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Expert Opinion

CME

Persistent Idiopathic Facial Pain

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The new term, "persistent idiopathic facial pain," is an improvement over the older vague one, "atypical facial pain," which was introduced by Frazier and Russell in 1924.[1]

CLINICAL HISTORY

A 57-year-old woman was seen for a third neurological opinion with a 1 to $1\frac{1}{2}$ year history of facial pain. She described a dull ache or burning around the medial canthus of the left eye, the left side of the nose, the left naso-labial fold, and the left side of the lip. The pain typically started about 8 AM and had an intensity of 9/10 all day. Amitriptiline, nortriptyline, gabapentin, zonisimide, levetiracetam, duloxetine, and tramadol all in moderate-to-high doses have proven ineffective. Hydrocodone, it was reported, moderately decreased the pain. A left upper molar extraction did not help. Cryoablation of the left infraorbital nerve was of no benefit.

An MRI scan of the brain with and without contrast was normal including views of the trigeminal nerve. A CT scan of the sinuses revealed mild hypertrophy of the inferior turbinates and a leftward projecting bony nasal septal spur. A second CT scan of the sinuses 11 months later was interpreted as normal. An erythyrocyte sedimentation rate, antinuclear antibody, RA factor, and Sjogren's antibodies were all normal. She has been evaluated by 3 neurologists, 3 ENT surgeons, a plastic eye surgeon, an anesthesia pain specialist, a general dentist, and an endodontist. One ENT physician told her that sinus surgery had a 50:50 chance of improving her symptoms but another advised against further surgery.

Questions.—What is the diagnosis? What are the treatments available? What is the prognosis?

EXPERT OPINION

This case should be considered within the group of chronic orofacial pain (COP) syndromes, a common disorder affecting more than 10% of the adult population.[2] In spite of its frequent occurrence, the etiology and pathophysiology of these disabling disorders are still poorly understood and differential diagnosis is a demanding challenge that often involves several specialists, like neurologists, odontologists, and ENT surgeons. In fact, the definition itself of COP is broad and includes several different entities: atypical odontalgias, masticatory pain, temporomandibular joint disorders, oral dysesthesias, and atypical facial pain,[3] recently

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redefined by the International Headache Society (IHS) as persistent idiopathic facial pain (PIFP).[4]

The differential diagnosis is often challenging as these conditions often coexist[5] as other disorders such as trigeminal neuralgias and primary headaches need to be considered. Aside from PIFP, however, all these pathologies are associated with other signs or symptoms that may help the clinician. Temporomandibular joint syndrome is associated with tenderness of the jaw joint or muscles and is aggravated by chewing or prolonged talking; incomplete jaw opening or clicking is often found. In atypical odontalgia the pain is strictly localized in the teeth or tooth socket without any organic cause. Pain in trigeminal neuralgia is short lasting, localized in the distribution of a trigeminal branch and triggered by stimulation of trigger zones while the clinical picture of primary headaches is quite different from COP and should not represent a difficult task for neurologists. Also other disorders, such as Tolosa Hunt syndrome, should be considered in the differential diagnosis, but these are associated with neurological signs, which are always missing in COP.

Within the group of COP syndromes, PIFP certainly represents the hardest diagnostic problem. The IHS provides 4 diagnostic criteria for this diagnosis: the pain must be present every day and for most of the day,[3] it must be deep, dull, and not well localized in a limited zone on the side of the face, it should not be associated with focal neurological signs or loss of sensibility and, most importantly, no abnormality should be found with laboratory and neuroradiologic investigations, which must include imaging studies of the face and jaw.[5] The case described may then be diagnosed as affected by PIFP, as the characteristics of the pain fulfill the criteria, there are no associated signs and clinical and laboratory investigations seem to have been broad and accurate enough to rule out other conditions. In addition, the demographic features are consistent with the diagnosis, since PIFP is more frequent in postmenopausal women. The finding of septal deviation is irrelevant and does not rule out the diagnosis. The diagnostic work-up, however, should be completed by a radiologic examination of the chest, since in rare occasions PIFP may be the presenting symptom of a lung cancer. Digital clubbing, increased erythrocyte sedimentation rate, and hypertrophic osteopathy are associated signs, but facial pain may be the only manifestation of the disease.[6]

Unfortunately, the treatments available for this condition are not always effective, as highlighted by the case described. Surgical procedures, including trigeminal microvascular decompression, are ineffective and should be avoided. Indeed, one of the main goals of a prompt diagnosis is to spare patients from futile and potentially harmful surgical treatments. Although the pathophysiology of the pain is still unknown, empiric pharmacological treatment with tricyclic antidepressants, antiepileptic drugs such as lamotrigine and selective serotonin reuptake inhibitors (SSRIs) have all been attempted. Amitriptyline at a dose of 25 to 100 mg/day was shown to be effective and should be considered as the first choice.[7] Positive results have been reported also with venlafaxine,[8] sumatriptan, and intranasal cocaine.[9] Although no trials with opioid analgesics are available, the response to hydrocodone reported in this case is interesting, as it suggests a central origin of PIFP. Indeed, recent work by Kanpolat and colleagues[10] showed pain relief after percutaneous trigeminal tract and nucleus ablation, also suggesting that central, rather than peripheral mechanisms are dominant in this disorder.

For patients who fail to respond to all drugs available, the prognosis is poor. The pain may spontaneously subside for some time but is very likely to recur in the same area. Other pain relief strategies, such as hot and cold compresses, acupuncture, biofeedback, and dental splints may be attempted alone or in association with tricyclic antidepressants or venlafaxine. Should all these treatments fail, when the pain is responsible for a very significant disability and all other procedures are ineffective, central analgesic surgery could be proposed in highly selected centers. The patient presented in this case might be a candidate for such an attempt, considering the relatively young age and the intensity and duration of facial pain. The time of follow-up should probably be increased, however, and more therapeutic strategies, both pharmacologic and nonpharmacologic, should be tested before suggesting such an invasive procedure.

Conflict of Interest: None declared

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