

A Review of Atypical Facial Pain

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Background: Atypical facial pain occurs in the territory of the trigeminal nerve, but the discomfort is not typical of trigeminal neuralgia. It may be as severe as trigeminal neuralgia, but its pattern and quality are different. Whereas trigeminal neuralgia is characterized by quick episodes of jabbing or lancinating pain, atypical facial pain is usually burning, aching, dull, or crushing. Moreover, an individual atypical facial pain attack always lasts longer than a few seconds and usually lasts minutes or hours (if not continuous). The distinction is important for making treatment decisions, because surgery, usually rhizotomy or vascular decompression, is highly effective for trigeminal neuralgia, whereas surgery is not appropriate for atypical facial pain.

A variation of atypical facial pain is discomfort similar to that of trigeminal neuralgia (e.g., lancinating) but atypical in location (e.g., far lateral on the face or in the occipital area). Some writers classify these as neuralgias of other nerves (in these examples, superficial temporal or occipital).

Frequency:

In the US: Although accurate figures are not possible because of the lack of agreement on criteria for classification, atypical facial pain is approximately one fifth as frequent as trigeminal neuralgia. The incidence of trigeminal neuralgia is 4-5 per 100,000 per year.

Race: No racial predilection is known.

Sex: Atypical facial pain affects both sexes with approximately equal frequency.

Age: The disorder mainly affects adults and is rare in the young.

History: Atypical facial pain must be distinguished from trigeminal neuralgia. It also must be distinguished from temporomandibular joint (TMJ) syndrome, migraine, and cluster headache.

- Trigeminal neuralgia is characterized by severe bursts of pain in one or more branches of the trigeminal nerve.
 - The bursts are quick, repetitive jabs of pain (lancinations). Each pain episode is only an instant in duration; the episodes recur irregularly many times a minute.
 - Trigeminal neuralgia episodes are not in synchrony with the heartbeat or pulse (usually slower).
 - The patient may wince, twitch, or cry out when a series of pain jabs occurs.
 - The pain is excruciating.
- Atypical facial pain may be as severe as trigeminal neuralgia, but its pattern and quality are different.
 - Pain episodes are always longer than a few seconds, lasting minutes to hours, and are sometimes continuous. The duration of the individual episode helps to make the distinction.
 - The pain is dull, aching, crushing, or burning.
 - Occasionally, patients use terms such as "sharp" or "knifelike" to describe the pain.
- TMJ syndrome is recognized by the following:
 - The location and quality of pain may be similar to those of atypical facial pain.
 - TMJ syndrome is characterized by focal tenderness of one or both TMJs and aggravation of Pharmacologic treatment is usually unsuccessful.

- Treatment of TMJ syndrome (by an oral surgeon) often is directed at either the arthropathy of the joint itself or at fatigue and spasm of the pterygoid and temporalis muscles.
- The following signs and symptoms recognize migraine. Diagnosis of migraine may be acceptable if some of these features are absent, but generally at least 3 should be present. The essential feature is recurrent, severe headache with normal neurologic examination and blood pressure.
 - o Female-to-male ratio is 3:1.
 - Pain is throbbing and synchronous with the heartbeat or pulse, usually on only one side of the head.
 - o Headache may be accompanied by scalp tenderness.
 - o Photophobia and sonophobia are common associated symptoms.
 - o Nausea often accompanies migraine.
 - o Patients often describe an aura at the onset.
 - o Sleep (if it can be achieved) can abort a migraine.
 - o Pharmacologic therapy, typically with ergotamine, isometheptene, or serotonin receptor agonists of the triptan family, is highly effective for migraine, almost to the point of diagnostic specificity. These agents are ineffective in atypical facial pain.
- Cluster headache, in terms of effective pharmacologic therapy, is closely related to migraine. Pain location and quality sometimes may be confused with atypical facial pain. Characteristics of cluster headaches include the following:
 - o Male-to-female ratio is 6:1.
 - o Pain awakens patient from sleep.
 - o Onset of pain is sudden.
 - o Episodes typically last minutes (but can last hours).
 - o Pain is intense, crushing, or burning.
 - o Pain recurs frequently up to several times a day in clusters of several days to weeks.
 - o Strictly unilateral, the pain often is associated with ipsilateral conjunctival injection and nasal congestion.
 - o Unlike migraine, cluster headache is very responsive to inhaled oxygen (i.e., abortive) therapy.

Physical:

- Neurological and physical examinations are usually normal.
- Trigger points, where pain is evoked by stimulation, are rare.
- The supraorbital or infraorbital foramen may be locally tender.
- Significant tenderness at one or both TMJs would favor the diagnosis of TMJ syndrome.

Causes:

- Atypical facial pain is usually without a specific cause. However, injury of any peripheral or proximal branch of the trigeminal nerve due to facial trauma or basal skull fracture can produce the disorder.

Imaging Studies:

- A brain MRI scans with gadolinium is the imaging modality of choice, although most studies will be normal. Examples of possible lesions include neuromas or cysts involving the trigeminal root or its distal branches outside the brain and malignancies of the skull base compressing or invading a branch of the nerve.
- CT scan of the head with contrast has a lower yield than MRI because of its poorer resolution of the brain stem and cranial nerves.

Medical Care:

- Medical treatment of atypical facial pain is less satisfactory than that of trigeminal neuralgia, reflecting the etiologic, symptomatic, and nosologic heterogeneity of the two conditions. Of the non narcotic drugs, tricyclic antidepressants give best results; phenytoin is of intermediate effectiveness, and carbamazepine is least effective. Any of these may be best in a particular patient.
- Of the new anticonvulsants, gabapentin and lamotrigine show promise, but their efficacy relative to the older drugs has not been established.
- In patients whose pain is typical of trigeminal neuralgia (e.g., lancinating) but atypical in location, carbamazepine is usually most effective, phenytoin of intermediate effectiveness, and tricyclics the least effective; gabapentin and lamotrigine retain their broad-spectrum positions.
- For neuropathic pain in any location, the same principle applies. For lancinating pain, the usual order of effectiveness is carbamazepine first, phenytoin intermediate, and tricyclics last. The reverse order is true for burning and other pain types.
- Aching joint pain and abdominal cramps are least likely to respond to these drugs. In all situations, the quality of pain is more important than its location.
- Narcotic treatment is sometimes appropriate as an adjunct but requires careful supervision, usually in collaboration with the patient's pharmacist. The most effective program is to provide a monthly ration of narcotic that is strictly enforced and documented. Older patients fare better with narcotics since the risk of addiction is low in patients older than 70 years.

Interventional Therapy:

- Maxillary, Infraorbital nerve, and sphenopalatine ganglion block is sometimes effective, which may be followed by a neurolytic procedure, usually pulse radiofrequency.
 - Because of the risk of anesthesia dolorosa, surgery should be reserved for cases of medical intractability with repeated demonstration of pain interruption by nerve block.
 - o Nucleus Caudalis stimulation with a laminotomy lead may be effective if initial trial is successful in suitable patient.

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