**Atypical facial pain**

**A diagnostic challenge**

**BACKGROUND** Facial pain is a relatively frequent cause of presentation to both general medical and dental practitioners. Although in the vast majority of cases the cause is dental disease or temporomandibular joint dysfunction, the remaining patients are often difficult to diagnose and treat.

**OBJECTIVE** This article discusses the differential diagnosis of facial pain and presents three cases of atypical facial pain.

**DISCUSSION** A detailed history, clinical examination, imaging and laboratory investigations may be required to establish the cause of atypical facial pain. An assessment of the patient’s mental state is mandatory, as depression or psychological overlay is common. In a small number of cases, the diagnosis remains unclear despite detailed investigation. These patients may have their symptoms ameliorated by empirical pharmacological therapy.

Facial pain, especially pain around the ear, is a relatively common surgical problem. It is frequently misdiagnosed, subjecting the patient to prolonged and costly treatment. Undoubtedly dental and oral diseases including temporomandibular joint (TMJ) dysfunction are the commonest causes, and atypical facial pain should be regarded as dental in origin until proven otherwise. However, there are many other causes (Table 1). Usually, the aetiology can be elicited by completing a careful history and physical examination. An assessment of the patient’s mental state is mandatory as depression or psychological overlay is common, especially in elderly women, and may be the primary cause or amplify an existing problem causing facial pain such as inadequate dentures.

Atypical facial pain is commonly constant, unremitting and centred over the maxilla. Nasopharyngeal carcinoma, vascular disease and thalamic infarcts can all produce pain around the face, but are frequently not identified by a standard physical examination. It is in these cases, and in cerebral neoplasms, that magnetic resonance imaging (MRI) and computerised tomography (CT) imaging with enhancement have proved most valuable.

There is a group of patients in whom the diagnosis remains elusive, despite exhaustive investigations. In such cases an empirical trial of carbamazepine or amitriptyline may ameliorate the symptoms.

**Anatomical aspects**

The face is richly endowed with pain fibres, and nowhere is the phenomenon of referred pain better illustrated than in this region of the body. Pain is frequently localised to the side of the face from a number of sites (Table 2) including upper cervical spine (C2, C3) lesions via the greater occipital, lesser occipital, and great auricular nerves. Therefore, a careful examination of the cervical spine is mandatory.

Dental caries in a lower molar tooth or ulceration of the pharynx, tongue or buccal mucosa may be referred to the ear via the auricular-temporal branch of the trigeminal nerve, tympanic branch of the glossopharyngeal nerve, or the auricular branch of the vagus nerve. In addition, in paranasal sinus disease, nasopharyngeal carcinoma, and cerebrovascular disease, pain may be experienced in the face or forehead. Otic causes are usually readily elucidated with the exception of baro trauma where recent exposure to atmospheric pressure change may not be volunteered during questioning. As facial pain...
does not cross the midline, a complaint of bilateral pain should alert the clinician to the possibility of a psychosomatic cause.

Case studies

The need for a high index of clinical suspicion is illustrated in the following cases studies.

**Case 1 – Mr AB**

Mr AB, aged 73 years, is a retired engineer. He had been in good health until he developed pain in both ears, worse in the left ear. The pain became quite severe while holidaying in the Northern Territory. He consulted a general practitioner who diagnosed ‘tropical ear’ and prescribed combined antibiotic/steroid/antifungal drops. The pain worsened, so he sought a second opinion, which matched the first. Mr AB became anorexic and depressed as he was unable to sleep due to the pain and inability to lie comfortably on either side. He returned home prematurely.

His family doctor referred him to a plastic surgeon who considered the problem was TMJ dysfunction and arranged a dental consultation. The dentist confirmed the diagnosis and fitted an occlusal oral splint. However, Mr AB’s condition continued to deteriorate and he was referred for another opinion. Questioning elicited that the pain was continuous, and in addition, his jaw ached when chewing. He had also noticed recent visual deterioration that he attributed to aging.

Examination revealed a normal tympanic membrane and canal. There was no evidence of TMJ dysfunction, oral, paranasal sinus or cervical spinal disease. However, central nervous system examination elicited hyperaesthesia over the temporal region bilaterally and there was some tenderness over the left superficial temporal artery (STA). Investigations (erythrocyte sedimentation rate [ESR] of 85, and a positive STA biopsy) confirmed the diagnosis of temporal arteritis.

Mr AB was immediately started on prednisolone and a week later the pain was much relieved.
Discussion

Case 1 – Mr AB

Pain around the ear is a relatively common condition, and while many cases have an aural cause, temporal arteritis should always be considered, especially in elderly patients as it occurs at a rate of 79 per 10,000 persons aged over 50 years.\(^3\) Temporal arteritis is a vasculitis involving the media of medium sized arteries causing necrosis of the media and fragmentation of the internal elastic lamina. The onset can be abrupt and is characterised by symptoms – as in this case – and fever. Pain on chewing results from claudication of the masticatory muscles and can be confused with pain emanating in the TMJ.

Eye symptoms are common and vasculitis of the ophthalmic artery can occur leading to ischaemic optic atrophy and blindness if the condition is untreated. Symptoms respond dramatically to steroids, which should be started on clinical suspicion and a raised ESR, as delay until a positive temporal artery biopsy is obtained may result in permanent visual impairment.\(^4\)

A diagnosis of TMJ dysfunction is frequently made in cases of pain around the ear, but should only be made if there is pain with mandibular movements, tenderness over the joint or masticatory muscles, or a history of jaw locking or limited opening.

Case 2 – Mr JW

Mr JW, aged 76 years, is a retired executive with a past history of hypertension and noninsulin dependent diabetes (controlled by diet), referred by his local doctor for assessment of numbness of the left side of his lower lip and lancing left sided facial pain. The provisional diagnosis was trigeminal neuralgia.

Mr JW stated that he had been edentulous for 50 years and had worn the same dentures for 15 years. He is a nonsmoker and drinks little alcohol. He explained that the pain only occurred when eating, although the numbness had been continuous for 3 months and appeared to be increasing. He was concerned that he might have a serious underlying disease as his mother had died from pharyngeal cancer.

Examination revealed an edentulous mouth with full dentures being worn. On their removal, marked alveolar bone resorption was evident. A tender area was elicited on palpation of the mandible in the region of the left mental foramen. Neurological examination was normal apart from revealing paraesthesia in the distribution of the left mental nerve. An orthopantomogram showed no bony lesion, however, as a result of alveolar bone resorption, the left mental foramen had come to lie close to the crest of the alveolar ridge.

A diagnosis of nerve compression due to pressure of the denture on the mental nerve was made and Mr JW was instructed to leave his lower denture out for 4 weeks. His symptoms resolved during this time and he was referred to his dentist for new dentures.
Discussion

Case 2 – Mr JW

The integrity of alveolar bone depends on the presence of teeth. When extracted, the supporting bone resorbs so that in time the mental foramen, initially approximately 1 cm inferior to the crest of the alveolus, comes to lie under the denture flange. Impingement of the denture on the mental nerve can occur causing a neuropathy with pain each time the nerve is compressed.

Trigeminal neuralgia causes lancing pain in the second or third division of the nerve distribution. It is precipitated by touching or moving the trigger area of the face. However, it differs from the pain in this case, as there is commonly a refractive period after nerve stimulation when repeated movement causes no pain. Chronic paroxysmal hemicrania is a rare cause of severe intermittent pain in the upper face, forehead, and temple. It is frequently accompanied by nasal stuffiness or lacrimation, can occur spontaneously many times a day, and last up to 15 minutes.

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Case 3 – Mrs JA

Mrs JA, aged 73 years, is a retired nurse who was referred for assessment of left sided facial pain involving the tongue, lower jaw, cheek and lip. The pain was increasing and in addition, she had numbness of the above structures. Mrs JA's medical history was long and complex, and included carcinomas of the breast and ovary, Crohn disease, temporal lobe epilepsy, and two minor strokes. She experienced frequent headaches, had restless hot legs, pain in both feet when wearing shoes, and complained of increasing deafness. Mrs JA was confined to a wheelchair and her social situation was unsatisfactory. She was, understandably, suffering from depression.

Neurological examination revealed paraesthesia involving the sensory distribution of the mandibular division of the left trigeminal nerve, although motor function was not impaired. Both feet and tibia were warm and there was some tenderness of the left tibia.

A provisional diagnosis of Paget disease involving the base of the skull and possibly the leg was made. The diagnosis was supported by a raised alkaline phosphatase and abnormal imaging findings. A lateral skull radiograph showed thickening of the calvarium with widening of the medullary space and blunting of the apices of the petrous temporal processes.

A CT of the skull revealed multiple cerebral infarcts, and an MRI revealed narrowing of the left foramen ovale and thickening of the calvarium. Radiographs of the lower limbs were normal.

It appeared that the narrowing of the foramen ovale may have caused mandibular nerve compression and therefore dysfunction.
Discussion

Case 3 – Mrs JA

This case illustrates the diagnostic dilemma of facial pain where there may be a number of confounding factors. Although Paget disease must be considered the most likely cause, there are a number of significant factors in this case. Paget disease a relatively common condition in the elderly and is frequently asymptomatic (being first noted as an incidental finding on a radiograph). The pelvis is the commonest site, however it is frequently seen in the femur where increased vascularity related to high bone turnover may produce a feeling of increased warmth of the limb, as in Mrs JA’s case. In the skull, increased bone activity can result in expansion of the calvarium (a ‘cotton wool’ appearance) making hats tight, and in the skull base, foramina narrow leading to compression neuropathy (cranial nerve dysfunction) such as deafness (symptoms complained of by Mrs JA).

Imaging revealed resorption of the apices of petrous temporal bilaterally and narrowing of the foramen ovale. This in turn could produce sensory nerve dysfunction by compression of the left mandibular division of the trigeminal nerve. Bone pain in Paget disease is frequently aching in nature. In this case, although the calvarium was thickened, its appearance was that seen in long term anti-epileptic treatment rather than Paget disease (Mrs JA had taken carbamazepine for temporal lobe epilepsy for many years, so the significance of this finding is unclear).

Alternatively, Mrs JA’s symptoms may result from her neurovascular lesions. Cerebral infarcts in the thalamic region, and aneurysms of the internal carotid or posterior inferior cerebellary artery, can produce altered sensation in cranial nerve distribution, with thalamic pain often being felt around the angle of mouth, leg and foot on the affected side. Magnetic resonance imaging identified a number of thalamic infarcts. As Mrs JA had a past history of ovarian and breast cancer, a careful search was made for evidence of cerebral metastases, the presence of which could have produced neurological symptoms.

Finally, Mrs JA was suffering from depression. This may have amplified her pain – psychosomatic factors commonly being found in patients with chronic facial pain. After evaluating all findings, it appeared that Paget disease was the most likely cause of her symptoms and Mrs JA was referred to an endocrine unit for further assessment and management, as patients with symptoms of Paget disease frequently respond to bisphosphonate therapy even when imaging and bone scan findings are equivocal.

Conclusion

The diagnosis of facial pain is usually readily elicited. However, there remains a group of patients with an atypical presentation who require considerable time and skill to arrive at the correct diagnosis. Depression or other psychogenic causes should always be considered. Despite an exhaustive investigation, there are some patients in whom the diagnosis remains elusive.

Summary of important points

- In the majority of cases, facial pain is caused by dental disease or temporomandibular dysfunction.
- Usually the cause can be determined by careful history and examination.
- Atypical facial pain may require more extensive investigation with imaging and laboratory investigation as causes such as nasopharyngeal carcinoma, vascular disease and thalamic infarcts may not be identified by clinical assessment.
- Depression or other psychogenic causes should always be considered.
- As facial pain does not cross the midline, bilateral pain raises the possibility of a psychosomatic cause.
- Temporal arteritis may cause pain around the ear, eye symptoms, and pain on chewing, and is an important differential diagnosis, particularly in the elderly.

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References