe headache and nuchal rigidity, but the course is more indolent than with bacterial meningitis.

The severity of headache associated with encephalitis depends on the type of virus involved. For example, the headache is usually mild with mumps encephalitis. With herpes simplex infection, however, the headache is abrupt and severe and frequently is associated with confusion, fever, altered level of consciousness, seizures, and focal neurologic signs.

Patients with brain abscess often have headache as their presenting complaint.[103] As the infection progresses, vomiting, focal neurologic signs, and depressed level of consciousness typically develop.

Headache is a frequent complaint in patients with human immunodeficiency virus infection and can be caused by a number of conditions, including aseptic meningitis, toxoplasmosis, cryptococcal or tuberculous meningitis, and cytomegalovirus encephalitis.

In a majority of cerebral infections, the mechanism of head pain includes meningeal irritation and increased ICP. In addition, headache may be a general reaction to fever or the toxic products of the infecting agent.[104]

Hypertensive Headache

Contrary to common belief, hypertension is not an important cause of headache, and the occurrence of headache and hypertension in the same patient is often coincidental.[105] Whether some patients with mild to moderate hypertension suffer from headache caused by elevated blood pressure is uncertain. The rate of blood pressure increase is more important as a cause of headache than the absolute blood pressure value. Diastolic pressures lower than 130 mm Hg are rarely the cause of headache.[72]

Nonetheless, the association of headache with severe hypertension is well documented. Acute, severe headache is a prominent symptom of hypertensive encephalopathy, and most patients have blood pressure readings in the range of 250/150 mm Hg. Other conditions include headache secondary to toxic agents (e.g., drug-induced hypertension), pheochromocytoma, and eclampsia.

The headache of severe hypertension typically is diffuse and is worse when the patient awakes in the morning and gradually subsides over the course of the day.[105] Treatment is directed at lowering the blood pressure; in most cases, the headache is relieved within 24 hours. In patients with hypertensive encephalopathy, the headache may persist for days until brain edema has resolved.

Cervicogenic Headache

Cervicogenic headache refers to headache originating from disorders of the neck. Diagnosis is based on the presence of one of the following three distinct sets of symptoms[106]:

1. Unilateral headache triggered by movements of the head or neck or certain head positions
2. Unilateral headache triggered by pressure on the neck
3. Unilateral headache spreading to the neck or possibly the ipsilateral shoulder or arm

Many of these headaches are reported after a whiplash injury. Even though neck structures play a primary role in the pathophysiology of some headaches, clinical patterns indicating a neck-headache relationship have not been adequately defined.

Medication-Induced Headache

Medication use, abuse, or withdrawal can be a cause of headache, and the term *medication-induced headache* is used to describe these conditions. Medication-induced headache is underdiagnosed and often difficult to manage.[107] Although not well understood, it tends to occur in patients with a primary headache disorder (e.g., migraine, tension-type) who use immediate-relief medications, often in excessive quantities.[108] Medications that have been implicated include NSAIDs, aspirin or acetylsalicylic acid (ASA), acetaminophen, barbiturate-analgesic combinations plus caffeine with or without codeine, opioids, caffeine, and ergotamine. A key factor in medication overuse headache is preemptive use of drugs, in anticipation of—rather than for—headache.[33] Women are affected more commonly than men, and the most frequently affected age group is that of persons between 30 and 40 years.[109] The headache itself is variable and may be accompanied by asthenia, nausea, anxiety, depression, and difficulty with concentration. Typically, it is worse on awakening in the morning and after physical exertion.

The symptomatic medication that leads to the development of this disorder initially provides some pain relief to the patient, but over time tolerance develops, and larger doses are required to obtain symptomatic improvement.[109]

Treatment typically requires complete withdrawal of the medication being overused, to achieve long-term results. In addition, these patients require a comprehensive education and follow-up program with pharmacologic, dietary, and behavioral components.[109]

Trigeminal Neuralgia

Trigeminal neuralgia is a painful unilateral affliction of the face, characterized by brief electric shock–like (lancinating) pains limited to the distribution of one or more divisions of the trigeminal nerve. Pain is commonly evoked by trivial stimuli (e.g., washing, shaving, smoking, talking, brushing the teeth) but also may occur spontaneously.[110] Individual attacks are brief, lasting a few seconds to less than 2 minutes, and are stereotypic in the individual patient. The lightning-like pains and unilateral grimaces characteristic of trigeminal neuralgia led to the designation of the term *tic douloureux*.[111] The diagnosis is straightforward in most patients on the basis of clinical criteria. However, because these symptoms also can be caused by an underlying mass lesion, CT or MRI is indicated in previously undiagnosed patients and when sensory loss or motor dysfunction is present.

Several drugs have been effective in treating trigeminal neuralgia, including carbamazepine, phenytoin, and baclofen; however, approximately 30% of patients fail to respond to medical therapy.[111] In these patients, surgical management, by alcohol or glycerol injection or microvascular decompression, may be indicated.[110,112]

Cough and Exertional Headache

In some patients, severe headache can be provoked by rapid increase in intra-abdominal pressure such as coughing, sneezing, laughing, heavy lifting or exertion, and the Valsalva maneuver. The pain starts within a few seconds of the precipitant and typically is brief when associated with cough but can last as long as 24 hours when associated with exertion. The headache is bilateral and throbbing in nature and in a majority of patients resolves spontaneously without persistent neurologic symptoms (e.g., neck stiffness or photophobia). In some patients, the headache may be secondary to structural lesions, especially in the posterior fossa[113]; therefore, all previously undiagnosed patients require CT, or preferably MRI, followed by lumbar puncture to rule out intracranial disease including SAH. For patients with recurrent benign exertional headache, treatment includes avoidance of the underlying triggering mechanism and use of analgesics as necessary. For patients with exertional headache, NSAIDs including indomethacin have been effective.[114]

Coital Headache

Coital cephalgia is a recurrent, benign headache associated with sexual activity and is more common in men than in women. Different types have been described, including headaches that occur before, during, or immediately after orgasm. They typically are occipital in location and may increase in severity with mounting sexual excitement. Their duration can be from minutes to hours. Occasionally, some patients experience a sudden, explosive headache that occurs during orgasm. In these patients, SAH should be ruled out.[113–115]

High-Altitude Headache

Headache is one of the cardinal manifestations of acute mountain sickness and can occur at altitudes higher than 5000 feet above sea level in unacclimatized persons. The headache is throbbing in nature, located in the temporal or occipital areas, and probably is caused by a mild increase in ICP secondary to brain swelling.[116] It is worse at night or in the early morning and exacerbated by the Valsalva maneuver or bending forward.[117] Other findings associated with high-altitude illness include fatigue, nausea, vomiting, dizziness, insomnia, and an altered mental status. Pulmonary edema and cerebral edema develop in severe cases. The treatment for these conditions includes supplemental oxygen and descent to a lower altitude.

KEY CONCEPTS

- Headache is a common presenting complaint in the ED. The goal of evaluation in this setting is to distinguish between benign primary headache disorders and the more serious and potentially life-threatening secondary causes of headache.
- A majority of patients do not have abnormal neurologic findings; therefore, the key to a successful diagnosis is a thorough and systematic history.
- Patients with the following headache presentations are at risk for serious underlying disease: sudden explosive headache; first or "worst-ever" headache; new-onset headache after age 50 years; headache associated with papilledema, alteration in or loss of consciousness, or focal neurologic symptoms; headache after head trauma; subacute headache with increasing frequency or severity; headache associated with fever, cancer, or immunosuppression; and headache triggered by exertion, sexual activity, or Valsalva maneuver.[118]
- The need for diagnostic studies is dictated by the suspected secondary cause of headache.

The references for this chapter can be found online by accessing the accompanying Expert Consult website.