ABSTRACT
Imaging for headaches is one of the most frequent and at times challenging decisions a neurologist has to make in the clinical practice of neurology and is complicated by today’s medical-legal milieu and overseeing regulatory organizations. Although the final decision should always be made by the treating physician, the American Academy of Neurology has developed practice parameters for the imaging of headaches. One strong criticism of the overzealous use of neuroimaging in patients with headache is the frequency of incidental findings, which may in turn increase a patient’s anxiety and potentially cause an exacerbation of symptoms. This may subsequently lead to unnecessary testing as well as neurosurgical intervention. The clinical interpretation of incidental findings and headaches is best made by a neurologist who is able to correlate the imaging findings with the patient’s symptoms and examination. One of the principal precepts in medicine, “primum nil nocere” or “first, do no harm” to patients, should be considered in the interpretation of these so-called abnormalities. The development of programs to certify competence in physicians who have completed accredited training programs in neuroimaging as well as headache medicine by the United Council for Neurologic Subspecialties is an important advancement that allows clinicians the opportunity to enhance quality of patient care.

INTRODUCTION
The most common complaint in the practice of neurology is headaches. The International Headache Society (IHS) classifies headaches (International Classification of Headache Disorders, 2nd Edition) into 14 major categories, subdivided into three broad groups: primary headache (categories 1 to 4); secondary headache (categories 5 to 12); and cranial neuralgias, central and primary facial pain, and other headaches (categories 13 and 14) (Headache Classification Subcommittee of the International Headache Society, 2004).

Although the paradigm in the diagnosis and treatment has shifted toward primary headaches, there continues to be a need for clinicians to rule out organic, or so-called secondary headaches. Therefore, a never-ending tension exists between the knowledge that primary headache disorders are far more frequent and the urge to order neuroimaging studies out of fear of missing an uncommon malady. Although a “therapeutic scan” would be greatly appreciated by patients and their families, it can also complicate and confuse the situation. Secondary headaches represent a symptom of
pathologic organic process and are associated with more than 316 disorders and illnesses. Although practice guidelines have been developed, these guidelines are not designed to supersede sound clinical judgment when treating individual patients. The cost-to-benefit ratio in performing a study is tempered by the medical and legal implications of omitting the study. As clinicians, neurologists need to understand the merits of neuroimaging and its effect in the management of patients with headache.

**NEUROIMAGING**

In 1994, the American Academy of Neurology (AAN) approved a guideline for the use of neuroimaging in patients with headaches and a normal neurologic examination. The consensus of the AAN was that “In adult patients with recurrent headaches that have been defined as migraine, including those with visual aura, with no recent change in pattern, no history of seizures, and no other focal neurological signs or symptoms, the routine use of neuroimaging is not warranted” (Quality Standards Subcommittee of the American Academy of Neurology, 1994). Since headaches account for approximately 4% of all outpatient visits, the final decision to image is left to the physician on a case-by-case basis.

In 2000, the United States Headache Consortium Report was unable to make any evidence-based recommendations with regard to the relative sensitivity of magnetic resonance imaging (MRI), compared with computerized tomography (CT) in evaluating migraine or other nonacute headaches (Silberstein, 2000). In the past, the advantage of CT was its cost; over the last several years, however, a gradual decrease has occurred in the reimbursement rate for MRI. Irrespective of cost, CT is the study of choice in acute trauma, acute subarachnoid hemorrhage (within 24 hours), and when MRI is contraindicated. In contrast, MRI is more sensitive than CT in detecting neoplastic disease, cervicomedullary lesions, pituitary disorders, and vascular diseases, including subdural hematomas, arteriovenous malformations, ischemic disease, venous infarctions, dissections, aneurysms, and subarachnoid hemorrhages (after 72 hours). Intracranial hypotension and hypertension are also better evaluated by MRI with and without gadolinium. MRI is also more sensitive in secondary headaches of pregnancy, which may be caused by cardiovascular disease, sinus thrombosis, pituitary apoplexy, disseminated intravascular coagulation, or emboli, keeping in mind that contrast should be avoided. The use of CT in adults and children has increased sevenfold in the past 10 years. CT scans contribute approximately 65% of effective radiation dose from all medical x-ray examinations. Radiation exposure is a special concern in children, who are considerably more sensitive to radiation than adults and have a longer life expectancy, resulting in a potentially higher likelihood to develop cancers. Clinicians must continually think about reducing exposure as much as reasonably achievable by (1) reducing radiation dose, (2) decreasing serial studies, and (3) utilizing other imaging modalities, such as MRI and ultrasonography (Brenner and Hall, 2007).

Since the first commercial MRI scanner was manufactured in 1980, MRI technology has continued to advance. Resolution and sensitivity in MRI have improved because of larger magnet size, availability of paramagnetic contrast, and the selection of acquisition sequences for specific pathologic indications. Newer vascular imaging packages, such as MR angiography (MRA) and MR venography (MRV), have also improved the sensitivity of MRI.

MRA is not routinely done unless there is a history of thunderclap head-
ache, a family history of aneurysms, or headaches that are continuously ipsilateral or progressive in nature. In these cases, a time-of-flight study MRA would be appropriate. In circumstances in which the history may include head trauma, thunderclap headache, or family history of vascular malformations or aneurysms, a gradient-echo sequence, which is sensitive for hemosiderin and calcification, should also be done.

Gadolinium-containing contrast agents are often used in MRI to enhance the quality of images. Because gadolinium is renally excreted, care must be taken to screen patients for renal disease or dysfunction. While the use of low-dose gadolinium-based contrast agents (0.1 mmol/kg body weight) in patients with impaired renal function has been shown to be non-nephrotoxic, doses greater than 0.2 mmol/kg have been associated with a rare disorder called nephrogenic systemic fibrosis (NSF) in patients with advanced kidney failure. Factors associated with NSF include dialysis, moderate/end-stage renal disease (glomerular filtration rate less than 30 mL/min/1.73m²), and repeated or higher-than-recommended doses of gadolinium contrast. NSF is a debilitating and sometimes fatal disease affecting the skin, muscle, and internal organs (Broome et al, 2007). Gadolinium is routinely used in patients with headache consistent with the criteria in Table 5-1. It is important for routine imaging protocols to be established and applied to ensure the quality and consistency of each image. A sample routine protocol for patients with headache that is currently instituted at the Dent Neurologic Institute is outlined in Table 5-2.

Neuroimaging ordered by a neurologist or headache specialist will often have a higher diagnostic yield than images ordered in the primary care setting. Sempere and colleagues (2005) reported a study in 1876 consecutive patients with nonacute headaches referred to a neurology clinic. Neuroimaging detected significant lesions in 1.2% of patients. Wang and colleagues (2001) retrospectively reviewed 402 adult patients who had been evaluated for chronic headaches

**KEY POINTS:**
- Gradient-echo images are sensitive to blood flow, calcification, and hemorrhage.
- High doses of gadolinium-based contrast agents have been associated with a rare disorder called nephrogenic systemic fibrosis in patients with advanced kidney failure.
- Nephrogenic systemic fibrosis is a rare and potentially fatal syndrome that involves fibrosis of the skin, joints, muscle, and internal organs.
- Neuroimaging detected significant lesions in 1.2% of patients with nonacute headaches.

**TABLE 5-1** Recommended Routine Use of Gadolinium in Patients With Headache

- Patients with abnormal neurologic examination
- Patients with positional headaches
- Patients with exertional or Valsalva maneuver–exacerbated headaches
- Patients with cluster or neuralgia-type headaches or facial pain
- Patients with known history of cancer, AIDS, or infectious disease

**TABLE 5-2** Standard Protocol for Patients With Headache at the Dent Neurologic Institute

- T1-weighted axial 5 mm
- T2-weighted axial fast spin echo 5 mm
- Fluid-attenuated inversion recovery (FLAIR) axial 5 mm
- Diffusion-weighted imaging axial
- FLAIR and/or T1-weighted sagittal
- T2-weighted coronal 3 mm through circle of Willis
(duration of 3 months or greater) and had no other neurologic symptoms or findings. Major abnormalities that were felt to be the actual cause of the headache were found in 15 patients (3.7%). Clinical suspicion is increased when certain “red flags” (warning signs) are present as outlined in Table 5-3.

Neuroimaging in Pediatric Headaches
Mazzotta and colleagues (2004) performed a prospective study at multiple pediatric headache centers and found that 9.3% of the 1485 patients had incidental findings. Findings that led to the diagnosis of secondary headache were observed in 9.1% of total patients, including sinusitis in 57.0% and intracranial mass in 17.4%. The report of the Quality Standards Subcommittee of the American Academy of Neurology and the Practice Committee of the Child Neurology Society (Lewis et al, 2002) made the following recommendations:

1. Obtaining a neuroimaging study on a routine basis is not indicated in children with

<table>
<thead>
<tr>
<th>TABLE 5-3 “Red Flags” in Adult Patients With Headache</th>
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<tbody>
<tr>
<td>The “first or worst” headache (“thunderclap”)</td>
</tr>
<tr>
<td>Subacute headaches with increasing frequency and severity</td>
</tr>
<tr>
<td>Progressive or new daily persistent headache</td>
</tr>
<tr>
<td>Chronic daily headache</td>
</tr>
<tr>
<td>Headaches always on the same side</td>
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<tr>
<td>Headaches resistant to treatment</td>
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<tr>
<td>New onset of headaches in high-risk population</td>
</tr>
<tr>
<td>- Patients with cancer</td>
</tr>
<tr>
<td>- Patients who are HIV positive</td>
</tr>
<tr>
<td>- Patients with dementia</td>
</tr>
<tr>
<td>- Patients who are taking an anticoagulant</td>
</tr>
<tr>
<td>- Patients with neurocutaneous syndrome</td>
</tr>
<tr>
<td>New onset of headaches after age 50</td>
</tr>
<tr>
<td>Patients with headaches and seizures</td>
</tr>
<tr>
<td>Headaches associated with symptoms such as fever, neck stiffness, nausea, and vomiting</td>
</tr>
<tr>
<td>Headaches with focal neurologic deficits not meeting the International Headache Society criteria of migraine with aura</td>
</tr>
<tr>
<td>Headaches associated with papilledema, cognitive impairment, or personality change</td>
</tr>
<tr>
<td>Headaches precipitated by exertion, Valsalva maneuver, or positional changes</td>
</tr>
<tr>
<td>Atypical cranial neuralgias poorly responsive to treatment</td>
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</tbody>
</table>

KEY POINT:
- In children with a family history of neurocutaneous syndrome (i.e., neurofibromatosis), a headache that is poorly responsive to treatment is a “red flag” and may warrant neuroimaging.

Neuroimaging should be considered in children with an abnormal neurologic examination (e.g., focal findings, signs of increased intracranial pressure, significant alteration of consciousness), the coexistence of seizures, or both.

Neuroimaging should be considered in children in whom there are historical features to suggest the recent onset of severe headache, change in the type of headache, or if there are associated features that suggest neurologic dysfunction.

Just as in the adult population, clinical suspicion is increased when certain red flags are present as outlined in Table 5-4 (Figure 5-1).

Headaches in Older Adults

Secondary headaches in patients over the age of 50 present a challenge to the clinician. Only 2% of migraineurs have their first headache over the age of 50. In addition, as the incidence of primary headaches declines with age, the occurrence of secondary headaches increases with advancing age. Although neoplasms may cause headaches in all age groups, they are far more prevalent in patients over the age of 50. The common causes of headaches beginning late in life include primary and secondary neoplasm, temporal arteritis, drug-induced headaches, trigeminal neuralgia, cervicogenic cervical myofascial pain, Parkinson disease, cerebrovascular disease, postherpetic neuralgia, and associated systemic diseases (Mechtler and Stiles, 2005) (Figure 5-2).

Neuroimaging of Peripartum Headaches

Headaches are one of the most common symptoms that occur in the postpartum period and are most frequent on days 3 to 6 postpartum. Postpartum women often have sleep deprivation along with irregular food intake and dehydration. The most common types of postpartum headaches include tension-type headaches, preeclampsia/eclampsia, spinal headache, and migraine. Secondary causes include cerebral venous thrombosis, cerebral vasculopathy, subarachnoid hemorrhage, or intraparenchymal hemorrhage. The use of MRI during pregnancy is considered safe, as mag-

<table>
<thead>
<tr>
<th>TABLE 5-4 “Red Flags” in Children</th>
</tr>
</thead>
<tbody>
<tr>
<td>Persistent headaches of less than 6-months’ duration that do not respond to medical treatment</td>
</tr>
<tr>
<td>Headache associated with abnormal neurologic findings, especially if accompanied by papilledema, nystagmus, or gait or motor abnormalities</td>
</tr>
<tr>
<td>Persistent headaches associated with a negative family history of migraine</td>
</tr>
<tr>
<td>Persistent headaches associated with substantial episodes of confusion, disorientation, or emesis</td>
</tr>
<tr>
<td>Headaches that repeatedly awaken a child from sleep or occur immediately on awakening</td>
</tr>
<tr>
<td>Family and medical history of disorders that predispose to CNS lesions and clinical laboratory findings that suggest CNS involvement</td>
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</tbody>
</table>

FIGURE 5-1  A 15-year-old girl presented with positional headaches and normal examination. A, FLAIR sagittal imaging confirms a “tight” posterior fossa, tonsillar herniation, and cervical syrinx. B, T2-weighted and (C) T1-weighted gadolinium sagittal sequences rule out intramedullary mass. The diagnosis was Arnold-Chiari I malformation.

FIGURE 5-2  A 60-year-old man with a chronic frontal headache and growing “bump” on his forehead. A, T1-weighted contrast images show a large enhancing frontal mass that erodes the calvarium and uplifts the scalp. B, Magnetic resonance venography confirms infiltration and occlusion of the superior sagittal sinus. Headaches resolved after resection of a meningioma.
The most common abnormalities found on MRI in migraineurs are white matter lesions localized in the subcortical or periventricular white matter. These white matter lesions have been variably reported in 12% to 48% of migraineurs compared with 2% to 11% of control subjects (Evans et al, 2007). These multiple, small punctate hyperintensities are seen best on fluid-attenuated inversion recovery (FLAIR) images. Several neuroimaging series have suggested an increase in prevalence of T2-weighted foci of hyperintensities in patients who have migraines with aura. Kruit and colleagues (2004) evaluated 295 patients with proton density and FLAIR images in a population of migraineurs compared with well-matched controls (140 patients). The results show that infarcts were seen most often in the posterior circulation of patients who experienced migraines with aura and when migraine attacks occurred more often than once a month. Deep white matter lesions were also more common in female migraineurs and in those whose attacks occurred more than once a month, but no differences were seen between the migraineurs with and without aura. Meta-analysis from seven case control studies determined that patients with migraines had a 3.9 times greater risk for white matter lesions compared with controls (Swartz and Kern, 2004). Recent findings have shown an increase in prevalence of infratentorial, mostly pontine, T2 hyperintensities in migraineurs compared with the general population (Kruit et al, 2006).

The introduction of 3-tesla MR systems into the clinical arena has improved the sensitivity for detecting CNS changes, thanks to an increased signal-noise ratio and spatial resolution. Voxel-based morphometry is a fully automated whole-brain technique that is being used as an unbiased method for the assessment of differences in brain volume. Migraineurs with T2 white matter lesions had significant reduction of gray matter density when compared with healthy controls. Such changes were located in the cortex of the frontotemporal lobe and in the cingulum (Rocca et al, 2006). Three-tesla imaging has also shown abnormalities in iron hemostasis within the periaqueductal gray matter. Iron deposits were increased within the periaqueductal gray matter and progressed with the severity of the headache (Welch et al, 2001).

An increased prevalence of patent foramen ovale (PFO) was found in patients with migraine with aura but not in those without aura. PFOs were found in approximately 20% to 25% of the general population and about 50% of patients who had migraine with aura. A moderate- or large-sized shunt was found more often in the migraineurs than in the controls. Shunting may occur at rest or with elevation in the right atrial pressure during Valsalva maneuver. PFO may be detected by transesophageal echocardiogram, transthoracic echocardiogram, or transcranial Doppler of the middle cerebral artery with bubble injection of agitated saline (Spencer et al, 2004). Cardiac

**KEY POINTS:**
- There is no known risk to performing an MRI during pregnancy, but gadolinium contrast is not recommended.
- FLAIR imaging is the most sensitive sequence for the evaluation of migraine-related white matter lesions.
- Transcranial Doppler with agitated saline bubble is a sensitive test for the detection of patent foramen ovale, which occurs in up to 50% of patients who have migraines with aura.

**MRI IN MIGRAINES**

The most common abnormalities found on MRI in migraineurs are white matter lesions localized in the subcortical or periventricular white matter. These white matter lesions have been variably reported in 12% to 48% of migraineurs compared with 2% to 11% of control subjects (Evans et al, 2007). These multiple, small punctate hyperintensities are seen best on fluid-attenuated inversion recovery (FLAIR) images. Several neuroimaging series have suggested an increase in prevalence of T2-weighted foci of hyperintensities in patients who have migraines with aura. Kruit and colleagues (2004) evaluated 295 patients with proton density and FLAIR images in a population of migraineurs compared with well-matched controls (140 patients). The results show that infarcts were seen most often in the posterior circulation of patients who experienced migraines with aura and when migraine attacks occurred more often than once a month. Deep white matter lesions were also more common in female migraineurs and in those whose attacks occurred more than once a month, but no differences were seen between the migraineurs with and without aura. Meta-analysis from seven case control studies determined that patients with migraines had a 3.9 times greater risk for white matter lesions compared with controls (Swartz and Kern, 2004). Recent findings have shown an increase in prevalence of infratentorial, mostly pontine, T2 hyperintensities in migraineurs compared with the general population (Kruit et al, 2006).
MRIs are also being utilized more often for the evaluation of PFOs (Horton and Bunch, 2004; Mechtler, 2004).

**MELAS and CADASIL**

MELAS is the term used for the syndrome clinically characterized by mitochondrial myopathy, encephalopathy, lactic acidosis, and stroke-like episodes. MRI findings include abnormal signal intensity in the basal ganglia and infarctions. In patients with this disorder, magnetic resonance spectroscopy (MRS) shows an elevation of lactate and may be more sensitive than MRI in the detection of MELAS-associated abnormalities. Clinically, MELAS begins with migraine, then progresses to hemiplegic migraine and eventually to stroke. Approximately 90% of the patients are symptomatic by age 40, and many of these “stroke-like” events cross usual vascular territories. MRS may potentially be used as a screening device for mitochondrial cytopathy and as a tool to follow treatment response. MRS does show a lactate doublet peak at 1.3 ppm in 65% of cases, and changes may precede diffusion-weighted image (DWI) abnormalities.

On routine MRI, these acute stroke-like cortical lesions have a so-called shifting spread of appearance, disappearance, and reappearance elsewhere. The lesions cross typical vascular territories, and the lactate level in the CSF is elevated (Kaufmann et al, 2004).

CADASIL (cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy) is seen in young adults and presents with migraines with and without aura, mood disturbance, focal neurologic deficits, strokes, and dementia. Most patients will show symptoms by the age of 60, usually beginning with a migraine and a prolonged aura. CADASIL is caused by a NOTCH3 mutation on chromosome 19 that can be autosomal dominant or sporadic. With dynamic contrast-enhanced MRI, a significant reduction of cerebral blood volume within subcortical T2 white matter lesions is found, but not in normal-appearing white matter. Typical white matter changes in CADASIL involve the anterior temporal poles and the external capsule. There are two types of lesions: (1) large coalescent lesions in the white matter, isointense on T1-weighted imaging (T1WI) and bright on T2-weighted imaging (T2WI); and (2) small well-delineated lacunaelike lesions that spare the cortex and are hypointense on T1WI and hyperintense on T2WI. CADASIL is underrecognized and underdiagnosed and should be considered in patients with the following history:

- One or more recurrent subcortical ischemic stroke, especially before the age of 60 and in the absence of vascular risk factors
- Migraines with aura, especially with atypical and prolonged auras
- Early cognitive decline with subcortical dementia (Markus et al, 2002)

**INCIDENTAL FINDINGS ON MRI**

Incidental findings are previously undetected abnormalities with potential clinical relevance that are unexpectedly discovered and unrelated to the purpose of the examination. One of the difficulties with ordering MRIs in migraineurs or patients with headache is the relatively high frequency of incidental findings. Depending on the experience of the physician ordering the tests, these findings may be misinterpreted as the cause of the headache. Some of the more common incidental findings seen on MRI are outlined in Table 5-5. These findings are often categorized based on their classification as a normal anatomic variant, parenchymal lesion, or cystic lesion.

Headaches are rarely, if ever, associated with the above-noted findings. Exceptions are enlarged pineal cysts or...
Arachnoid cysts that are associated with mass effect or obstructive hydrocephalus or when developmental venous anomalies or telangiectasias are associated with other arteriovenous malformations or hemorrhage. Telangiectasias, or developmental venous anomalies, when associated with other vascular malformations (ie, cavernous hemangioma) can rarely cause hemorrhage. In general, far too much emphasis is placed on these incidental findings in regard to the pathophysiology of the patient’s pain syndrome and furthermore may cause unwarranted patient anxiety as well as further diagnostic testing, which may be unnecessary and potentially harmful (Figure 5-3).

**Normal Anatomic Variants**

Variations of CSF-containing spaces are commonly seen as incidental findings. These include ventricular asymmetry, cavum septi pellucidi, cavum vergae, and cavum veli interpositi.

Ventricular asymmetry of the frontal horns and bodies of the lateral ventricles is common, with the left generally larger than the right. With lateral ventricular asymmetry, close inspection of the foramina of Monro is needed to rule out masses in this region, such as colloid cysts, giant cell astrocytomas, or intraventricular tumors.

Present in 2% to 3% of normal adults, *cavum septi pellucidi* is a variable-sized CSF collection between the two layers of the septum pellucidum, anterior to the foramina of Monro. It has been reported more often in closed-head injuries as well as in schizophrenics but has not been reported with an increase in frequency among patients with headache. Portions of cavum septum pellucidi extending posteriorly to the columns of the fornix are known as the *cavum vergae*. *Cavum veli interpositum* is a triangle-shaped CSF space between the bodies of the lateral ventricles, below the fornices, and above the third ventricle (Epelman et al, 2006).

*Arachnoid granulations* are small protrusions of the arachnoid through the dura. They extend to the venous sinuses of the brain, allowing CSF to exit the subarachnoid space and enter the bloodstream. These well-defined filling defects are seen most often around the transverse and sigmoid sinuses. Arachnoid granulations have also been called pacchionian bodies. On high-resolution MRIs utilizing thin slices, up to 70% of patients were found to have arachnoid granulations.
DWIs showed isointensity to normal brain tissue, which is higher than the reported signal intensity of arachnoid cysts and lower than that of epidermoids (Kan et al, 2006).

A large cisterna magna, also called mega cisterna magna, is the space between the medulla oblongata and the inferior surface of the cerebellum. Mega cisterna magna is a cystlike enlargement of the cisterna magna seen in 1% to 3% of the population. Mega cisterna magna communicates with the subarachnoid space and the fourth ventricle.

An enlarged Meckel cave is located just inferolateral to the cavernous sinus and is hyperintense on T2WIs. It contains the gasserian ganglion. Occasionally, Meckel caves may be quite enlarged but are usually symmetric. The enhancement pattern is never homogenous, but circumferential.

**Parenchymal**

*Developmental venous anomalies* (DVAs), also called venous angiomas, are congenital cerebrovascular malformations with angiogenically mature venous elements. The veins that compromise a DVA usually form a little cluster called a *caput medusae*. They are most often seen adjacent to the frontal horn of the lateral ventricle or within the cerebellum. There is no arterial component to these malformations. The incidence of DVAs has been reported to be 2.6%. DVAs are poorly visualized on T1WIs, and the study of choice is a T1-weighted contrast sequence. Large DVAs can possibly present as a flow void on T2WIs. Fifteen percent to 20% of DVAs occur with a coexisting cavernous malformation and have also been associated with telangiectasias. Although there have been case studies of patients
who have had seizures or brain hemorrhages, DVAs do not cause any symptoms and are felt to remain dormant or silent throughout life. Most symptomatic patients have associated cavernous malformations, which are best observed on gradient-echo images. In a patient diagnosed with a headache or migraine, a DVA is a probable incidental finding unless there is evidence of an associated cavernous malformation or blood products.

Capillary telangiectasias have been recognized with increasing frequency on MRI studies. Most likely localized in the pons, capillary telangiectasias may be hyperintense on T2WIs and hypointense on gradient-echo imaging. They are not seen on noncontrast TIWI, but are best visualized with gadolinium-contrast TIWI studies. There is no mass effect, and they are generally asymptomatic. About two-thirds of capillary telangiectasias reveal an enlarged draining vessel. The overwhelming majority of capillary telangiectasias are clinically silent, but when associated with hemorrhage, they are usually also associated with other vascular malformations (Castillo et al, 2001) (Figure 5-4).

Intracranial lipomas are malformations believed to result from abnormal differentiation of the meninx primitiva, the undifferentiated mesenchyma that surrounds the developing brain. CNS lipomas are congenital malformations, not true neoplasms. These are predominantly located in the subarachnoid space: 80% are in the midline, 80% are supratentorial (of which 40% to 50% are in the interhemispheric fissure over the corpus callosum), 15%

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**KEY POINT:**
- Gradient echo may show a hypointense blooming artifact in developmental venous anomalies if a coexisting cavernous malformation is present.

**FIGURE 5-4**

A 35-year-old patient with lifelong history of episodic severe headaches consistent with International Headache Society criteria for migraines without aura. MRI shows evidence of an enhancing lesion within the pons consistent with a capillary telangiectasia. C, D, The incidental finding was not seen on T1-weighted imaging noncontrast or T2-weighted imaging sequences, except for subtle changes on FLAIR. A, B. Patient responded well to pharmacotherapy.
to 20% are suprasellar, 10% to 15% are infratentorial. Blood vessels and cranial nerves almost always course through them; therefore, surgical treatment has a high morbidity and is rarely indicated. Interhemispheric lipomas are almost always associated with hypogenesis or agenesis of the corpus callosum. On CT, lipomas are sharply demarcated areas of hypodensity. The MR appearance of a lipoma is that of a hyperintense mass on T1-weighted sequences and hypointense on long repetition time (T2WI). Large lipomas will reveal chemical shift artifact resulting from the different chemical shifts of water and fat. Chemical shift artifact results in the appearance of a dark rim at one edge of an object (eg, lipoma) and a bright rim at the opposite edge. Application of a fat-saturation pulse (short tau inversion recovery) will make the lipoma hypointense to gray matter, confirming the diagnosis and distinguishing it from a hemorrhage (Bakshi et al, 1999).

Large perivascular spaces, or Virchow-Robin spaces, are fluid-filled spaces that appear to contain CSF and surround penetrating arteries predominantly in the basal ganglia. Other common locations are midbrain, deep white matter, and the subinsular cortex. They can be differentiated from lacunar infarcts by their typical location, CSF signal in all pulse sequences, and lack of surrounding gliosis (FLAIR hyperintensity). Numerous prominent Virchow-Robin spaces in the basal ganglia are called état criblé.

Ependymitis granularis is characterized by small foci of increased T2-signal intensity and is commonly seen along the anterolateral aspect of the frontal horns. This represents a focal pooling of interstitial fluid. Ependymitis granularis should not be mistaken for demyelinating plaque, microangiopathy, or transependymal CSF flow. **Cysts**

*Pineal cysts* on high-resolution MRI are seen in 23% of the normal population. Although the prevalence of pineal cyst in autopsy series has been reported to be between 25% and 40%, pineal cysts are non-neoplastic, intrapineal, homogeneous fluid-filled cavities clearly distinct from the tectum. Sixty percent of pineal cysts are slightly hyperintense to CSF on T1WI, while on FLAIR they are mildly hyperintense. A T1WI sagittal image will reveal a lack of mass effect on the adjacent structures, such as the tectum and corpus callosum, and 60% will enhance either completely or partially. Most pineal cysts are clinically silent, discovered incidentally, and have a gender ratio of 3:1 favoring females. Benign pineal cysts are usually less than 3 cm in size, homogenous, and cause no perilesional edema or hydrocephalus. When not associated with mass effect, pineal cysts rarely cause headaches (Pu et al, 2007).

*Arachnoid cysts* are the most common congenital cystic abnormalities in the brain that do not communicate with the ventricular system. Fifty percent to 60% are found in the middle cranial fossa, while other locations include the cerebellopontine angle (10%), suprasellar (10%), and miscellaneous (convexity, quadrigeminal). They constitute approximately 1% of all intracranial masses. Even if they are large, arachnoid cysts are usually asymptomatic and have a male-to-female ratio of 4:1. It is uncommon for middle fossa arachnoid cysts to be associated with headaches. An MRI will typically show a sharply demarcated round/ovoid extraaxial cyst that follows CSF signal. Diffusion-weighted imaging is hypointense, while in epidermoids it is hyperintense.

**KEY POINTS:**
- Chemical shift artifact and fat-suppression pulse sequence help differentiate a lipoma from a hemorrhage.
- Short tau inversion recovery is used to suppress fat signal.
- An arachnoid cyst is a well-demarcated, round/ovoid, extraaxial cyst that follows CSF signal. Diffusion-weighted imaging is hypointense, while in epidermoids it is hyperintense.
A choroid plexus cyst is the most common cyst seen intracranially in adults. Most cases are hyperintense on T1WI as well as T2WI. A rim or nodular enhancement with contrast is present. A high signal or restricted diffusion is seen on DWI in 65% of patients. Choroid plexus cysts are prevalent in approximately 40% of older patients, and they are usually bilateral, rarely causing symptoms.

Choroidal fissure cysts are relatively common incidental findings. They are typically ovoid, 5 mm to 30 mm in diameter, lateral to the cisterna ambiens, and superior medial to the hippocampus within the choroidal fissure. They follow a CSF signal in all
pulse sequences and are asymptomatic (Osborn and Preece, 2006).

A Tornwaldt cyst is a benign developmental lesion located on the posterior wall of the nasopharynx on the midline, with an incidence of 0.06%. The content of the cyst is generally high in protein and anaerobic bacteria. As a result, Tornwaldt cysts appear to be hyperintense on T1WI and T2WI and do not enhance. Patients with these cysts are generally asymptomatic but rarely develop halitosis (Moody et al, 2007).

The detection of incidental findings poses various practical and ethical issues. Performing MRIs at high-field strength and with more sensitive sequences may lead to the detection of subtle brain abnormalities that would not have been detected previously. Medically significant findings in asymptomatic patients undergoing imaging occur in 1% to 2% of cases. A recent prospective, population-based cohort study of 2000 patients concluded that asymptomatic brain infarcts were present in 7.2% of patients, cerebral aneurysms in 1.8%, and benign primary tumors in 1.6%. White matter lesions were found in all but 5.4% of participants aged 45 to 59 and in all but 2% of those aged 75 and older (Vernooij et al, 2007).

Incidental findings in a healthy pediatric population were detected in 21% of patients (Kim, 2002). Of these, 8% of the total subject population required either routine or urgent clinical referral. The most common nonreferable finding was chronic sinusitis. In patients who required referral, findings included acute sinusitis, focal white matter lesions of uncertain etiology, and tonsillar ectopia. Of the children who presented to the emergency department with headaches, 77% had a secondary cause. This is significantly higher compared with adults who had a 16% to 20% incidence of secondary headaches on presentation. Of these children, 45% suffered from secondary headaches with neurologic causes.

In general, patients with headache, especially migraineurs, have a relatively high incidence of anxiety. For children and adults, the presence of comorbid anxiety or depression was associated with significantly higher medical costs when compared with migraine alone (Pesa and Lage, 2004). An incidental finding can place added psychological burden on these patients, which would unfortunately magnify their symptoms. Most incidental findings do not cause headaches; still correlation between headache and incidental findings should be made by a neurologist or neurosurgeon. Finally, it is important to convey to the patient that only a clinical expert can label neuroimaging findings as “symptom related,” “incidental, but without consequences,” or “incidental with consequences.”

**RHINOSINUSITIS**

Rhinosinusitis consists of a group of disorders characterized by inflammation of the nasal mucosa and the paranasal sinuses. The IHS criterion for acute frontal sinus headache is pain located directly over the sinus that can radiate to the vertex or behind the eyes. An acute maxillary sinus headache is located over the antral area and can radiate to the upper teeth and the forehead. Acute ethmoiditis is a headache that is located between and behind the eyes and can radiate to the temporal area. Acute sphenoiditis headache is located in the occipital area, the vertex, the frontal region, or behind the eyes (Silberstein, 2004). Imaging characteristics of acute sinusitis include air-fluid levels, partial or complete opacification of a sinus, and mucosal membrane thickening of at least 4 mm. Findings suggestive of chronic sinusitis include mucosal thickening, bony remodeling, polyposis, and mucous retention cysts. Incidental sinus inflammation is present on MRI in more than 32% of asymptomatic patients. Polyp and retention cysts are a rare cause of headaches (Cady et al, 2005).
Imaging findings of mucosal thickening fluctuate with the normal nasal cycle. There is a cyclical passive congestion and decongestion of each side of the nasal turbinates, nasal septum, and ethmoid air cells mucosa that rotates from side to side over the course of 1 to 8 hours. Chronic sinusitis is not validated as a cause of headaches or facial pain unless it relapses into the acute stage. Although most headache specialists agree that chronic recurrent headaches are rarely caused by sinus disease, the exception to the rule would be sphenoid sinusitis. This is an uncommon infection and accounts for approximately 3% of all cases of acute sinusitis. Typically, this headache is aggravated by standing, walking, bending, or coughing and is the most common symptom of acute sphenoid sinusitis.

IDIOPATHIC INTRACRANIAL HYPERTENSION

Idiopathic intracranial hypertension (IIH), also known as pseudotumor cerebri, is characterized by elevated CSF pressure without ventriculomegaly. The condition occurs most commonly in obese women of childbearing age. The incidence may be at least 19 per 100,000 in women between the ages of 20 and 44 who are at least 20% above their ideal body weight. Diagnostic criteria of IIH include: (1) increased intracranial pressure greater than or equal to 250 mm H₂O; (2) symptoms or signs of increased intracranial pressure without any localizing features; no mass or lesion; no evidence of hydrocephalus, structural or vascular lesion; normal CSF contents; and (3) no clinical or neuroimaging suspicion of venous sinus thrombosis. Symptoms most commonly reported in a case-control study of outpatients with IIH include headaches (94%), transient visual obscuration (68%), pulse-synchronous tinnitus (58%), photopsia (54%), and retrobulbar pain (44%) (Friedman, 2004). The headache profile is consistent with a severe daily pulsatile headache that gradually increases in intensity. There may be associated nausea but rarely vomiting. Papilledema is the hallmark sign of IIH. Horizontal diplopia occurs in approximately one of three patients with IIH, and sixth nerve palsies are present in 10% to 20% of patients.

On the TIWI, a partially empty sella turcica is seen quite often. In general, partial empty sellas are evident in 30% of the normal population. Empty sellas without headaches is most likely an incidental finding (Binder et al, 2004). Enlarged tortuous optic nerve sheaths and posterior sclerae flattening, as well as small venous sinuses and veins, may also be seen in IIH. After appropriate treatment, which may include weight loss, carbonic anhydrase inhibitors, serial lumbar punctures, or lumbar puncture shunt/optic nerve sheath fenestration, normalization of the size of the cerebral sinuses and veins may occur, as well as reversal of the partial empty sella. In this way, imaging can be used to follow the patient’s response to the treatment.

SPONTANEOUS INTRACRANIAL HYPOTENSION

Intracranial hypotension is a frequently misdiagnosed syndrome of headache caused by reduced intracranial CSF pressure. CSF volume depletion may occur after trauma, after craniotomy or lumbar puncture, and after trivial trauma. Intracranial hypotension also may be spontaneous, especially in patients with connective-tissue disorders such as Marfan syndrome. The typical clinical profile is a young or middle-aged adult with orthostatic headaches. MRI abnormalities most often seen on sagittal and axial TIWI include brain descent in 40% to 50% of cases, caudal displacement of tonsils in 25% to 75% of cases, diffuse pachymeningeal dural enhancement in 85% of cases, and bilateral subdural fluid collections in 15% of cases, of which the majority are

KEY POINTS:
- Sinus retention cysts and/or polyps are a rare cause of chronic headaches unless they involve the sphenoid sinus.
- Empty or partially empty sella is nearly always an incidental and asymptomatic finding, but it may be associated with idiopathic intracranial hypertension.
- Imaging characteristics of idiopathic intracranial hypertension include empty sella, enlarged tortuous optic nerve sheaths, posterior sclerae flattening, and small venous sinuses.

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hygromas. Other cerebral findings include crowding of the posterior fossa, engorgement of the cerebral venous sinuses, and obliteration of some of the subarachnoid cisterns (prepontine or peri-chiasmatic cisterns) (Farb et al, 2007). Within the spine, evidence of extra-arachnoid fluid collections, meningeal diverticula, spinal pachymeningeal enhancement, and engorgement of the spinal epidural venous plexus can be seen. At times, identification of the actual leak may be seen (Schievink, 2006). In patients with chronic positional headaches, a T1-weighted contrast study is mandatory if one wants to diagnose spontaneous intracranial hypotension syndrome (Mokri, 2004) (Table 5-6) (Figure 5-6).

CEREBRAL VENOUS THROMBOSIS

Headaches are a prominent symptom in 75% to 95% of patients with cerebral venous thrombosis (CVT) (Cumurciuc et al, 2005). The headaches of CVT are persistent and can be worsened by a Valsalva maneuver or recumbency. Local causes of CVT include mastoiditis, sinusitis, trauma, or neurosurgical procedures. More systemic causes include septicemia, herpes, HIV, cancer, prothrombotic conditions (factor V Leiden mutation), protein S deficiency, pregnancy, use of oral contraceptives, and vasculitis. Other symptoms and signs of CVT include seizures, papilledema, altered consciousness, and focal neurologic symptoms. Only 15% to 30% of the patients experience headaches alone (Leach et al, 2006).

MRI and MRV are recommended for the appropriate evaluation of patients with suspected CVT. Transverse sinus hypoplasia and atresia are normal variants. Asymmetric transverse sinuses are seen in up to 50% of patients, with partial or complete absence of one transverse sinus in 20% of the cases. In most cases, the right sinus is larger than the left (Alper et al, 2004). On T1WI, acute thrombus is isointense while subacute thrombus becomes hyperintense. On T2WI, initially the thrombus is hypointense and then becomes hyperintense when subacute. The T2 gradient-
KEY POINT:
- “Location, location, location” is the best clue to locating colloid cysts; 99% are wedged into the foramen of Monro (anterior superior third ventricle).

echo image shows that the thrombus is hypointense but “blooms” (Idliah et al, 2006). In 40% of cases, a DWI shows a hyperintense clot and an occluded vessel. With contrast, a peripheral enhancement around the acute clot is usually present, similar to the “empty delta sign” on contrast-enhanced CT. On MRV, there is an absence of flow and an occluded sinus with abnormally enlarged collateral channels (Figure 5-7).

COLLOID CYST
Third ventricular colloid cysts may be a cause not only of cough and exertional headaches, but also of thunderclap headaches. Headaches are the most common symptom of a third ventricular colloid cyst and are reported in 68% to 100% of diagnosed patients. The headache is generally located in the bilateral frontoparietal or frontooccipital region. Pain is typically quite severe and relieved by recumbency. Loss of consciousness, change in cognition, seizures, coma, and death may occur. The mean size of colloid cysts is 15 mm, and 99% wedge near the foramen of Monro, attached to the anterior, superior, third ventricular roof. Two-thirds are hyperintense on T1WI, while the majority are isointense to the brain on T2WI. DWI does not restrict, and on FLAIR it is hyperintense. MR is also predictive of the ease with which the colloid cyst can be aspirated; if the signal intensity is dark on T2WI, signifying a high viscosity (high-proteinaceous or cholesterol-laden cysts), it will be quite difficult to aspirate (Armao et al, 2000). A patient with positional headaches associated with a colloid cyst, especially accompa-
nied by hydrocephalus, should be immediately referred to a neurosurgeon (Hellwig et al, 2003) (Figure 5-8).

**CHIARI I MALFORMATION**

Cough, exertional, and sexual headaches may be classified as primary, but the clinician must search for secondary causes that include Arnold-Chiari type 1 malformation, subarachnoid hemorrhage, intracranial metastasis, posterior fossa masses, carotid or vertebral artery dissection, and idiopathic.

Chiari I malformation is a congenital malformation with cerebellar tonsillar herniation of at least 5 mm below the foramen magnum. When symptomatic, crowding of the craniocervical junction, obstructive hydrocephalus, and syringomyelia may occur. The fundamental problem appears to be underdevelopment of the posterior fossa. Symptoms most commonly reported in patients with Chiari I malformation include suboccipital headaches, retroorbital pressure or pain, clumsiness, dizziness, vertigo, tinnitus, paresthesias, muscle weakness, and lower cranial nerve symptoms (Arnett, 2004).

The imaging characteristics of Chiari I malformation include tonsils that extend more than 5 mm below the line connecting the basion with the opisthion. This is best seen on sagittal T1WI. Tonsillar tips that extend less than 5 mm below the foramen magnum are normal and may be called tonsillar ectopia. Cerebellar tonsils normally ascend with age; an extension of 6 mm is normal until the age of 10, 5 mm for patients aged 10 to 30 years, and 4 mm for those aged 30 to 80 years. In the typical Chiari I malformation, the low-lying tonsils are pointed in a “peglike” manner and have a vertical (not horizontal) orientation. There is usually a compressed or absent cisterna magna. Syrinx can occur in 15% to 75% of patients. Phase-contrast MRI reveals pulsatile systolic tonsillar descent and obstructed CSF flow through the foramen magnum (Hofkes et al, 2007). The workup for a Chiari I malformation

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**KEY POINTS:**

- Symptoms of Arnold-Chiari I malformation include suboccipital headaches, retroorbital pressure or pain, clumsiness, dizziness, vertigo, tinnitus, paresthesias, weakness, and cranial nerve symptoms.
- Cerebellar tonsils, which are displaced by more than 5 mm to 6 mm below the level of the foramen magnum with “peglike” (triangular) appearance confirm the diagnosis of Chiari I malformation.
should include a brain MRI to rule out a secondary Chiari I malformation and a complete spine MRI to rule out syrinx and tethered cord (Case 5-1).

**CRANIAL NEURALGIA**

Neuralgias are defined as intense burning and stabbing pain caused by irritation or damage to a nerve. Neuralgia may be idiopathic or secondary. The prototype of cranial neuralgia is trigeminal neuralgia, which is characterized by a brief electric shocklike pain, most frequently in the maxillary and mandibular divisions of cranial nerve V. Secondary causes of cranial neuralgias include skull-base tumors, vascular compression, neuritis, multiple sclerosis, herpes zoster, sinus disease, dental disease, and trauma. Glossopharyngeal and occipital neuralgia are well-defined, but far less common, cranial neuralgias. Secondary causes of trigeminal neuralgia include leptomeningeal disease (ie,
Carcinomatosis) and primary tumors, such as adenoid cystic carcinoma and squamous cell carcinoma with retrograde perineural spread of tumor into the cavernous sinus, gasserian ganglion, and trigeminal nerve. Patients who have tumor-related trigeminal neuralgia usually experience progressive or continuous pain, although bouts of intensification may occur. In contrast, patients having idiopathic trigeminal neuralgia are afflicted by short attacks of severe pain with intervals of painless periods. Cranial neuralgias can also be caused by neurofibromatosis type II. Glossopharyngeal neuralgias cause pain over the

Case 5-1

A 38-year-old man presented for evaluation of positional headaches, provoked by looking upward and coughing. The pain was a disabling headache associated with photophobia, sonophobia, and nausea without vomiting and had been progressive over the past year. The pain was worse on awakening and occipital in location. When he looked upward, the pain was immediate and caused him to drop to his knees. Laughing, sneezing, and straining also aggravated his headache. No visual symptoms were noted. The patient also reported a daily low-grade headache for which he took six tablets of acetaminophen plus aspirin plus caffeine a day. Complicating the history was a diagnosis of a “benign” intraventricular tumor that had been diagnosed 30 years earlier but had never been biopsied or treated. Serial MRIs in the past had been stable. Neurologic examination was essentially normal except for diffuse hyperreflexia.

MRI of the brain showed a large heterogenous mass within the lateral ventricle measuring 5.2 cm × 6 cm × 4.3 cm. Minimal progression had occurred when compared with the study done 5 years earlier. Evidence of a partially empty sella and significant Chiari I malformation measuring 15 mm was present but had not been discernable on the previous MRI. The moderate hydrocephalus was not associated with transependymal flow.

The diagnosis was (1) intraventricular ganglioma causing a secondary Chiari I malformation, (2) analgesic rebound headache, and (3) positional headache secondary to Chiari I malformation.

After subtotal resection of an intraventricular ganglioglioma, the headache completely resolved. Postoperative MRI of the brain confirmed postoperative changes, resolution of the tonsillar herniation, and improvement of the empty sella (Figure 5-9).

Comment. This is a case of acquired secondary Chiari I malformation. The most common cause of secondary cough headaches is a Chiari I malformation. Sagittal T1WI is recommended in patients with exertional coughing, sneezing, and laughing headaches.
ninth cranial nerve distribution, which may be lancinating in nature. Pain in the throat, tonsillar region, posterior third of the tongue, larynx, and nasopharynx is quite common. Secondary causes include leptomeningeal metastases or masses in or around the jugular foramen. In all patients with cranial neuralgias, standard imaging sequences are T1-WIs with and without contrast with thin slices through the skull base and thin-slice T2WI through the posterior fossa for the evaluation of aberrant vasculature. Thin slices, 2 mm or less, through the posterior fossa will allow precise imaging of the exiting cranial nerves as well as adjacent vessels. Leptomeningeal metastasis is also a common cause for cranial neuralgias as well as headaches. It is best seen with magnetization transfer or postcontrast T1WIs. Postcontrast FLAIR images are also being used for the evaluation of leptomeningeal disease (Mechtler, 2007) (Figure 5-10).

**FIGURE 5-10** A 35-year-old patient with left-sided trigeminal neuralgia that is becoming more frequent and is associated with continuous paresthesias in the maxillary and ophthalmic branch of cranial nerve V. A, B, T2-weighted images (T2WIs) show a relative hypointense extraaxial mass within the Meckel cave and cerebellopontine angle on the left (arrows). C, D, T1WI contrast study shows homogeneous enhancement (arrows). Imaging and pathology were consistent with meningioma. The diagnosis was secondary trigeminal neuralgia.
CONCLUSION
In 2006, the United Council of Neurologic Subspecialties (UCNS) adopted a neuroimaging core curriculum and examination to prepare neurologists for the independent practice of neuroimaging as a subspecialty of neurology. The primary purpose of the UCNS is to provide for accreditation and certification with the goal of enhancing the quality of training in neurologic subspecialties, such as headaches and neuroimaging, and improve quality of patient care. Headache specialists and neurologists now have a unique opportunity to be certified in both subspecialties and advance the diagnosis, treatment, and research of the most common ailment in society, headaches.

REFERENCES


