Chapter 7
Diagnosis of Major Secondary Headaches, Nonvascular Disorders

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Introduction

The second chapter on diagnosis of secondary headaches includes nonvascular disorders.

The organization of the chapter is by unrelated secondary disorders, linked by their propensity to cause headache, and their ICHD-3 classification are listed in Table 7.1.

Secondary Idiopathic Intracranial Hypertension (Pseudotumor Cerebri)

Normal cerebrospinal fluid (CSF) pressure ranges from 70 to 250 mm of H_2O. Elevated intracranial hypertension may be idiopathic or due to secondary causes. Secondary causes for increased intracranial pressure (ICP) are listed in Table 7.2.

Once secondary causes of raised ICP are excluded, the diagnosis of idiopathic intracranial hypertension (IIH, pseudotumor cerebri) can be made on the basis of headache, documented raised ICP, and headache either developing along with the increased pressure and/or reported improvement of headache with lowering of ICP following removal CSF with lumbar puncture (LP). Although once considered the hallmark of IIH, evidence of papilledema is no longer a necessary criterion, given

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Table 7.1 ICHD-3 organization of secondary nonvascular headache disorders

1. Increased CSF pressure (idiopathic intracranial hypertension, IIH)
2. Low CSF pressure
3. Brain tumor
4. Infections such as HIV
5. Chiari malformation type 1
6. Homeostasis disorders
7. Toxic substances
8. Cervicogenic
9. Temporomandibular disorder
10. Trigeminal neuralgia and painful trigeminal neuropathies
11. Other cranial neuralgias

Table 7.2 Secondary causes of intracranial hypertension

- Venous sinus thrombosis
- Mass lesion/cerebral edema
- Meningitis
- Radical neck dissection
- Hypothyroidism/hypoparathyroidism
- Vitamin A intoxication/deficiency
- Renal disease
- Obesity
- Anemia from iron deficiency
- Drugs (tetracycline, minocycline, tretinoins, human growth hormone, corticosteroid withdrawal, oral contraceptives, lithium)

Table 7.3 Idiopathic intracranial hypertension (IIH, pseudotumor cerebri), ICHD-3 criteria

1. Diagnosed by CSF pressure greater than 250 mm CSF with LP in a lateral decubitus position without sedation, or by monitoring
2. ≥2 of:
   a. Headache developed with IIH
   b. Headache is relieved by decreasing intracranial CSF pressure
   c. Headache is worse with increased CSF pressure

the fact that cases of IHH without papilledema does infrequently occur. The finding of papilledema on examination still remains strong supporting evidence for the diagnosis.

As noted above, IIH was previously referred to as benign intracranial hypertension as well as pseudotumor cerebri. Diagnostic criteria by the ICHD-3 for IIH are listed in Table 7.3.

The disorder tends to affect obese females (body mass index >30). Patients most often report a constant, daily, pressure-like headache pain that may be frontal, retro-orbital or diffuse in location, and at least moderate in severity. The headache is aggravated by Valsalva-type maneuvers. Other signs and symptoms include papilledema as well as cranial nerve dysfunction. It is not uncommon for the patient

Table 7.4 MRI findings suggestive of idiopathic intracranial hypertension (IIH, pseudotumor cerebri)

- Empty sella turcica or flattening of pituitary gland
- Distension of the optic nerve sheaths
- Vertical tortuosity of the optic nerves
- Flattening of posterior globes
- Protrusion of the optic nerve heads
- Transverse cerebral venous sinus stenosis

Table 7.5 Clinical pearls for diagnosing idiopathic intracranial hypertension (IIH, pseudotumor cerebri)

- Obese women, age 20–50
- Dull, constant, daily, nonthrobbing headache
- Papilledema
- Diplopia
- Transient visual obscurations (TVOs)
- Tinnitus
- Neck or back pain
- Enlarged blind spot
- Shoulder and arm pain
- Unusual noises in the head can be heard by patient; sometimes bruits by examiner
- Empty sella or normal MRI

The clinical pearl for IIH diagnosis:

- *The diagnosis of IIH cannot be made without an LP done in the lateral decubitus position*!

to report visual changes such as blurring or transient visual obscurations (TVOs). Diplopia related to cranial nerve VI palsy and pulsatile tinnitus are additional common complaints. Persistently elevated CSF pressures can lead to permanent visual loss.

The patient should be evaluated with magnetic resonance imaging (MRI) and magnetic resonance venography (MRV) to rule out venous thrombosis, which as noted, is the most common secondary cause other than obesity. Other findings suggestive of IIH can be seen on MRI as listed in Table 7.4. A neuro-ophthalmologic examination, including visual field testing, is required to monitor visual acuity. An LP is necessary to document raised ICP. *The diagnosis of IIH cannot be made without an LP performed in the lateral decubitus position*! (Table 7.5).

An opening pressure of greater than 250 mm H_2O in adults, and greater than 280 mm H_2O in children is confirmatory of the diagnosis. The previous ICHD-2 classification allowed for the diagnosis of IIH if the opening pressure was greater than 200 mm H_2O in nonobese individuals, but this criterion has been changed in the ICHD-3 criteria of 2013. Patients respond favorably after the withdrawal of CSF, but unfortunately the response is short lasting, and further treatment will be described in Chap. 17.

Table 7.6 Common clinical manifestations of intracranial hypotension

- Headache that worsens immediately after assuming the upright position and improves within a minute of lying down; delayed responses to postural changes are also possible
- Headache is bilateral, throbbing, located frontally, or occipitally
- Tinnitus
- Impairment in hearing (muffled, echoed, ear fullness)
- Photophobia
- Nausea, vomiting
- Vertigo, dizziness
- Pain and stiffness in the neck, interscapular region, arm
- Cranial nerve dysfunction (commonly horizontal diplopia from impaired function of CN VI, III, or the MLF)
- Gait imbalance
- Anorexia
- Blurry vision
- Phonophobia, hyperacusis, change in hearing
- Facial numbness
- Galactorrhea
- General malaise

CN cranial nerve, *MLF* medial longitudinal fasciculus

Low Cerebrospinal Fluid Pressure Headache

Headache caused by low CSF pressure is either the result of a previous LP, a CSF fistula, or idiopathic in etiology. The clinical manifestations are similar despite the etiology of the intracranial hypotension (Table 7.6).

Classically, patients report a headache in the upright position with relief of symptoms when recumbent. CSF opening pressure is measured at below 60 mmHg H_2O. Studies of the CSF may reveal a normal to slightly elevated protein level and even a mild lymphocytic pleocytosis.

Approximately a third of patients will develop headache following LP. The postdural puncture headache (previously termed postlumbar puncture headache) generally occurs within 5 days after the dural puncture. Spontaneous improvement typically occurs within 2 weeks of the onset of symptoms. In cases without spontaneous improvement, an epidural lumbar blood patch can provide prompt relief.

The symptoms are most likely related to a persistent dural tear caused by the LP needle, resulting in fistula formation. Female gender, younger age (31–50 years), and prior history of postdural puncture headache are risk factors.

Methods to try to reduce the risk of postdural puncture headache include inserting the LP needle bevel parallel to the longitudinal axis of the dural fibers, using a smaller needle size, replacement of the stylet before the needle is withdrawn, and using noncutting needles such as the Sprotte needle. The duration of recumbency following an LP or the recommendation to increase fluids does not seem to influence the occurrence of postdural puncture headache.

Idiopathic low CSF pressure headache or CSF fistula headache also produces symptoms of low-pressure headache, although the response to positional changes

Table 7.7 Neuroimaging findings and low CSF pressure headache

Computed tomography
• Subdural hematomas, hygromas
Radioisotope cisternography
• No evidence of radioactivity beyond the basal cisterns with a paucity or absence over the cerebral convexities
• Parathecal radioactivity episode of CSF leak
• Early (<4 hours) appearance of radioactivity in the kidneys and bladder
MRI brain
• Diffuse pachymeningeal enhancement
• Descent or "sagging" of the brain (cerebellar tonsils herniation, crowding of the posterior fossa, obliteration of the prepontine or perichiasmatic cisterns)
• Flattening of the optic chiasm
• Enlargement of the pituitary
• Subdural hematomas or hygromas
• Ventricular collapse
• Engorgement of cerebral venous sinuses
MRI spine/MR myelography/CT myelography
• Extra-arachnoid fluid collection
• Extradural extravasation of fluid/contrast
• Spinal pachymeningitis/paraspinal enhancement
• Engorgement of the spinal venous plexus
• Meningeal diverticula/dilated nerve root sleeves
• Contrast extravasation of a single nerve root

is less impressive than with postdural puncture headache. If the headache develops into a chronic condition, the classical features of orthostatic headache often diminish and may even be present in the lying position.

In the case of a CSF fistula, there is sometimes a known trauma or iatrogenic cause such as a neurosurgical procedure. More commonly, fistulas may occur spontaneously without a known precipitating event, as in the case of idiopathic low CSF pressure headache. Spontaneous CSF leaks are most commonly identified in the cervical or thoracic region.

An MRI of the brain with and without gadolinium is often diagnostic of low CSF pressure headache, demonstrating evidence of brain sag and diffuse pachymeningeal enhancement without evidence of leptomeningeal involvement. Other imaging findings are listed in Table 7.7.

Unfortunately, despite the number of diagnostic imaging studies which can be utilized, finding the actual site of the leak is often quite difficult and in some cases impossible. Radioisotope cisternography is no longer recommended given poor sensitivity and more useful modalities such as MRI with fat suppression and computed tomography (CT) myelography. CT myelography may be the most reliable diagnostic approach to utilize.

Headache Attributed to Intracranial Neoplasm

Headache may be the initial presentation in approximately 20% of patients with brain tumors. The incidence of headache increases to 50–70% of patients later in the course of their illness.

Most individuals with an underlying brain tumor who present with headache will also have other focal neurologic symptoms such as seizures, confusion, or hemiparesis. Brain tumor headache is characterized as progressive, diffuse, nonpulsating, and associated with nausea and/or vomiting. The headache may be constant or intermittent. The headache worsens with physical activity, Valsalva-type maneuvers, and tends to be most severe in the morning and after napping.

Both mass effect of the tumor and hydrocephalus contribute to the headache, causing local pressure and/or traction on pain-sensitive structures of the brain. Headache is more frequent with infratentorial tumors than supratentorial tumors. Finally, patients with primary headache disorders before developing a brain tumor will often have some features of their preexisting headaches with their brain tumor headache.

Headache Attributed to Infectious Diseases

Any underlying infection may produce a headache or worsen a preexisting primary headache condition. The infection may be systemic or intracranial. Patients with headache related to systemic infection generally have fever, malaise, and diffuse myalgias.

Headache is common in HIV-infected patients at any stage of the illness and has been noted to occur with HIV seroconversion related to primary infection. Later in HIV illness, any presentation of headache or change in pattern of headache should be assumed to be secondary (Table 7.8).

Intracranial infections are most often bacterial or viral, but various opportunistic infections may occur, particularly in immunosuppressed patients. In general, the greatest risk for opportunistic infections is in HIV patients with CD4 counts below 200 cells/mm^3, and those with CD4 counts greater than 500 cells/mm^3 are not considered to be at risk. Evaluation for intracranial infections should be performed in individuals presenting with new-onset or worsening headache associated with fever, meningismus, altered mentation, or focal neurologic deficits.

Headaches associated with infection can be caused by meningitis, encephalitis, brain abscess, or subdural empyema. Antibiotic therapy should be initiated immediately if there is concern for intracranial infection, after which the clinician can proceed with diagnostic testing with urgent CT, LP, and MRI.

The headache attributed to infection should resolve within 3 months of successful treatment. A *persistent* headache pattern may develop in up to 1/3 of patients following a past episode of meningitis despite adequate treatment. Another subset,

Table 7.8 The clinical pearl on HIV headache

• *Any presentation of headache or change in pattern of headache in HIV-positive patients should be assumed to be secondary*

chronic headache attributed to infection, refers to headaches lasting for more than 3 months when the underlying infection remains active.

Headache Attributed to Chiari Malformation Type I

Chiari malformation type 1 (CM1) is most often congenital although acquired cases may occur, most commonly as a result of intracranial hypotension, excessive CSF drainage or injury. CM1 is diagnosed on MRI if there is greater than a 5-mm inferior displacement of the cerebellar tonsils below the foramen magnum; only 3 mm is required if there is associated crowding of the subarachnoid space at the craniocervical junction as evidenced by obstruction of CSF flow seen on MRI CINE flow studies.

Patients with CM1 commonly report headache along with a number of other symptoms related to compression of the cerebellum, brainstem, and cervical cord (Tables 7.9 and 7.10). It is important to remember that not all patients with evidence of CM1 on imaging are symptomatic.

Commonly associated with this condition are tethered cord, IIH, syringomyelia, and scoliosis. The headache is located occipitally and is brief in duration, lasting less than 5 min. It is often triggered by Valsalva-type maneuvers. This secondary cough headache is further discussed with primary cough headache in Chapter 3. Although a patient's symptoms can be effectively treated with various medications, especially indomethacin, suboccipital decompression surgery may be indicated for those with headache with significant neurological signs and symptoms.

Headaches Associated with Disorders of Homeostasis

There are a number of systemic disorders and metabolic conditions frequently associated with headache (Table 7.11). The patient will exhibit signs and symptoms related to the underlying condition in addition to the headache. Diagnostic testing is required to confirm the diagnosis. Upon treatment of the underlying condition, the headache will resolve.

Table 7.9 Headache attributed to Chiari malformation type 1, ICHD-3 criteria

A. Diagnosis of Chiari malformation type 1 by imaging
B. ≥2 of:
a. History consistent with >1 of:
i. Headache started with CM1
ii. Headache stopped within 3 months after successful treatment of CM1
b. Headache has ≥1 of:
i. Triggered by Valsalva, such as cough
ii. Posterior location
iii. Duration less than 5 min
C. Headache occurs along with other symptoms or signs of posterior fossa or cervical spinal cord dysfunction

Table 7.10 Commonly reported symptoms in patients with Chiari malformation type I

• Occipital or suboccipital headache induced by cough or Valsalva maneuver (secondary cough headache)
• Dizziness, vertigo, disequilibrium, impaired coordination
• Ears: pressure, loss of hearing, hyperacusis, tinnitus
• Eyes: nystagmus, oscillopsia, photopsia, visual blurring, visual field deficits, diplopia
• Dysphagia, hoarseness
• Nausea and vomiting
• Neck pain
• Muscle weakness
• Numbness and paresthesias of extremities
• Insomnia, sleep apnea
• Depression

Toxic Headaches

A number of substances may produce headache either due to exposure or withdrawal (Table 7.12). Typically, once the exposure ends, the headache resolves. Headache is a commonly listed adverse effect of multiple medications. Therefore, a review of the patient's list of medications noting their start date can be helpful in pinpointing any correlation with the headache.

Cervicogenic Headache

Headache may be a referred pain originating from the neck. This type of headache must be distinguished clinically from those patients with neck pain as an associated symptom of a primary headache disorder.

Patients at risk for cervicogenic headache include those with a history of arthritis with known cervical spondylosis and degenerative disc disease, or those with a history of neck trauma, particularly whiplash-type injuries. An examination may

Table 7.11 Headaches related to disorders of homeostasis, ICHD-3

- Headache secondary to hypoxia/hypercapnia
  - High altitude headache
  - Headache attributed to airplane travel
  - Diving headache
  - Sleep apnea headache
- Dialysis headache
- Headache secondary to arterial hypertension
  - Headache associated to pheochromocytoma
  - Headache attributed to hypertensive crisis without hypertensive encephalopathy
  - Headache attributed to hypertensive encephalopathy
  - Headache attributed to preeclampsia oreclampsia
  - Headache attributed to autonomic dysreflexia
- Headache associated with hypothyroidism
- Fasting headache
- Cardiac cephalgia

Table 7.12 Some of the substances known to provoke headache

- Nitric oxide donor (nitroglycerin, nitrates, and nitrites of cured meats)
- Phosphodiesterase inhibitor (e. g., sildenafil, vardenafil for erectile dysfunction)
- Carbon monoxide
- Alcohol
- Food components and additives (MSG, aspartame, tyramine)
- Cocaine
- Cannabis
- Histamine induced
- Calcitonin gene-related peptide (CGRP)
- Medications including herbal remedies

MSG monosodium glutamate

reveal tenderness, muscle spasm of the cervical paraspinal and neck muscles, and limitations in cervical range of motion.

Cervical myofascial pain alone without evidence of degenerative changes in the cervical spine should be diagnosed as tension-type headache. Degenerative change in the spine is a very common finding in individuals without symptoms of headache and neck pain, and therefore this finding in isolation cannot be used for definitive diagnosis of cervicogenic headache.

The pain is most often unilateral, typically starts in the occipital region, and radiates frontally. The unilaterality must be stressed as a key clinical symptom, along with the primary neck pain complaint, and the report that neck movement precipitates or aggravates the pain. Even when pain is reported bilaterally, there tends to be a one-sided predominance.

Relief after cervical anesthetic blockade can confirm the diagnosis. The head pain likely originates from stimulation of the upper cervical roots leading to the

Table 7.13 Cervicogenic headache, ICHD-3 criteria

A. Clinical, lab, or imaging evidence for a lesion of the cervical spine or cervical neck tissues known to cause headache
B. Proof of causation with ≥2 of:
a. Headache developed with the onset of the neck lesion
b. Headache improved with treatment of the neck lesion
c. Headache is made worse by provocative neck maneuvers, and neck range of motion is reduced
d. Headache is abolished by diagnostic cervical blocks

Table 7.14 Clinical pearls on cervicogenic headache

• *Symptoms*: must have neck pain as a key complaint and must not fit ICHD-3 beta criteria for migraine or hemicrania continua. There should be no autonomic features, and usually no migrainous features of photophobia, phonophobia, or nausea
• *Risk factors*: arthritis, trauma to neck, whiplash injury
• *Pain*: unilateral (key!), occipital, frontal
• *Triggers*: movement of neck (key!), coughing, sneezing, pressure on upper cervical or occipital region, prolonged upright position
• *Exam*: cervical range of motion limitations, awkward head position
• Imaging evidence of a disorder or lesion within the spine or muscles of the neck
• Abolished by diagnostic cervical blockade

activation of the trigeminal nucleus caudalis located within the upper segment of the cervical spinal cord.

The ICHD-3 criteria for cervicogenic headache are somewhat limited in utility. They are included in Table 7.13, but clinically useful pearls follow in Table 7.14.

Temporomandibular Disorder

Temporomandibular joint (TMJ) dysfunction is a fairly common problem. Patients may present with headache which is localized to the preauricular region, mandible, masseter, and temporal region. In addition to frontotemporal headache, patients often complain of otalgia, tinnitus, and dizziness. Clinical history may elicit symptoms of bruxism during sleep and reported jaw locking or popping. Limited jaw opening and tenderness of the masticatory muscles may be noted during examination. TMJ dysfunction leads to myofascial pain contributing to the symptoms of headache. Symptoms are often self-limited, but in persistent cases, referral to a TMJ specialist may help to correct the problem.

Headache attributed to TMJ disorder should have pain in conjunction with the development of the disorder and should remit as the problem is treated. If this is still uncertain, provocative maneuvers including active or passive movement of the jaw should be able to provoke the headache. ICHD-3 beta points out that there is an overlap between this disorder and tension-type headache, and when there is uncertainty about TMJ dysfunction as a cause, coding should lean toward the diagnosis of tension-type headache.

Table 7.15 Trigeminal neuralgia/trigeminal neuropathy, ICHD-3 classification system

I. Classical trigeminal neuralgia
a. Classical trigeminal neuralgia, purely paroxysmal
b. Classical trigeminal neuralgia, with concomitant persistent facial pain
II. Painful trigeminal neuropathy
a. Painful trigeminal neuropathy attributed to acute herpes zoster
b. Postherpetic trigeminal neuropathy
c. Painful posttraumatic trigeminal neuropathy
d. Painful trigeminal neuropathy attributed to multiple sclerosis (MS) plaque
e. Painful trigeminal neuropathy attributed to space-occupying lesion, e.g., neoplasm
f. Painful trigeminal neuropathy attributed to some other disorder

Trigeminal Neuralgia

Trigeminal neuralgia (TN) is a disorder involving one or more of the sensory divisions of the trigeminal nerve that often produces brief but severe lancinating pain. The disorder was previously referred to tic douloureux and typically affects older individuals in its classical form.

TN used to be divided into classical or symptomatic subtypes. The 2013 ICHD-3 beta dropped the symptomatic form, and TN is divided into two broad categories, classical TN and what is really not TN and is now called painful trigeminal *neuropathy* (PTN; see Table 7.15).

Classical TN is further subdivided into a purely paroxysmal form, with lightning-like, electric shock-like pains lasting seconds, and a form with concomitant persistent facial pain of moderate intensity in the same affected area, usually the second and third division of the trigeminal nerve (V2, 3). Classical TN is often related to neurovascular compression of the trigeminal nerve root near the dorsal root entry zone, usually by the superior cerebellar artery.

PTN encompasses the trigeminal neuropathic pain syndromes caused by other disorders such as multiple sclerosis, postherpetic neuralgia, trauma, and that caused by a tumor or lesion. Thus, all TN is really secondary.

Diagnosis of Classical Trigeminal Neuralgia

The diagnosis of TN requires recognition of the well-described, excruciating lightning-like paroxysms of pain in one or more of the divisions of the trigeminal nerve, with triggers, without radiation, without autonomic features, and with latency periods. Pain is brief but tends to have successive recurrences, with refractory periods.

The location of TN is in V2 and V3; < 5 % of TN is located in V1. Bilateral cases are rare, except for PTN related to multiple sclerosis.

Chewing, talking, or touching the face may trigger pain, although paroxysms can occur spontaneously as well. The triggers are characteristically described as innocuous, often occurring in a stereotypical location, and are sometimes so severe

Table 7.16 Classical trigeminal neuralgia, ICHD-3 overall diagnostic criteria

I.	Pain is strictly located in ≥one branch of V, with no additional radiation
II.	≥3 of:
	a. Paroxysms of pain lasting from less than 1 sec to 2 min
	b. Severe
	c. Quality is electric shock-like, shooting, stabbing, sharp
	d. Provoked by triggers of innocuous stimuli to the ipsilateral face

Table 7.17 Clinical features of classical trigeminal neuralgia

- Unilateral facial pain limited to the distribution of the trigeminal nerve (mandibular (V3) or maxillary (V2) divisions > ophthalmic (V1) division
- Affects older patients, > age 50 years
- Women > men
- Attacks are of brief, less than a second to 2 min in duration
- Pain often provoked by triggers, but after repeated triggers, there is often a refractory period
- Constant, dull pain can develop between bouts of acute pain
- Rarely occurs during sleep
- Attacks become more common over time
- Remissions are possible
- Neurologic examination is normal except in cases in which there is an underlying lesion
- No autonomic features

that patients stop eating and lose weight. After repeated triggering, there is often a refractory period of relief in TN.

The ICHD-3 criteria for the diagnosis of TN overall are listed in Table 7.16.

From a diagnostic standpoint, the differential will be between TN and primary stabbing headaches, as well as the shorter trigeminal autonomic cephalalgias (TACs), such as paroxysmal hemicrania (PH) and short-lasting unilateral neuralgiform headache attacks (SUNHAs). Table 7.17 lists the major clinical features of TN and Table 7.18 some pearls on distinguishing TN from the short-lasting TACs and primary stabbing headaches. Rarely, the short-lasting TACs and TN can occur in the same individual, requiring separate concomitant treatments.

As noted, classical TN comes in two forms, a purely paroxysmal form and a form with the shock-like paroxysms, but a persistent moderate facial pain in between the stabs. The ICHD-3 criteria for these two forms, simply distinguished by the absence or presence of the continuous facial pain, are listed in Table 7.19.

PTN, formerly called symptomatic TN, is the result of an underlying structural lesion (Table 7.20). Common secondary causes include herpes zoster, multiple sclerosis, aneurysms, syringomyelia, post medullary infarction, sarcoidosis, and various tumors including meningiomas, schwannomas/acoustic neuromas, cholesteatomas, epidermoids, and metastases. PTN can also be posttraumatic. The ICHD-3 types of PTN are listed in Table 7.21.

Clinical features of PTN are listed in Table 7.22 and clinical pearls on diagnosing classical TN versus PTN are listed in Table 7.23. Patients should undergo an MRI/MRA of the brain with and without gadolinium to determine if they have a lesion where repair or treatment could lead to cure.

Table 7.18 Clinical pearls on distinguishing trigeminal neuralgia (TN) from short-lasting TACs, such as paroxysmal hemicrania or SUNHA, and primary stabbing headaches

- Location:
  - TN is in V2–V3
  - Paroxysmal hemicrania (PH) attacks are in V1, with some ear symptoms
  - SUNHAs are in V1, with some ear symptoms
  - Primary stabbing headaches can occur anywhere on the head, including out of a trigeminal distribution and tend to vary and migrate
- Duration:
  - TN attacks are less than a second to 2 min
  - PH attacks have a mean duration of 14 min and do not overlap with TN
  - SUNHA lasts from 1 to 600 s and can overlap with TN
  - Primary stabbing headaches last for a few seconds and can overlap with TN
- Autonomic features
  - TN has no autonomic features
  - PH and SUNHA have autonomic features, and both have, as diagnostic features, a "sensation of fullness in the ear"
  - Primary stabbing headaches have no autonomic features
- Triggers
  - TN has triggers and trigger zones
  - PH and SUNHA also can have triggers, but without a refractory period. The triggers can overlap or coexist with TN
  - Primary stabbing headaches do not have triggers
- Treatment
  - TN responds to antiepilepsy drugs (AEDs), carbamazepine, oxcarbazepine, lamotigine, gabapentin, and baclofen
  - PH is an indomethacin-responsive syndrome
  - SUNHA responds to AEDs such as gabapentin, and lamotrigine, and so can overlap with TN
  - Primary stabbing headaches, when they occur in volleys, are often indomethacin responsive
- Primary stabbing headaches:
  - These ice-pick pains tend to occur in single jabs, occur irregularly across time, can occur outside a trigeminal distribution, and do not have triggers
  - They occur in 40 % of migraineurs
  - They can herald attacks of migraine
  - Volleys of ice-pick pains are indomethacin responsive
- SUNHA:
  - The autonomic features are key, as is the V1 location
  - SUNHA occurs as single stabs, series of stabs, or in a sawtooth pattern. The latter would distinguish from TN. The single stab can herald a longer attack with autonomic features, which would distinguish from TN
  - SUNHA and PH have, as a diagnostic feature, a "sensation of fullness in the ear." TN does not
- Hemicrania continua:
  - The exacerbations in HC tend to be long, not lightning-like, so the differential from TN with concomitant persistent facial pain is straightforward, especially because HC is indomethacin responsive and TN is not

Table 7.19 The two forms of classical trigeminal neuralgia, ICHD-3 criteria

I.	Classical trigeminal neuralgia, purely paroxysmal
	a. Meets criteria for classical TN
	b. No interictal persistent facial pain
II.	Classical trigeminal neuralgia with concomitant persistent facial pain
	a. Meets criteria for classical TN
	b. Persistent moderate intensity facial pain in the affected area interictally

Table 7.20 Summary of the differences between trigeminal neuralgia (TN) and the pain trigeminal neuropathies (PTN)

- Classical TN: typically caused by a neurovascular anomaly resulting in compression of the trigeminal nerve
- Painful trigeminal neuropathies (PTN, formerly called symptomatic TN): caused by an underlying structural lesion (e.g., herpes, trauma, MS, or space-occupying lesion such as neoplasm)
- Both have a clinical response to carbamazepine/oxcarbazepine. Response to medication does not determine diagnosis

Table 7.21 Painful trigeminal neuropathies (PTN), ICHD-3 criteria

I.	Painful trigeminal neuropathy attributed to acute herpes zoster
II.	Postherpetic trigeminal neuropathy
III.	Painful trigeminal neuropathy attributed to multiple sclerosis (MS) plaque
IV.	Painful trigeminal neuropathy attributed to space-occupying lesion
V.	Painful trigeminal neuropathy attributed to another disorder

Table 7.22 Clinical findings suggestive of painful trigeminal neuropathy (PTN), ICHD-3 criteria (formerly known as symptomatic trigeminal neuralgia)

- Bilateral pain
- Neurologic abnormalities: sensory loss, masticatory weakness
- Pain in the ophthalmic division (V1)
- Onset below the age of 50 years
- Unresponsiveness to medical treatment
- Abnormal trigeminal reflex testing

Table 7.23 Clinical pearls on diagnosing classical trigeminal neuralgia (TN) versus painful trigeminal neuropathy (PTN)

- In young patients, PTN is far more common, and MS should be considered in the differential diagnosis
- In older patients, classical TN is more likely and is usually related to neurovascular compression, typically from the superior cerebellar artery overlying a trigeminal root
- If a patient has pain in V1, it is probably not TN
- If a patient does not have triggers, it is probably not TN
- If a patient does not have refractory periods, it is probably not TN
- If a patient has autonomic features, first consider one of the trigeminal autonomic cephalalgias
- If a patient has continuous pain without lancinating paroxysms, it is not TN

Table 7.24 Other facial neuralgias

Classification	Clinical features
Persistent idiopathic facial pain (atypical facial pain)	Pain: bilateral, > 2 hrs/day or constant dull ache or nagging pain Location: may involve entire face May be seen as part of a more diffuse chronic pain syndrome Triggers: stress or chronic pain syndrome Age: < 50
Glossopharyngeal neuralgia	Pain: paroxysmal, unilateral, jabbing Location: angle of jaw, base of tongue, tonsillar fossa, or ear Triggers: swallowing, talking, coughing Age: tends to be younger than classic TN Bilateral cases do occur May have TN as well Syncope, bradycardia, and asystole (especially with glossopharyngeal–vagal neuralgia) Pathology: vascular compression
Nervus intermedius neuralgia	Pain: a brief stabbing pain or long duration Location: deep within the internal auditory canal Trigger point: located within posterior wall of auditory canal
Postherpetic trigeminal neuropathy	Known herpetic eruption or CSF varicella zoster virus detected Pain: constant, severe, burning Associated hyperpathia Location: follows dermatomal distribution of prior skin eruption V1 most common trigeminal distribution

Other Facial Neuralgias

There are a number of other facial pain syndromes and neuralgias which are listed in Table 7.24. Many of these neuralgias have very specific diagnostic features, such as the swallowing trigger of glossopharyngeal neuralgia and the deep ear pain location for nervus intermedius neuralgia. A careful imaging workup for these rare neuralgias looking for secondary causes such as neoplasm is always mandatory.

Patients who do not fit a typical pattern of TN and for whom no secondary causes could be found were previously referred to as having atypical facial pain and are now said to have *persistent idiopathic facial pain*. They tend to describe the pain as being more diffuse, often bilateral, and more constant in nature.

These individuals tend to be younger in age and often were believed to have underlying psychiatric illness due to the fact that stress can exacerbate the condition. Atypical facial pain may also be part of a more diffuse chronic pain syndrome that involves other parts of the body.

Conclusions on Secondary Nonvascular Headaches

- IIH is often a disease of obese women, aged 20–50
- The diagnosis of IIH cannot be made without an LP
- Any presentation of headache or change in pattern of headache in HIV-positive patients should be assumed to be secondary
- Low CSF pressure headache is confirmed with an MRI without and with contrast that shows pachymeningeal enhancement, sometimes with brain sag
- Classic TN is a disease of the elderly and generally due to a vascular anomaly; PTN is a disease of younger patients and often due to MS

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