Chapter 109

Unruptured Vascular Malformation and Subarachnoid Hemorrhage

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International Headache Society (IHS) code and diagnoses:
- 6.2.2. Headache attributed to subarachnoid hemorrhage
- 6.3. Headache attributed to unruptured vascular malformation
- 6.3.1. Headache attributed to saccular aneurysm
- 6.3.2. Headache attributed to arteriovenous malformation (AVM)
- 6.3.3. Headache attributed to dural arteriovenous fistula
- 6.3.4. Headache attributed to cavernous angioma

World Health Organization (WHO) codes and diagnoses: G44.810
- 644.810 Headache attributed to subarachnoid hemorrhage
- 644.811 Headache attributed to saccular aneurysm
- 644.811 Headache attributed to arteriovenous malformation
- 644.811 Headache attributed to dural arteriovenous fistula
- 644.811 Headache attributed to cavernous angioma

Short description: Subarachnoid hemorrhage (SAH) occurs when blood leaks between the layers of the pia-arachnoid membrane. Arteries and veins passing through this potential space are both possible sources of bleeding. The majority of SAHs arise from ruptured saccular aneurysms or AVMs (38). Miscellaneous causes include cavernous and venous malformations and capillary telangiectasias. The headache of SAH typically is abrupt in onset, developing within seconds, and is often described as the worst ever experienced. The headache is usually followed by pain radiating into the occipital or cervical region and is often accompanied by blunting of consciousness, vomiting, phono- and photophobia, and neck stiffness. The headache remains severe for hours and then clears over several days to a few weeks.

Patients with AVMs usually present with either an intraparenchymal hemorrhage (50 to 60%) or seizures (30%) (5,21,71). Patients with perimesencephalic or pretruncal SAH, in whom a saccular aneurysm or AVM cannot be demonstrated, may develop an explosive headache similar to that described by patients with an aneurysm, but loss of consciousness is exceptional and the course of the illness is generally benign (62,70).

Unruptured saccular aneurysms often remain asymptomatic for many years but may suddenly produce warning symptoms, including headache, because of either impending rupture (25,47) or progressive enlargement, leading to compression of neighboring structures (47). Unruptured AVMs may mimic migraine (8,31). Epilepsy is the most common clinical presentation of cavernous malformations (54), followed by signs and symptoms of cerebral hemorrhage. Venous malformations are incidental findings in patients presenting with seizures or headache. In the majority, a causal relationship between the angioma and the presenting symptoms is not established (49).

EPIDEMIOLOGY

The true prevalence of saccular aneurysms and other intracranial vascular malformations is not precisely known. Venous malformations and capillary telangiectasias have a low risk of bleeding or causing symptoms by other mechanisms and are often found incidentally at autopsy (71). They occur in approximately 2.6% of subjects (71).

The prevalence of cavernous malformations is 0.5% in autopsy series and 0.4 to 0.9% in serial magnetic resonance (MR) studies (55). They affect both sexes with equal frequency. Most cavernous malformations appear as solitary lesions; however, they have been reported in association with other vascular lesions such as capillary telangiectasias, AVMs, and venous malformations (55). The

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prevalence of cavernous malformations range between 0.02% in autopsy series and 0.9% in MR studies (40).

One retrospective MR angiography study indicated that unsuspected aneurysms are seen in 2.8% of studies (24). Conversely, their frequency in prospective studies is 3 to 6% (53). Postmortem studies of consecutive autopsies indicate that approximately 5% of the population may harbor one or more saccular aneurysm (9,60). In these studies, more than half of the demonstrated aneurysms were unruptured and unrecognized prior to death. Long-term follow-up studies of patients harboring incidentally discovered cerebral aneurysms suggest that the vast majority never rupture or cause any symptoms (68).

The annual risk of rupture of incidental aneurysms less than 7 mm in diameter is low (0.1%), but the risk is higher (1.5% with aneurysms between 7 and 12 mm). The annual risks for additionally discovered aneurysms are 0.4% and 0.8%, respectively (41).

Twenty to 30% of patients with cerebral aneurysms have multiple lesions (aneurysms), usually two or three (47,53). Arterial hypertension and the presence of multiple aneurysms highly positively correlate and, considering both experimental and epidemiologic studies, arterial hypertension is considered a risk factor for aneurysm formation (47). The significance of hypertension for aneurysmal rupture, however, is less conclusive (47).

Overall, aneurysmal SAH is more common in women than in men (39,65). However, in considering the gender difference by decade, a 4:1 male:female ratio is encountered in the first decade of life, which becomes 1:1 by the fifth decade of life (67). The increased incidence of aneurysmal SAH in women is most probably related to their greater susceptibility to aneurysm formation, rather than an increased risk of aneurysmal rupture (47). In contrast, there is a modest male preponderance among patients with AVMs (71).

An AVM and a saccular aneurysm coexist in approximately 10% of patients. Because the associated aneurysms have a predilection to the AVM feeding arteries, it is believed that increased blood flow is a major factor in the formation of the saccular aneurysm (76). When bleeding occurs, it is more often from the aneurysm than from the AVM (62).

Estimates of the annual incidence of SAH depend on the population surveyed, the methods used for analysis, and the accuracy and extent of the investigations. Reports can be divided into epidemiologic surveys and referral centers or large-scale cooperative studies. Epidemiologic studies include not only hospitalized patients, but also those 15 to 20% of individuals with SAH who died before receiving medical treatment. In the Western countries, the average annual incidence of SAH is estimated at 11 per 100,000 population, with variations for age, sex, and geographic locations (56,62). The incidence of SAH has remained stable over the last 30 years. Saccular aneurysms account for approximately 85% of SAH; nonaneurysmal perimesencephalic hemorrhage about 10%; and AVMs, cavernous, venous malformations, or capillary telangiectasias the rest (62). Patients with angiographically negative SAH (e.g., perimesencephalic or pretruncal hemorrhage) have an excellent prognosis and are unlikely to rebleed (62,70). Rupture of a dilated vein or venous malformation in the prepontine or interpeduncular cistern is believed to be responsible for the majority of these cases (53).

Up to 50% of patients with saccular aneurysms who are admitted to neurosurgical departments experience warning symptoms in the form of minor bleeding episodes, days or even several months before a major hemorrhage (25,47,65). Headache is the most common symptom of this warning leak (47), occurring in 9 of 10 patients. Minor leaks occur with AVM as well, as evidenced by pathologic documentation of hemosiderin adjacent to the malformation (71). At surgery, at least 10% of AVMs show evidence of minor bleeding episodes. Small AVMs are more likely to cause minor bleeding than large ones (71).

GENETICS

There is no significant genetic predisposition to the development of AVMs (71). In contrast, a familial form of cavernous malformations, which is characterized by multiple lesions and an autosomal-dominant inheritance pattern, is caused by mutations in the CCM1–CCM3 genes on chromosomes 7 and 3, respectively (11,14).

Evidence supporting the role of genetic factors in the pathogenesis of intracranial aneurysms stems from the association of intracranial aneurysms with inherited connective-tissue disorders (e.g., autosomal polycystic kidney disease, Ehlers-Danlos syndrome type IV [vascular EDS]) and their familial occurrence (56). In contrast to sporadic aneurysms, familial ones (a) occur less often on the anterior communicating artery; (b) rupture at a younger age; and (c) are smaller in size at rupture (34). In a segregation analysis of published pedigrees, several possible patterns of inheritance of saccular aneurysms were identified, with autosomal transmission being the most likely (57). This suggests that genetic heterogeneity is an important feature of intracranial saccular aneurysms (56).

ANATOMY AND PATHOLOGY

After a hemorrhage, the subarachnoid space contains a variable mixture of cerebrospinal fluid (CSF) and clotted and liquid blood. The extent of dissemination varies considerably. Bleeding on the surface of the brain spreads out and collects later at the base, whereas a hemorrhage at the base initially fills the cisterns. In cases of severe SAH, the blood spreads within minutes over the convexities of the cerebral hemispheres. The average SAH releases 7 to 10 mL of blood into the CSF. A red blood cell (RBC)

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count of 105 per mm^3 indicates that 3 mL of blood entered the CSF. Following the initial hemorrhage, the RBC count decreases, and RBCs remain detectable in the fluid only for 4 to 21 days. The average survival of RBCs in CSF is much shorter than in the circulation. It has been suggested that RBCs in the subarachnoid space lose their cellular integrity, resulting in an immune-mediated hemolysis (50).

RBC lysis liberates pigments (oxyhemoglobin, methemoglobin, and bilirubin), causing the supernatant of the centrifuged CSF to stain yellow (xanthochromia). By spectrophotometry, oxyhemoglobin can be detected as early as 2 hours after the bleeding, but usually it takes a few hours or more for RBCs to lyse and for xanthochromia to develop. In a large series of patients with SAH, it was shown that xanthochromia could be detected in all patients in whom the CSF was examined within the initial 2 weeks and at least 12 hours after the hemorrhage (62,63).

In the subarachnoid space, extravasated blood causes an aseptic inflammatory reaction (22). The meningeal reaction is evident within 2 hours of the hemorrhage and begins as an outpouring of polymorphonuclear leukocytes, followed by the appearance of lymphocytes and large mononuclear phagocytes. This cellular reaction is transient and persists only as long as blood or products of the breakdown of blood are demonstrated in the subarachnoid space. Thickening and pigmentation of the pia and arachnoid occur, and hemosiderin-containing adhesions are forced among these membranes, the blood vessels, the nerves, and the brain. In case of aneurysmal SAH, the process is most marked at the base of the brain. If the exit foramina of the fourth ventricle are affected, obstructive hydrocephalus may occur. Hydrocephalus is more common after SAH in the territory of the anterior communicating artery, probably because blood is directed into the basal subarachnoid space. Hydrocephalus is also common in patients with multiple episodes of SAH because of the functional impairment of the arachnoid villi and increasing leptomeningeal fibrosis.

Cerebral vasospasm is one of the most important causes of death and disability in patients surviving the first critical days of SAH. Cerebral vasospasm is a syndrome of ischemic consequences of an angiographically proven, time-dependent, transient cerebral arterial narrowing. It is rarely pronounced before day 4 following the initial hemorrhage and peaks at approximately day 7. At that time, 40 to 70% of patients will have some reduction in the caliber of one or more of the arteries of the circle of Willis or its branches (10,28). The clinical symptoms of delayed cerebral ischemia are characterized by an insidious onset of confusion and decreased level of consciousness, followed by focal motor and speech impairment (10,28). Manifest neurologic deficits related to delayed cerebral vasospasm occur in 20 to 30% of patients with aneurysmal SAH, whereas they occur much less frequently in patients with SAH due to AVMs (28).

PATHOPHYSIOLOGY

The initial pain that follows SAH results from local distention, distortion, and stretching of the cerebral vessel and its adjacent arachnoid. It is a referred pain due to stimulation of arteries in the circle of Willis, which derive their innervation from the fifth, ninth, and tenth cranial nerves, and the upper cervical spinal nerves. Sensory fibers are directly stimulated by subarachnoid blood with the resultant release of neuropeptides such as substance P and initiation of head pain (17,37,44). Levels of calcitonin gene–related peptide (CGRP) are low in patients who die after SAH (18,19). Furthermore, there is increased release of CGRP following SAH (26), analogous to what is demonstrated in migraine. Increased intracranial pressure and the later development of hydrocephalus or delayed cerebral ischemia also may contribute to the genesis of headache.

Distention of an aneurysmal sac may produce pain through pressure on the free edge of the tentorium, with pain referred through trigeminal nerve fibers, or by direct pressure on the first branch of the fifth cranial nerve. Enlargement of aneurysms arising from the internal carotid artery (ICA) near the take-off of the posterior communicating artery (PComA) or the distal part of the basilar artery (BA) may produce incomplete or complete oculomotor nerve palsy. This is the most common nonhemorrhagic presentation of an intracranial aneurysm and is almost always accompanied by pain (51). Because cranial nerve III carries no pain-sensitive fibers, distention of the arterial wall is the most likely pain mechanism. The pain is almost always ipsilateral to the aneurysm and usually is peri- or retroorbitally felt (2).

Aneurysms of the intracavernous ICA, which constitute approximately 2 to 3% of all intracranial aneurysms, rupture only rarely. If they do so, they do not bleed into the subarachnoid space but produce a carotid–cavernous fistula with exophthalmos, congestion of conjunctival and fundus veins, and often a bruit audible over the ipsilateral eye and the skull. Carotid–cavernous fistulas are dural arteriovenous shunts that cause predominantly orbital and periorbital pain and discomfort, largely related to pressure buildup and compression of neighboring pain-sensitive structures. When not associated with a carotid–cavernous fistula, intracavernous saccular aneurysms can produce pain along the first and second divisions of the trigeminal nerve (probably from direct compression of these structures or their branches) and palsies of the third, fourth, or sixth cranial nerves (65). Sometimes, if the pressure in the cavernous sinus is sufficiently increased, patients develop an acute cavernous sinus syndrome with paresis of muscles of ocular movement, secondary to compression of cranial nerves III, IV, or VI, and severe pain (64).

Generalized or localized headache episodes may be due to warning symptoms of impending SAH. Minor leakage of blood into the subarachnoid space stimulates the sensory nerve endings both mechanically and chemically.

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Alternatively, the pain could be caused by hemorrhage confined to the aneurysmal wall or a sudden expansion of the aneurysm (47).

Unruptured AVMs are sometimes discovered during investigations of patients complaining of headache (7). The literature is abundant with isolated cases and small case series implicating AVMs in migraine (43). However, because the majority of people have headaches, it is difficult to determine the true relationship (coincidental, contributory, or causative) between an AVM and headache. Bruyn argued that, based on prevalence and incidence figures of AVMs and migraine, the relationship between both conditions is causal (8). The mechanisms of AVM-related migraine headaches were not discussed.

Clusterlike headaches have recently been linked to unruptured middle cerebral artery AVMs (45). Muñoz and colleagues argued that the relationship between the headache and the AVMs is likely causal because both patients experienced no further cluster headache symptoms after excision of the lesions. Furthermore, they speculated that blood flow alteration in the intracavernous ICA sets a pericarotid neurogenic focus, leading to cluster headache.

Patients with headache and documented cavernous or venous malformations are considered asymptomatic (40,49). A recent report challenges that concept (13). De Benedittis reported the case of a 62-year-old man who presented with a cluster headache variant (SUNCT syndrome [short-lasting, unilateral, neuralgiform headache with conjunctival injection and tearing]) and later was found to have a cavernous angioma on MR imaging (MRI). De Benedittis postulated that the pontine lesion could involve the trigeminal root entry zone and irritate fibers of the first division of the trigeminal nerve as well as the greater superficial petrosal nerve, thus causing head and facial pain with dysautonomic symptoms (13).

CLINICAL FEATURES

Headache and Subarachnoid Hemorrhage

The IHS diagnostic criteria for headache attributed to SAH (6.2.2) (Revised International Classification for Headache Disorders [ICHD-II]) are as follows:

A. Severe headache of sudden onset fulfilling criteria C and D.
B. Neuroimaging (computed tomography [CT] or MRI T2 or FLAIR) or CSF evidence of nontraumatic hemorrhage with or without clinical signs.
C. Headache develops simultaneously with hemorrhage.
D. Headache resolves within 1 month.

A sudden headache that has never been previously experienced and that is accompanied by depressed consciousness and neck stiffness is the hallmark of SAH. Descriptive terms of the headache include "worst ever," "tremendous," "bursting," "exploding," and "unbearable." Although it could be initially focal and lateralized, SAH-related headaches rapidly generalize and radiate into the occipitonuchal region. With blood seeping into the spinal subarachnoid space, back pain, meningismus, and radicular symptoms follow (30).

The headache of aneurysmal SAH commonly, although not always, peaks instantly. Indeed, a prospective study of 102 patients with sudden severe headache showed that only 50% of the 42 patients with aneurysmal SAH complained of an instantly severe headache (35). The researchers also found that some patients with pretruncal nonaneurysmal hemorrhage (35%) and many with benign thunderclap headache (68%) complained of instantly severe headache. The headache of pretruncal nonaneurysmal hemorrhage may be more gradual, that is, develope over minutes rather than seconds, but the predictive value of this feature is poor (62).

The duration of SAH-related headache varies from 2 to 3 days, usually with the minor hemorrhages, and up to several days (average 8 days) with large hemorrhages. The excruciating headache that usually drives the patient to seek medical care is shorter lived, however, lasting 1 to 2 hours.

Various neurologic signs and symptoms accompany the headache of SAH (Table 109-1). Neck stiffness, disorientation, photophobia, nausea, and altered mentation are common. Unconsciousness for greater than 1 hour is associated with a high mortality rate. If consciousness is lost slowly or supervenes after a lucid period, then secondary hemorrhage or acute hydrocephalus should be suspected. Photophobia is part of the meningism that occurs in up to 60% of patients (65). Neuro-ophthalmologic findings include homonymous hemianopsia, papilledema, and

TABLE 109-1 Neurologic Symptoms and Signs of Subarachnoid Hemorrhage

Signs and Symptoms	*Frequency (%)*
Headache	85–95
Neck stiffness	74–84
Disorientation	48
Nausea, vomiting	45
Altered mentation	43
Focal motor deficits	20
Seizures, convulsions	15
Coma	14
Cranial nerve palsy	13
Papilledema	13
Ocular hemorrhage	12
Homonymous hemianopsia	9
Paresthesia	5

Adapted from refs. 29 and 65.

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intraocular hemorrhage (subhyaloid, vitreous, subretinal). Finally, systemic signs and symptoms that are often encountered in patients with SAH include elevated temperature, hypertension, chest pain, arrhythmias, abnormalities on electrocardiogram (ECG), and rarely cardiopulmonary arrest (29,65). In some patients, the time course of the elevated temperature has a close correlation with the onset of signs of delayed ischemia (66).

Prior to a definitive aneurysmal SAH, about half of patients experience warning symptoms, particularly headache, due to minor leakage of blood into the subarachnoid space or caused by aneurysmal expansion without actual subarachnoid hemorrhage (4,12,32,47,65). This so-called warning leak occurs days to several months before the major hemorrhage (47). In about two thirds of patients, the headache has associated signs and symptoms such as nausea and vomiting (20%), neck stiffness or pain (30%), visual disturbances such as blurred or decreased vision and visual defects (15%), and motor or sensory disturbances (15 to 20%) (47). Photophobia rarely occurs. The headache is most often characterized as unusual in severity and location, being similar to but less intense than that of the major bleed. It usually subsides over 1 or 2 days, but in some cases it is unremitting for as long as 2 weeks or until a subsequent major hemorrhage occurs (32). Overall, the site of the headache seems to be a poor localizing symptom for the aneurysm, with the exception of those on the PComA, in which the headache is often ipsilateral and retroorbital. The warning headache of aneurysmal origin is so unusual that 40 to 75% of patients seek medical advice (16,32). Unfortunately, the attacks of headache are too often misinterpreted as migraine, tension headache, the flu, or sinusitis, and patients are discharged without appropriate investigations (27,47). In a 5-year study from Denmark, 15% of over 1000 patients with SAH volunteered a history of sudden headache with neck pain, dizziness, vomiting, or drowsiness (23). Almost two thirds of the patients were misdiagnosed by physicians, and 43% were dead in 2 years. A study from the United Kingdom indicates that only the minority of patients (one third) with warning leaks are referred for appropriate care (61). Recently, the existence of minor "warning headache episodes" was challenged, arguing that may represent a recall bias (36).

Day and Raskin compared the abrupt, intense, and instantly peaking warning headache of cerebral aneurysms to a clap of thunder and called it thunderclap headache, even in the absence of SAH (12). Bleeding within the aneurysm wall or vasospasm was considered a potential cause of that warning headache pattern. A history of thunderclap headache should always prompt an investigation for cerebral aneurysm, although other conditions can present similarly (Table 109-2).

Thunderclap headaches can be classified into asymptomatic (with or without neurologic signs or symptoms) and symptomatic (of an intracranial disorder) varieties

TABLE 109-2 Some Conditions That Can Cause Thunderclap Headache

Condition	*References*
Aneurysmal SAH	29
Nonaneurysmal pretruncal SAH	37,53,71
Warning headache of unruptured cerebral aneurysms	3,12,16,23,25,32,46,47
Intracerebral hemorrhage	42
Pituitary apoplexy	67
"Crash migraine"	20
Sexual headaches	48
Cough and exertional headaches	48
Benign thunderclap headaches	16,37,70

SAH, subarachnoid hemorrhage.

(58), and many argue that they are benign when appropriate neuroinvestigational studies (CT scan with or without lumbar puncture [LP]) exclude any evidence for SAH (16,58,69).

Duffy studied 71 patients who were admitted to a local neurosurgical department with severe headache of sudden onset suggestive of SAH (16). All patients underwent cranial CT, angiography, and LP if SAH was not demonstrated. Of the 71 patients, 63 (89%) had a proven SAH, with a demonstrable aneurysm in 46 patients (73%). Eight patients (11%) had no evidence for SAH on either CT or LP and the angiogram did not reveal any aneurysms or other vascular malformations. None of these eight patients had recurrence of their symptoms during follow-up periods of at least 6 months (16). A more recent prospective study indicated that benign thunderclap headache is even more common: 65 of 102 patients (64%) had SAH with an aneurysmal demonstration in 42 and pretruncal nonaneurysmal SAH in 23; the rest had benign thunderclap headache (35).

HEADACHE AND UNRUPTURED VASCULAR MALFORMATION

The IHS diagnostic criteria for headache attributed to unruptured vascular malformation (ICHD-II) are as follows:

6.3.1 Headache attributed to saccular aneurysm:

- **A.** Any new headache including thunderclap headache and/or painful third nerve palsy fulfilling criteria C and D.
- **B.** Neuroimaging evidence of saccular aneurysm.
- **C.** Evidence exists of causation by the saccular aneurysm.
- **D.** Headache resolves within 72 hours.
- **E.** Subarachnoid hemorrhage, intracerebral hemorrhage, and other causes of headache ruled out by appropriate investigations.

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6.3.2 Headache attributed to AVM; 6.3.3 Headache attributed to dural arteriovenous fistula; 6.3.4 Headache attributed to cavernous angioma:

- **A.** Any new headache fulfilling criteria C and D.
- **B.** Neuroimaging evidence of AVM (6.3.2), dural arteriovenous fistula (6.3.3), cavernous angioma (6.3.4).
- **C.** Evidence exists of causation by AVM (6.3.2), fistula (6.3.3), cavernous angioma (6.3.4).
- **D.** Headache resolves within 72 hours (6.3.2).
- **E.** Subarachnoid hemorrhage, intracerebral hemorrhage, and other causes of headache ruled out by appropriate investigations (6.3.2) and (6.3.3) and (6.3.4).

Headache is reported in approximately 18% of patients with unruptured cerebral aneurysms (59); cerebral ischemic symptoms and seizures are less frequent. No specific clinical features of headaches are related to unruptured cerebral aneurysms. They could be focal or diffuse; unilateral or bilateral; frontal, occipital, or ocular; and acute in onset or gradual (3,51). Cranial nerve palsies from aneurysmal compression can accompany the headaches of unruptured cerebral aneurysms. Palsy of eye movement and facial pain mimicking trigeminal neuralgia can occur with aneurysms of the carotid–cavernous sinus (33). On the other hand, aneurysms of the posterior communicating artery cause cranial nerve III palsy, whereas those located on the anterior inferior cerebellar artery or the basilar artery can result in abducens nerve palsy (33,65).

Patients with unruptured AVMs can present with headaches that mimic migraine (43) or cluster headache (45). The argument whether these headaches are directly caused by the vascular malformation or are merely coincidental is still open. Headache is a frequent complaint of patients who harbor cavernous or venous angiomas, but the lesions are not believed to be pathophysiologically related (40,49). Epilepsy and then hemorrhage are the two most common clinical presentations of cavernous angiomas (40). Venous angiomas are largely asymptomatic lesions that are found on autopsy or during radiologic investigations of patients with various neurologic complaints, including headache (49).

DIAGNOSIS

SAH or the warning leak of a saccular aneurysm or AVM should be suspected in any patient who presents with a severe, unusual, and unremitting head or face pain of abrupt onset, particularly if hemicranial or hemifacial. The suspicion of SAH should be heightened when the headache is associated with vomiting, neck stiffness, or altered mentation. In those patients presenting with only sudden and

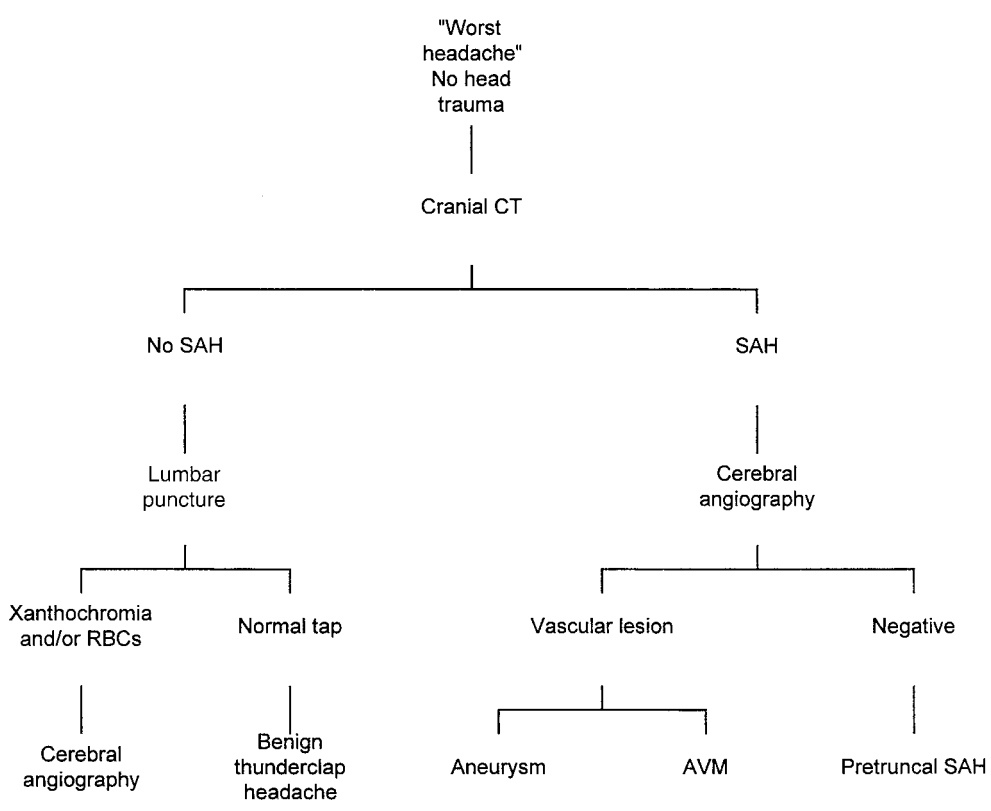

FIGURE 109-1. Diagnostic approach to patients with worst headache.

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severe headache, the probability of SAH is increased when the pain is associated with exertion or when the headache is preceded by transient focal symptoms, an episode of loss of consciousness, vomiting, or seizures (35).

Operational diagnostic procedures in patients with sudden onset of a severe and unusual headache ("the worst headache") aid in reaching a correct diagnosis (Fig. 109-1). Modern CT scanning is the first-line investigation for these patients because it excludes 97% of suspected SAH cases when performed within 12 hours of headache onset (44). However, the CT may fail to detect SAH resulting from a minor leak or if it is performed more than 24 hours following SAH. Consequently, an LP is recommended if SAH is still suspected. If xanthochromic staining of the CSF is seen or if there are RBCs on CSF microscopy after an atraumatic puncture, then cerebral angiography should be performed. If, on the other hand, the CT scan and the CSF are normal, the headache can be regarded as a benign symptom, and cerebral angiography is not indicated (15).

MRI is of less use in the acute stage of SAH (29) because leakage of blood into CSF causes only small changes in MRI signal characteristics. When the initial SAH is missed or if the diagnosis is delayed, an MRI may indicate evidence of prior bleeding (an abnormally high signal intensity due to conversion of oxyhemoglobin into methemoglobin) or could detect the aneurysm or the vascular malformation.

Dural arteriovenous fistula should be suspected in patients who present with headache and a pulsatile tinnitus. These lesions are best visualized on MR studies or during conventional cerebral angiography.

PROGNOSIS

Nearly one in five patients with SAH dies before reaching the hospital (6,29), and the prognosis of those who reach the hospital generally is dismal. Overall, approximately 50% die following an SAH, and 50% of the survivors are left more or less disabled (1,6,52,62). These morbidity and mortality data have not changed in recent decades despite recent advances in the management of complications of SAH (e.g., vasospasm). This is primarily because the diagnosis is still usually established only after a major hemorrhage. Among survivors of SAH, a disabling headache may occur in every fourth patient (52).

MANAGEMENT

Headache and restlessness should be controlled with appropriate analgesics and moderate sedation. Aspirin is relatively contradicted because it could increase the risk of bleeding by (a) enhancing the fibrinolytic activity of the blood, (b) increasing the bleeding time, and (c) impairing platelet aggregation. Instead, acetaminophen or codeine is recommended. Narcotics can be used judiciously. In many centers, nimodipine, a calcium channel antagonist, is administered intravenously and later orally for a total of 14 days to reduce the delayed cerebral ischemia of SAH (1,29).

Patients who are discovered to have multiple cerebral aneurysms or cavernous malformations should be counseled for possible familial clustering, and screening of direct family members with MRI may be advisable. Also, patients with familial connective tissue disease may benefit from screening MRI looking for asymptomatic cerebrovascular malformations.

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