Trigeminal Neuralgia Caused by Intracranial Epidermoid Tumour: Report of a Case

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SOMMAIRE

La névralgie faciale est une complication reconnue chez les patients qui présentent des tumeurs intracrâniennes touchant le nerf trijumeau. Cette étude de cas décrit une tumeur épidermoïde siégeant dans l’angle ponto-cérébelleux chez un homme d’âge moyen présentant une névralgie faciale unilatérale par ailleurs classique. Ce cas fait ressortir les difficultés du diagnostic, ainsi que l’importance d’adopter une démarche multidisciplinaire lorsque la névralgie faciale est associée à une tumeur cérébrale.

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rigeminal neuralgia, or tic douloureux, is a well-recognized disorder that most commonly occurs in people over 50 years of age and slightly more commonly in women than in men.\textsuperscript{1} Trigeminal neuralgia is characterized by paroxysms of unilateral facial pain, most commonly in the second and third trigeminal divisions, that are severe and lancinating.\textsuperscript{1,2} Low-intensity mechanical stimulation of the trigger zones by speaking, washing or eating can provoke pain.\textsuperscript{1} The condition is usually treated with carbamazepine or other anticonvulsant medications, although surgical management has also been used in recalcitrant cases.\textsuperscript{1,3}

Diagnosis of trigeminal neuralgia is based primarily on a history of characteristic pain attacks that are consistent with specific, widely accepted research and clinical criteria. For most patients with trigeminal neuralgia, the results of their clinical examination, imaging studies and laboratory tests are unremarkable. This type of trigeminal neuralgia is called “classical trigeminal neuralgia” and its cause is unknown, although vascular compression of the trigeminal nerve roots is often suggested.\textsuperscript{3} In a smaller group, the signs and symptoms of trigeminal neuralgia are secondary to those of another disease process that affects the trigeminal system, such as multiple sclerosis or a cerebellopontine-angle tumour. This type of trigeminal neuralgia is called “symptomatic trigeminal neuralgia.”

The case report presented here describes a cerebellopontine-angle epidermoid tumour in a patient with otherwise classical unilateral trigeminal neuralgia.

Case Report

A 43-year-old man with no significant medical history was referred to the department of dentistry at Toronto’s Mount Sinai Hospital with an 18-month history of right-sided facial pain. The patient had previously sought treatment from 4 dentists who were unable to provide the patient with a diagnosis. Various pharmacological remedies were prescribed, including codeine, oxycodone and meloxicam, but the patient continued to have pain. The patient had also visited the emergency department of a local
hospital 3 months previously and was given antibiotic treatment, but again without resolution of his symptoms.

At the outset of the consultation and history, the patient described his pain as a dull aching pain in the area of tooth 46 that was exacerbated with chewing. Clinical evaluation demonstrated sensitivity to percussion in association with tooth 46. The tooth responded normally to temperature. Infrequently, pain of short duration radiated through the right side of the face. The patient stated that the pain occurred spontaneously at times. He also described discomfort in the gingival tissue surrounding tooth 46. The results of clinical and panoramic radiological examinations provided no evidence of contributory dentoalveolar or other craniofacial disease that might explain the patient’s symptoms.

In light of the patient’s history, and clinical and radiographic findings, a provisional diagnosis of neuropathic pain was made. Despite these findings, the patient continued to believe that his pain was related to pulpal or gingival disease in tooth 46. Further investigation of pain of nondental origin was carried out, including a computed tomography (CT) scan and a consultation with a neurologist. The head and neck CT scan demonstrated a lesion within the right cerebellopontine-angle cistern that measured 2.9 cm × 1.2 cm, which was thought to represent either an arachnoid cyst or an epidermoid tumour. As a result of his CT findings, the patient was referred to a neurosurgeon, and in the interim, he was placed on carbamazepine (200 mg, 3 times a day) to ameliorate his pain; however, he had only minimal relief.

Despite the clinical and radiographic findings (in particular, the CT findings), the neurosurgeon continued to believe that the problem was caused by dentoalveolar disease. It was his impression that the patient was prone to magnify symptoms of other somatic complaints and that he was exaggerating his oral symptoms. The neurosurgeon thought that there must be underlying, but as yet undeclared, dental disease (particularly because he was able to evoke pain after percussion of tooth 46), so he referred the patient back to the Dental Unit at Mount Sinai Hospital for additional evaluation.

In the interim, and against advice, the patient sought additional dental treatment and had endodontic therapy done on tooth 46 (Fig. 1). Predictably, he found no relief, but in spite of this, continued to insist that his pain was of dental origin. The fact that the patient’s pain was not eliminated after endodontic treatment was thought to add credence to the diagnosis and to the probability that his pain was nonodontogenic.

With this new information, the patient was referred to the neurosurgeon again for investigation with magnetic resonance imaging (MRI). Results of the MRI showed displacement of the brainstem, and a lesion abutting and compressing the trigeminal nerve, findings that could cause the symptoms similar to those the patient described (Fig. 2).
Trigeminal Neuralgia

The patient was treated surgically and the lesion was excised with a retrolabyrinthine and retrosigmoid approach. The histopathological report indicated that the lesion was an epidermoid tumour.

The patient recovered without complications or post-operative neurological deficits, and most importantly, with no further reports of facial pain.

Discussion

Trigeminal nerve neuropathy can underlie symptomatic trigeminal neuralgia and can involve any segment of the nerve, from its central origins to its peripheral branches. Multiple sclerosis and infarct are the most common abnormalities of the brainstem that cause trigeminal neuralgia. Rarely is it attributed to an intracranial tumour (incidence < 0.8%), which is one of the factors that makes diagnosis of the disorder so difficult. Also, trigeminal neuralgia can mimic dentoalveolar disease, as it did in this patient’s case. Most of the tumours that are known to lead to the development of trigeminal neuralgia (acoustic and trigeminal schwannomas, meningiomas, epidermoid cysts, lipomas and metastases) are generally located in the posterior fossa.

Epidermoid tumours are benign cysts that have well-characterized histology, namely a stratified squamous-lined cyst, and luminal contents consisting of desquamated keratin, cell debris and cholesterol clefts. Epidermoid tumours are often located at the pontocerebellar angle, but may occur in the fourth ventricle and suprasellar region. Epidermoid tumours rarely cause trigeminal neuralgia, but are thought to cause trigeminal nerve neuropathy by either compressing the nerve directly or eliciting an inflammatory reaction indirectly by the release of its contents.

Intracranial epidermoid tumours are slow growing, but usually adhere to critical structures, such as the brainstem, cranial nerves and vascular structures. Clinical symptoms, in addition to trigeminal neuralgia, may include headache, dizziness, unstable gait and hemifacial spasm. Surgical excision of the tumours generally yields excellent results, although a conservative approach is often advised since aggressive surgery may result in transient but significant severe cranial nerve dysfunction during the postoperative period.

Because clinical findings do not differentiate classical (basically idiopathic) trigeminal neuralgia from trigeminal neuralgia that might be caused by neoplastic disease, diagnostic brain imaging studies should be part of the initial evaluation of any patient with symptoms of trigeminal neuralgia. In fact, CT scans of the brain are usually adequate screening for a tumour, whereas MRI is capable of demonstrating the anatomic landmarks around the trigeminal ganglion and the pontocerebellar angle.

This patient’s experience underscores the importance of a multidisciplinary and transdisciplinary approach to the diagnosis and management of orofacial pain. Because several forms of dentoalveolar disease, including musculoligamentous conditions, can cause dental pain, these diseases must first be ruled out with a thorough history and clinical examination. Then investigations for other conditions known to cause orofacial pain, such as those described here, should be done. Finally, precise and open communication is necessary not only between the patient and health care givers, but also among the health care givers themselves. The importance of this requirement simply cannot be overstated.

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