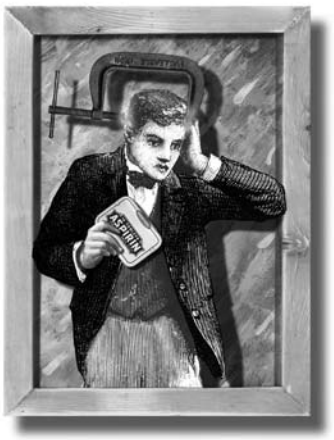

Headache as a manifestation of otolaryngologic disease

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Headache can be caused by a multitude of factors, but experienced physicians accustomed to treating patients with headache are adept at making an accurate diagnosis. Occasionally, however, a patient has an unusual presentation of headache or facial pain. In these cases, it can be difficult to classify the etiology of the headache despite the performance of a thorough physical examination and the acquisition of appropriate diagnostic tests. Awareness of some of the otolaryngologic diseases that can manifest as facial pain or headache may help the physician better diagnose and treat this complex problem.

(Key words: headache, atypical facial pain, otolaryngologic disease, sinusitis, neuralgia, temporomandibular joint disorders, arteritis)

The otolaryngologist is not excluded from seeing patients with complaints of chronic headaches or facial pain. Headache is a constituent of many otolaryngologic ailments, generally resulting from one of four basic mechanisms: traction, distension, inflammation, or direct pressure. Headaches resulting from diseases directly affecting structures of the head and neck are described in this article.

Patient evaluation

History

The physician must bear in mind that some headaches signal serious and urgent conditions and that a properly obtained history will usually lead to the correct diagnosis. It is recommended that information be obtained about the following:

☐ the characteristics of the pain;
☐ duration of headache, associated signs and symptoms;
☐ rate and mode of onset;
☐ presence of trigger points or provocation by facial or jaw movement;
☐ frequency;
☐ site;
☐ status of the overall condition, whether it is getting better, worse, or the same;
☐ precipitating factors;
☐ family history; and
☐ measures that provide relief.

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Examination

Initial assessment should include evaluation of the patient's mood, affect, and speech, and measurement of the patient's height, weight, and blood pressure. The scalp and skull should be examined, and, in younger patients, the head circumference should be measured and plotted. Palpation and percussion of the head and auscultation for bruits over the head, eyes, and neck should be done. Passive and active flexion, extension, and rotation of the neck must be assessed. A full cranial nerve and ophthalmologic examination is done. The motor, reflexes, sensory, and cerebellar systems should be examined and attention paid to inspection and palpation of the spine. The skin should also be examined, particularly for any midline skin defects, neurocutaneous lesions, or unusual pigmented lesions. Ear, nose, and throat structures, including dentition and lymphatics, should be assessed. Limited or asymmetric jaw movement or temporomandibular joint (TMJ) tenderness, as well as enlarged or tender temporal scalp arteries should be noted. Trapezius or posterior cervical trigger points or small vesicular eruptions in the distribution of CN V, including the external ear or portions of the hair-bearing scalp, must be identified.

Significant deviations in stature can indicate hypothalamic or pituitary disorders. Head enlargement can be familial or raise the possibility of hydrocephalus or occult tumor. Vascular anomalies can produce asymmetric bruits. Localized tenderness about the scalp, face, or neck can indicate trauma or local inflammation. Papilledema, retinal hemorrhage, or pallor of the optic disks can be clues to serious central nervous system (CNS) disease. Neck stiffness suggests meningeal inflammation and sometimes early cerebellar tonsillar herniation. Focal neurologic signs can signify a localized intracranial pathologic lesion. Posterior fossa disease or hydrocephalus can cause cerebellar ataxia. Examination of the skin can reveal hyperpigmented or hypopigmented areas, skin cancer, trauma, or midline lesions.

Laboratory tests

The physician must be selective and order laboratory tests with high diagnostic yield. Computed tomography of the head is the test of choice for excluding most structural or space-occupying lesions. Magnetic resonance imaging (MRI) is used when images of deeper soft tissue structures of the brain or spinal cord are desired.

Simple laboratory tests such as those listed in *Figure 1* are helpful in some cases. Lumbar puncture with analysis of spinal fluid and recording of pressure is the test of choice in determining the presence of CNS infection. Cerebrospinal fluid should be sent for the analyses that are listed in *Figure 2*.

Clinical syndromes

Clinical syndromes of otolaryngologic diseases that can manifest as facial pain or headache are outlined in *Figure 3* and described in the following text. Treatment options are suggested, as well.

Atypical facial pain

Atypical facial pain is described as a chronic burning or aching pain without focal findings or any discernible etiology and is considered a diagnosis of exclusion. There appears to be no trigger point that initiates the pain cycle. Rather, the pain is characteristically bilateral and changes locations frequently over weeks to months. The intensity fluctuates slowly over time and is rarely entirely absent. Typically located in the face, the pain can spread to the cranium. Women are more commonly affected than men in an age group from 30 to 50 years.[1] Significant psychiatric findings, most commonly depression, somatization, or adjustment disorders, are found in 60% to 70% of the patients.[1] Psychiatric evaluation is recommended. Treatment consists of antidepressants. Many of these patients become the victim of iatrogenic problems resulting from multiple invasive evaluations and excessive medication. Patients with atypical facial pain respond less frequently to carbamazepine and neurosurgical decompression of vascular structures than do patients in whom trigeminal neuralgia is diagnosed.

Cluster headache

Cluster headache is a headache of vascular etiology, characterized by an explosive onset of severe, unilateral, periorbital or retro-orbital pain without nausea or photophobia. Ipsilateral nasal stuffiness and lacrimation are prominent, and individual headaches last less than 2 hours, occurring over weeks or months. Treatment options include nonsteroidal anti-inflammatory drugs (NSAIDs), ergotamine, oxygen inhalation, calcium channel blockers, and histamine-2 blockers.

Eagle's syndrome

Also known as stylohyoid syndrome, Eagle's syndrome results from elongation of the styloid process or from calcification of the stylohyoid ligament. Elongation of the styloid process is estimated to occur in 4% to 28% of the general population, although only a small percentage of these patients are symptomatic.[2] Patients with Eagle's syndrome describe a moderate pain deep in the tonsillar fossa and upper neck that can radiate to the ear and that can be aggravated by swallowing. Representative pain produced by transoral palpation of the styloid process through the tonsillar fossa is diagnostic. Excision of the styloid process proves curative in the majority of cases.

Glossopharyngeal neuralgia

Glossopharyngeal neuralgia is a neuritic-type pain initiated by nonnoxious stimulus of a trigger point and similar in all aspects to that of trigeminal neuralgia. The trigger point is in the oropharynx, and the pharyngeal stimulus can be swallowing or coughing. Sharp and lancinating pain tends to be unilaterally located in the posterior pharynx, soft palate, base of the tongue, ear, mastoid, or side of the head. There may be associated hiccuping, nausea, vertigo, tinnitus, aural fullness, hearing loss, or dysgeusia. The pattern of the attacks is unpredictable. Treatment involves carbamazepine or intracranial nerve section.

Gradenigo's syndrome

The classic tetrad of retro-orbital pain, suppurative otitis, abducens palsy, and diplopia defines this syndrome. The intense orbital pain and persistent aural discharge are caused by extradural abscess involving the petrous apex of the temporal bone. Lateral rectus palsy results from irritation of cranial nerve VI as it passes under the petrosphenoid ligament in Dorello's canal. Treatment involves antibiotics and surgery in refractory cases.

Herpetic facial pain

Herpetic papulovesicular skin eruption can occur in a dermatomal distribution of the trigeminal nerve presenting with burning, itching, and lancinating pain. This disorder is caused by the varicella-zoster virus, which infects the trigeminal nerve in childhood as chickenpox. The trigeminal nerve is involved in 18% of patients with herpes zoster, but other cranial nerves and facial dermatomes might also be involved. The virus remains dormant in the ganglion for decades and can be reactivated by trauma or stress or if the patient is immunocompromised. The reactivated virus is transported distally in the axon and produces small, crusting pustules on the skin. Treatment is a 10-day course of oral prednisone. The addition of antiviral agents improves outcome and relieves acute pain; however, opioid pain medications are used in severe cases. Advanced age portends a worse response to the medication and decreased chance of spontaneous recovery, presumably because of decreased cell-mediated immunity.

Checklist

- ☐ Complete blood cell count
- ☐ Antibody tests
- ☐ Erythrocyte sedimentation rate
- ☐ Electrolytes
- ☐ Blood urea nitrogen level
- ☐ Serum glucose level
- ☐ Urinalysis

Figure 1. *Laboratory tests that may be helpful in diagnosing cause of headache.*

Checklist

- ☐ Cryptococcal antigen
- ☐ Venereal Disease Research Laboratory (VDRL) test
- ☐ Cytology
- ☐ Routine blood cell counts
- ☐ Analysis for protein
- ☐ Analysis for glucose
- ☐ Tuberculosis
- ☐ Bacterial cultures

Figure 2. *Recommended analyses of cerebrospinal fluid for determining presence of central nervous system infection.*

Checklist

- ☐ Atypical facial pain
- ☐ Cluster headache
- ☐ Eagle's syndrome
- ☐ Glossopharyngeal neuralgia
- ☐ Gradenigo's syndrome
- ☐ Herpetic facial pain
- ☐ Posttraumatic neuralgia
- ☐ Rhinopathic headaches
- ☐ Superior laryngeal neuralgia
- ☐ Temporal arteritis
- ☐ Temporomandibular disorders
- ☐ Tolosa-Hunt syndrome
- ☐ Trigeminal neuralgia
- ☐ Trotter's syndrome

Figure 3. *Clinical syndromes of otolaryngologic diseases that can manifest as facial pain or headache.*

A diagnosis of postherpetic neuralgia is made when the pain persists more than 2 months. Tricyclic antidepressants, anticonvulsants, or baclofen is used for pain relief as NSAIDs and opioids are ineffective in this stage.

The most common infectious cause of facial nerve paralysis is herpes zoster. Papulovesicular lesions around the auricle in conjunction with ipsilateral facial nerve paralysis are the hallmarks of the Ramsay Hunt syndrome (herpes zoster oticus). Associated findings include sensorineural hearing loss, vertigo, tinnitus, intense otalgia, and vesicular eruptions that can extend to the tympanic membrane. Rising titers of varicella zoster virus antibodies and enhancement of the facial nerve can confirm diagnosis by MRI using gadolinium as the contrast medium. Systemic corticosteroids may help to reduce pain, minimize the incidence of postherpetic neuralgia, and decrease vertigo.

Additionally, herpes zoster can involve the auriculotemporal nerve, producing papulovesicular skin lesions along the nerve's distribution. These lesions can be obscured by scalp hair, and the pain can mimic that of a TMJ disorder.

Posttraumatic neuralgia

Neuroma or neuritic pain in the parietal or occipital regions is occasionally associated with head trauma. Predisposing factors include poor wound closure, local infection, retained foreign materials, skull fracture, extensive subcutaneous swelling or hematoma, diabetes mellitus, and coexisting peripheral neuropathy elsewhere in the body. A variable interval exists between the onset of pain and the initial injury, although a time span of 2 to 6 months is most common. At first, pain may be mild and nonneuritic, evolving into a more typical pattern over several months. Although trigger areas may be evident at examination, the pain is often spontaneous and poorly correlated with the patient's activities. Treatment includes the same medications as for trigeminal neuralgia. Recurrent infiltration of trigger areas with a local anesthetic such as bupivacaine hydrochloride can be surprisingly effective, providing extended relief for many months after a series of three to four injections.

Injuries to the trigeminal nerve occur as the result of facial trauma and surgery. Perhaps the most common injury is to the mandibular branch of the trigeminal nerve as a result of extraction of third molars. The reported incidence of trigeminal nerve injuries is 3% to 5% for third molar removal and 90% to 100% for mandibular osteotomies.[3] Most injuries produce anesthesia; pain is a rare consequence. The mandibular nerve is most commonly injured by compression of the nerve in the mandibular canal, either by direct pressure on the root tip or local swelling and edema after tooth removal. Most cases present as persistent anesthesia or paresthesia after the local anesthetic has worn off. Complete recovery occurs in more than 90% of the cases in 3 to 4 months. Fewer than 10% of patients have permanent anesthesia. Anesthesia persisting after this period portends a poor prognosis. Antiepileptic drugs such as carbamazepine, phenytoin, or baclofen are most useful for shooting, shocklike pain, and tricyclic antidepressants such as amitriptyline hydrochloride are used for burning, aching dysesthetic pain. When medication fails, surgical repair can be attempted. Microsurgical repair produces 50% to 60% pain reduction in patients with hyperalgesia and hyperpathia and 15% to 20% pain reduction in patients with anesthesia dolorosa and sympathetically maintained pain.[4]

Rhinopathic headaches

Headache is a common feature of acute sinusitis. The diagnosis is usually obvious because of the associated nasal symptoms of congestion, obstruction, and purulent discharge. The pain of acute sinusitis is typically more severe than that associated with chronic sinus disease, and chronic sinusitis often presents without other obvious nasal symptoms. Patients with acute ethmoiditis have symptoms of nasal obstruction, purulent rhinorrhea, and headache that can be bilateral or unilateral with associated pain located in the medial canthal region. Acute maxillary sinusitis typically presents as pain and pressure over the involved sinus along with nasal congestion and purulent nasal discharge. The pain often radiates to the teeth, zygoma, or periorbital or temporal regions. The pain of acute frontal sinusitis is often severe, located over the forehead and associated with tenderness on percussion of the involved sinus. The pain is exacerbated with bending over, a Valsalva's maneuver, or changes in barometric pressure. Acute sphenoid sinusitis has a variable presentation of symptoms, but postnasal drip, purulent rhinorrhea, and nasal congestion are the most common. The associated headache is usually poorly localized, but patients complain most often of retroorbital or vertex pain.

Acute sinusitis is treated medically with antibiotics and, in some cases, mucolytics, decongestants, and antihistamines. Maximal medical therapy for chronic sinusitis includes topical nasal steroids, mucolytics, decongestants, antihistamines, and antibiotics. Patients with refractory acute sinusitis are treated surgically.

Some patients have chronic headaches caused by sinus abnormalities other than acute purulent sinusitis. These patients have sinonasal anatomic variations or abnormalities or occult inflammatory disease that either causes or exacerabates their headaches. In these patients, the presence or absence of nasal symptoms is often unreliable as an indication of an underlying pathologic condition. Treatment includes steam inhalation and saline irrigations, topical corticosteroid sprays, or topical decongestants for mild or intermittent headaches. Systemic decongestants are also effective, although tachyphylaxis generally develops. Experimental therapy consists of capsaicin, substance-P antagonists, or neutral endopeptidase agonists. Surgical therapy is indicated if the diagnosis of rhinopathic headache is well established and if more conservative measures fail.

Superior laryngeal neuralgia

Superior laryngeal neuralgia is analogous to trigeminal neuralgia or glossopharyngeal neuralgia. It is confined to severe lancinating pain in the small region near the thyrohyoid membrane innervated by the superior laryngeal nerve. There may be a trigger area in the throat stimulated by talking, coughing, or swallowing. Treatment is carbamazepine or peripheral nerve section.

Temporal arteritis

Also known as giant cell arteritis or Horton-Magath syndrome, temporal arteritis results from inflammation of the superficial temporal artery. This finding is actually a harbinger of more widespread involvement of the arteries of the head and neck. More than 95% of these patients are older than 60 years.[5] These patients display a constellation of signs and symptoms, including fever, sweating, weakness, generalized fatigue, anorexia

with weight loss, and jaw claudication. Many patients complain of moderate to severe persistent throbbing or a dull aching pain in the region of the superficial temporal artery either unilaterally or bilaterally. Occasionally, patients report brief instances of sharp, shooting head pain or pain that is located exclusively in the occipital region. Palpation reveals a tender nodular pulseless superficial temporal artery with adjacent sensitivity of the scalp. Biopsy shows intimal thickening and necrosis with a foreign body giant cell reaction. Blindness is the most feared consequence of this disease and occurs in 30% to 50% of untreated cases, with bilateral blindness occurring in 25% of these patients.[3]

The erythrocyte sedimentation rate (ESR) is the "gold standard" diagnostic test and is consistently elevated in all but the rarest cases. Diagnosis is confirmed by arterial biopsy although skip lesions in the vessel necessitate a specimen of greater than 1 cm in diameter and occasional biopsy of the contralateral temporal artery.

Treatment is high-dose prednisone started promptly before biopsy confirmation of the diagnosis and resulting in dramatic reduction of pain within days. Lack of a definite clinical response to prednisone within several weeks makes the diagnosis much less secure without a positive biopsy finding. The ESR usually falls within weeks of starting therapy with steroids. The prednisone is tapered to a maintenance schedule of every other day over a period of weeks after the headache has resolved and the ESR has returned to normal. The disease lasts 1 to 2 years, during which maintenance steroid therapy is continued to prevent loss of vision.

Temporomandibular disorders

Temporomandibular disorders encompass multiple disorders associated with the TMJ presenting as headache, earache, or facial pain (singly or in combination) with limited opening of the jaw and joint noise. The TMJ undergoes both gliding and hinge motions and is thus termed a *ginglymoarthrodial joint*. The etiology and natural history of these disorders are poorly understood and the source of considerable debate. The current classification of temporomanidibular disorders includes internal derangements, degenerative joint disease, and myofascial pain. True TMJ pain is characterized by tenderness to palpation over the condyle and by pain on jaw movement. Acutely inflamed TMJs also produce a steady aching when the jaw is not moving. The characteristic pain and trismus that patients with TMJ disorders have is due to displacement of the articular disk.

A history of prior facial injury, jaw fracture, or stressful life situations can be significant contributing factors. Other important findings include unilateral or bilateral distribution of pain, duration of clicking or popping, and jaw position when the click or pop occurs. Treatment includes interarticular corticosteroid injections, dental splints, or orthodontic therapy, either singly or in combination.

Forty percent of patients with a TMJ disorder have been found to have histologic evidence of degenerative joint disease (DJD).[4] The incidence of DJD is known to increase with age.[5] Crepitus, pain on jaw movement, and the presence of grating or grinding sounds characterize DJD. Usually self-limiting, the painful stage of DJD typically lasts less than 1 year. There exists a characteristic radiographic appearance resulting from remodeling of the entire joint by osteophytes that form on the anterior surface of the condyle, causing flattening of the condyle. Degenerataive joint disease results from either gross trauma or infection of the joint or from long-term abuse in the form of bruxism, malrelationships of articular disks, or from other chronic repetitive manipulations of the jaw made by many patients as a behavioral response to stress. Treatment consists of NSAIDs, physical therapy, intra-articular corticosteroid injections, or joint reconstruction surgery.

Myofascial pain dysfunction syndrome is considered the most common painful disorder of the TMJ. Eighty percent of patients with this syndrome have painful muscle spasm of the TMJ.[6,7] The cause of this disorder is thought to be muscle fatigue produced by chronic oral habits that are often an involuntary tension-relieving mechanism. Positive findings include pain of unilateral origin, muscle tenderness, clicking or popping noise in the TMJ, and limitation of jaw movement. There should be no clinical, radiographic, or biochemical evidence of organic TMJ changes.

Treatment includes NSAIDs, muscle relaxants, application of moist heat, massage, soft diet, and acrylic splints, thereby decreasing muscle spasm and pain.

Tolosa-Hunt syndrome

A deep retro-orbital pain associated with ophthalmoplegia characterizes Tolosa-Hunt syndrome. The etiology is associated with an inflammatory lesion of the cavernous sinus with involvement of cranial nerves III, IV, and VI. Corticosteroids are the treatment of choice; however, recurrences can occur months to years later.

Trigeminal neuralgia

Trigeminal neuralgia is also known as tic douloureux or trifacial neuralgia. This syndrome is characterized by progressive or fluctuating repetitive brief paroxysms of severe, sharp, jabbing or lancinating pain lasting seconds to 30 minutes and occurring in the distribution of one or more divisions of CN V. However, facial numbness or weakness, loss of corneal reflex, changes in taste or smell, or impairment of other cranial nerve function is inconsistent with the diagnosis of trigeminal neuralgia. The maxillary division is the most common branch of the trigeminal nerve to be affected, followed by the mandibular and the ophthalmic branches. Some external stimulus such as touching, rubbing, or even cold air blowing across a specific trigger point precipitates these attacks. The most frequent location of the trigger point is the lateral aspect of the nose or at the corner of the mouth. Attacks may occur many times in rapid succession and many times a day with spontaneous remissions lasting many months or even years. The nasal sinuses, oral mucosa, periodontal ligaments, and muscles of facial expression or mastication can also serve as triggers. Diagnosis includes MRI of the head in patients of any age with symptoms of trigeminal neuralgia. In patients with signs and symptoms in addition to the classic presentation and when MRI is inconclusive, a lumbar puncture with analyses of CSF as listed in *Figure 2* is warranted.

Medical treatment includes carbamazepine, baclofen, phenytoin, sodium valproate, or chlorphenesin carbamate. Each drug should be tried for at least 2 weeks; combinations of the two drugs may also be used. Adjunctive therapy includes tricyclic antidepressants and nonsteroidal anti-inflammatory drugs. Surgical treatment is considered when medications cannot control the pain. Surgical management options include microvascular decompression of the nerve root via

a posterior fossa surgical approach, alcohol block of the involved division of the trigeminal nerve, and percutaneous radiofrequency thermocoagulation of the trigeminal sensory root as it exits the gasserian ganglion.

Trotter's syndrome

Nasopharyngeal tumors can produce facial pain and ipsilateral conductive hearing loss. Facial pain results from pressure on cranial nerves V, VII, IX, or X (singly or in combination). The conductive hearing loss is due to obstruction of the eustachian tube and resultant serous otitis media. All patients with these symptoms warrant nasophayrngoscopy to rule out a nasopharyngeal mass.

Comment

As in many other medical fields, the otolaryngologist is often confronted with the daunting task of delineating the etiology of a patient's complaint of chronic cephalgia. Laboratory and radiographic data are helpful; however, it requires a thorough history and physical examination correlated with good clinical judgment to make the appropriate diagnosis. Awareness of some of the otolaryngologic diseases that can manifest as facial pain or headache may help the physician in the diagnosis and treatment of this complex problem.

References

1. Mann J D, Lundeen TF. Headaches and facial pain. In: Bailey BJ, ed. *Head & Neck Surgery—Otolaryngology*. Philadelphia, Pa: JB Lippincott Company; 1993:761-773.

2. Kaufman SM, Elzay RP, Irish, EF. Styloid process variation. Radiologic and clinical study*Arch Otolaryngol* 1970;91:460-463.

3. Olesen J. Classification and diagnostic criteria for headache disorders, cranial neuralgias and facial pain. *Cephalalgia* 1988;8(suppl 7):28.

4. Clough C. Non-migrainous headaches. *Br Med J* 1989;299:70-72.

5. Saper JR. Chronic headache syndromes. *Neurol Clin* 1989;7:387-412.

6. Behrens MM. Headaches associated with disorders of the eye. *Med Clin North Am* 1978;62:507-521.

7. Blackwood HJJ. Arthritis of the mandibular joint. *Br Dent J* 1963;115:317-326.

8. Schwartz LL, Marbach JL. Changes in the temporomandibular joint with age. *J Am Soc Periodont* 1965;3:184-189.

9. Laskin DM. Etiology of the pain-dysfunction syndrome. *J Am Dent Assoc* 1969;79:147-153.

10. Griffin CJ, Sharpe CJ. The structure of the adult temporomandibular joint. *Aust Dent J* 1960;5:190-195.

Suggested reading

Lee KJ, ed. *Essential Otolaryngology—Head and Neck Surgery,* 7th ed. Stamford, Conn: Appleton & Lange; 1999.

Paparella MM, Shumrick DA, Gluckman JL, Meyerhoff WL, eds. *Otolaryngology*. Philadelphia, Pa: WB Saunders Co; 1991.