Facial pain

After the dentist and ENT have finished

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Summary

A 63-year-old woman is referred for neurologic consultation because of electrical shocks of pain in a left V3 distribution. The attacks are very brief, but occur many times a day, triggered by cold, touch, and chewing. In between attacks, she is pain free. Her neurologic examination is normal. This straightforward presentation of trigeminal neuralgia leaves the consulting neurologist wondering what to do with the other 45 minutes of the visit. The current article is vignette-based and deals with patients who may not fit into such a neat category. It is meant for the general neurologist, not the headache specialist. Aside from routine scenarios, it includes discussions of atypical facial pain, temporomandibular joint disorder, burning mouth syndrome, and other vexing types of facial pain.

The International Headache Society (IHS) classification of headaches is important for headache specialists. For the general neurologist, it is clinically useful to divide facial pain syndromes into 4 major categories: neuralgiform disorders, non-neuralgiform headache disorders, facial pain as part of a systemic disease, and facial pain as a “medically unexplained syndrome.” The table (page 177) lists the clinical syndromes and the corresponding IHS codes.

Key point: Benign disorders present with a normal neurologic examination. Trigeminal sensory loss almost invariably indicates an underlying lesion. Numbness matters.¹

Primary neuralgiform disorders

Trigeminal neuralgia is the prototype for the primary neuralgiform disorders, even though compression by an aberrant loop of artery is often implicated in its pathogenesis. Pain is almost invariably unilateral. Bilaterality or demonstrable sensory loss should raise the suspicion of a structural lesion. The mandibular division is most commonly involved, followed by the maxillary branch. Pain limited to the supraorbital branch is unusual. The individual jabs of pain are extremely brief, but may occur in bursts that last for a minute or two. Bursts may be followed by a refractory period, during which the usual triggers no longer produce pain. Pain is so severe that it causes wincing, hence “tic douloureux.” Although patients are usually pain-free between the episodes, some will complain of a vague aching pain. Imaging may reveal causative abnormality in up to 15% of patients presenting with relatively typical trigeminal neuralgia.

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Intermedius neuralgia is a rare disorder that is believed to be due to abnormal discharges in the nervous intermedius, which runs in between the seventh and eighth cranial nerves.

Symptomatic treatment of the individual episodes is nearly impossible and therapy is aimed at prophylaxis. The American Academy of Neurology Practice Parameter² states that “Carbamazepine or oxcarbazepine should be offered for pain control while baclofen and lamotrigine may be considered useful. For patients with trigeminal neuralgia refractory to medical therapy, Gasserian ganglion percutaneous techniques, gamma knife, and microvascular decompression may be considered.” Gabapentin and pregabalin have also been used. Because TN is primarily a disease of middle and late age, the axiom of “start low and go slow” should be followed.

Vignette A 73-year-old man reports repetitive occurrences of throat pain and fainting. Upon swallowing, he develops a severe pain in the base of the tongue, radiating to the left ear. It is followed by a feeling of lightheadedness and then loss of consciousness. He awakens almost immediately, without sequelae.

Glossopharyngeal neuralgia (GN) is characterized by severe, unilateral pain in the base of the tongue, tonsillar fossa, angle of the jaw, or the ear (subserved by cranial nerves IX and X). In about 10% of cases, pain is followed by syncope. In cases of “swallow syncope,” the progression is severe pain-bradycardia-syncope. An imaging procedure is indicated, to rule out an underlying structural lesion. GN is analogous to trigeminal neuralgia. Because it is far less common, there are a few studies on which to base recommendations for treatment, and the treatments for TN are extrapolated to GN.³

Vignette A 43-year-old woman has recurrent bouts of severe stabbing pain, deep in the external auditory canal. ENT examinations have been normal. Neurologic examination is normal.

Intermedius neuralgia is a rare disorder that is believed to be due to abnormal discharges in the nervous intermedius, which runs in between the seventh and eighth cranial nerves. As with other cranial neuralgias, structural causes must be excluded. Recognizing the syndrome may spare the patient the anxiety of being labeled “psychosomatic.”

Vignette A 55-year-old man presents with severe pain in the left eye and temple. The pain is sudden in onset and lasts from a few seconds to a couple of minutes. It is accompanied by ipsilateral conjunctival injection, lacrimation, and rhinorrhea. The attacks can occur up to 20 times a day.

The pain sounds very much like cluster headache. However, the onset is later in life, the attacks are of shorter duration, and the associated autonomic features are more pronounced. It is short-lasting unilateral neuralgiform headache with conjunctival injection and tearing (SUNCT). As in cluster headache, there is a male predominance. SUNCT is notoriously unresponsive to both indomethacin and many anticonvulsants. There have been reports of acute responses to lidocaine and successful prophylaxis with lamotrigine or topiramate.⁴ Raeder paratrigeminal neuralgia was a term applied to similar pain accompanied by varying degrees of sympathetic dysfunction. It is no longer in common usage.

Secondary (symptomatic) neuralgias

Vignette A 50-year-old man was punched in the left eye, sustaining a blowout fracture of the orbit. Surgical repair corrected vertical diplopia from entrapment of the inferior rectus. Postoperatively, he complained of numbness of the left cheek, followed by spontaneous burning and shooting pain, and tactile and thermal allodynia. Spontaneous pain and allodynia are the hallmarks of neuropathic pain. In this case, this syndrome can be traced to traumatic injury of the inferior orbital nerve. The key to diagnosis is sensory mapping. Although pain syndromes may spread, the area of true sensory loss corresponds to the injured nerve. Similar syndromes can be seen with injuries to the inferior alveolar nerve (“numb chin syndrome”) and to the superior orbital fissure (although other cranial nerves are invariably involved in the latter). If pain does not subside spontaneously, this neuropathic pain can be treated with antidepressants and anticonvulsants. Surgical exploration for a neuroma may be required.

Vignette A 28-year-old woman presented with electrical jolts of pain in the left lower jaw and a normal neurologic examination. She was symptom-free for about 6 months, when she began to notice similar pains in the right cheek.

Trigeminal neuralgia in a young person, particularly when bilateral, should prompt a search for MS. While it usually appears late in the disease, it is the most common painful presentation and eventually affects up to 4% of patients with MS (about 400 times the general prevalence).⁵ Surgical treatment is rarely an option and pharmacologic management is the same as in primary trigeminal neuralgia.

Facial pain due to cerebrovascular disease is more diffuse in localization. There is a dysesthetic quality to the pain. Lesions are most commonly found in the thalamus or in the trigeminothalamic pathways.

Vignette A 63-year-old man has undergone Mohs procedures for recurrent basal cell carcinoma of the face. He presents with 1 month of progressive numbness and pain across the right cheek, to the labial commissure. Examination shows sensory loss from the zygoma to the labial commissure, including the gums and the upper front teeth on the right. MRI scan of the brain is normal.

An MRI scan with thin cuts through the orbits revealed tumor extension into the inferior orbital foramen. It was due to direct extension of basal cell carcinoma along the nerve. More commonly, metastases to the base of the skull produce multiple cranial nerve syndromes, with a component of facial pain. Five classical syndromes⁶ include:

- Orbital syndrome: numbness or pain in the ophthalmic division of the trigeminal nerve, sparing the second and third division, and accompanied by varying degrees of oculomotor, trochlear, and abducens palsies. Proptosis is common.
- Parasellar syndrome: similar to the orbital, but the second division of the trigeminal nerve is involved and proptosis does not occur.
- Middle fossa syndrome: pain in any or all of the divisions of the trigeminal nerve, followed later by abducens and facial palsies.
- Jugular foramen syndrome: ipsilateral retroauricular, throat, or occipital pain, hoarseness, and dysphagia.
- Occipital condyle syndrome: severe pain in the occipital region. Even though it does not produce facial pain, it is included to complete the series and because it is often missed. All of these syndromes are most commonly seen in breast or prostate cancer patients and focal radiation provides palliation.

Vignette A 45-year-old woman complains of 2 days of severe and stabbing retroauricular and facial pain. She hears a clicking sound on swallowing and develops labial dysarthria. Over the next 48 hours, she develops a complete left facial palsy.

Aside from a good neurologic examination, this warrants a careful examination of the external auditory canal, looking for vesicles. Ramsay Hunt syndrome is herpes zoster infection of the geniculate ganglion and needs to be treated aggressively. Etiologic therapy includes a 7–10 day

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course of acyclovir 800 mg 5 times a day or famciclovir 500 mg 3 times a day (there is unclear evidence for benefit of any one antiviral or route of administration over another). A 3–7 day course of prednisone 60 mg per day reduces pain and inflammation and may have an influence on facial nerve functional recovery. Opioids may be needed for acute pain control and oxycodone can be titrated to effectiveness. Gabapentin can also be titrated rapidly and may provide some relief.

Vignette A 19-year-old man with recurrent bouts of otitis media presents with retro-orbital pain and diplopia. On examination, there is patchy sensory loss in the first 2 distributions of the trigeminal nerve and he has trouble abducting the ipsilateral eye.

The clinical triad of facial pain, diplopia, and otorrhea is indicative of infection of the petrous apex, or Gradenigo syndrome, an unusual complication of otitis media, previously treated with radical surgery that sacrifices the labyrinth. There are reports of successful conservative management with antibiotics, but recognition of the syndrome is key to emergency management. A similar and even more dangerous syndrome occurs just anterior to that site, when the cavernous sinus is infected and thrombosed (similar to the parasellar syndrome, cited above).

Non-neuralgiform headache disorders

Vignette A 45-year-old woman has had 5 years of severe bouts of pain on the left side of her head, eye, and cheek. They last from a few minutes to half an hour and are accompanied by tearing, eyelid swelling, and rhinorrhea. She can have up to 5 or 10 severe attacks during the day. She is occasionally awakened from sleep by these attacks.

Chronic paroxysmal hemicrania is a trigeminal autonomic cephalalgia (TAC) and an indomethacin-responsive syndrome. Events are invariably unilateral and rarely change sides. It differs from SUNCT in that the pain is of longer duration and there is a clear response to indomethacin. Relief from 150 mg or less per day is essentially diagnostic.⁷ Episodic paroxysmal hemicrania is a much less common form of the same syndrome. When the syndrome is unaccompanied by autonomic features, it is called idiopathic stabbing pain (previously ice-pick headache). Cluster headache, another headache/facial pain with autonomic features, is even longer lasting than CPH, with individual bouts lasting up to 90 minutes. They often occur at the same time of day, each day of the cluster. There may be a few attacks per day. Attacks are precipitated by alcohol intake, are unilateral, and rarely change sides. They resolve after a number of weeks (the cluster) and patients are headache-free for months to years. During the attack, the pain is so severe that, unlike migraine patients, cluster sufferers are unable to sit still and tend to pace around. Treatment with injectable sumatriptan reliably provides relief for the individual attack. High flow oxygen has also had its supporters. Prophylactic measures include calcium channel blockers and lithium. In severe or chronic cases, a tapering course of methylprednisolone can be effective to break the attack, followed by prophylactic measures.⁸

Question for rounds: What is the difference between the Horner syndrome seen in facial pain with autonomic symptoms and the Horner seen in Pancoast syndrome? Answer: a true Horner has ptosis, miosis, and anhidrosis. In these syndromes, there is hyperhidrosis.

Vignette A 36-year-old woman has chronic pain on the left side of her face. It radiates to the ear and temple. It is present upon awakening, subsides in the early part of the day, but returns

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after eating. Neurologic examination is normal, but there is tenderness to palpation on the cheek, and forced mouth opening produces pain in the ear and a loud click.

Dysfunction of the temporomandibular joint (TMJ) or the surrounding muscles produces unilateral pain and tenderness. It can be caused by bruxism during sleep or teeth clenching. In mechanical cases, a simple mouthguard can provide relief. It also occurs as a complication of rheumatoid arthritis and osteoarthritis. However, it is commonly associated with depression and other somatic symptoms requiring psychopharmacologic or psychotherapeutic interventions.

Vignette A 75-year-old woman complains of pain in the left face, jaw, and temple. It is markedly exacerbated by chewing and she often has to stop eating, because of the pain.

Giant cell arteritis (GCA) is a systemic disease that can produce facial pain. Jaw claudication is pathognomonic. An elevated sedimentation rate and temporal artery biopsy confirm the diagnosis. It is a medical emergency that must be treated with high doses of methylprednisolone.

Vignette A 30-year-old female gymnast presents with pain radiating up the left jaw, to the temple. It is throbbing in nature and exacerbated by neck movement.

The first task is to rule out structural disease of the carotid artery, such as dissection or aneurysm. Once that is eliminated, pain that is exacerbated by pressure over the carotid artery is called carotidynia. It can be treated with membrane stabilizing anticonvulsants or with migraine prophylactic drugs.

Vignette A 27-year-old woman presents with left maxillary pain, following a dental extraction. The pain is present all day, every day, and does not follow a nerve distribution. She had previously undergone numerous root canals and apicoectomies for dental or facial pain. Her examination is normal. Atypical facial pain (primary idiopathic facial pain) is a particularly vexing syndrome. Patients commonly undergo numerous dental procedures and unsuccessful treatments aimed at trigeminal neuralgia before the appropriate diagnosis is reached. The main job of the neurologist is to avoid any further destructive procedures. Antidepressant medications and supportive psychotherapy are useful interventions.

Vignette A 50-year-old woman presents with 6 months of burning pain in the mouth, gums, and lips. There are no clear exacerbating or relieving factors and it is present all day, everyday. Burning mouth syndrome can be caused by a number of systemic or local processes, such as xerostomia, candidiasis, gastric reflux, and inadequate oral hygiene.⁹ Rarely, there are vitamin or mineral deficiencies that can be easily corrected. However, by the time these patients reach a neurologist, most of those issues have been ruled out. There are no well-controlled studies to inform treatment. Antidepressant medications may paradoxically worsen the condition, because of dry mouth. Local rinses and artificial saliva may be helpful, along with behavioral and supportive psychotherapy.

Vignette A 32-year-old woman complains of constant pain across the entire right side of the face, from just above the hairline to the neck. Examination shows numerous tender points on both sides of the neck, paraspinal regions, thighs, and elbows. She has a history of dysmenorrhea, irritable bowel syndrome, insomnia, chronic fatigue syndrome, fibromyalgia, and multiple chemical sensitivity syndrome. Extensive medical and neurologic workups have been unrevealing. When asked about depression, she states, "Wouldn't you be depressed with all of these problems?" Somatoform disorder is one of the medically unexplained syndromes.

Table Facial pain syndromes

Syndrome/IHS	Location	Character	Duration	Triggers	Signs	Treatment
TN/13.1.1	V3>V2>V1	Electrical	Seconds	Cold, touch	None	AED, baclofen, surgery
GPN/13.2.1	Throat/ear	Same	Same	Swallowing	Syncope	Same
Intermedius/13.3	Ear/face	Same	Same	None	None	Same
SUNCT/3.3	Eye/temple	Same	Same	None	Autonomic	Refractory
Traumatic/13.1.2	Single nerve	Numb, burning	Variable	Cold, touch	Sensory loss	
Central/13.8.1	Any	Burning	Variable	Cold, touch	Sensory loss, allodynia	AED, TCA, SNRI
Malignant/13.12						
Orbital	V1	Variable	Constant	Variable	Proptosis, V1, ocular paresis	Primary lesion
Parasellar	V1 & V2	Variable	Constant	Variable	Ocular paresis, V1 & V2	Primary lesion
Middle	V1-V3	Variable	Constant	Variable	VI & VII paresis	Primary lesion
Jugular	Throat	Stabbing	Seconds	Swallowing	IX, X, XI	Primary lesion
Occipital	C2	Variable	Variable	Movement	Tenderness over condyle	Primary lesion
Ramsay-Hunt/13.15.1	Ear, V1-V3	Electrical, burning	Seconds- constant	Touch	Zoster eruption: V, VII, VIII	Antivirals, steroids
Gradenigo/13.12	V2-V3	Aching	Constant	None	VI paresis	Antibiotics
CPH/EPH/3.2.2/3.2.1	Eye, temple	Stabbing	2-20 min	None	Autonomic	Indomethacin
Cluster/3.1.1	Eye, temple	Stabbing	20-90 min	Alcohol	Autonomic	O ₂ , Triptans
ISP/4.1	Eye, temple	Stabbing	Seconds	None	None	Indomethacin +/-
TMJ/11.7	Diffuse	Aching	Constant	Chewing	Tenderness	Appliance, TCA, SNRI
GCA/6.4.1	Temple/jaw	Aching	Constant	Chewing	Tenderness	Steroids
Carotidynia/5.6	Diffuse	Aching	Constant	Carotid press	None or Horner	AED, migraine meds
PIFP/13.18.4	Multifocal	Variable	Constant	Variable	None	Chronic pain meds
BMS/13.18.5	Mouth	Burning	Constant	Anxiety	None	Local, chronic pain
Somatoform/12.1	Diffuse	Variable	Constant	Anxiety	None	Psychopharmacology, psychotherapy

Abbreviations: AED = antiepileptic drug; BMS = burning mouth syndrome; CPH/EPH = chronic paroxysmal hemicrania/episodic paroxysmal hemicrania; GCA = giant cell arteritis; GPN = glossopharyngeal neuralgia; IHS = International Headache Society classification number; ISP = intermittent stabbing pain; PIFP = primary idiopathic facial pain; SUNCT = short-lasting unilateral neuralgiform headache with conjunctival injection and tearing; TCA = tricyclic antidepressant; TMJ = temporomandibular joint dysfunction; TN = trigeminal neuralgia; SNRI = serotonin nonspecific reuptake inhibitor.

Patients have difficulty expressing emotional issues in appropriate terms and convert them into physical complaints. The issue here is to recognize the syndrome, but avoid confronting the patient with a diagnosis and to form a collaborative therapeutic bond.¹⁰

Management of facial pain requires attention to historical detail, meticulous sensory examination, and sensitivity to psychological factors.

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