# Trigeminal Neuralgia Due to an Acoustic Neuroma in the Cerebellopontine Angle

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*This case report first reviews the intracranial tumors associated with symptoms of trigeminal neuralgia (TN). Among patients with TN-like symptoms, 6 to 16% are variously reported to have intracranial tumors. The most common cerebellopontine angle (CPA) tumor to cause TN-like symptoms is a benign tumor called an acoustic neuroma. The reported clinical symptoms of the acoustic neuroma are hearing deficits (60 to 97%), tinnitus (50 to 66%), vestibular disturbances (46 to 59%), numbness or tingling in the face (33%), headache (19 to 29%), dizziness (23%), facial paresis (17%), and trigeminal nerve disturbances (hypesthesia, paresthesia, and neuralgia) (12 to 45%). Magnetic resonance imaging with gadolinium enhancement or computed tomography with contrast media are each reported to have excellent abilities to detect intracranial tumors (92 to 93%). This article then reports a rare case of a young female patient who was mistakenly diagnosed and treated for a temporomandibular disorder but was subsequently found to have an acoustic neuroma located in the CPA.*
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**Key words:** trigeminal neuralgia, temporomandibular joint disorders, acoustic neuroma, cerebellopontine angle, multiple sclerosis, neoplasms

Trigeminal neuralgia (TN) is one of the most debilitating pain syndromes known to humans.[1] This condition is often secondary to central lesions, multiple sclerosis (MS), or vascular compression, but almost 50% of cases are of unknown etiology (idiopathic). Although the exact cause of idiopathic TN is still unknown, various conditions have been reported to induce TN, including tumors,[2,3] MS,[4] and vascular contact or compression of the trigeminal root.[2,4–6]

## Intracranial Tumors and Trigeminal Neuralgia

The percentage of intracranial tumors found in patients complaining of TN-like symptoms is not high. Yang and colleagues[4] assessed the value of magnetic resonance imaging (MRI) in evaluating TN in 51 patients who failed a trial of conservative treatment. They reported that 8 patients (16%) had cerebellopontine angle (CPA) tumors, 5 patients (10%) were diagnosed with MS, 2 had sphenoid or ethmoid sinusitis, 1 had meningitis, 1 had

trigeminal neuritis, and 27 (53%) had vascular compression of the trigeminal root. Nomura and colleagues[6] investigated 164 patients who presented with TN as their initial symptom. They reported that 14% of the patients had intracranial lesions and 35% had microvascular compression. Sindou et al[7] studied 350 consecutive TN patients and reported that, in 6% of the cases, the cause of the TN was a tumor or a vascular malformation in the CPA. Other investigators have also reported the prevalence of TN symptoms in brain tumors. Puca et al[8] reported that 7% of patients with extra-axial tumors of the posterior and middle cranial fossae presented with symptoms resembling TN. Additionally, Matthies and Samii[9] reported that 1 to 3% of patients with schwannoma (acoustic neuroma) presented with TN-like symptoms. Earlier, Puca et al[10] described a group of patients in which 10% of those with extra-axial tumors of the posterior and middle cranial fossae had TN symptoms. Hence, TN symptoms are commonly reported by patients subsequently diagnosed with intracranial tumors. However, Selesnick et al[11] reported that in a group of 126 patients with acoustic neuroma, none had TN symptoms. The incidence of subjective hearing loss reported in that study is less than in other studies and may be due to inclusion of a larger proportion of small (< 1 cm) tumors. Small acoustic neuroma tumors present with subtle and more limited symptoms than their larger counterparts.

In middle-aged patients with TN symptoms, intracranial tumors, MS, acoustic neuromas, and meningiomas are frequently observed.[4,12] In patients younger than 29 years of age with TN symptoms, the prevalence of intracranial tumor or MS is reported to be virtually 100%.[4] In the age group from 29 to 39 years of age with TN symptoms, 45% had a tumor or MS. In the age group from 40 to 59, the prevalence of tumors is 20%, and for those older than 60 it is 18%. Based on the above percentages, imaging should be performed for all patients who have TN-like symptoms, since all age groups have a risk of brain tumors.

Hearing deficits (60 to 97%) are the most commonly reported clinical symptom of the acoustic neuroma, followed by tinnitus (50 to 66%), vestibular disturbances (46 to 59%), numbness or tingling in the face (33%), headache (19 to 29%), dizziness (23%), facial paresis (17%), and trigeminal nerve disturbances (hypesthesia, paresthesia, and neuralgia) (12 to 45%).[11–13] The most commonly reported symptoms associated with other types of intracranial tumors are hyperesthesia, reduced or absent corneal reflex, facial palsy, masticatory weakness, hearing loss, and/or ataxia.[6,10]

Computed tomography (CT) and MRI are reported to have excellent detection ability for intracranial tumor. A CT scan without contrast is reportedly able to detect a lesion in 73 to 76% of patients. If a contrast medium is used, this rate increases to 93%.[6,13] Magnetic resonance imaging, with its better soft tissue resolution and multiplanar capability, is able to depict the intracranial course of the trigeminal nerve[14] and has detected the mass in 92% of patients with known lesions.[6] Furthermore, MRI provides superior definition of the tumor's boundary and of its relationship with adjacent structures.[15]

The treatment outcome and prognosis for acoustic neuroma with TN symptoms are very favorable. In one study, 89% of patients with extra-axial tumors of the posterior and middle cranial fossae experienced postoperative pain relief from surgery,[8] and gamma knife for acoustic neuroma patients has also provided pain relief postoperatively.[16] Other researchers have reported that the gamma knife[17] and stereotactic radiosurgery (X-knife) are effective in the treatment of acoustic neuroma.[18]

Some investigators have reported on the morbidity from surgery for acoustic neuroma. Harner et al[19] performed surgery for acoustic neuroma through a retrosigmoid suboccipital craniectomy. They reported that the facial nerve was preserved in 86% of patients and that delayed or partial paresis developed in 50% of the patients. Foote et al[17] reported that 72% of patients who received gamma knife surgery developed a new or progressive facial or trigeminal neuropathy (or both). Andrews and associates reported on posttreatment morbidity after stereotactic radiotherapy. In their study, trigeminal neuropathy was seen in 3% of patients, vestibulocochlear neuropathy in 29% of patients, scalp pain or tic pain in 27% of patients, and dizziness in 23% of patients.[18]

## Case Report

A 27-year-old woman visited the UCLA Graduate Orofacial Pain Clinic with a complaint of pain in the right masseter muscle that radiated upward to the cheek and forehead. The pain was triggered by swallowing and licking of the lips.

One year prior to the initial visit, the patient had developed pain after a dental procedure that radiated to the right ear and chin. Her pain was exacerbated by eating. She was given 400 mg

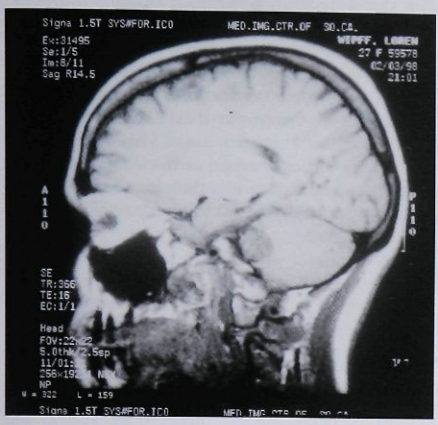


Fig 1 T1-weighted sagittal MRI showing 2.0-cm mass at the cerebellopontine angle.

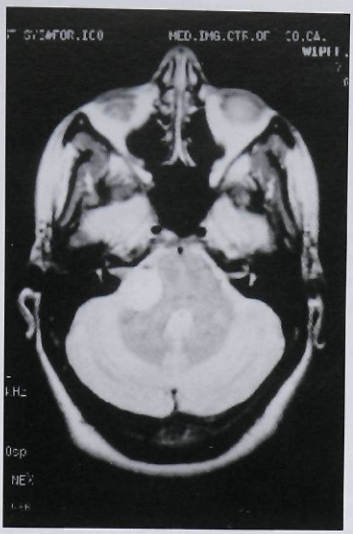


Fig 2 Axial gadolinium-enhanced MRI shows a mass suggestive of an acoustic neuroma or meningioma at the right cerebellopontine angle.

ibuprofen on the assumption that she was having an inflammatory response to the dental treatment. However, she returned 1 month later, indicating that the pain was worsening. At that point, she was referred to a temporomandibular disorder (TMD) specialist for evaluation. She apparently became pain-free for 5 months after treatment for TMD, then returned to her physician, indicating that the pain had returned again 3 days prior. The physician referred her back to the dentist to have a nightguard made.

When the patient came to the UCLA Orofacial Pain Clinic, she complained of the same pain that she had reported to her physician. She indicated that the pain was continuous (24 hours/day) and was variable in intensity, without a temporal pattern. It was exacerbated by brushing her teeth and washing her face, pushing on her teeth, talking, licking her lips, and touching the right face or right palate. The pain occasionally awakened her during the night. She described it as electrical shock–like, with occasional soreness in the right temporomandibular joint (TMJ) and the angle of the mandible. There were no associated autonomic symptoms.

Neurologic and stomatognathic examinations revealed no abnormalities. The range of jaw motion was unrestricted and pain-free. No pain was elicited during examination of the TMJ. Some tenderness was noted in the masticatory muscles, but the pain complaint was not duplicated or aggravated by the muscle assessment. Because of the lack of abnormal musculoskeletal findings, it was the authors' impression that the patient did not have a TMJ problem and would require further assessment, including brain MRI. Since the patient was in pain at the time of the examination and the pain had a neuropathic quality and presentation, gabapentin was initially prescribed for her, and arrangements were made for further assessment. When she increased the gabapentin to 900 mg per day, the pain decreased significantly. She was subsequently referred for a neurologic evaluation and MRI to rule out an intracranial tumor or MS. The MRI showed a large 2.0-cm mass at the right CPA, suggestive of an acoustic neuroma (Figs 1 and 2), and the patient was scheduled for surgery to remove the tumor.

## Discussion

Acoustic neuromas are generally slow-growing, and patients are reported to experience clinical symptoms for long periods of time before seeking medical treatment (0.6 to 5 years).[9,11–13] The tumor discussed in this report was undoubtedly present during the time the patient was being seen by the physician and dentist, who assumed she had a TMJ problem, although her developing symptoms at that time were consistent with neuropathy and not with a TMD.

This case suggests several important points for consideration regarding orofacial pain patients. First, the doctor should understand the signs and symptoms of the various orofacial pain conditions, including TMD and TN, since this is critical for appropriate diagnosis and management. It has been our experience that often, when an orofacial pain condition is not clearly understood, the patient is referred for TMJ evaluation and treatment. In such a case, TMD treatment may be undertaken unnecessarily, and the condition causing the pain is not addressed.[1] Based on the findings of our examination, the patient did not have a TMD, but TN. The fact that she did not fit the age profile for TN was of great concern; hence she was sent for further evaluation to rule out a central lesion or a demyelinating disease.

To evaluate an orofacial pain condition, it is necessary to undertake a complete assessment, including a neurologic examination. However, knowledge of the various conditions that can cause orofacial pain is imperative, and data from an examination will be useless without this knowledge. This patient presented to the physician, the dentist, and to the authors with the same complaints. Musculoskeletal problems such as TMD do not usually show the temporal pattern that is characteristic of TN, with alternating complete remission and exacerbation. It was our concern with this young patient who presented with neuralgia-like symptoms that she had a tumor or MS, since this risk is high in young patients who present with similar symptoms.[4] A thorough neurologic assessment should be conducted routinely to assess for hearing loss, vestibular disturbances, numbness or tingling of the face, or trigeminal disturbances that are characteristic of acoustic neuromas.[11–13] Other types of intracranial tumors may also show some clinical symptoms, including hypesthesia, reduced or absent corneal reflex, facial palsy, masticatory weakness, hearing loss, or ataxia.[6,10] Interestingly, in this case, the patient did not show any abnormal findings in neurologic examinations by either the orofacial pain specialist or the neurologist. An additional and obligatory part of the evaluation for TN, therefore, is the MRI or CT scan with contrast, since these modalities have high sensitivity for the detection of intracranial tumors.[6,13] Magnetic resonance imaging is reported to be slightly more sensitive than the CT scan with contrast for detection of intracranial lesions.[15,20]

Tumors in the posterior fossa are more likely to cause TN-like symptoms than are tumors in any other location. This area encompasses the trigeminal nerve path as it emerges from the pons, runs anteriorly and superiorly through the prepontine cistern and finally over the petrous ridge to the trigeminal ganglion in Meckel's cave.[3]

The neoplasm most likely to produce TN is a benign, slowly growing, extra-axial tumor that compresses the trigeminal root.[2,3] The increasing pressure on the trigeminal root or ganglion may induce loss of myelination in the trigeminal sensory root.[5] It has been proposed that this loss of myelination results in ephaptic short-circuiting within the nerve root, resulting in facial pain and sensory deficits.[21] Acoustic tumors represent the most common type of tumor associated with TN.[3,4] Some authors believe that tumors push the trigeminal nerve root against the superior cerebellar artery, producing a neurovascular conflict similar to the vascular compression theory proposed for classic TN.[5]

Bullitt and coworkers[3] reported that carbamazepine is an effective drug in the temporary treatment of TN with intracranial tumors. The patients in the study tended to respond to the drug at least temporarily, with improvement of symptoms; however, no patient was relieved of pain for more than 1 year. Importantly, the authors concluded that an initial response to carbamazepine cannot be used to exclude the diagnosis of tumor. Gabapentin has also been reported to be effective for TN.[22,23] In the present case, gabapentin (900 mg) decreased the pain while the patient was in the process of receiving a more thorough evaluation for her neuralgia-like symptoms.

Fortunately, the prognosis for acoustic neuroma or benign tumor surgery is good, although all of the surgical procedures are associated with some risk of morbidity.[16–18]

## Conclusions

This case report describes a 27-year-old female who presented to the UCLA Orofacial Pain Clinic with a condition previously diagnosed and treated

as a TMD by her primary care physician and subsequent TMD specialist. The patient described the same symptoms that she had reported to the previous doctors. These symptoms included sharp, shooting, episodic pain that seemed to resolve temporarily with TMJ treatment. A careful examination excluded any TMJ component to her pain complaint and she was given a preliminary diagnosis of TN, with an intracranial tumor or MS left as possible causes of her symptoms. Subsequent evaluation with MRI found a 2-cm tumor in the CPA. This case emphasizes the need for careful evaluation of orofacial pain, taking into account the description of the pain as well as its temporal patterns.

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