HEADACHE and FACIAL PAIN

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I) Introduction

Headache disorders are among the most prevalent medical problems worldwide

The World Health Organization estimates that 50% to 75% of all adults between the ages of 18 and 65 years have headaches

This high prevalence results in significant disability and lost productivity: headache disorders are the 3rd highest cause of years lost to disability worldwide

Accordingly, headaches are one of the most common reasons patients present to physicians in primary care settings, in the emergency department (ER), or in neurologists' offices

For clinicians evaluating a patient with headaches, the 1st responsibility is to diagnose the type of headache correctly, and then treat appropriately , because most headache disorders have excellent treatments available, which can reduce the burden of disability

Headache disorders fall into 2 categories, primary headache disorders (those caused by the headache disorder itself, not due to other causes) and secondary headache disorders, those caused by (or "symptomatic of") another underlying medical problem

The pain can be due to the involvement of pain-sensitive structures in the head , including cranial nerves, cervical nerve roots, blood vessels, meninges, scalp, temporomandibular joint (TMJ), teeth, pericranial and cervical muscles, and paranasal sinuses

Patients may also have multifactorial headaches, so a detailed history and examination are necessary to identify the contributing factors

Headache disorders may remain refractory to treatment or have an insufficient response to treatment if these comorbidities are not identified and addressed

II) Diagnosis

A detailed history and examination are vital in understanding the headache's cause

There are no biomarkers currently available for primary headache disorders

Key points from the history

When obtaining the history the following information must be elicited:

- Onset
- Precipitants and triggers
- Duration
- Location(unilateral or bilateral; frontal, lateral, vertex ,or occipital)
- Quality and severity
- Frequency
- Alleviating and exacerbating factors
- Positional influences (better or worse when supine)
- Waking the patient from sleep, or occurring upon awakening
- Associated with menses
- Associated symptoms

Additional aspects of the history important in evaluating a patient with headache are:

- Analgesic use
- Caffeine use
- Medical history
- Current or recent pregnancy
- Medications(including asking specifically about contraceptive use, over-the –counter treatments, and supplements)
- Social history, including detailed screening for illicit drugs
- Family history
- Sleep, including a history of insomnia and snoring, symptoms suggestive of obstructive sleep apnea

The semiology of the headache helps to differentiate a primary from a secondary headache disorder

The history also allows a clinician to identify red flags that suggest a secondary headache disorder

- Acute onset or progressive worsening from baseline
- New or different headache
- Systemic symptoms:
  - Fever, weight loss
- Risk factors:
  - Malignancy
  - Immunosuppression
  - IV illicit substance use
  - Hypercoagulability, including pregnancy
- Smoking
- Age>50, or no prior headache history
- Features of increased ICP:
  - Waking patient from sleep
  - Worsening with Valsalva maneuver
  - Supine worsening of pain
- Focal features:
  - Seizures
  - Mental status abnormality
  - Cranial nerve deficits
  - Weakness
  - Sensory changes (loss of sensation, paresthesias; location and pattern of spread)
- Precipitants:
  - Trauma
  - Newly prescribed medications
  - Infection

RED FLAGS

- Acute onset or progressive worsening from baseline
- New or different headache
- Systemic symptoms: fever, weight loss
- Risk factors: malignancy, immunosuppression, IV illicit substance use, hypercoagulability, including pregnancy
- Smoking
- Age more than 50 years, or no prior headache history

- Features of increased ICP: waking patient from sleep, worsening with Valsalva maneuver, supine worsening of pain

- Focal features: seizures, mental status abnormality, cranial nerve deficits, weakness, sensory changes(loss of sensation, paresthesias, location of pattern of spread)

- Precipitants: trauma, newly prescribed medications, infection

Key points for the neurological examination

Patients with primary headache disorders usually have normal general medical and neurological examinations, although an acutely symptomatic patient with an autonomic cephalalgia may have signs suggesting that disorder

Some patients with chronic headaches have findings of temporomandibular joint (TMJ) tenderness on palpation, evidence of dental wearing, or pain with palpation of the cervical muscles or the occipital ridge to suggest comorbid causes of headache such as cervicalgia

In the era of the smartphone, patients may bring pictures of themselves to a clinician for review if they have paroxysmal symptoms and signs(such as ptosis or lacrimation), which can aid in the diagnosis

Patients should have a general medical and neurological examination to assess for secondary causes of headache

Attention to vital signs is important: patients with significant hypertension may be susceptible to developing certain secondary headaches outlined below

Fever may suggest an underlying infection, including a CNS infection

A cardiovascular exam can evaluate for arrhythmia or carotid stenosis, which can cause secondary headache syndromes

A detailed head and neck exam includes evaluating for nuchal rigidity, cervical myofascial pain, occipital Tinel sign(evaluated by eliciting tenderness or tingling when palpating near the occipital protuberance along the occipital nerve), and palpation of the TMJ, assessment of dental wearing or chipping to suggest bruxism, and observing the oropharynx for narrowing that could suggest obstructive sleep apnea

A full neurological exam should also be performed, with emphasis on the funduscopic exam to assess for papilledema

The cortical sensory exam can suggest cortical dysfunction that may occur with venous sinus thrombosis

Focal neurological deficits, including field cuts, cranial nerve palsies, weakness, or sensory symptoms, often suggest a secondary headache

III) Primary headache disorders

Primary headache disorders are those not due to another medical condition

Diagnosis is established by history and exam

Migraine is by far the most prevalent primary headache disorder

The table outlines common primary headache disorders based on key features of the history

The figure shows the common locations of pain in the primary headache disorders, compared to that of headaches caused by sinus disease

TABLE 10-1. Key Features of Primary Headache Disorders

	Episodic Migraine	Episodic Tension	Episodic Cluster	Paroxysmal Hemicrania	SUNCT and SUNA
Sex	Female > male	Female > male	Male > female	Female > male	Male > female
Location	Unilateral > bilateral	Bilateral (band around the head)	Unilateral (behind or around the eye)	Unilateral (behind or around the eye)	Unilateral (behind or around the eye)
Quality	Throbbing, pulsatile	Dull pressure or tightening (vice-like)	Stabbing, burning, boring	Stabbing, burning, throbbing	Stabbing, burning
Severity	Moderate to severe	Moderate	Severe	Severe	Severe
Attack duration	4–72 h	30 min–7 d	15–180 min	2–30 min	1 s–10 min
Attack frequency	Variable	Variable	From 1 every other day to 8/d	>5/d to 40/d	From 1/d to 200/d
Autonomic features	No	No	Yes	Yes	Yes

SUNCT, short-lasting unilateral neuraliform headache attacks with conjunctival injection and tearing; SUNA, short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms.

withdrawal and overuse)

FIGURE 10-1. Location of pain associated with primary headache disorders. *Left* to *right*: sinus headache, cluster headache, tension headache, migraine headache. (Used with permission of A.D.A.M.)

* MIGRAINE

A) Migraine without aura

It is estimated that 1 in 7 adults worldwide has migraine

It impacts women more than men in a 2:1 ratio

Migraine may start in childhood and manifest occasionally with abdominal symptoms(" abdominal migraine")

Motion sickness in children is a risk factor for the development of migraine

Migraine headaches are most likely to develop in adolescence and early adulthood

They can be episodic or chronic

The disability and lost productivity from migraine are substantial, because it impacts people in their prime working years

Migraine has numerous identified triggers, including weather changes, menses and caffeine(both withdrawal and overuse)

Many patients identify foods and drinks such as alcohol(most commonly red wine), soft cheeses, and nitrite-heavy foods, such as processed meats, as precipitants, but data are sparse in this area and many migraine attacks occur without identifiable triggers

To diagnose migraine, a patient must have at least 5 attacks with the following characteristics:

1. The headache lasts for 4 to 72 hours if untreated
2. It must include at least 2 of the following features:
    a. Throbbing
    b. Unilateral headaches
    c. Worsening with activity, such as walking
    d. Moderate to severe pain
3. It must be associated with at least one of the following:
    a. Nausea, vomiting, or both
    b. Photophobia and phonophobia

B) Migraine with aura

Migraine headaches are often preceded by focal neurological symptoms known as auras

These are also called classic migraine or complicated migraine

Auras are defined as fully reversible neurologic symptoms with a gradual onset, usually followed by a headache

The aura usually lasts 5 to 60(often 20) minutes and is typically unilateral

It usually resolves without lingering neurologic deficits

Patients are diagnosed with this disorder when they have an aura followed by a headache that meets the criteria for migraine, as above

Some auras occur without a headache (" acephalgic migraine"), but these symptoms usually require additional investigation for a definite diagnosis

Visual auras are by far the most common

Some include a" fortification" spectrum(zigzag lines off the central vision, usually spreading gradually) or a scintillating (or flickering) scotoma (an area of decreased visual acuity surrounded by preserved vision)

Migraine auras can also involve the sensory symptoms, most commonly paresthesias (tingling or pins-and-needles sensation)

The paresthesias often “ march” or spread gradually over the course of several minutes along a limb or extend from an arm to the leg or face

Migraine auras can also include a gradual onset of weakness, a variant known as hemiplegic migraine when severe

Hemiplegic migraine may be sporadic but there is also a syndrome of familial hemiplegic migraine, sometimes associated with well-characterized genes

Migraine auras are believed to be due to “ cortical spreading depression” in which there is a spread of hyperpolarization of the cortex followed by a wave of depolarization

Imaging studies have shown decreased regional cerebral blood flow in the cortex during migraine aura, but not to the level of worrisome ischemia

C) Complications associated with migraine

*Status migrainosus

When migraine lasts for more than 72 hours, the condition is known as status migrainosus

This is often caused by abortive medication overuse (often referred to as rebound headache) and frequently requires intravenous (IV) treatment or a brief course of oral steroids to break the headache cycle

* Stroke risk associated with migraine

Patients with migraine with aura have an increased cardiovascular risk when compared to healthy controls

The use of oestogen-based contraceptives is therefore contraindicated in patients with migraine with aura, as the combination results in a substantially increased stroke risk

* Migraine and menses

Women of reproductive age frequently have exacerbation of migraine during menses, most commonly 1 to 2 days prior to onset of bleeding, often persisting for up to 3 days into bleeding

This is thought to be due to the withdrawal of estrogen that occurs with menses

Some women have migraine at the time of menstruation only, a condition known as pure menstrual migraine

Most , however, have a few episodic headaches at other times of the month, or menstrually related migraine

It is important to identify the relationship of menses to migraine because there are specific treatments that may be helpful for patients with a clear exacerbation around their menses

* Chronic migraine

Patients who have a headache more than 15 days/month for more than 3 months are diagnosed with chronic migraine

Some patients with chronic migraine do not have typical features of migraine with all headaches, but they must have at least 8 days of headache consistent with migraine to be diagnosed with chronic migraine

If the headaches are not consistent with migraine, other diagnoses must be considered

Patients often describe a history of gradually progressive episodic migraines that increase in frequency to the point of meeting criteria for chronic migraine

With frequent headaches, many patients with chronic migraine have some component of medication overuse headache (MOH)

Importantly, patients with chronic migraine can revert to episodic migraine after effective treatment

D) Migraine treatments

1. Abortive treatments

Abortive treatments, also called rescue medications, are medications used to stop a migraine at the onset

All abortive treatments are most effective if the patient is treated at the onset of the headache

Delay in treatment results in more prolonged disability, so patients must be counseled on the appropriate use of abortive treatments

Nonsteroidal anti-inflammatory drugs (NSAIDs) and triptans (serotonin 1b/1d agonists) are the mainstay of abortive treatments

Many patients respond to NSAIDs alone

For some patients, however, they are insufficient; some patients have contraindications to using NSAIDs. In these cases, triptans can be highly effective

There are numerous different types of triptans , with different rates of onset of action and half-lives

There are 2 long-acting triptans (naratriptan and frovatriptan) and 5 fast-acting triptans (almotriptan, eletriptan, sumatriptan,rizatriptan, and zolmitriptan)

There are also numerous different formulations, including oral pills, disintegrating tablets, nasal sprays, and injectables

Historically, ergotamines were prescribed as abortive treatments, but they carry a higher cardiovascular risk and have been replaced by triptans

Triptans and NSAIDs can be combined when needed and may have a synergistic effect in treating migraine pain

Caffeine is also often added to many migraine treatments because it can help abort the pain; many over-the-counter “ migraine preparations” contain caffeine

Triptans are currently not known to be safe in pregnancy and have a cardiovascular risk

Triptans also interact with selective serotonin reuptake inhibitors and serotonin-norepinephrine reuptake inhibitors , with a low risk of serotonin syndrome (recent medications: “ditans” which are 5HT1F agonists and “gepants” which are CGRP blockers)

Patients must be counseled on side effects of all treatments

Identifying the right abortive treatment requires careful consideration of the patient’s headache features , comorbidities, concurrent medication use, cost, and family planning goals

Using abortive treatments on a chronic basis more than twice a week can result in medication overuse headache (MOH), so patients should be counseled to not use any of these treatments chronically more than twice a week to prevent this complication

2. Adjuvant treatments

Because nausea and emesis are frequently associated with migraine, many patients benefit from antiemetics

Interestingly, prochlorperazine and metoclopramide are more effective than ondansetron, both in alleviating the nausea and in reducing the severity of the pain

Antiemetics may also be useful in preventing patients from vomiting their abortive therapies

They are frequently used in emergency room (ER) and urgent care settings for patients with severe or refractory migraine

They are often combined with ketorolac and diphenhydramine for patients with status migrainosus

3. Preventive treatments

Preventive treatments, also called prophylactic treatments, are used for patients with chronic migraine or frequent and disabling headaches that do not respond sufficiently to abortive treatments

Preventive therapy aims to reduce the frequency and severity of migraine, although patients are unlikely to become completely headache-free and should be counseled accordingly

All prophylactic treatments take some time to have an effect ;patients should remain on treatment for at least a month (barring significant side effects or other concerns) before assuming that the treatment is ineffective

There are 3 primary categories of preventive oral medications: antihypertensives, antiseizure medications , and antidepressants

Within each category, there are specific drugs with the most evidence of efficacy(table)

In addition to oral therapies, onabotulinum toxin A (often referred to simply as Botox) was also approved as migraine prophylaxis for chronic migraine in 2010

are more

TABLE 10-2. Migraine Prophylaxis Oral Medications

Antihypertensives	Antiseizure Drugs	Antidepressant
Metoprolol	Sodium valproate	*Amitriptyline*
Propranolol	Topiramate	*Venlafaxine*
Timolol		

Medications in **bold** have level A evidence for efficacy. Medications in *italics* have level B evidence for efficacy.

In 2018, a new class of preventative therapy for chronic migraine , Calcitonin Gene –Related Peptide (CGRP) antagonists was approved by the FDA

Erenumab is an injectable human monoclonal antibody that antagonizes CGRP receptor function

As with abortive treatments, selecting the right prophylactic medication requires careful consideration of the patient's comorbidities , concomitant medications, cost , and family planning goals

Patients must be counseled about treatment options and side effects, including teratogenicity and impact on contraceptives

Patients who require preventive therapies also require abortive treatments

Some abortive treatments interact with prophylactic medications (such as antidepressants and triptans) which should be taken in consideration

4. Lifestyle modifications

Lifestyle factors are important to identify

A comorbid sleep disorder (insomnia, obstructive sleep apnea …) makes patients more susceptible to migraine

Skipping meals, insufficient fluid, excessive caffeine intake, and lack of exercise make susceptible patients more prone to migraine attacks

Patients should be counseled on these factors

* TENSION-TYPE HEADACHE

Tension-type headache (often referred to as tension headaches, stress headaches, or ordinary headaches) are the next most prevalent primary headache disorders, occurring in 30% to 70% of adults worldwide

Pain is usually bilateral and described as pressure or tightness

It is usually mild to moderate and lasts for under an hour to several days

Unlike migraine, it is not associated with photophobia, phonophobia, nausea, or vomiting

The examination is generally normal , but some patients have pericranial tenderness to palpation of the scalp, neck, or shoulder muscles

Tension-type headaches can be episodic or chronic(occurring more than 15 days/month)

Interestingly, patients with infrequent tension-type headaches generally do not seek medical attention, because they do not have significant disability from their symptoms

Patients with frequent or chronic tension-type headaches benefit from treatment

Tension-type headache treatment

1. Abortive treatments

Many patients with tension headaches do not require abortive treatments because the pain is generally mild and does not interfere with the patient's functioning

For those with moderate to severe pain, NSAIDs are the mainstay of treatment

Aspirin and acetaminophen may also be used, but the latter is often less effective than NSAIDs

Patients should be counseled about the development of MOH and advised to not use analgesics more than twice a week for long periods

2. Preventive treatments

Antidepressants are the first-line preventive therapy for chronic tension headache

The tricyclic amitryptiline is the most studied to date and has good evidence for efficacy

Other antidepressants , including mirtazapine and venlafaxine are second-line therapies

Muscle relaxants such as tizanidine are helpful sometimes, particularly in patients with a cervicogenic component

3. Adjuvant treatments

Tension headaches are often reported to be triggered by stress (physical or emotional); addressing these triggers, if chronic, is important

Biofeedback (a mind-body technique used to teach patients greater body awareness and how to control some physical reactions to pain and stress) can be effective

Poor posture and neck muscle spasm are also frequent contributors to chronic tension-type headaches, and physical therapy can help

* TRIGEMINAL AUTONOMIC CEPHALALGIAS

Trigeminal autonomic cephalalgias (TACs) are the 3rd major category of primary headache disorders

They are characterized by unilateral pain associated with cranial autonomic symptoms

The diagnosis is made by careful evaluation of the pattern of the pain and its associated features (table shown on a previous slide)

A) Cluster headache

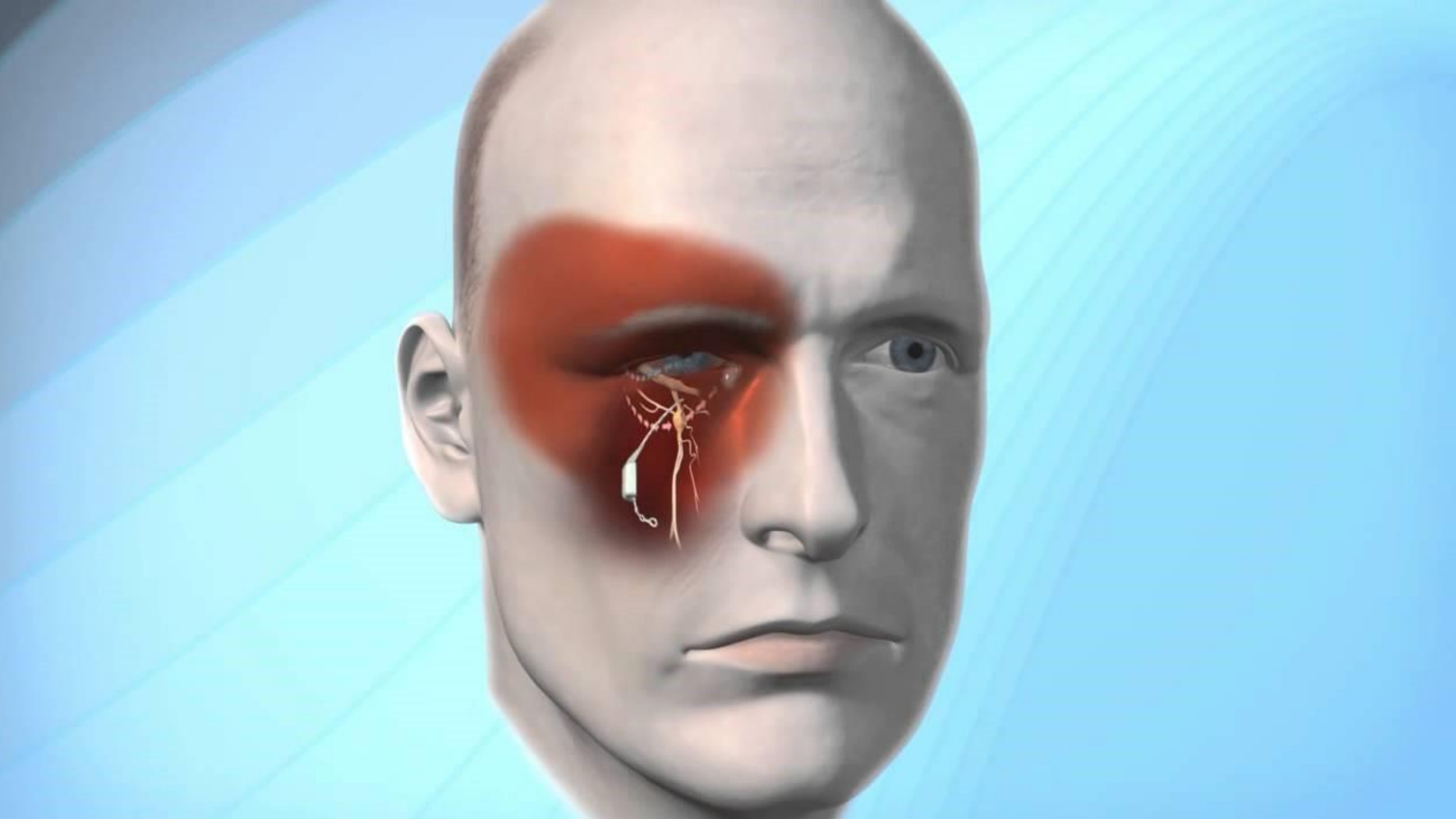
Cluster headaches are severe headaches characterized by unilateral pain involving the orbit, supraorbitally, at the temple, or combination of these(previous figure)

Cluster periods are bouts of recurrent attacks of pain, generally lasting weeks to months

These periods are followed by remission lasting anywhere from months to years

The pain is often excruciating

During an attack, patients are often restless and pacing, unlike in migraine where activity exacerbates the pain



The pain must be associated with one of the following cranial autonomic symptoms:

- Conjunctival injection, lacrimation, or both
- Nasal congestion, rhinorrhea, or both
- Eyelid edema
- Forehead and facial sweating or flushing
- Sensation of fullness in the ear
- Miosis, ptosis, or both

Cluster headaches typically last between 15 and 120 minutes

During a cluster period, headaches can occur several times a day or as infrequently as every other day

Cluster headaches are relatively uncommon but are 3 times more likely to occur in men

The age of onset is typically in early to mid-adulthood (20-40 years)

The cause is unknown, but activation of the posterior hypothalamic gray matter has been seen in some patients during attacks

Alcohol, histamines, and nitroglycerine are triggers in susceptible patients

A Horner syndrome caused by carotid dissection may mimic a cluster headache (figure) but does not usually have the pain characteristics of this primary headache disorder

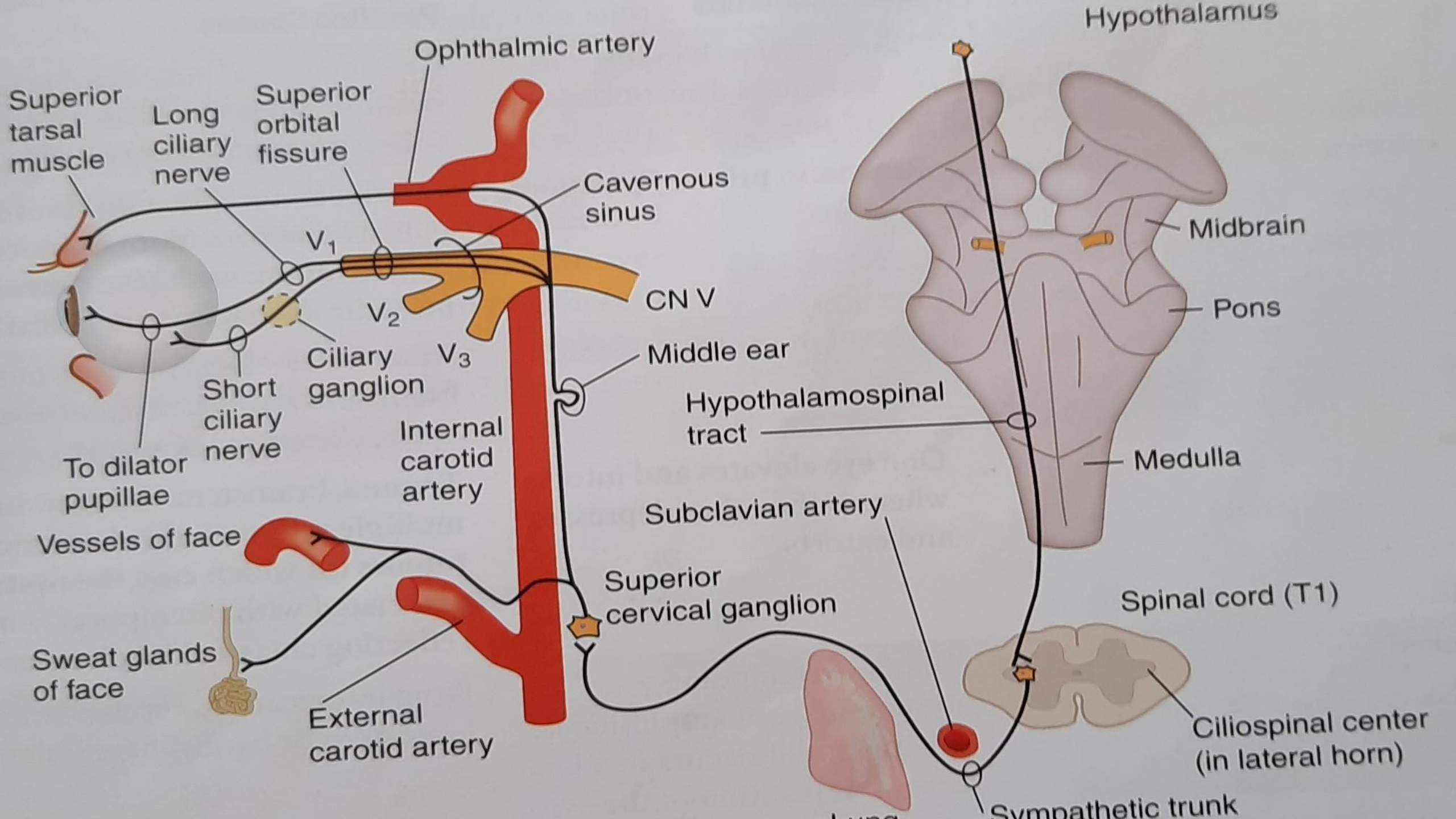
Cluster headaches may be episodic or chronic

Chronic cluster headache is defined as intractable cluster headaches with less than 1 month of remission before the recurrence of symptoms

Fortunately, less than 15% of cluster patients have chronic cluster

dissection may mimic a

FIGURE 10-2. Cluster headache. Horner syndrome. Note mild *unilateral* ptosis (on the patient's right side), anisocoria with a *smaller* pupil on the side of the ptosis, and redness from associated cluster headache. The common "upside-down" ptosis (*i.e., elevation*) *of the lower* lid is masked by coexisting eyelid laxity and blepharitis, which can be confounding factors in older adults.


Hypothalamus
Ophthalmic artery
Superior tarsal muscle
Long ciliary nerve
Superior orbital fissure
Cavernous sinus
Midbrain
V_1
V_2
V_3
CN V
Pons
Ciliary ganglion
Middle ear
Short ciliary nerve
Hypothalamospinal tract
Internal carotid artery
To dilator pupillae
Medulla
Subclavian artery
Vessels of face
Superior cervical ganglion
Spinal cord (T1)
Sweat glands of face
Ciliospinal center (in lateral horn)
External carotid artery
Sympathetic trunk

First-order (or central):
- Hypothalamic infarcts, tumor
- Mesencephalic stroke
- Brainstem: ischemia (Wallenberg syndrome), tumor, hemorrhage
- Spinal cord: syringomyelia, trauma

Second-order (or preganglionic):
- Cervicothoracic cord/spinal root trauma
- Cervical spondylosis
- Pulmonary apical tumor: Pancoast tumor

Third-order (or postganglionic):
- Superior cervical ganglion (tumor, iatrogenic)
- Internal carotid artery: dissection, trauma, thrombosis, tumor
- Base of skull: tumor, trauma
- Middle ear problems
- Cavernous sinus: tumor, inflammation (Tolosa–Hunt syndrome), aneurysm, thrombosis, fistula

Cluster headache treatments

1. Abortive treatments

A first-line abortive treatment for cluster headaches is 100% oxygen, delivered at 12 to 15L/min

Patients may receive this treatment in an urgent care or ED setting and if effective, be prescribed a home oxygen tank

For patients who do not respond , or who do not have access to home oxygen, triptans are prescribed

Sumatriptan and zolmitriptan are effective as abortive therapies

In the past, dihydroergotamine (DHE) was prescribed as an abortive treatment , but triptans are favored over DHE given their safety profile

Patients with cluster headache are very susceptible to MOH and must be counseled appropriately

Occipital nerve block can also be effective to abort a cluster cycle

2. Preventive treatments

Preventive treatments for cluster headache are similar to those used for migraine and include antihypertensive,antiseizure, and psychiatric medications

Verapamil is the first-line therapy for cluster headache prophylaxis

If not tolerated or if there are contraindications, glucocorticoids

(prednisone or dexamethasone) are also effective

Lithium and topiramate are often used as second-line agents or as add-on therapy when needed

B) Short-lasting unilateral neuralgiform headache attacks

Short-lasting unilateral neuralgiform headaches are unilateral, moderate to severe headaches

Pain is around the orbit or temple but may also occur in the trigeminal distribution and therefore be mistaken for trigeminal neuralgia

The headache is a stabbing pain or recurrent stabbing sensation lasting from 1 second to 10 minutes

Patients with a lesion in the posterior fossa may present with symptoms suggestive of short-lasting neuralgiform headache so brain imaging with MRI is important to establish that this is a primary and not secondary headache disorder

There are 2 forms, differentiated by the types of associated autonomic symptoms (which always occur on the same side as the headache):

- Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing(SUNCT): Autonomic symptoms include both conjunctival injection and lacrimation
- Short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms(SUNA):

  Autonomic symptoms include at least one of the following:

  * forehead or facial sweating or flushing
  * Ptosis or pupillary miosis
  * Eyelid edema
  * Nasal congestion, rhinorrhea, or both
  * Ear fullness
  * Either conjunctival injection or lacrimation , but not both

Both SUNCT and SUNA can be episodic or chronic

The chronic forms are diagnosed by persistent symptoms lasting more than a year, or for less than a year but less than 1 month of remission

1. Abortive treatment

SUNCT and SUNA are challenging disorders to treat, given the brevity of symptoms

Intravenous lidocaine has helped abort the cycle in some patients

2. Preventive treatment

Antiseizure medications including topiramate, gabapentin, and lamotrigine are used as preventive therapy in patients with frequent or recurrent symptoms

Occipital nerve blocks can also be helpful, especially when systemic medications are contraindicated or not tolerated

C) HEMICRANIA

The final TAC is hemicrania, a unilateral headache , differentiated from the other TACs both by the duration of symptoms and by its unique response to indomethacin

Patients present with a unilateral orbital or temporal headache associated with one or more autonomic symptoms on the same side as the headache(the symptoms and signs are the same as SUNCT and SUNA)

There are 3 variants of hemicranias, differentiated by the duration of symptoms:

- Episodic paroxysmal hemicranias:
    * Recurrent attacks separated by at least one pain-free month
    * Attacks last between 2 and 30 minutes but can recur within a day
- Chronic paroxysmal hemicranias:
    * Recurrent attacks without remission, or less than 1 month of remission before recurrence
    * Attacks last between 2 and 30 minutes but can recur within a day
- Hemicrania continua: Intractable pain and autonomic symptoms consistent with hemicranias, lasting for more than 3 months

All 3 forms respond to indomethacin, and this response to treatment is required to make the diagnosis

Hemicrania is more common in women and typically occurs in mid-adulthood (30-40 years of age)

As with other TACs , MRI to exclude a lesion in the posterior fossa is also advised to exclude a secondary headache syndrome

Abortive and preventive treatment

Indomethacin is the definitive treatment for hemicranias

An indomethacin trial is both diagnostic and therapeutic

The dose is titrated gradually over 10 days to a maximum of 225 mg a day, divided into 3 doses, until the patient has a therapeutic response

If there is no response, the diagnosis is not consistent with hemicranias and other etiologies must be considered

IV) Opioids in headache treatment

Opioids are not more effective than alternative therapy and are generally strongly discouraged for use in headache medicine

Most headache conditions are recurrent disorders , so to use opiods in this setting risks development of a secondary opioid use disorder

Opioids also tend to cause MOH

V) SECONDARY HEADACHE DISORDERS

Secondary headache disorders are headaches caused by a medical condition or medication

They have a broad differential for causes, ranging from preeclampsia and pheochromocytoma to fever and medication side effects

Most secondary headache disorders are associated with other features in the history, examination, or laboratory assessment, which aid in the diagnosis

Treatment is based on addressing the underlying disorder

There are 6 major categories of secondary headaches that may present with headache only and must be considered

1. Vascular causes

There are numerous vascular causes of headache

All cerebral hemorrhages can cause headache

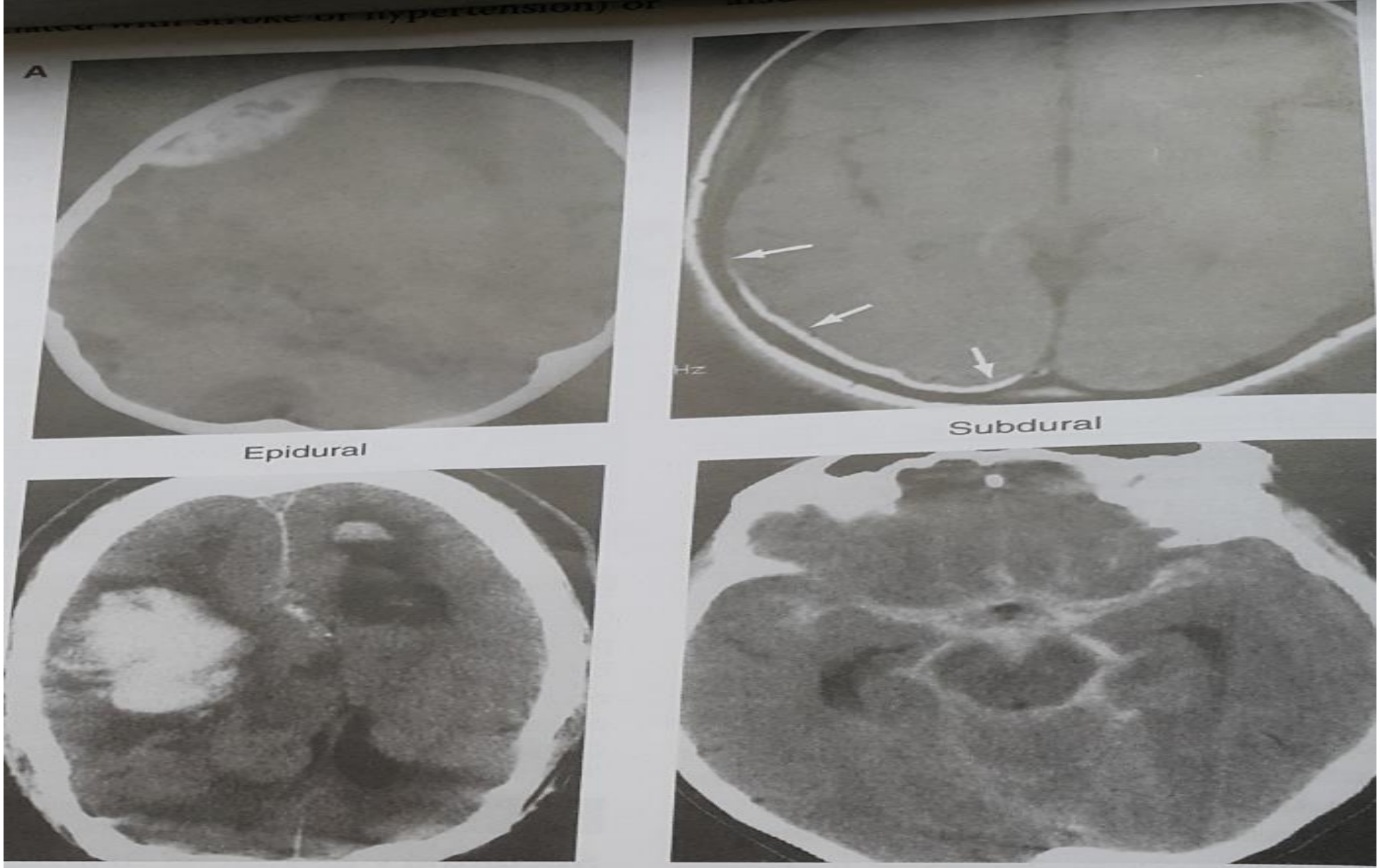
This includes subarachnoid hemorrhage(SAH) , intraparenchymal hemorrhage, and subdural and epidural hematomas

These hemorrhages may be spontaneous (associated with stroke or hypertension) or traumatic

Patients with intracerebral hemorrhages typically present with what is referred to as thunderclap headache; the onset is abrupt and severe

Emergency imaging , usually with a noncontrast CT Brain scan, is needed to evaluate abrupt-onset headache(figure)

Cerebral vessel imaging is also warranted if a SAH is identified , to assess for an aneurysm



s of intracerebral hemorrhages on CT scans. **(A)** Epidural hemorrhage. **(B)** Su
hemorrhage. **(D)** Subarachnoid hemorrhage. Arrows point to the subdural he

Ischemic strokes are often associated with headaches

Their semiology is nonspecific, but typically abrupt in onset

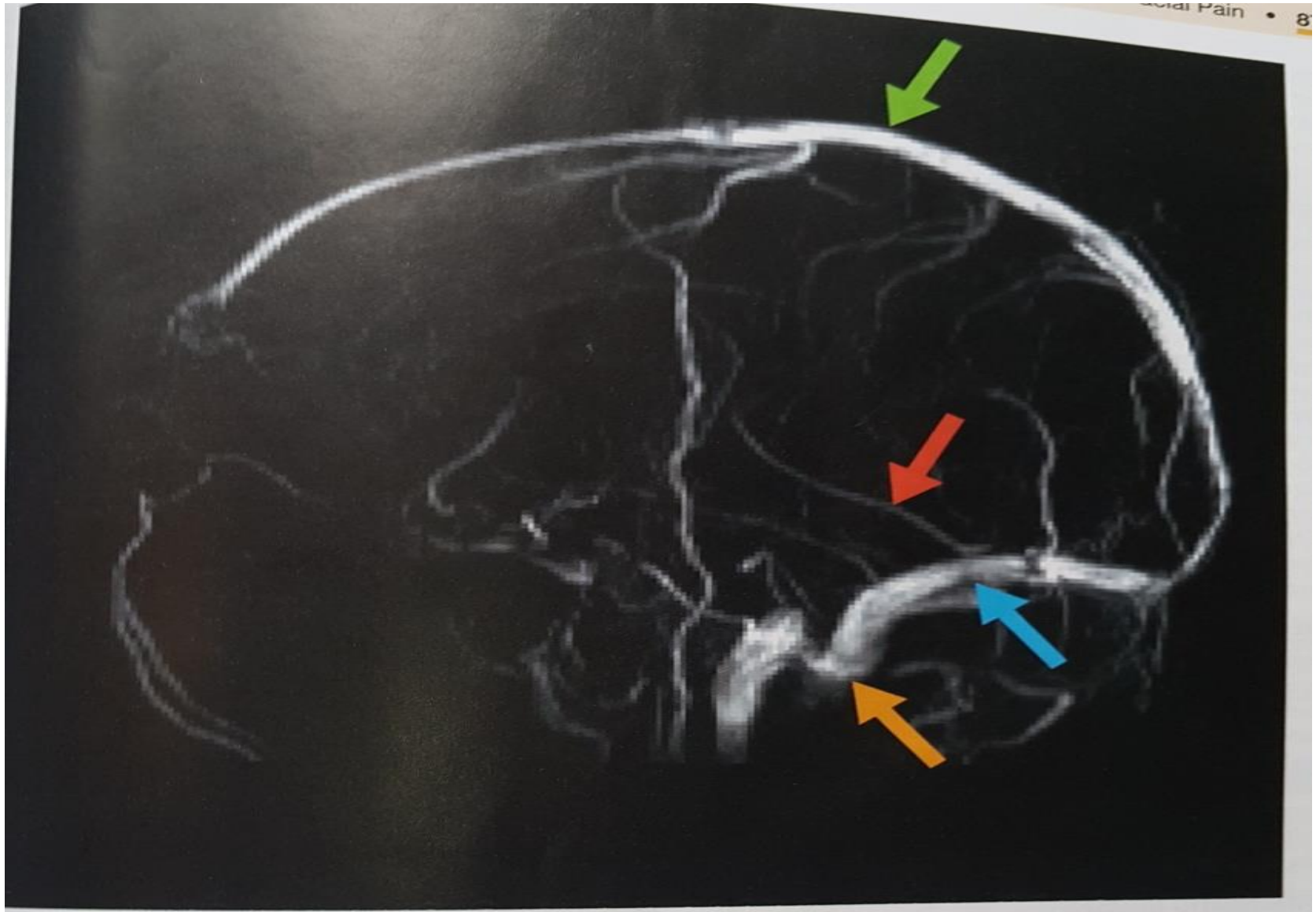
Patients may have focal neurologic deficits which aid in the diagnosis

Cerebral thrombosis, either arterial or venous, can also cause headache

Patients with venous sinus thrombosis often have headaches with features of increased intracranial pressure(ICP)

Thrombosis should be considered particularly in patients with hypercoagulability states, including pregnancy

The diagnosis is made on imaging, including that of cerebral vessels (figure)



E 10-4. Deep venous sinus thrombosis in a pregnant woman. Sagittal image from a magnetic resonance ...ram (MRV) demonstrating occlusion of the deep venous system, including the straight sinus (red arrow). The ...or sagittal sinus (green arrow) and right transverse sinus (blue arrow) and sigmoid sinus (orange arrow) are paten...

Cerebral vasculitis frequently causes a nonspecific headache

When part of a systemic vasculitis, it is considered a secondary angiitis

If the vasculitis occurs in the cerebral vessels alone, it is referred to as primary CNS angiitis

In addition to headache, patients often have paroxysmal focal neurological deficits

Cerebral arterial vessel imaging and lumbar puncture (LP) are often required to make this diagnosis

Giant cell arteritis (GCA) , also called temporal arteritis, is a peripheral cranial arterial vasculitis that often presents with unilateral headache

Patients are generally above the age of 50 and report additional symptoms including vision changes(amaurosis fugax), jaw claudication, fever, and scalp tenderness

Involvement of the branches of the external carotid artery, and occasionally the ophthalmic artery, can result in blindness if not readily identified and treated promptly

Patients usually have elevated inflammatory markers(erythrocyte sedimentation rate and C-reactive protein)

Empiric steroids should be started in any patient with a high clinical concern

Temporal artery biopsy is the gold standard, but GCA can cause “ skip lesions” and may require serial biopsies to identify the pathology

2. Infectious or inflammatory causes

Intracranial infections , such as encephalitis and meningitis, usually present with headache and often fever

They may also have nuchal rigidity and altered mental status

As the infection progresses , seizures and focal neurological deficits may occur

Infections can be bacterial, viral , fungal, or parasitic; the headache semiology does not help to differentiate the underlying cause

LP is the crucial diagnostic test and necessary in any patient for whom there is concern for a CNS infection

There are often other signs of infection, and the headache has a temporal correlation to the infection

CNS inflammatory and autoimmune conditions such as sarcoidosis and lupus frequently present with headache

3. Neoplastic causes

Intracranial neoplasms may present with headaches, especially when there is significant mass effect

The headache semiology may be nonspecific but may have features of intracranial hypertension, including wakening the patient from sleep , being worse when supine, and worse with Valsalva maneuver

The headache may occur early or late with neoplasms and with any type of primary cancer

4. Traumatic causes

Head and neck injuries often result in headache

To be attributed to trauma, the headache must develop with a temporal association with the injury

The severity of the injury does not necessarily correlate with the severity of the headache; even minor head injuries or whiplash may cause headaches

There is no specific headache semiology that helps with the diagnosis

In patients with a history of significant head or neck injury, it is important to assess for intracranial hemorrhage or dissection of cervical vessels- which may require additional treatment

5. Intracerebral pressure disorders

Intracranial hypertension and hypotension can cause headaches, but with markedly different semiologies

A) Intracranial hypertension

Intracranial hypertension may be " idiopathic"(most common in obese young woman) or due to medications or systemic disorders

The headache is often described as worse when *supine* or sleeping

(awakening the patient from sleep) , or with Valsalva maneuver

Patients often have associated features , including papilledema, pulsatile tinnitus, or visual symptoms

Patients should have imaging to exclude a mass lesion or venous sinus thrombosis

If the imaging is unrevealing , the diagnosis is made with an LP when the patient is in the lateral decubitus position with legs extended

Intracerebral pressure (ICP) is elevated above 200 mm CSF

Acetazolamide is the first-line treatment for idiopathic intracranial hypertension (IIH)

Patients require monitoring of their visual fields, and treatment for obesity is warranted when present

B) Intracranial hypotension

Intracranial hypotension may be spontaneous or traumatic

The headache improves when the patient is supine but worsens with standing

The pain is often most severe at the vertex and can be associated with neck pain or tinnitus

It is frequently traumatic- occurring after an attempted epidural puncture or LP

Generally, the dural leak causing the headache heals gradually, without intervention,but when symptoms persist or when the headache etiology is unclear, brain MRI may be helpful ;it may show evidence of sagging(figure)

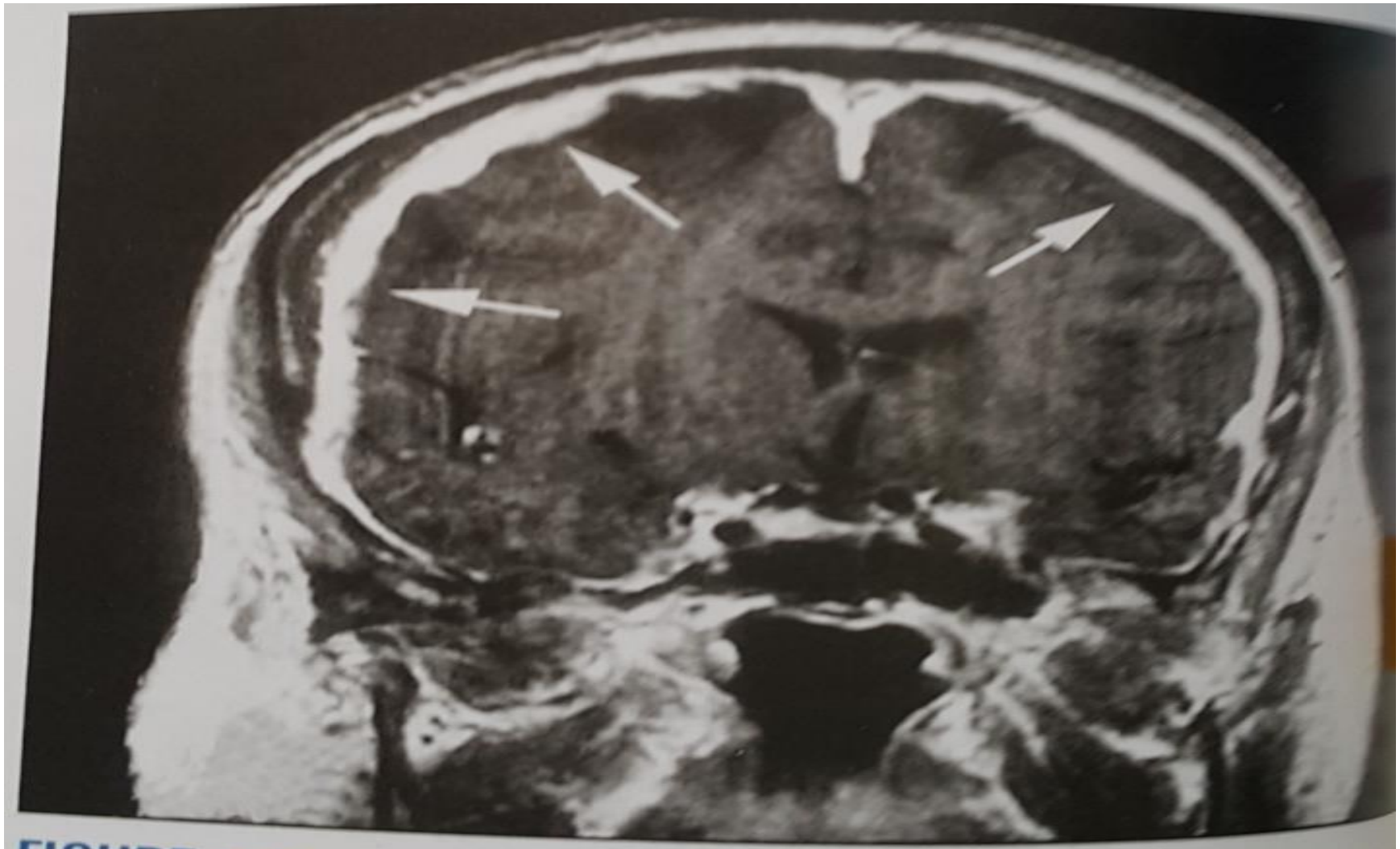


FIGURE 10-5. Intracranial hypotension. Gadolinium-enhanced MRI scan of a patient with intracranial hypotension. There is widespread, symmetric meningeal enhancement (*arrows*).

The definitive diagnosis is made with an LP showing an opening pressure below 60 mm CSF

If a patient remains symptomatic from intracranial hypotension, a blood patch may be attempted to cover the dural leak(if it can be found)

6. Medication causes

Medications associated with headaches are numerous

They range from hormonal therapies, including contraceptives, to nitric oxide

Withdrawal of a medication or other treatment may also cause a headache; the most common example is a caffeine withdrawal headache

Headache semiology is nonspecific, but the temporal association to medication change helps establish the diagnosis

Medication overuse headache (MOH), also referred to as rebound headache or drug-induced headache, is a chronic headache occurring in patients with a primary headache disorder

The baseline headache disorder is typically markedly exacerbated i.e., more frequent, severe, or both) when the medication is overused

MOH is diagnosed when patients use an abortive therapy 10 or more times per month for more than 3 months with an increase in headaches

VI) Head and neck disorders

There are many different structural disorders that can cause headache

Treatments are based on the specific causes identified, and these disorders may occur simultaneously with other primary or secondary headache disorders

Sinusitis is commonly associated with a headache

It may be bifrontal or unilateral

Acute sinusitis is often associated with other symptoms of a respiratory tract infection

Temporomandibular joint disorder (TMD) is another common cause of headache and may be unilateral or bilateral

On exam, there may be evidence of dental wearing(chipped and flattened teeth) and discomfort on palpation of the joint

Cervicogenic headaches are also common and may be identified by palpation of myofascial trigger points in the neck

Cervical range of motion is often reduced

VII) Trigeminal neuralgia

The pain of trigeminal neuralgia is shock-like, occurring in one or all branches of the trigeminal nerve

The pain is usually paroxysmal and recurrent

It may be triggered by common activities such as brushing hair or teeth

It may be idiopathic or due to structural causes such as a mass or vascular lesion, or a demyelinating lesion of multiple sclerosis

Carbamazepine is a common first-line treatment

VIII) SUMMARY

Migraines, tension headaches, and TACs are the 3 most common types of primary headache disorders

The International Headache Society maintains an evidence-based categorization of primary and secondary headache disorders

If a headache history and pattern is not consistent with a primary headache disorder, the clinician should consider a secondary headache disorder, review the rare types of primary headache disorders, or consider that the headache may be a combination of more than one headache disorder