Introduction

Each of us is likely to have experienced headache either sporadically or chronically. Indeed, it is estimated that 40% of the worldwide population suffers with severe, disabling headache at least annually. It is well that such a common ailment usually has a benign course, but headache may be the presenting symptom of life-threatening disease. Given this, and the frequency that the complaint is encountered in ENT practice, we as otolaryngologists should be comfortable with the evaluation, diagnosis and treatment of headache and facial pain.

Patterns and Distribution of Pain

Brain parenchyma itself is not pain sensitive, but the meninges and supporting structures are heavily innervated. Pain may be elicited when these structures are inflamed or distorted. In the case of migraine, it is believed that activation of trigeminovascular structures is the source of the headache. Processes inside the cranial vault may produce pain referred to areas of the jaw, neck, face and scalp. Pathology within the anterior or middle cranial fossae may elicit pain that is referred to the scalp or face anterior to the coronal suture. Lesions in the posterior fossa cause pain in the more posterior portions of the head and upper neck. Pain arising from processes in the sphenoid or sella is commonly referred to the vertex. Simultaneous anterior and posterior headache may arise from lesions of the incisura, but are more commonly seen with generalized intracranial disease such as meningitis or subarachnoid hemorrhage.

Much attention has been to given to the sinuses and nose as a source of acute and chronic head and face pain. It is therefore worth a brief review of sinonasal innervation. The general sensory innervation of the mucosa of the nose and paranasal sinuses is from the ophthalmic and maxillary branches of the trigeminal nerve, with minor contributions from the greater superficial petrosal branch of the 7th nerve. Pain arising from processes within the sinuses is therefore frequently referred to the corresponding cutaneous dermatome innervated by the 5th nerve, or to the auricular or periauricular region due to the contributions of the 7th nerve. The mucosa of the sinuses is rather insensitive to pain, and the turbinates are more sensitive than the nasal septum. The region of the ostia of the sinuses is the most sensitive of any of the nasal structures.

Frontal sinus innervation is derived from the ophthalmic branch of the trigeminal nerve. Irritation of the mucosa of the frontal sinus therefore produces pain in the forehead or anterior cranial fossa related to the dural and cutaneous distribution of the nerve. The ophthalmic division of the 5th cranial nerve also innervates the anterior ethmoid air cells via the anterior ethmoidal nerve, a branch of the nasociliary. The anterior
ethmoids may also receive some innervation from the small supraorbital branch that supplies the frontal sinus. The anterior ethmoid nerve has the largest territory of innervation within the nasal cavity and supplies the anterior septum and lateral nasal wall, including the superior, middle and inferior turbinates and middle meatus.

The posterior ethmoid air cells and sphenoid sinus derive most of their sensory innervation from the maxillary division of the 5th cranial nerve via the inconsistently present posterior ethmoid nerve. The maxillary division supplies the posterior septum and a large portion of the superior and middle turbinates as well. Some of the innervation of the territory of the posterior ethmoid and sphenoid sinuses is derived from branches of the greater superficial petrosal branch of the 7th cranial nerve, and the ophthalmic branch of the trigeminal nerve. The maxillary sinuses are innervated by the posterior superior alveolar, infraorbital and anterior superior alveolar nerves, all of which are branches of the maxillary division of the trigeminal nerve.

Otalgia is a frequent complaint among patients with head and face pain. While ear pain may be due to processes arising within the ear, it may be referred from a distant process and manifest as ear pain. This is a well-known association, but it should be kept in mind when evaluating patients with headache or facial pain. A brief review of the innervation of the ear is warranted. Sensory innervation of the ear is derived from five sources – cranial nerves V, VII, IX and X as well as the cervical plexus. Pain arising from the distributions of any of these nerves may be referred to the ear. Pathology of the anterior tongue or oral cavity (including the temporomandibular joint) may cause otalgia via the third division and auriculotemporal branch of the trigeminal nerve. Lesions of the tongue base and tonsillar fossa can cause ear pain through the petrosal ganglion and Jacobson’s branch of the glossopharyngeal nerve. Lesions of the hypopharynx and supraglottic larynx may cause ear pain by stimulation of fibers of the jugular ganglion and Arnold’s branch of the vagus nerve.

Evaluation

The most important information in the accurate diagnosis of headache and facial pain comes from the patient’s history. This may require a redirected or more targeted history after more common causes are excluded. Indeed, the otolaryngologist frequently must first reverse a self-diagnosis of “sinus headache” reached by the patient before a more thorough history can be attained and more accurate diagnosis reached. As Acquadro and Montgomery point out, the evaluation of headaches can be quite time-intensive, requiring diagnostic and treatment modalities foreign to most otolaryngologists. In fact, there is a growing number of physicians who focus their practice on the diagnosis and treatment of headache. For many diagnoses however, the otolaryngologist may elect to treat the patient himself, remain a consultant with recommendations to the primary care physician or refer the patient to a headache specialist. Regardless of who eventually leads the patient’s healthcare team, there is an increasing emphasis on a multidisciplinary approach. The more experience a clinician gains in obtaining a history for headache, focusing on pertinent symptoms, the more skilled the clinician will become in diagnosis.

A complete history begins with determining the age when the patient first experienced the headache. The events occurring to the present should be recorded as well. The rate of onset and offset, intensity, quality, location, duration and response to treatments should be recorded. The frequency and timing of attacks may hold important information. The patient’s own description of the quality of the pain may aid determining its source. Pressure-like pain is common in viscous or chamber-derived pain, while sharp and shooting pains are more characteristic of neuritic pain. Throbbing pain is typical in vascular headaches. Burning and aching suggests muscular pain.

Any associated symptoms – nausea and vomiting, fever, diplopia, syncope, hiccupping, lacrimation, nasal congestion, photophobia, phonophobia, diarrhea, polyuria – should be noted. Does the patient experience any warning or aura prior to the attack? Any precipitating factors such as head movements, eating, relation to
sleep, foods, stress, menses, weather, medications or alcohol should be recorded. Factors providing relief may not only guide diagnosis, but treatment as well.

A thorough past medical history should include inquiries as to prior head injuries, intracranial infections or processes and past surgeries. Any past or present medical and psychiatric illnesses should be documented. Inquiry should be made as to current medications including the type and frequency of use of over-the-counter analgesics, oral contraceptives, herbal medicines and topical agents used on the head and face. Special attention should be paid to antihypertensives and vasodilators. A complete family and social history with amount and duration of tobacco, alcohol and recreational drug use should be noted. Life stressors should be assessed. A complete review of systems is mandatory in the complete evaluation of head and face pain.

The physical examination in patients with headache or facial pain is guided by the history. A complete head and neck and neurological examinations are mandatory. Special attention should be placed on the evaluation of the cranial nerves, with an evaluation of the eyes including fundoscopy. Limited or asymmetric jaw opening should be noted, as should any crepitus in the temporomandibular joint (TMJ) with opening and closing of the mouth. The teeth are carefully inspected for abrasions and the tongue for scalloped edges indicative of bruxism or tongue compression against the dentition at night. The muscles of mastication should be carefully palpated for tenderness. The temporal arteries should be palpated for tenderness or nodules. Trigger points within the trapezius or posterior cervical triangle should be sought. The patient’s posture and range of motion of the neck are evaluated. The occipital notch and scalp are palpated and percussed for differential tenderness indicative of occipital neuralgia or underlying subdural process. A positive Romberg test is a relatively common finding in this population. In the event of a positive test (instability on standing with the eyes closed), the test is repeated after asking the patient to try to recognize a number traced on the forehead with the eyes closed. Improved performance may indicate that the positive test is of “psychogenic” rather than neurologic origin.

After the complete history and physical examination, laboratory tests may be ordered. The necessary and sufficient evaluation of the headache and facial pain patient is the matter of some debate. In general, the tests requested should be guided by the history and physical examination findings. In the patient with recent onset headache of moderate to severe intensity and findings on the neurologic examination, EEG may be the test of first choice. EEG is also helpful in the patient with muscle contractions in whom epilepsy is suspected. Radiographic studies (CT and MRI) are useful to evaluate new onset moderate to severe headaches, or to rule out intracranial pathology in patients with long-standing, chronic headache or facial pain. Lab tests directed to rule out autoimmune disease or other systemic disease should be ordered if the history suggests a generalized process. EMG may be useful if a diagnosis of primary muscle disease or neuropathy is suspected. Tenderness of the TMJ may be further evaluated by radiographic imaging of the joint. Postural disorders with muscle pain and trigger points in the neck and shoulder should be evaluated with cervical spine radiographs.

Psychometric testing may be of benefit in the evaluation and treatment of the patient with headache and facial pain. Many tests have been applied, but probably the most widely used is the Minnesota Multiple Personality Inventory (MMPI). While especially useful in the evaluation of the chronic headache and facial pain patient, a thorough discussion of psychometric testing is beyond the scope of this discussion and mentioned here only for completeness.

** Syndromes **

** Tension-Type Headache **

Tension-type headache (TTH) is the most common type of headache. It occurs in 69% of men and 88% of women over a lifetime and the annual prevalence is 63% in men and 88% in women. TTH can be further distinguished as “episodic” TTH (ETTH) or “chronic” TTH (CTTH). The distinction
is made largely on frequency of occurrence – less than 15 days a month for ETTH and greater than 15 days a month for CTTH. The diagnostic criteria that distinguish TTH from other headache syndromes are largely related to the quality, intensity, location and duration of the pain. Headaches last from 30 minutes to 7 days. They are often described as pressing or tightening (non-pulsating) in quality. The intensity is mild to moderate and may limit but not prohibit activities. Its location may be bilateral or variable. There is no aggravation with physical activity, nausea and vomiting is rare, and photophobia or phonophobia may occur though not simultaneously.

Patients who acknowledge the role of stress in the etiology of their headaches, especially those with ETTH, are frequently well managed by biofeedback and stress reduction techniques. Posture correction and physical exercises should be prescribed as indicated. Patients with bruxism may benefit from a dental splint. For patients with ETTH, medications may be avoided, but when needed, the patient may do well with low dose benzodiazepines or amitriptyline once daily in a short course spanning several weeks. Pharmaceuticals are more likely to be necessary in the patient with CTTH. Abortive medications include aspirin, acetaminophen, aspirin-caffeine-butalbital or phenacetin combinations or short half-life non-steroidal anti-inflammatory medications (NSAIDs). Preventive medications include daily antidepressants, muscle relaxants and long half-life NSAIDs. Opiates and benzodiazepines may be effective but prolonged use is contraindicated. Daily NSAID use should be limited to less than one week. The treatment regimen employed must be individualized based upon the triggering factors elucidated in the history and physical exam findings.

**Migraine**

Migraines are perhaps the most studied of the headache syndromes. This is due in part to the high incidence and significant loss of productivity and limitation on quality of life suffered by those with the syndrome. It is estimated that 17% of females and 6% of males have migraine headaches. Onset is usually in the second or third decade. Migraines are characterized by headaches of moderate to severe intensity located unilaterally with a pulsating quality. Attacks last from 4-72 hours (2-48 hours in children less than 15 years old) and are aggravated by routine physical activities. In order to meet diagnostic criteria, there must be nausea, vomiting, photophobia or phonophobia. Migraines may occur with or without aura. Migraine with aura is less common. Vision complaints are the most common manifestation of aura, but patients may experience paresthesia, aphasia, nausea and vomiting prior to the onset of headache. These findings are completely reversible and precede the headache by no more than 60 minutes.

Migraines seem to have a triggering event that precipitates a sterile inflammatory response around intracranial vessels that is mediated by the trigeminovascular system. Triggering factors may include stress, menses, pregnancy and oral contraceptive pills, infection in the head and neck, trauma or surgery, red wine, aged cheeses, vasodilating medications, strong odors, irregular diet or sleep and bright sunlight or flickering lights. Recent studies have discovered serotonin receptor subtypes in the central nervous system that play significant roles in the neurologic changes and intracranial blood vessel change. Newly available treatments such as sumatriptan target these receptors. Several other neuropeptides have been identified as pro-inflammatory and are believed to play a significant role in migraine development. Further investigation is hoped to provide treatment alternatives with fewer side effects.

The treatment of migraine headaches may be approached using several strategies: aborting the attacks at their onset, controlling the pain once is fully evolved and reducing the frequency of attacks. Therapies aimed at aborting an attack should be started as soon as the premonitory or warning signs are noted. Abortive therapy has been revolutionized with the introduction of 5-hydroxytryptamine (5-HT) receptor agonists. These include sumatriptan (*Imitrex*) available in oral, subcutaneous injection
or nasal spray forms, naratriptan (Amerge), rizatriptan (Maxalt) and zolmitriptan (Zomig) all available in oral preparations. These medications have allowed the migraine sufferer to quickly and effectively treat attacks several times a month with minimal side effects. Other medications used to abort headaches include ergotamine tartrate administered sublingually, or in combination with caffeine by mouth. Dihydroergotamine 45 is administered in a nasal spray. Butorphanol is a mixed narcotic agonist/antagonist available by nasal spray. It does have potential for abuse and chronic use is contraindicated. Midrin is an orally administered compound of acetaminophen, isomethypene mucate, a sympathomimetic amine and dichloralphenazone, a mild sedative. It has a low side effect profile and may be used until relief is attained. Many NSAIDs have been shown to be effective in migraine headaches. The short-rise time, short-acting medications such as naproxen, ketorolac, ibuprofen and choline magnesium trisalicylate have the greatest usefulness. Lidocaine administered intranasally in 4% spray, either singly or in combination with nasal decongestants, has been shown to be effective, although of short duration. Intravenous lidocaine with diphenhydramine may also be effective.

If abortive therapy fails, management should be aimed at reducing the intensity of the pain and controlling associated symptoms such as nausea and vomiting. It is desirable to avoid opiates for the treatment of migraine. Finkel et. al. recommend several treatment regimens: (1) prochlorperazine (Compazine) IV push that may be repeated in 20 minutes if no effect, (2) dihydroergotamine IV push followed by IV prochlorperazine, (3) chlorpromazine (Thorazine) IV push, may repeat in 20 minutes if needed and (4) haloperidol IV push followed by lorazepam IV push. Options (1) and (3) should not be combined, but may be followed by (2) or (4) if necessary.

Patients experiencing 2 or more attacks per month should be started on a prophylactic regimen. Appropriate first steps are to limit the activities or factors that trigger the headaches. This may be effective by itself, but medical prophylaxis is often needed as well. Multiple antidepressant medications have been shown to be effective in the prevention of migraine headache. These include amitriptyline, nortriptyline, doxepin, trazodone, imipramine and desipramine. The newer selective serotonin re-uptake inhibitors (SSRI) including Prozac and Zoloft, have not been shown to be effective in migraine therapy. Bellergal, a low dose, sustained relief ergotamine may be useful in preventing attacks. NSAIDs have some usefulness in the prevention of attacks as well as the treatment of the acute headache. β-blockers, specifically propranolol, nadolol, atenolol, timolol and pindolol have been used with some success but are contraindicated in patients with depression, asthma or diabetes. Calcium channel blockers (verapamil, nifedipine, nimodipine) have shown some effectiveness preventing migraine attacks as well.

Binder et al. found 51% of migraine sufferers obtained complete prophylaxis for an average of 4.1 months duration after the injection of botulinum toxin type A (BOTOX) into the facial and scalp musculature. An additional 38% obtained a partial response for an average duration of 2.7 months. The investigators also reported a 70% complete response rate among patients treated acutely for migraine headache within 1-2 hours post-treatment. These results hold promise for a novel treatment modality for the migraine sufferer.

Needless to say, the treatment of migraine can be a time-consuming and frustrating proposition. Lifestyle changes with the avoidance of the triggering event must be stressed to the patient. Medication changes should be adequately evaluated before dismissed as ineffective, and all medications should be started one at a time at the lowest dose. It is often necessary to combine medications for acute pain or abortive therapy with those used for prophylaxis, however some interactions do occur and this should be done with caution. The reader is referred to the manufacturers’ data regarding recommended dosages, contraindications and complete list of interactions and side effects for all the medications listed.
Cluster Headache

Cluster headache (CH) is characterized by intensely severe pain (sometimes termed *suicide headache*) with boring or burning qualities located unilaterally in the orbit, supraorbital or temporal area. Attacks last from 15 to 180 minutes. The headache is associated with at least one symptom of autonomic hyperactivity: conjunctival injection, lacrimation, nasal congestion, rhinorrhea, forehead and facial sweating, miosis, ptosis or eyelid edema. Attacks occur between one every two days to eight per day. At least 5 such attacks must occur to meet the diagnostic criteria. Nausea and vomiting is uncommon and there is no aura. Onset is usually in the second to fifth decades. Cluster headache is the only headache syndrome with a male preponderance. It is associated with alcohol use and intolerance, and during an active phase or “cluster”, alcohol may precipitate an attack.

There are both episodic and chronic types. Episodic CH has periods of activity alternating with periods of inactivity. Active periods vary in frequency from two or more per year, to one every two or more years and tend to occur in regular intervals. The duration of active periods ranges from seven days to a year. In chronic forms, the remission phases last less than 14 days while the prolonged ones are absent for at least one year.

Treatment is aimed at preventing an attack during a cluster. Once an effective therapy is discovered, it is continued for 6 to 8 weeks and then gradually tapered. Options for treatment include calcium channel blockers (nifedipine, nimodipine, verapamil), low dose daily ergotamine (*Bellergal*) and lithium carbonate (especially in chronic forms of CH). Methysergide has been found to be effective but use is limited to 4 months as prolonged continuous use may cause retroperitoneal fibrosis. Trials with valproic acid are ongoing. Some have used antihistamines, both H1 and H2 blockers with limited success. The role of steroids is controversial, but they are frequently used in prophylaxis during an active period.

Some treatments have been found to be effective in the acute treatment of an attack. Oxygen inhalation, 6-10 liters per minute administered by face mask seems to be particularly effective in young patients with attacks primarily at night. 5-HT receptor agonists are effective in shortening an attack if given at the first indication of pain. Intranasal lidocaine administered either 4% topical or 2% viscous at the posterior aspect of the inferior turbinate affecting a sphenopalatine block may be effective in terminating an acute attack.

Temporal Arteritis

Temporal arteritis is characterized by daily headaches of moderate to severe intensity, scalp sensitivity, fatigue and various non-specific complaints with a general sense of illness. 95% are over 60 years old. The pain is usually unilateral, although some cases of bilateral or occipital pain do occur. The pain is a continuous ache with superimposed sharp, shooting head pains. The pain is similar to and may be confused with that of CH, but CH tends to occur in younger patients. The two may also be distinguished on physical exam when dilated and tortuous scalp arteries are noted. The erythrocyte sedimentation rate (ESR) is markedly elevated in temporal arteritis as well.

Definitive diagnosis is made by artery biopsy from the region of the pain, although negative biopsy may be due to the spotty nature of the disease and does rule out the diagnosis. High dose steroid therapy usually precipitates a dramatic decrease in head pain. Failure to respond to steroid therapy with a negative biopsy should call the diagnosis into question. If the diagnosis seems likely based on history and physical examination, steroids should be started immediately to avoid vision loss, the most common complication of the disorder occurring in 30% of untreated cases. The biopsy remains
positive for 7-10 days from starting steroid therapy. Steroids may be tapered to an every other day maintenance schedule when the pain resolves and ESR normalizes. The disease is usually active for 1-2 years, during which time steroids should be continued to prevent vision loss.

**Chronic Daily Headache**

Chronic daily headache (CDH) is described as headache occurring at least 6 days a week for a period of at least 6 months. The pain is usually present throughout the day with little time spent pain-free. The head pain is typically bilateral, frontal or occipital, non-throbbing and moderately severe. The syndrome is associated with the overuse and abuse of many common over-the-counter pain medications (aspirin, acetaminophen, ibuprofen, etc.), barbiturates and opioid analgesics. A carefully taken history will reveal an increasing need for medications and the emergence of a chronic headache that is qualitatively distinct from the headache for which it was originally taken. This led to the idea of CDH being a “transformed migraine”.

The treatment centers on the withdrawal of the causative medication. In order to be successful, several points should be followed: (1) the patient must understand the syndrome, (2) the offending medication should be tapered over 10 days and completely ceased for a minimum of 2 months, (3) the substitution of other agents that may perpetuate the disorder must be avoided (4) antidepressant medications prescribed at gradually increasing dosages aid in withdrawal of the offending medication, (5) adjuvant therapy such as physical therapy or biofeedback should be employed, and (6) in refractory cases, consultation with a neurologist with inpatient management to assure complete abstinence from the medication and control withdrawal symptoms may be necessary. Withdrawal symptoms may be prominent, usually occurring in the first 4 days, but sometimes occurring up to 3 weeks after cessation of the causative medication. These include nervousness, restlessness, increased headaches, nausea and vomiting, insomnia, diarrhea and tremor.

**Trigeminal Neuralgia**

Trigeminal neuralgia (formerly also known as tic doloureux) is characterized by paroxysmal pain attacks lasting from a few seconds to less than two minutes. The pain is severe and distributed along one or more of the branches of the trigeminal nerve with a sudden, sharp, intense stabbing or burning quality. Between attacks the patient is completely asymptomatic without neurological defects (no facial numbness, loss of corneal reflex or change in taste or smell). The pain may be precipitated from trigger areas or with certain daily activities such as eating, talking, washing the face or brushing the teeth. Attacks are the same in an individual patient. Structural causes of facial pain should be excluded. The syndrome is most common in patients over 50. The course may fluctuate over many years and remissions of months or years are not uncommon.

Medical treatment of the disorder includes carbamazepine, gabapentin, baclofen, phenytoin, or sodium valproate. Tricyclic antidepressants (TCA) and NSAIDs may be used as adjuvant therapy. Opiates are usually ineffective. Surgical treatment is occasionally necessary when medical therapy fails to control the pain attacks.

**Glossopharyngeal Neuralgia**

Glossopharyngeal neuralgia is characterized by pain attacks similar to those in trigeminal neuralgia, but located unilaterally in the distribution of the glossopharyngeal nerve. Pain is most common in the posterior pharynx, soft palate, base of tongue, ear, mastoid or side of the head. Swallowing, yawning, coughing or phonation may trigger the pain. Management is similar to that for trigeminal neuralgia.
Atypical Facial Pain

Atypical facial pain is a diagnosis of exclusion for pain not meeting the diagnostic criteria of other facial pain syndromes. Mongini refers to the term atypical facial pain as outdated and includes its description in psychogenic facial pain. Indeed, the description of the pain may be inconsistent with bilateral pain that often changes locations over weeks to months. The pain is not triggered and not electrical in quality. Intensity fluctuates but the patient is rarely pain-free. Pain is typically located in the face and seldom spreads to the cranium in contradistinction to TTH. It is more common in women aged 30 to 50 years old. Sixty to 70% of these patients have significant psychiatric findings, usually depression, somatization or adjustment disorders, therefore psychiatric evaluation is indicated. Treatment is with antidepressants, beginning with low dose amitriptyline at bedtime and increasing the dose until pain and sleep are improved.

Post-traumatic Neuralgia

Traumatic injuries may induce a pain syndrome due to the development of neuroma. This occurs most often on the occipital and parietal regions of the scalp. The diagnosis of post-traumatic neuralgia is made based upon the history of trauma temporally correlated with the development of the characteristic neuritic pain. Poor wound closure, infections, foreign material in the wound, hematoma, skull fracture, diabetes mellitus or peripheral neuropathy elsewhere in the body predispose to neuroma development. Pains commonly begin 2 to 6 months after the injury. Medical therapy is similar to that used in trigeminal neuralgia. Bupivacaine injection of trigger areas may be effective, relieving pain for months. A well-defined trigger point may be amenable to surgical excision.

Post-herpetic Neuralgia

Herpetic skin eruption is caused by the reactivation of latent varicella-zoster virus from the sensory nerve ganglia. The reactivated virus is carried via the axons distally to the skin where it produces a painful rash with crusting vesicles in a dermatomal distribution. The trigeminal nerve is the second most commonly affected after nerves in the thoracic region. Steroids are often used for the acute eruption in otherwise healthy individuals, while antivirals, NSAIDs and opiates are often used in immunocompromised patients.

Pain that persists 2 or more months after the acute eruption is known as post-herpetic neuralgia. NSAIDs and opiates are of little use in the treatment of the neuralgia. Anticonvulsants in conjunction with TCA or baclofen are most useful for the control of shooting neuritic pains.

Temporomandibular Disorders

Temporomandibular disorders (TMD) include a heterogeneous group of processes all with a similar clinical presentation. Common symptoms of TMD include temporal headache, earache, facial pain, limited jaw opening or joint noise. The majority of TMD originate spontaneously with only 40% able to recall a specific event, usually trauma, preceding the onset of pain. This suggests that there is a significant role of emotional and psychological factors in the etiology of spontaneously occurring TMD. As a result of relatively recent advances in the understanding of the pathogenesis of TMD, they may be further classified as internal derangements, degenerative joint disease (DJD) and myofascial pain.

Pain that truly originates in the TMJ is rare and characterized by tenderness to palpation of the condyle and pain with joint movement. Internal derangements are characterized by anterior and medial displacement of the articular disk. This produces a “click” as the disk with reduced with
mood opening. This clicking may be benign, with minimal clinical symptoms or discomfort. When symptomatic, pain occurs just before or during the click and the joint is mildly tender to palpation. Patients may have limited mouth opening as a result of attempts to avoid pain. DJD has a similar presentation, with pain at joint movement and crepitus over the joint. The painful stage usually lasts less than a year. Long-standing DJD causes flattening of the condyle and osteophyte formation making it easily recognizable radiographically. The vast majority of patients, 60%-70% have combined muscle and joint pain with muscle pain dominating the clinical picture. These patients usually have tenderness to palpation of the muscles of mastication.

NSAIDs and physical therapy are the mainstays of treatment for TMD. Similar to tension headache, biofeedback and trigger point injection may be beneficial. Benzodiazepines are useful for muscle pain, but chronic use may lead to dependence and tolerance. Muscle relaxants are of little benefit. In chronic muscle pain, antidepressants may be more useful that analgesics or anxiolytics. TCA are useful in those patients with sleep disturbance, or SSRIs may be used for patients intolerant of TCA.

Pseudotumor Cerebri (Benign Intracranial Hypertension)

Pseudotumor cerebri presents with intermittent headache of variable intensity. The patient has a normal neurological examination, although a sixth nerve palsy is rarely found. There is papilledema with no evidence of hydrocephalus or mass on CT scan. Cerebrospinal fluid (CSF) pressures are greater than 200 mm H2O with normal CSF chemistries and cultures. The history may also be characterized by one or several of the following: (1) mastoid or ear infection, (2) menstrual irregularity or other endocrine disorder, (3) weight gain of greater than 10% over baseline weight over 6 months, (4) exposure to steroids (especially withdrawal), vitamin A, tetracycline or nalidixic acid, (5) retro-orbital or vertex headache, especially with empty sella syndrome (6) fluctuations in vision, (7) recurrent unilateral or bilateral tinnitus, (8) constriction of visual fields lasting longer than several months. Medical therapy is aimed at reducing CSF production with acetazolamide and furosemide. Weight loss and low-salt diet are also beneficial. In chronic cases with loss of visual fields that do not respond to weight loss and medical therapy, lumboperitoneal or ventricular shunting with incision of the optic nerve sheath may be necessary.

Intracranial Processes

Primary or metastatic tumors of the brain produce headache 30% of the time. 15% have headache as the presenting symptom. The pain is described as dull or aching, lateralized and relatively mild initially. The pain may crescendo with increasing intensity and frequency with progression of the disease. Initial response to non-narcotic analgesics is common, becoming resistant as the pain intensifies. Early morning headache is present only in 10% but is strongly suggestive when present, especially when accompanied by vomiting without significant nausea. The pain may increase with coughing, Valsalva or changes in body position. Focal neurological signs may be a late finding despite significant head pain.

Subdural hematoma (SDH) presents with a fluctuating level of consciousness with moderate to severe headache after trauma. The pain is usually lateralized to the side of the hematoma with tenderness to percussion over the hematoma. The patient may also have Battle’s sign (ecchymosis over the mastoid) and hemotympanum. In chronic SDH, the traumatic occurrence may be remote or not remembered at all.

Subarachnoid hemorrhage (SAH) presents with the sudden onset of severe, generalized headache (“the worst headache of my life”). Early in the process the pain may be localized. Nausea, vomiting and stiff neck are common, progressing to back pain as blood circulates into the spinal subarachnoid
space. Imaging may not show the blood collection or a mass effect, in which case a lumbar puncture should be performed to establish the diagnosis.

Central Nervous System Infection

There is little diagnostic dilemma when headache is associated with fever and stiff neck. However, many inflammatory conditions of the CNS may produce headache with a paucity of associated symptoms. These include epidural abscess, fungal, tuberculous or luetic meningitis, acquired immunodeficiency syndrome (AIDS) of the CNS, and meningeal sarcoidosis. Diagnosis is dependent on history and physical with lumbar puncture for CSF studies after CT or MRI has ruled out intracranial mass.

Hypertension

Chronic, untreated hypertension is an occasional cause of headache. It is most likely to occur in patients with diastolic pressures over 115 mm Hg. The pain is often described as throbbing and may be associated with nausea. Acute headache associated with rapid rises in blood pressure may be found in pheochromocytoma, renal artery stenosis or hyperadrenalism.

Low Intracranial Pressure Headache

Headache resulting from the removal of CSF with subsequent decreases in intracranial pressure (ICP) is a well-known phenomenon. This most commonly occurs after lumbar puncture (LP), but may occur spontaneously or as a result of trauma. The headache is usually worsened by sitting or standing upright and improves or disappears with lying flat. Pain is steady, often described as pulling, and occurs most commonly in the vertex or occipital regions. Nausea is common and transient 3rd or 6th nerve palsies have been reported. Nearly all of these headaches will resolve spontaneously over several days. Bed rest and fluids are recommended but have not been conclusively shown to speed recovery. In cases occurring after LP that do not resolve with conservative management, autologous blood patching in the epidural space is indicated.

Sinus Headache

While acute sinusitis is widely accepted and recognized as a cause of headache with pain referred to the skin or intracranial structures also innervated by the nerve branches providing the sinuses, chronic sinusitis or sinonasal abnormalities as a cause of headache has been more controversial. According to the International Headache Society Manual for Headache Diagnosis published in 1988: “Other conditions which may cause headache such as nasal passage abnormality due to septal deflection, hypertrophic turbinate and atrophic sinus membranes are not sufficiently validated as causes of headache. Chronic sinusitis is not validated as a cause of headache unless relapsing into an acute phase. Migraine and tension-type headache are often confused with true sinus headache because of similarity in location.” Much investigation as to the treatment of headache and facial pain presumed to be of sinonasal origin has been performed, with multiple authors demonstrating good outcomes in carefully selected cases.

The pain associated with acute sinusitis is commonly described as constant, dull and aching. Occasionally the pain is sharp and may be worsened by jarring of the head, bending forward or stooping. Although the discomfort is most often located over the acutely inflamed sinuses, the pain may be referred to other areas of the head or face based upon the anatomic structures sharing innervation with affected sinuses as described previously. In most instances, the pain will be accompanied by such symptoms as purulent nasal discharge, malaise and congestion.
In 1988, Stammberger and Wolf described their experience with headaches and sinus disease. They proposed classifying headaches into 3 groups for diagnostic purposes: (1) those with headaches clearly attributable to sinus disease (i.e. acute sinusitis), (2) those with headaches clearly due to non-sinus causes such as migraine, neuralgias, etc, and (3) those with complex problems and in whom there seems to be no overt indication of sinus disease. It is this last group that presents the biggest diagnostic and treatment dilemma. With the use of nasal endoscopy and CT scanning in the coronal plane, a thorough evaluation of the sinus anatomy allows the identification of mucosal contact points that are hypothesized to produce head pain even in the absence of other sinonasal pathology.

Multiple neuropeptides have been found in the nasal mucosa that function as neurotransmitters. One of these is Substance P (SP), which functions to mediate pain impulses to the cerebral cortex. Receptors within the sinonasal mucosa are stimulated by chemical and caloric irritants, as well as by mechanical irritants such as pressure. These receptors are one component of local reflexes mediated by SP, and when stimulated produce vasodilation, plasma extravasation (neurogenic edema) and hypersecretion. Pressure exerted on the nasal mucosa by polyps or mucosal swelling can be enough to trigger an SP-mediated pain sensation. Due to the axonal reflex, relatively small lesions may lead to a vicious cycle and significant symptoms. Certain anatomic conditions predispose to the process. Not all of these conditions are disease states per se, but they are factors that may reduce the already narrow spaces of the anterior ethmoid and middle meatus, thereby giving rise to areas of mucosal contact, secretion retention and malventilation of the sinuses promoting polypoid degeneration of opposing mucosal surfaces. All of these conditions, even when small and well circumscribed, may have one dominating clinical symptom – headache.

Agger nasi cells lie anterior and superior to the insertion of the middle turbinate on the lateral nasal wall. Even in the absence of disease, these cells may narrow the frontal recess. When diseased, the agger nasi cells may completely obstruct the frontal recess.

Variations in the uncinate process may occur. The posterior margin may be delected anteriorly and displace into the middle meatus to such an extent that it contacts the lateral aspect of the middle turbinate. This contact is even more prone to occur if the middle turbinate has a paradoxical curve or is pneumatized (concha bullosa). The uncinate may be curved laterally and posteriorly as well, narrowing the hiatus semilunaris and ethmoid infundibulum and impeding drainage of the anterior ethmoid cells. Pneumatization of the uncinate itself has been described and may further narrow the middle meatus.

Patients with headaches of sinus origin do not necessarily present with the typical sinusitis history. The physical examination and plain radiographs may be negative as well. When in question, the combination of rigid nasal endoscopy and CT scanning provide the most complete information regarding sinus anatomy and disease. Multiple authors have reported good successes with sinus surgery targeted at these points of mucosal contact and anatomical variation for the treatment of chronic headache presumed to arise from the nose and sinuses. Clerico performed directed sinus surgery on 7 of 10 patients previously diagnosed with headache syndromes with improvement in all. Patients were offered surgery only if they had a clearly positive response...
to intranasal decongestion and topical anesthesia to the area of presumed mucosal contact. Theoretically, if mucosal contact is the cause of the patient’s symptoms, relief of the contact, whether it is medical or surgical, should provide headache relief. The surgical procedure in each case was individualized to each patient’s symptoms and anatomic findings. Parsons and Batra noted that 91% of 34 patients experienced a decrease in headache intensity after rhinologic surgery directed to the relief of contact points, and 85% reported a decrease in frequency. They still cautioned against the use of the presence of contact points as the sole indication for surgery, as the majority of patients in their series also had chronic sinusitis that served as the primary indication for surgical intervention.

While the evidence is inconclusive, it seems that there is at least some support for mucosal contact as a cause of headache. Any surgical intervention aimed at the relief of headache must be clearly limited to the area in question. It stands to reason that if indeed mucosal contact and obstruction are the cause of the headache, some objective evidence of abnormal sinus function, whether it is mucosal edema or polypoid degeneration, should exist on examination or imaging. Until further evidence is uncovered, using headache as the only indication for surgical intervention is to be discouraged. When it is performed, clear demonstration of improvement in the headache should be obtained with topical decongestants and anesthesia to the proposed site of origin of the pain.

Conclusion

Headache and facial pain are common ailments with many varied causes and are commonly encountered in otolaryngologic practice. The correct diagnosis can usually be reached by history and physical alone. A significant behavioral component is involved in most cases, therefore it is important that the correct diagnosis be reached quickly and appropriate therapy instituted for the greatest chance of successful treatment. Failure to properly identify the cause and treat accordingly may lead to patient frustration and distrust with a high likelihood of non-compliance and failure. Effective treatment is rewarding to the clinician as the patient’s productivity and quality of life are greatly improved.
Case Presentation

A 29 year old white male presents with complaint of severe headache. It is located unilaterally on the right in the peri/retro-orbital region. He describes the pain as excruciating. There is a constant boring pain that is punctuated by lancinating pain radiating posteriorly to the mastoid area and occasionally to the upper neck. He notices that it is difficult to keep his eyes open when having the pain. He denies any nausea or vomiting. He has had similar headaches everyday for the past three weeks. They tend to occur during the night, awakening him from sleep, or in the afternoons. Each headache lasts from 30 minutes to 2 hours without treatment. In the past he has used NSAIDs with some success, but now they have no effect. In addition, inhalational oxygen has been beneficial, but now provides little relief.

He is otherwise healthy and is not on any medications when not having the headaches. He has no known drug allergies. Past surgical history is remarkable only for hernia repair. There is no significant history of head trauma. He is a non-smoker and drinks alcohol sporadically. He does not use recreational drugs. When he has the headaches, he avoids alcohol as he has noticed that alcoholic beverages may precipitate an attack.

On further questioning, he has had these headaches several times before. The last was 3 years ago in the spring. The time prior to that was 5 years ago from the current episode. The first time he experienced the headache was at age 19 when he was awakened from sleep by a severe headache. It was not followed by any further headaches until several years later. These had a marginal response to NSAIDs. Each of these episodes lasted approximately one month. With the last episode, he was treated with Verapamil and a Prednisone taper with cessation of the headaches.

Examination is performed while he is not experiencing an attack and is normal.

The patient is diagnosed as having cluster headaches of the episodic type. He is begun on Prednisone taper and Verapamil. For abortive therapy, he is given a prescription for sumatriptan nasal inhalers and subcutaneous injection.

He returns one month later. The headaches have now resolved. They decreased in intensity and frequency shortly after starting the steroids and Verapamil. The sumatriptan provided relief almost instantaneously at first, then with slower onset as the frequency of use increased. It continued to provide relief however.
References


