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Atypical facial pain: clinical considerations and differential diagnosis

Abstract Atypical facial pain (ATFP), recently defined as persistent idiopathic facial pain by the revision of the Classification of the International Headache Society (IHS), is a poorly understood condition, which still lacks clear diagnostic criteria and proper treatment. The pain is described as “persistent facial pain that does not have the characteristics of cranial neuralgias and is not attributable to another disorder”. In general, however, according to the IHS criteria, a diagnosis of ATFP is possible when the pain in the face is present daily and persists for most or all of the day. The pain is confined at onset to a limited area on one side of the face, often in the nasolabial fold or side of the chin and may spread to the upper or lower jaw or a wider area of the face of neck and is deep and poorly localised. It is not associated with sensory loss or other physical signs. Laboratory investigations including X-ray of face and jaws do not demonstrate relevant abnormality. Pain may be initiated by operation or injury to face, teeth or gums but persists without any demonstrable local cause. But, the definition and the diagnostic criteria are over-simplified when

we face the reality of the clinical practice. Many different disorders may be included in this diagnostic category, making differential diagnosis very complex. Diagnosis of ATFP is therefore, usually, a process of elimination. A targeted history and an accurate examination are crucial to correctly classify this facial pain.

Key words Atypical facial pain • Persistent idiopathic facial pain

Chronic orofacial pain is a common and debilitating problem affecting at least 10% of the adult population and 50% of the elderly population [1]. It is a poorly understood condition that represents a challenge for clinicians in terms of diagnosis and treatment. This term refers to many different disorders, such as temporomandibular joint disorders, headaches, neuralgias, atypical facial pain (ATFP), pain of mucosal origin and dental pains. Patients suffering from orofacial pain may, therefore, seek and receive treatment from different practitioners, making this problem multidisciplinary. In 1999, Woda and Pionchon [2] proposed a unified concept of chronic idiopathic orofacial pain, including in this group ATFP, atypical odontalgia, masticatory pain, temporomandibular joint disorders and oral dysaesthesia. They stated that these conditions share a common clinical picture: they are more common in women, the pain does not follow a nervous pathway and is present for months, it returns periodically in the same form over several months or years, it has no major paroxysmal character, it is present through all or part of the day, it is absent during sleep and psychological factors are often present. Harris and Feinmann [3] suggested that the four syndromes of chronic idiopathic facial pain (ATFP, atypical odontalgia, temporomandibular joint disorders and oral dysaesthesia) often coexist or occur sequentially. Maier and Hoffmeister [4] included sympathetic dystrophy in the diagnosis of ATFP.

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One of the disorders included in the group of chronic orofacial pain is ATFP. The term “atypical” was first used in 1924 by Frazier and Russell [5] and applied to patients with facial pain that failed to respond to neurosurgical treatment. In the past years many clinicians have tried to describe ATFP in different ways, with the only result being confusion and lack of clarity. The aetiology of ATFP is so controversial that some early authors refused even to accept that the condition existed [6].

In general, patients with this condition complain of a steady, generally unilateral and not well localised pain. The quality of the pain is usually described as deep, constant, aching, pulling or crushing. There are not paroxysms of short duration; the pain is usually present all day and every day. Although patients complain of excruciating pain, they do not usually appear to be in severe pain. ATFP is not triggered by any of the precipitating factors typical of neuralgias. Most of the patients with ATFP complain of other symptoms, including headache, neck and backache, dermatitis or pruritis, irritable bowel and dysfunctional uterine bleeding [1].

ATFP still lacks proper diagnostic criteria. The International Headache Society, in its revised classification in 2004, included ATFP as a “previously used term” under the classification heading “persistent idiopathic facial pain” (code 13.18.4) [7]. The society described this as ‘persistent facial pain that does not have the characteristics of cranial neuralgias and is not attributable to another disorder’. According to these criteria, a diagnosis of ATFP is possible when the pain in the face is present daily and persists for most or all of the day. The pain is confined at onset to a limited area on one side of the face, often in the nasolabial fold or side of the chin and may spread to the upper or lower jaw or a wider area of the face of neck and is deep and poorly localised. It is not associated with sensory loss or other physical signs. Laboratory investigations including X-ray of face and jaws do not demonstrate relevant abnormality (but it does not mention specifically which types of investigations or radiographs should be used). Pain may be initiated by operation or injury to face, teeth or gums but persists without any demonstrable local cause. The pain around the ear or forehead may precede the diagnosis of an ipsilateral pulmonary neoplasm that causes a referring pain because of a vagus nerve invasion.

The society also described atypical odontalgia as a continuous pain in the teeth or tooth sockets in the absence of an identifiable cause, and also laid down criteria for a diagnosis of burning mouth.

The International Headache Society proposed these criteria but there is still uncertainty and no greater clarity. In the old classification of 1988 [8], the IHS described ATFP (code 12.8) as a persistent pain that does not have the characteristics of cranial neuralgias and is not associated with physical signs or a demonstrable organic cause. The diagnostic criteria were the same as the new one.

The aetiology of ATFP is still unknown. Some risk factors have been suggested as aetiologic factors, however any one can be considered as the causal. The role of female hormones has been implicated, as ATFP is much more common in women than in men and because of the physiologic and therapeutic modification of oestrogen levels in patients with these pain conditions. Osteoporosis, which appears with menopause, and neuralgia-inducing cavitation osteonecrosis have been linked to ATFP. The presence of psychosocial factors is also a common feature, but it is not known whether these are causal or whether the pain induces the psychosocial problem. In some cases, infection of the sinuses or teeth, or minor nerve trauma can also be considered as risk factors. However, none of the above factors can be considered as the sole aetiologic factor. Different neuropathic mechanisms may be at work: nociceptor sensitisation, phenotypic changes and ectopic activity from the nociceptors, central sensitisation possibly maintained by ongoing activity from initially damaged peripheral tissues, sympathetic abnormal activity, alteration of segmental inhibitory control, and hyper- or hypoactivity of descending controls [9].

ATFP must be distinguished from typical facial pains, primary headaches and dental pains. Table 1 summarises the characteristics of the most common disorders that have to be differentiated from ATFP. However, other rare conditions need to be considered when confronted by facial pain. In particular, trigeminal neuralgia is characterised by severe, quick bursts of pain in one or more branches of the trigeminal nerve. The bursts last only an instant and they recur irregularly during the day. The pain is described as excruciating and it is often triggered by facial movement, change of temperature or touching the face at a specific point (trigger point). Intensity is severe. It affects generally older people, with only a slight predominance in women. Sometimes patients with trigeminal neuralgia can have additional symptoms of ATFP [10]. The treatment consists, in general, of anticonvulsants.

Temporomandibular joint (TMJ) syndrome is characterised by focal tenderness of one or both TMJs and aggravation of pain by chewing, talking and lateral jaw movements. The quality of pain can be similar to that of ATFP; pain can be dull or stabbing. Intensity is moderate. The treatment consists of NSAIDs and surgery.

A relevant but not often considered condition is carotidynia. It is a chronic pain syndrome characterised by pulsating pain that arises from the carotid artery and radiates up the neck to include pain in the ipsilateral face, ears, jaws and teeth. Diagnosis is made by palpation of the carotid artery, which may elicit or increase the pain.

ATFP is usually without a specific cause. However, sometimes injury of branch of the trigeminal nerve due to facial trauma or basal skull fracture can produce the disorder. Imaging studies are normal but, on rare occasions, ATFP could be the presenting symptom of lung cancer. In

Table 1 Differential diagnosis of facial pain

Facial pain	Location	Quality	Intensity	Duration	Aggravating factors	Other characteristics
Trigeminal neuralgia	Second and third division of trigeminal nerve, unilateral	Lancinating, stabbing, burning, electric shock-like	Severe	Seconds	Touching or washing the face, eating, chewing, smiling, talking, blowing nose, yawning, brushing the teeth, shaving	–
Post-herpetic neuralgia	Usually ophthalmic or maxillary branch of fifth nerve, unilateral	Aching, burning, jabs	Severe	Constant	Contact, movement	–
Atypical facial pain	One side of the face, nasolabial fold or side, chin, jaw, neck; poorly localized	Deep, aching, pulling, crushing	Moderate to severe	Constant	–	–
Temporomandibular joint syndrome	Jaw, mandible, preauricular region	Dull, stabbing	Moderate	Minutes to hours	Palpation of the jaw joint or the muscles of mastication, mastication, prolonged talking	Incomplete jaw opening, clicking on lateral movements
Tolosa-Hunt syndrome	Mainly retro-orbital, unilateral	Aching	Severe	Constant	–	Opthalmoplegias, sensory loss over forehead, ptosis, pupil usually spared
Raeder paratrigeminal syndrome	Fronto-temporal and maxilla, unilateral	Deep, lancinating	Severe	Constant	–	Ptosis, miosis
Carotidynia	Face, ear, jaws, teeth, upper neck, unilateral	Throbbing	Moderate	Constant	Compression of common carotid	–
Cluster headache	Orbital, suborbital, and/or temporal, unilateral	Variable	Severe	Minutes to three hours	Alcohol, stress, heat, cold, bright light	Autonomic symptoms
Tension-type headache	Frontotemporal and/or parietal, bilateral	Pressing, tightening	Mild to moderate	Minutes to days	–	–
Migraine	Frontotemporal, orbital, usually unilateral	Pulsating, throbbing	Moderate to severe	Hours	Physical activity	Aura in migraine with aura
Pulpitis	Teeth, other parts of the face, not well localized	Throbbing	Slight to severe	Minutes to hours	Mechanical, foods, cold, heat, suit	–
Orofacial tumours	Variable	Variable (atypical)	Severe	Slight to severe	Jaw movement	Frequently neurological signs, WBC abnormalities

this case, the facial pain is almost always unilateral, and is most commonly localised to the ear, the jaws and the temporal region. The pain is frequently described as severe and aching, and may be continuous or intermittent. Aggravation and expansion of the pain, digital clubbing, increased erythrocyte sedimentation rate and hypertrophic osteopathy may contribute to the diagnosis. Referred pain, due to inva-

sion or compression of the vagus nerve, as well as paraneoplastic syndrome secondary to the production of circulating humoral factors by the malignant tumour cells, is implicated in the pathophysiology of facial pain associated with non-metastatic lung cancer. Radiotherapy and tumour resection with vagotomy are very effective in aborting the facial pain. Thus, lung cancer should be included in the dif-

ferential diagnosis of facial pain that is atypical and/or refractory to treatment [11].

Treatment of ATFP can be difficult and unsatisfactory. It consists mainly in patients' education and in pharmacotherapy with tricyclic antidepressants. Some anticonvulsants (phenytoin, carbamazepine, gabapentin, lamotrigine) can be less effective. Analgesics and surgical procedures such as microvascular decompression are not effective. Other pain relief strategies include hot and cold compresses, acupuncture, biofeedback and dental splint.

Diagnosing ATFP is not easy. It is not unusual that patients with ATFP undergo numerous dental procedures, see many doctors and undergo many medical tests before being successfully diagnosed and treated. A diagnosis of ATFP is usually a process of elimination. When a patient complains of constant facial pain restricted to one side of the face, the physician must first rule out any other conditions. A targeted history and an accurate examination are crucial to correctly classify this facial pain [12].

Despite the recent classification of the IHS [7], older terminology still seems to be widely used. A recent survey conducted in UK [13] aimed to compare views of UK specialists in dentistry and medicine who deal with chronic facial pain on their use of the term ATFP, how they reach this diagnosis or their alternative preferred equivalent diagnosis and what treatment they recommend. The study showed that 89% of practitioners from the various specialties in the UK who treat facial pain still use the term ATFP. To make the diagnosis of ATFP, 48% of specialists excluded other kinds of pain, 22% said they used certain criteria and 17% said that they used both methods. Although most of these specialists mentioned that they used the diagnosis-by-exclusion approach, it is not clear how they did this. Despite the fact that strong psychological factors may be present, there was no tendency among the various specialists to refer these patients to psychiatrists or for psychotherapy.

Management of ATFP requires a specific knowledge of the diagnostic criteria. This is extremely important in the process of differential diagnosis and in the choice of the most accurate and effective therapeutic treatment strategy. Facial pain has appreciable impact on the population. Many problems remain unsolved because causal mechanisms are

unclear. Compounding the problem, the nosology is complicated by liberal uses of the terms "atypical" and "idiopathic", which are vague and overlapping.

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