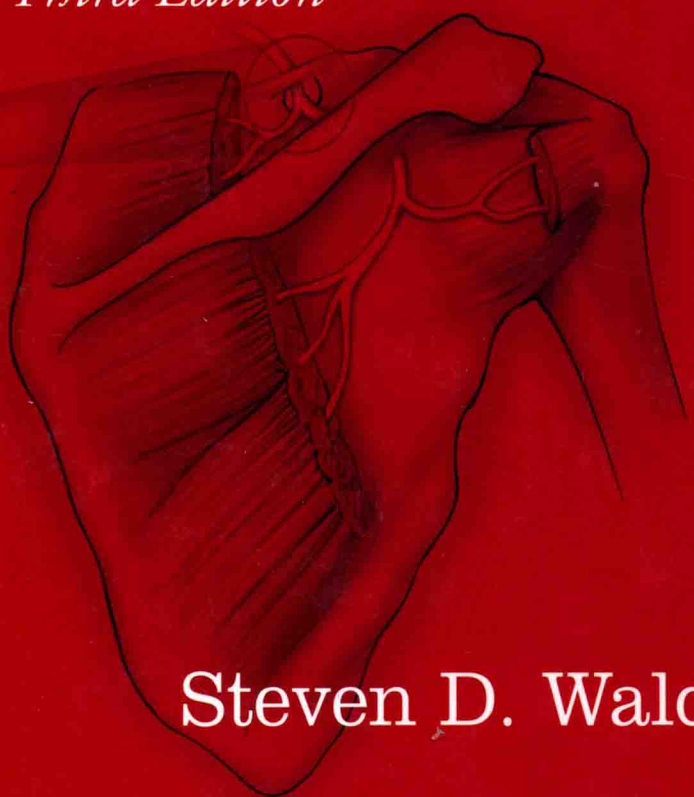
Atlas of Uncommon Pain Syndromes

Third Edition



Steven D. Waldman

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Atlas of UNCOMMON PAIN SYNDROMES

THIRD EDITION

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Atlas of UNCOMMON PAIN SYNDROMES

To Kathy—great wife, great mother, and technical support guru extraordinaire!

Steven D. Waldman, MD, JD

PREFACE

It has been said that the three most dangerous things in medicine are (1) a medical student with a sharp object, (2) a resident with a recently published study from the *New England Journal of Medicine,* and (3) an attending physician with an anecdote. One must suspect that point 2 was at play when in the 1940s while on rounds at the University of Maryland Hospital in Baltimore, Maryland, Theodore Woodward, MD, stated, "*If you hear hoof beats out on Green Street, don't look for zebras*"! How this admonition to aspiring physicians morphed into *when you hear hoof beats, look for horses, not zebras"* is anybody's guess. (My son, an ophthalmology resident in Baltimore, suggests that it was also just as likely that this sage piece of advice was accompanied by a long-winded and confusing anecdote—see point 3.)

On the surface, most of us would agree with Dr. Woodward's logic that the most common things are the most common. Occam agreed, when in the fourteenth century he put forth the philosophical tenant of parsimony, which proposes that simpler explanations are, all things being equal, almost always better than more complex ones. He used a razor to "shave away" unnecessary or extraneous data to get to the simplest solution. The razor was all the rage as a medical instrument in the fourteenth century, so it is not surprising that Occam chose it as his preferred medical device. Occam's razor certainly has a nice ring to it—better than Occam's MRI, which would no doubt be the name of his maxim if he had lived in the twenty-first century, given that currently the MRI is certainly our most popular medical device for "shaving away" extraneous data.

Which brings us to KISS—not the Gene Simmons rock band KISS, but the admonition "*Keep it simple, stupid.*" KISS was set forth by Lockheed aeronautical engineer, Kelly Johnson, when he handed his design team a few simple tools and challenged them to design combat jets that could be easily fixed with the simple tools that were available in combat situations. It is still not exactly clear to me who was "stupid," but I certainly hope it is not the guys who fix the jets I fly on. KISS makes sense when designing jet engines, but what does this have to do with the individual patient? The sick one? The scared one? The one you worry about in the middle of the night? Unfortunately, very little. Because for the individual patient with a difficult diagnosis, Hickam was probably more correct than Occam.

Harry Hickam, MD, while on teaching rounds at Duke University, admonished his students and residents that "*Patients can have as many diseases as they damn well please!*" (also see point 3). He correctly posited that when diagnosing the individual patient, using Occam's razor often provides the correct diagnosis. More often than we would care to admit, though, when dealing with a patient with a perplexing constellation of signs and symptoms, it can provide the wrong one. In fact, overreliance on Occam's razor can be downright dangerous for patient and physician alike. Often, the simplest, or in the case of medical diagnosis, the most common, illness is exactly what is causing the patient's symptoms. But sometimes, in our almost obsessive desire to make the diagnosis, simplicity is our enemy. In our haste to make the patient fit the diagnosis, we get it wrong. Uncommon diseases are called *uncommon diseases* because they are uncommon—they are not called *unknown diseases.* Since the beginning of time, healers have recognized that the correct diagnosis is the key to getting the patient well, and, as a corollary, they also realized that the wrong diagnosis is not a "practice builder." Which brings us to country music legend Mickey Gilley.

In 1976, Mickey Gilley recorded the classic country ballad *"Don't the Girls All Get Prettier at Closing Time,"* a plaintive lament about loneliness and late-night desperation and how one's perception of things can change as circumstances change. What turns an unknown disease into an uncommon disease is knowledge. What changes our perception of what a constellation of symptoms and physical findings mean when confronted with a perplexing diagnosis is knowledge. As we gain more clinical experience, things that were once unknown become known, even commonplace. The more we hone our clinical acumen, the easier it is to put the pieces together. Our perception of the diagnostic information our patients present us with changes from a jumble of disparate signs and symptoms to the certain diagnosis of an uncommon disease—one that we will never miss again! *Atlas of Uncommon Pain Syndromes,* Third Edition, seeks to accomplish three things: The first is to familiarize the clinician with a group of uncommon pain syndromes that occur with enough frequency that they merit serious study—not rare or orphan diseases, just uncommon ones that are often misdiagnosed. Second, this text is written with the goal of helping clinicians reinforce their knowledge of common pain syndromes to help in those situations when Occam is sort of correct—when the pieces of the puzzle do not quite fit the simple diagnosis. The third goal is more about the clinician and a little less about the patient. It is about what attracted many of us to medicine to begin with. It is the irresistible charm of being presented with a difficult clinical problem and getting it right. And what a great feeling that is! I hope you enjoy reading the third edition of *Atlas of Uncommon Pain Syndromes* as much as I did writing it.

Steven D. Waldman, MD, JD

CONTENTS

Chapter 1

ICE PICK HEADACHE

ICD-9 CODE **784.0**

ICD-10 CODE **R51**

The Clinical Syndrome

Ice pick headache is a constellation of symptoms consisting of paroxysms of stabbing jabs and jolts that occur primarily in the first division of the trigeminal nerve. These paroxysms of pain may occur as a single jab or a series of jabs that last for a fraction of a second followed by relatively pain-free episodes. The pain of ice pick headache occurs in irregular intervals of hours to days. Similar to cluster headache, ice pick headache is an episodic disorder that is characterized by "clusters" of painful attacks followed by pain-free periods. Episodes of ice pick headache usually occur on the same side, but in rare patients, the pain may move to the same anatomical region on the contralateral side. Ice pick headache occurs more commonly in women and is generally not seen before the fourth decade of life, but rare reports of children suffering from ice pick headache sporadically appear in the literature. Synonyms for ice pick headache include jabs and jolts headache and idiopathic stabbing headache.

Signs and Symptoms

A patient suffering from ice pick headache complains of jolts or jabs of pain in the orbit, temple, or parietal region (Figure 1-1). Some patients describe the pain of ice pick headache as a sudden smack or slap on the side of the head. Similar to patients suffering from trigeminal neuralgia, a patient suffering from ice pick headache may exhibit involuntary muscle spasms of the affected area in response to the paroxysms of pain. In contrast to trigeminal neuralgia, involving the first division of the trigeminal nerve, there are no trigger areas that induce the pain of ice pick headache. The neurological examination of a patient suffering from ice pick headache is normal. Some patients exhibit anxiety and depression because the intensity of pain associated with ice pick headache leads many patients to believe they have a brain tumor.

Testing

Magnetic resonance imaging (MRI) of the brain provides the best information regarding the cranial vault and its contents. MRI is highly accurate and helps identify abnormalities that may put the patient at risk for neurological disasters secondary to intracranial and brainstem pathological conditions, including tumors and demyelinating disease (Figure 1-2). Magnetic resonance angiography (MRA) also may be useful in helping identify aneurysms, which may be responsible for the patient's neurological findings. In patients who cannot undergo MRI, such as a patient with a pacemaker, computed tomography (CT) is a reasonable second choice. Radionuclide bone scanning and plain radiography are indicated if fracture or bony abnormality, such as metastatic disease, is considered in the differential diagnosis.

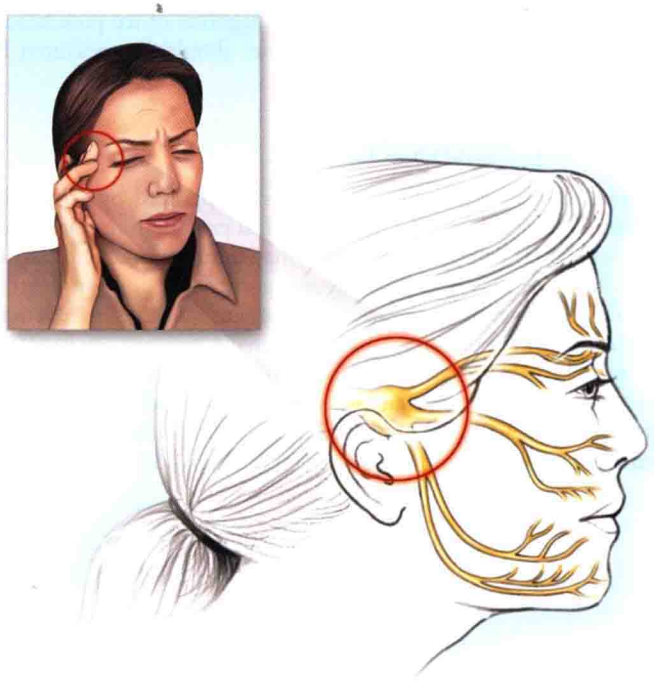

Figure 1-1 Ice pick headache is characterized by jabs or jolts in the orbit, temple, or parietal region.

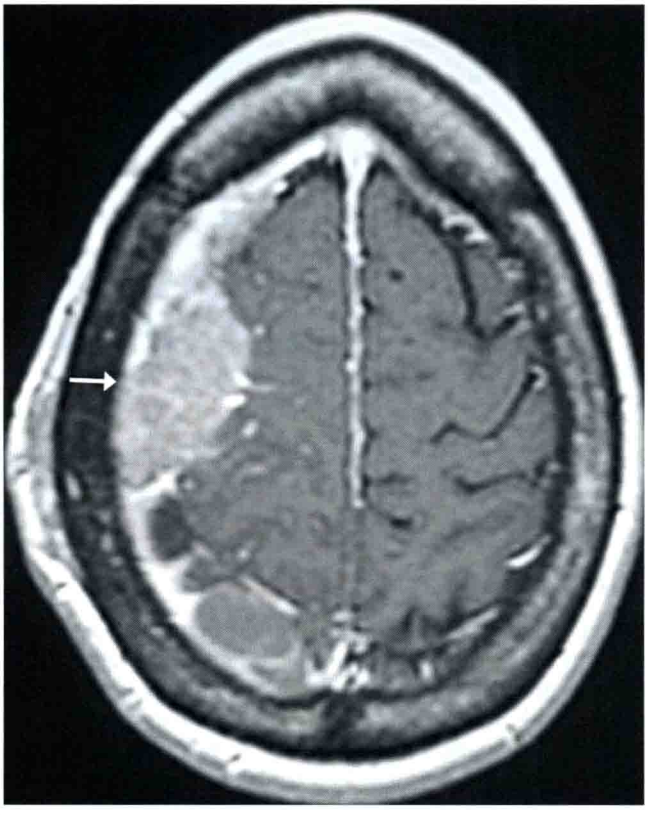

Figure 1-2 Diffuse pachymeningeal and calvarial metastasis from carcinoma of the breast. Axial T1-weighted postgadolinium MRI shows diffuse nodular and bandlike contrast-enhanced thickening of the dura over the high right frontoparietal convexity. *(From Haaga JR, Lanzieri CF, Gilkeson RC, editors:* CT and MR imaging of the whole body, *4th ed, Philadelphia, 2003, Mosby, p 198.)*

Screening laboratory tests consisting of complete blood cell count, erythrocyte sedimentation rate, and automated blood chemistry should be performed if the diagnosis of ice pick headache is in question. Intraocular pressure should be measured if glaucoma is suspected.

Differential Diagnosis

Ice pick headache is a clinical diagnosis supported by a combination of clinical history, normal physical examination, radiography, and MRI. Pain syndromes that may mimic ice pick headache include trigeminal neuralgia involving the first division of the trigeminal nerve, demyelinating disease, and chronic paroxysmal hemicrania. Trigeminal neuralgia involving the first division of the trigeminal nerve is uncommon and is characterized by trigger areas and tic-like movements. Demyelinating disease is generally associated with other neurological findings, including optic neuritis and other motor and sensory abnormalities. The pain of chronic paroxysmal hemicrania lasts much longer than the pain of ice pick headache and is associated with redness and watering of the ipsilateral eye.

Treatment

Ice pick headache uniformly responds to treatment with indomethacin. Failure to respond to indomethacin puts the diagnosis of ice pick headache in question. A starting dosage of 25 mg daily for 2 days and titrating to 25 mg three times per day is a reasonable treatment approach. This dose may be carefully increased to 150 mg per day. Indomethacin must be used carefully, if at all, in patients with peptic ulcer disease or impaired renal function. Anecdotal reports of a positive response to cyclooxygenase-2 (COX-2) inhibitors in the treatment of ice pick headache have been noted in the headache literature. Underlying sleep disturbance and depression are best treated with a tricyclic antidepressant compound, such as nortriptyline, which can be started at a single bedtime dose of 25 mg.

Complications and Pitfalls

Failure to correctly diagnose ice pick headache may put the patient at risk if intracranial pathological conditions or demyelinating disease, which may mimic the clinical presentation of chronic paroxysmal hemicrania, is overlooked. MRI is indicated in all patients thought to be suffering from ice pick headache. Failure to diagnose glaucoma, which also may cause intermittent ocular pain, may result in permanent loss of sight.

Clinical Pearls

The diagnosis of ice pick headache is made by obtaining a thorough, targeted headache history. Patients suffering from ice pick headache should have a normal neurological examination. If the results of the neurological examination are abnormal, the diagnosis of ice pick headache should be discarded and a careful search for the cause of the neurological findings should be undertaken.

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Chapter 2

SUPRAORBITAL NEURALGIA

ICD-9 CODE **350.1**

ICD-10 CODE **G50.0**

The Clinical Syndrome

The pain of supraorbital neuralgia is characterized as persistent pain in the supraorbital region and forehead with occasional sudden, shocklike paresthesias in the distribution of the supraorbital nerves. Sinus headache involving the frontal sinuses, which is much more common than supraorbital neuralgia, can mimic the pain of supraorbital neuralgia. Supraorbital neuralgia is the result of compression or trauma of the supraorbital nerves as the nerves exit the supraorbital foramen. Such trauma can be in the form of blunt trauma directly to the nerve, such as when the forehead hits the steering wheel during a motor vehicle accident, or repetitive microtrauma resulting from wearing welding or swim goggles that are too tight. This clinical syndrome also is known as swimmer's headache.

Signs and Symptoms

The supraorbital nerve arises from fibers of the frontal nerve, which is the largest branch of the ophthalmic nerve. The frontal nerve enters the orbit via the superior orbital fissure and passes anteriorly beneath the periosteum of the roof of the orbit. The frontal nerve gives off a larger lateral branch, the supraorbital nerve, and a smaller medial branch, the supratrochlear nerve. Both exit the orbit anteriorly. The supraorbital nerve sends fibers all the way to the vertex of the scalp and provides sensory innervation to the forehead, upper eyelid, and anterior scalp (Figure 2-1). The pain of supraorbital neuralgia is characterized as persistent pain in the supraorbital region and forehead with occasional sudden, shocklike paresthesias in the distribution of the supraorbital nerves. Occasionally, a patient suffering from supraorbital neuralgia complains that the hair on the front of the head "hurts" (Figure 2-2). Supraorbital nerve block is useful in the diagnosis and treatment of supraorbital neuralgia.

Testing

Magnetic resonance imaging (MRI) of the brain provides the best information regarding the cranial vault and its contents. MRI is highly accurate and helps identify abnormalities that may put the patient at risk for neurological disasters secondary to intracranial and brainstem pathological conditions, including tumors and demyelinating disease (Figure 2-3). Magnetic resonance angiography (MRA) also may be useful in helping identify aneurysms, which may be responsible for the patient's neurological findings. In patients who cannot undergo MRI, such as a patient with a pacemaker, computed tomography (CT) is a reasonable second choice. Radionuclide bone scan, CT, and plain radiography are indicated if sinus disease, fracture, or bony abnormality such as metastatic disease is considered in the differential diagnosis.

Screening laboratory tests consisting of complete blood cell count, erythrocyte sedimentation rate, and automated blood chemistry testing should be performed if the diagnosis of supraorbital neuralgia is in question. Intraocular pressure should be measured if glaucoma is suspected (Figure 2-4).

Differential Diagnosis

Supraorbital neuralgia is a clinical diagnosis supported by a combination of clinical history, normal physical examination, radiography, CT, and MRI. Pain syndromes that may mimic supraorbital

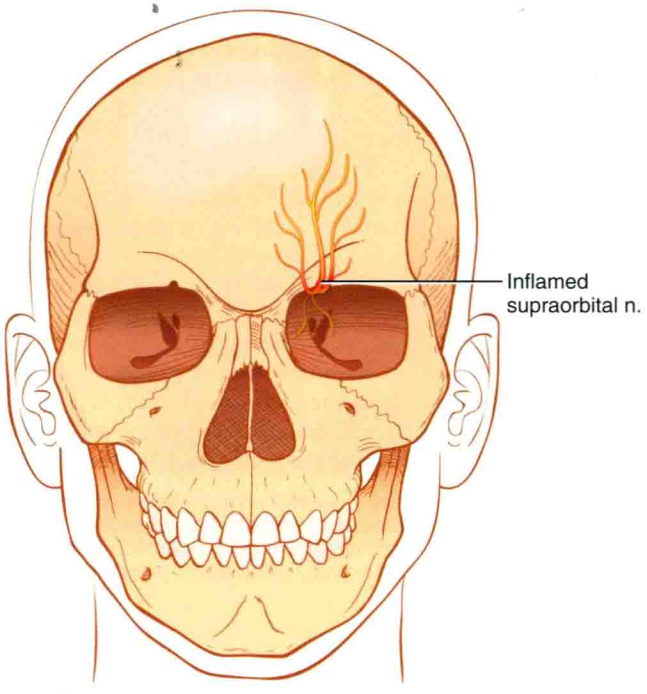

Figure 2-1 The supraorbital nerve sends fibers all the way to the vertex of the scalp and provides sensory innervation to the forehead, upper eyelid, and anterior scalp. *n,* Nerve.

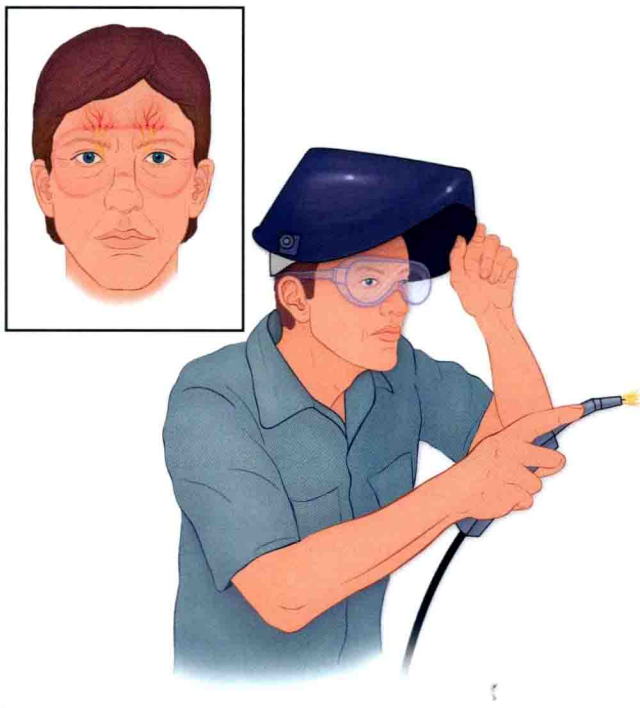

Figure 2-2 Occasionally, a patient with supraorbital neuralgia complains that the hair on the front of the head "hurts." The supraorbital nerve sends fibers all the way to the vertex of the scalp and provides sensory innervation to the forehead, upper eyelid, and anterior scalp.

neuralgia include ice pick headache, trigeminal neuralgia involving the first division of the trigeminal nerve, demyelinating disease, and chronic paroxysmal hemicrania. Trigeminal neuralgia involving the first division of the trigeminal nerve is uncommon and is characterized by trigger areas and tic-like movements. Demyelinating disease is generally associated with other neurological findings, including optic neuritis and other motor and sensory abnormalities. The pain of chronic paroxysmal hemicrania lasts much longer than the paroxysmal pain of supraorbital neuralgia and is associated with redness and watering of the ipsilateral eye.

Treatment

The primary treatment intervention for supraorbital neuralgia is the identification and removal of anything causing compression of the supraorbital nerves (e.g., tight welding or swim goggles). A brief trial of simple analgesics alone or in combination with gabapentin also should be considered. For patients who do not respond to these treatments, supraorbital nerve block with local anesthetic and a steroid is a reasonable next step.

To perform supraorbital nerve block, the patient is placed in the supine position. Using a 10-mL sterile syringe, 3 mL of local anesthetic is drawn up. When treating supraorbital neuralgia with supraorbital nerve block, 80 mg of depot steroid is added to the local anesthetic with the first block, and 40 mg of depot steroid is added with subsequent blocks.

The supraorbital notch on the affected side is identified by palpation. The skin overlying the notch is prepared with antiseptic solution, with care taken to avoid spillage into the eye. A 25-gauge, 1½-inch needle is inserted at the level of the supraorbital notch and is advanced medially approximately 15 degrees off the perpendicular to avoid entering the foramen. The needle is advanced until it approaches the periosteum of the underlying bone (Figure 2-5). A paresthesia may be elicited, and the patient should be warned of such. The needle should not enter the supraorbital foramen; if this occurs, the needle should be withdrawn and redirected slightly more medially.

Because of the loose alveolar tissue of the eyelid, a gauze sponge should be used to apply gentle pressure on the upper eyelid and supraorbital tissues before injection of solution to prevent the injectate from dissecting inferiorly into these tissues. This pressure should be maintained after the procedure to avoid periorbital hematoma and ecchymosis.

After gentle aspiration, 3 mL of solution is injected in a fanlike distribution. If blockade of the supratrochlear nerve also is desired, the needle is redirected medially and, after careful aspiration, an additional 3 mL of solution is injected in a fanlike manner.

Underlying sleep disturbance and depression associated with the pain of supraorbital neuralgia are best treated with a tricyclic antidepressant compound, such as nortriptyline. The tricyclic antidepressant can be started at a single bedtime dose of 25 mg.

Complications and Pitfalls

Failure to diagnose supraorbital neuralgia correctly may put the patient at risk if an intracranial pathological condition or demyelinating disease, which may mimic the clinical presentation of supraorbital neuralgia, is overlooked. MRI is indicated in all patients thought to have supraorbital neuralgia. Failure to diagnose glaucoma, which also may cause intermittent ocular pain, may result in permanent loss of sight.

The forehead and scalp are highly vascular, and when performing supraorbital nerve block the clinician should carefully calculate the total milligram dosage of local anesthetic that may be given safely, especially if bilateral nerve blocks are being performed. This vascularity gives rise to an increased incidence of postblock ecchymosis and hematoma formation. Despite the vascularity of this anatomical region, this technique can be performed safely in the presence of anticoagulation by using a 25- or 27-gauge needle, albeit at increased risk for hematoma, if the clinical situation dictates a favorable risk-to-benefit ratio. These complications can be decreased if manual pressure is applied to the area of the block immediately after injection. Application of cold packs for 20-minute periods after the block also decreases the amount of postprocedure pain and bleeding.

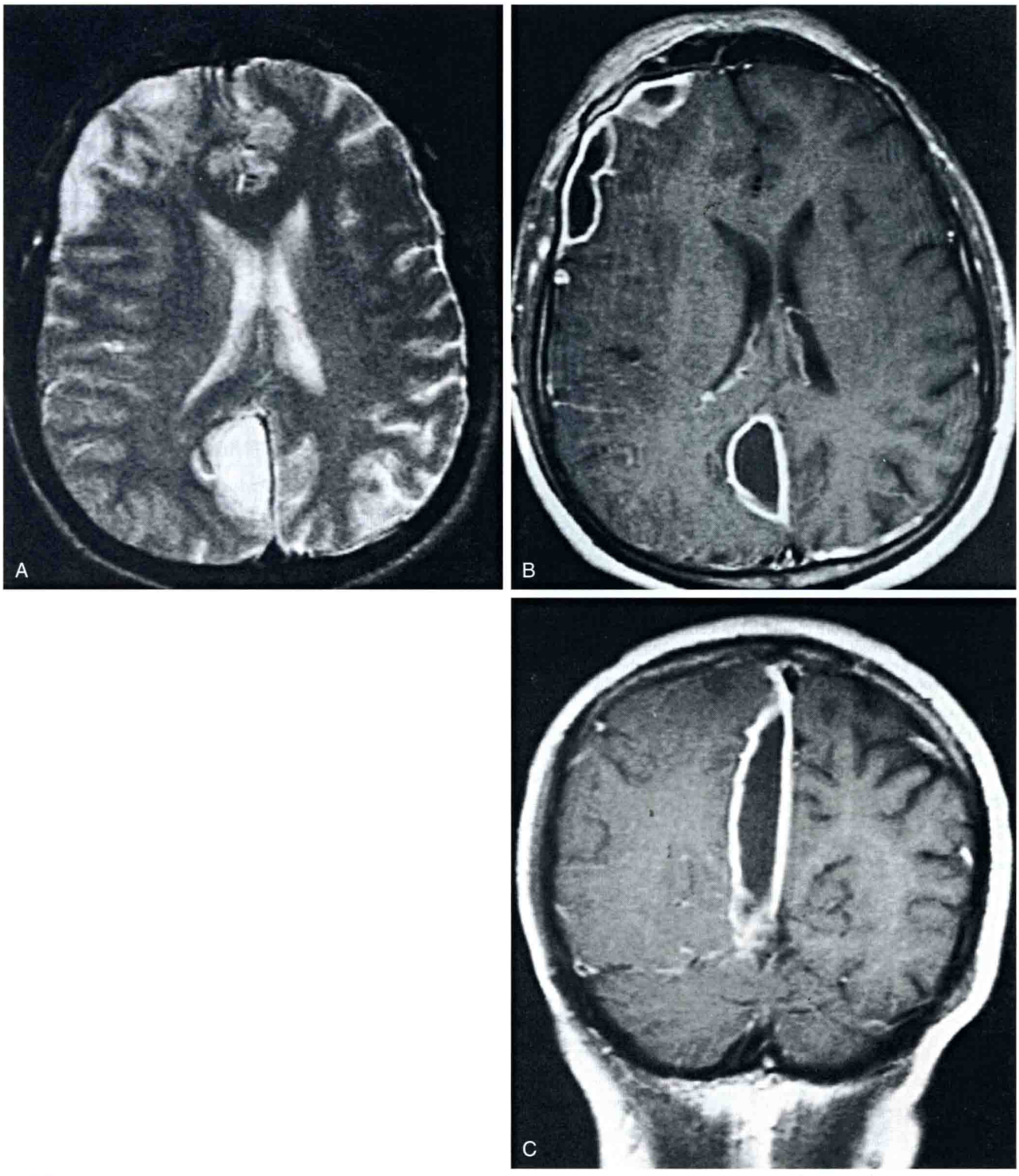

Figure 2-3 Subdural empyema in a patient with sinusitis. **A,** T2-weighted MRI shows high-signal-intensity extraaxial fluid collection in the right frontal convexity and along the falx on the right side. **B** and **C,** Gadolinium-enhanced MRI shows extraaxial fluid collections in the right frontal convexity and along the falx with intense peripheral enhancement. The signal intensity of the fluid collection is slightly higher than that of cerebrospinal fluid. *(From Haaga JR, Lanzieri CF, Gilkeson RC, editors:* CT and MR imaging of the whole body, *4th ed, Philadelphia, 2003, Mosby, p 209.)*

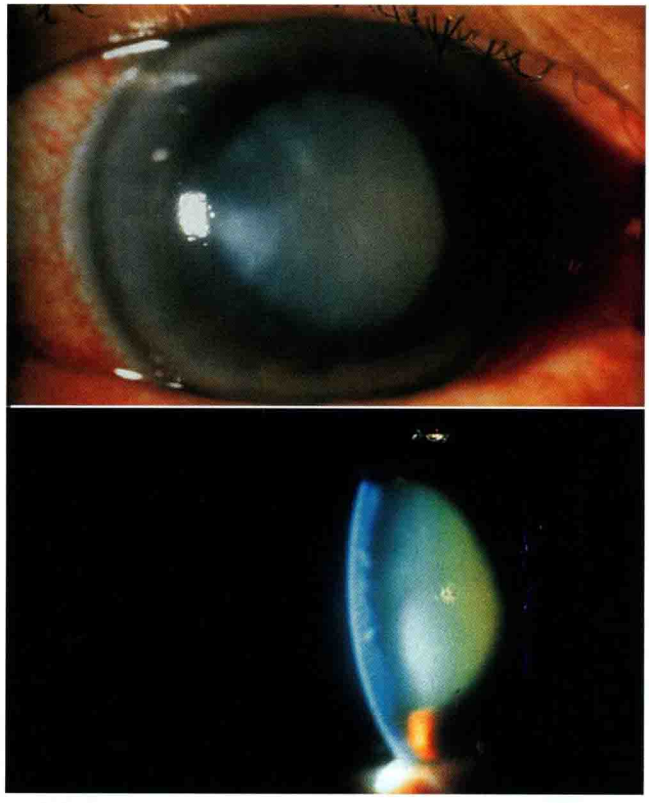

Figure 2-4 Acute angle closure resulting from an intumescent cataractous lens. The eye is red with a hazy view of the anterior segment from corneal edema, with a fixed, irregular, semidilated pupil from iris infarction. The slit image shows the corneal edema and a very shallow anterior chamber. Some uveitis may be present because of ischemia, and this must be differentiated from the larger accumulations of lens material and macrophages seen with phacolytic glaucoma. *(From Spalton DJ, Hitchings RA, Hunter P:* Atlas of clinical ophthalmology, *3rd ed, London, 2005, Mosby, p 225.)*

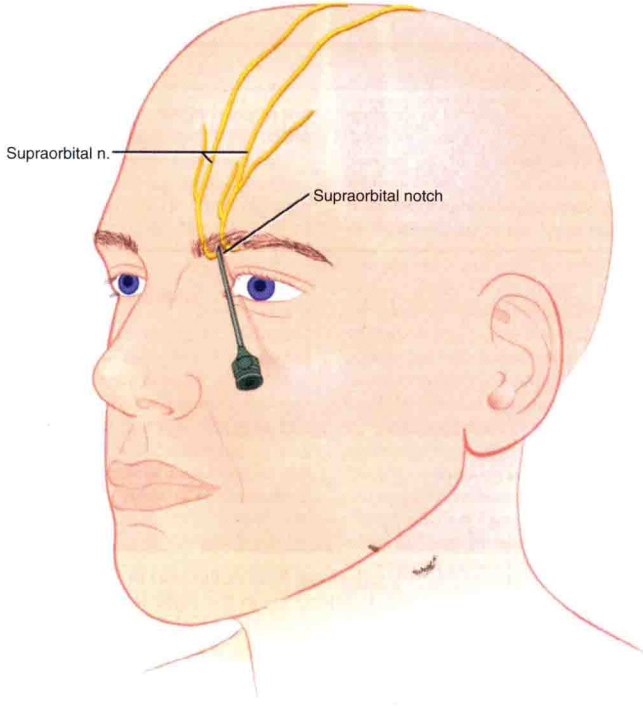

Figure 2-5 Injection technique for relieving the pain of supraorbital neuralgia. *(From Waldman SD:* Atlas of pain management injection techniques, *2nd ed, Philadelphia, 2007, Saunders.)*

Clinical Pearls

Supraorbital nerve block is especially useful in the diagnosis and palliation of pain secondary to supraorbital neuralgia. The first step in the management of this unusual cause of headache is the correct fitting of swimming goggles that do not compress the supraorbital nerves. Coexistent frontal sinusitis should be ruled out in patients who do not respond rapidly to a change in swim goggles and a series of the previously mentioned nerve blocks. Any patient with headaches severe enough to require neural blockade as part of the treatment plan should undergo MRI of the head to rule out unsuspected intracranial pathological conditions.

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Chapter 3

CHRONIC PAROXYSMAL HEMICRANIA

ICD-9 CODE **784.0**

ICD-10 CODE **R51**

The Clinical Syndrome

Chronic paroxysmal hemicrania shares many characteristics of its more common analogue, cluster headache, but has several important differences (Table 3-1). Similar to cluster headache, chronic paroxysmal hemicrania is a severe, episodic, unilateral headache that affects the periorbital and retroorbital regions. In contrast to cluster headache, which occurs 10 times more commonly in men, chronic paroxysmal hemicrania occurs primarily in women (Figure 3-1). The duration of pain associated with chronic paroxysmal hemicrania is shorter than that of cluster headache, lasting 5 to 45 minutes. This pain does not follow the chronobiological pattern seen in patients with cluster headache. Patients with chronic paroxysmal hemicrania usually experience more than five attacks per day. Chronic paroxysmal hemicrania uniformly responds to indomethacin, whereas cluster headache does not.

Signs and Symptoms

During attacks of chronic paroxysmal hemicrania, patients exhibit the following physical findings suggestive of Horner's syndrome on the ipsilateral side of the pain:

- Conjunctival and scleral injection
- Lacrimation
- Nasal congestion
- Rhinorrhea
- Ptosis of the eyelid

TABLE 3-1

Comparison of Cluster Headache and Chronic Paroxysmal Hemicrania

Comparison Factors	Cluster Headache	Chronic Paroxysmal Hemicrania
Gender predominance	Male	Female
Response to indomethacin	Negative	Positive
Chronobiological pattern	Positive	Negative
Alcohol trigger	Positive	Negative
Length of attacks	Longer	Shorter
Horner's syndrome	Present	Present

As in cluster headache, the patient may become agitated during attacks, rather than seeking dark and quiet as does the patient with migraine. In contrast to cluster headache, alcohol consumption does not seem to trigger attacks of chronic paroxysmal hemicrania. Between attacks, the neurological examination of a patient with chronic paroxysmal hemicrania should be normal.

Testing

Magnetic resonance imaging (MRI) of the brain provides the best information regarding the cranial vault and its contents. MRI is highly accurate and helps identify abnormalities that may put the patient at risk for neurological disasters secondary to intracranial and brainstem pathological conditions, including tumors and demyelinating disease (Figure 3-2). Magnetic resonance angiography (MRA) also may be useful in identifying aneurysms, which may be responsible for the patient's neurological findings. In patients who cannot undergo MRI, such as a patient with a pacemaker, computed tomography (CT) is a reasonable second choice. Radionuclide bone scanning and plain radiography are indicated if fracture or bony abnormality such as metastatic disease is considered in the differential diagnosis.

Screening laboratory tests consisting of complete blood cell count, erythrocyte sedimentation rate, and automated blood chemistry testing should be performed if the diagnosis of chronic

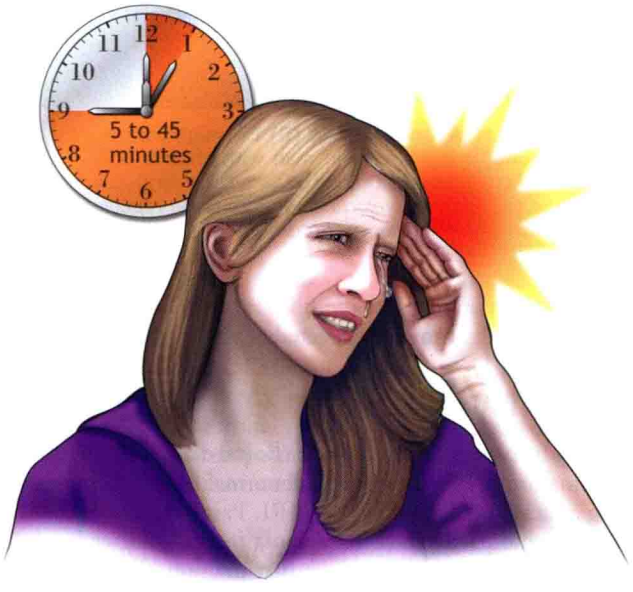

Figure 3-1 In contrast to cluster headache, which occurs primarily in men, chronic paroxysmal hemicrania occurs primarily in women.

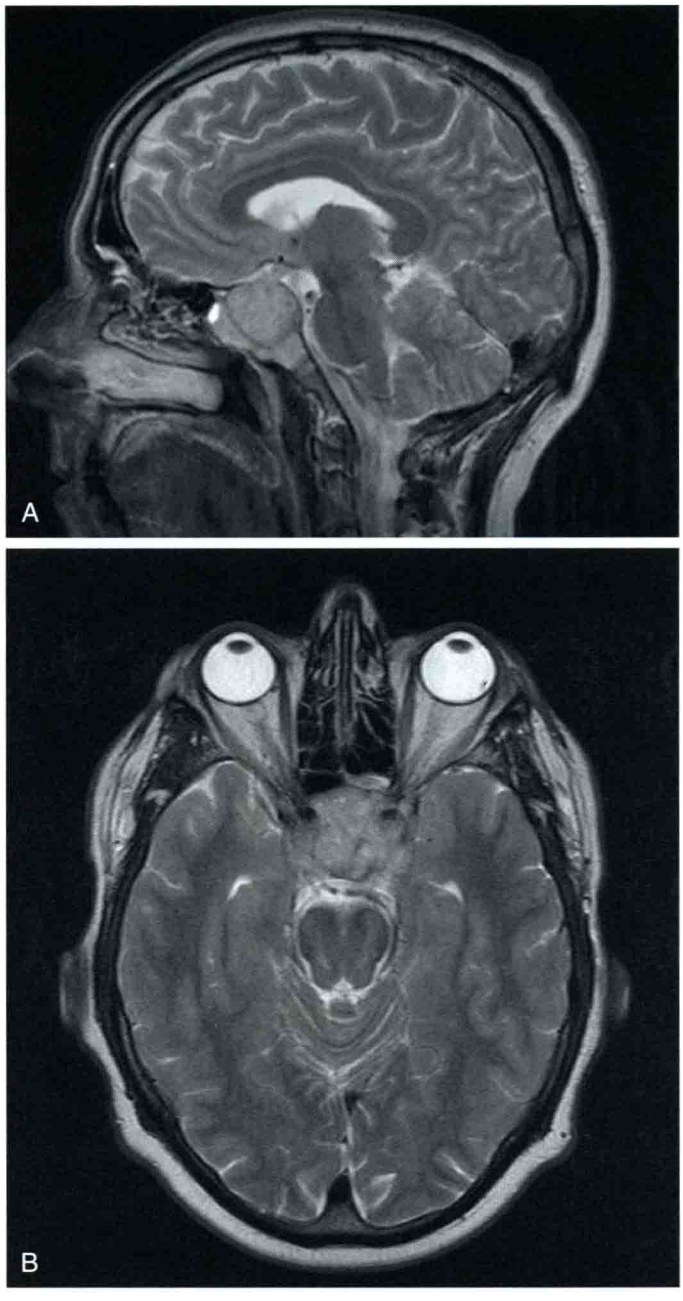

Figure 3-2 Sagittal **(A)** and semiaxial **(B)** T2-weighted images of a massive prolactinoma in a 41-year-old man with chronic daily headache. *(From Benitez-Rosario MA, McDarby G, Doyle R, Fabby G. Chronic cluster-like headache secondary to prolactinoma: uncommon cephalalgia in association with brain tumors,* J Pain Symptom Manage *37:271–276, 2009.*

paroxysmal hemicrania is in question. Intraocular pressure should be measured if glaucoma is suspected.

Differential Diagnosis

Chronic paroxysmal hemicrania is a clinical diagnosis supported by a combination of clinical history, abnormal physical examination during attacks, radiography, and MRI. Pain syndromes that may mimic chronic paroxysmal hemicrania include cluster headache, trigeminal neuralgia involving the first division of the trigeminal nerve, demyelinating disease, and ice pick headache. Trigeminal neuralgia involving the first division of the trigeminal nerve is uncommon and is characterized by trigger areas and tic-like movements. Demyelinating disease is generally associated with other neurological findings, including optic neuritis and other motor and sensory abnormalities. The pain of cluster headache lasts much longer than the pain of chronic paroxysmal hemicrania, and cluster headache has a male predominance, a chronobiological pattern of attacks, and a lack of response to treatment with indomethacin.

Treatment

Chronic paroxysmal hemicrania uniformly responds to treatment with indomethacin. Failure to respond to indomethacin puts the diagnosis of chronic paroxysmal hemicrania in question. A starting dose of 25 mg daily for 2 days and titrating to 25 mg three times per day is a reasonable treatment approach. This dose may be carefully increased up to 150 mg per day. Indomethacin must be used carefully, if at all, in patients with peptic ulcer disease or impaired renal function. Anecdotal reports of a positive response to cyclooxygenase-2 (COX-2) inhibitors in the treatment of chronic paroxysmal hemicrania have been noted in the headache literature. Underlying sleep disturbance and depression are best treated with a tricyclic antidepressant compound, such as nortriptyline, which can be started at a single bedtime dose of 25 mg.

Complications and Pitfalls

Failure to diagnose chronic paroxysmal hemicrania correctly may put the patient at risk if intracranial pathological conditions or demyelinating disease that may mimic the clinical presentation of chronic paroxysmal hemicrania is overlooked. MRI is indicated in all patients thought to have chronic paroxysmal hemicrania. Failure to diagnose glaucoma, which may cause intermittent ocular pain, may result in permanent loss of sight.

Clinical Pearls

The diagnosis of chronic paroxysmal hemicrania is made by obtaining a thorough, targeted headache history. Between attacks, patients with chronic paroxysmal hemicrania should have a normal neurological examination. If the neurological examination is abnormal between attacks, the diagnosis of chronic paroxysmal hemicrania should be discarded and a careful search for the cause of the patient's neurological findings should be undertaken.

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