

A CASE REPORT

TRIGEMINAL NEURALGIA CAUSED BY INTRACRANIAL EPIDERMOID TUMOR: REPORT OF A CASE AND REVIEW OF THE DIFFERENT THERAPEUTIC MODALITIES

Bayram Cirak, MD, Nejmi Kiymaz, MD, and Atilla Arslanoglu MD

Trigeminal neuralgia (TGN) is characterized by recurrent paroxysms of unilateral facial pain that typically is severe, lancinating, and activated with cutaneous stimulation. Paroxysms typically last for 1 to 2 seconds. Etiology includes compression of the trigeminal nerve by vascular structures, tumors and multiple sclerosis plaques in the medulla spinalis. TGN is rather rare as a presenting symptom with intracranial tumors. Epidermoid tumors comprise 1% of all intracranial tumors. The

majority of epidermoid tumors are located at the pontocerebellar angle, the third ventricle, and the suprasellar region. Symptomatology is not different from other tumors located at the same sites. Trigeminal neuralgia may be caused by tumor compressing the nerve itself, an inflammatory response to the epidermoid tumor or vascular compression of the nerve.

We describe the case of a 30-year-old female presenting with right-sided facial pain. Her past history revealed mul-

tle teeth extractions done to relieve facial pain. Radiological evaluation, including magnetic resonance imaging (MRI), showed a pontocerebellar angle (PCA) epidermoid tumor. TGN resolved completely after total tumor resection. A temporary facial paralysis consequent to surgical removal of the tumor improved 6 months postoperatively, and she remained pain-free, without medications.

Keywords: Brain tumor, epidermoid tumor, trigeminal neuralgia.

Trigeminal neuralgia (TGN), or tic douloureux, is a debilitating syndrome consisting of unilateral short bursts of lancinating pain in the distribution of one or more branches of the trigeminal nerve. Typically, touching specific trigger points, speaking, or eating can provoke pain. The incidence of TGN is reported to be 4.3 per 100,000 people and occurs predominately in females. Pathogenesis includes vascular compression of trigeminal nerve roots, intracranial tumors, intracranial infections and MS plaques in the medulla spinalis (1).

Any intracranial tumor causing 5th nerve compromise anywhere along its course may cause TGN. PCA epidermoid tumors, although of benign nature, are of considerable neurosurgical interest because of their close proximity and adherence to the cranial nerves and brain stem. Epidermoid tumors comprise 1% of all intracranial tumors. They arise from dorsal midline ectodermal remnant cells. In the

majority of cases, they are located at the PCA. Histopathologically, epidermoid tumors have one-layered stratified squamous epithelia. They are benign tumors; malignant transformation is rather rare (2-4).

CASE REPORT

A 30 year-old woman presented with the chief complaint of right facial pain of 8 years' duration. The pain was sharp, intermittent on the right cheek and provoked by eating and speaking. Evaluation of her past

history revealed multiple tooth extractions of the molar and premolar teeth bilaterally, based on the erroneous belief that pain was dental in origin. On neurologic examination, cranial nerve functions were normal. She had neither cerebellar nor brainstem signs. Her chewing, swallowing, hearing, facial motor and sensory functions were normal. The uvula was in the midline and the gag reflex was intact.

Since her complaint continued after teeth extractions, she was referred to

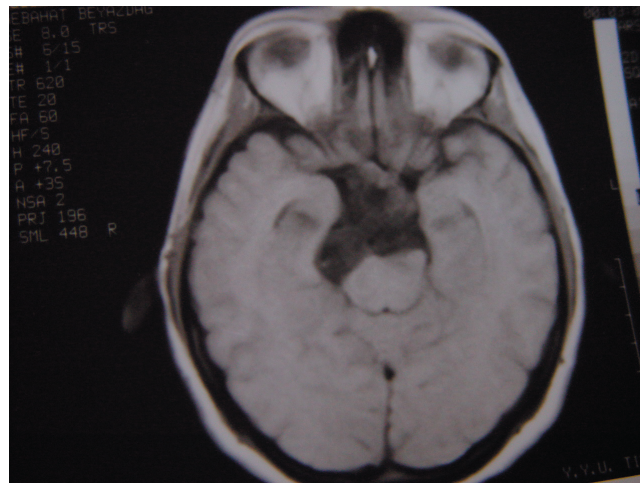


Fig 1. Cross section view of T1-weighted image of tumor.

From Department of Neurosurgery and Department of Neuroradiology, Johns Hopkins Medical Institutions, Baltimore, MD, and Department of Neurosurgery, School of Medicine, Yuzuncu Yil University, Van, Turkey. Address Correspondence: Bayram Cirak, MD, 103 Willow Bend Dr. 4-D, Owings Mills, MD 21117.

Email: bayramcirak@yahoo.com

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azepine and baclofen, can also be used. Most patients initially respond to medical treatment, but 30-75% do not obtain long-term pain relief or experience side effects of the drugs (14). For these patients, surgical intervention should be considered.

There is a continuing struggle between supporters of percutaneous ablation of the ganglion of Gasser and advocates of microvascular decompression of the trigeminal nerve root (15, 16). The idea that TGN might be caused by vascular compression is based on Dandy's observations in the 1930s. If no vascular compression is found at operation, the sensory root can be partially transected as described by Dandy in 1934. The surgical technique for microvascular decompression is simply to remove the offending vessel and insert a piece of Teflon as popularized by Jannetta in 1985.

The most widely used percutaneous procedure is percutaneous controlled differential thermocoagulation, as outlined by Sweet and Wepsic in 1974. It is an ambulatory procedure, requiring only brief anesthesia. A needle is placed transcutaneously through the foramen ovale into the Gasserian ganglion under direct X-ray control. Once the position of the needle has been verified in the conscious patient by electrical stimulation, the affected trigeminal division is coagulated. Altering the electrical current and duration of coagulation controls the amount of destruction. The aim of this procedure is to destroy unmyelinated and small myelinated pain conducting fibers while sparing the thick myelinated touch conducting fibers, thus preserving touch sensation.

Jannetta described microvascular decompression as sparing the nerve and causing a high proportion of immediate relief (90%), and those patients who are free of pain one year after the operation will suffer no recurrence (16). He also states that if the patient had a prior destructive procedure, decompression would be less effective. Both procedures achieve a high proportion of immediate relief in vascular compressive trigeminal neuralgia. The surgical results, in tumor cases, were excellent in patients with near-total tumor removal, as well as in patients with total removal. Aggressive surgical removal results in transient but significant cranial nerve dysfunction in the postoperative period. A conservative approach is indicated for patients in whom

the capsule is adherent to the brain stem and the cranial nerves. Perioperative administration of steroids and wound protection may be beneficial for preventing the development of postoperative aseptic meningitis and hydrocephalus (17).

Trigeminal nerve neuropathy may involve its full course, from nuclei in the brain stem to peripheral branches. The nerve can be divided into four segments for simplifying the differential diagnoses: extracranial, cavernous sinus, Meckel's cave and brain stem. Multiple sclerosis, infarct, and glioma are the most common abnormalities in the brain stem leading to trigeminal neuropathy. Neurovascular compression, followed by acoustic and trigeminal schwannomas, meningiomas, epidermoid cysts, lipomas, and metastases are the most common cisternal causes. Trigeminal neuropathy arising from Meckel's cave and cavernous sinus is frequently due to meningiomas, trigeminal schwannomas, epidermoid cysts, metastases, pituitary adenomas, and aneurysms. The most common extracranial causes are malignant tumors, which may demonstrate perineural tumor spread. Because the clinical findings may not help in differential diagnosis as in our case, MRI must be used to visualize the entire course of the fifth cranial nerve. The standard study should include T2-weighted images of the whole brain and high-resolution axial and coronal T1-weighted images of the skull base obtained with and without contrast material enhancement (3, 5, 8).

TGN is a syndrome caused by a variety of mechanisms, including vascular compression, intracranial infections, demyelinating diseases and intracranial tumors. Epidermoid tumor as a cause of TGN is very rare. In those cases with epidermoid tumor, trigeminal neuralgia can be caused either by tumor compression of the nerve or an inflammatory reaction to the tumor. Anatomical changes in the route of the vascular structures caused by tumor compression may also play a role in the development of trigeminal neuralgia. Microvascular decompression may be needed in addition to tumor removal in such cases.

In summary, we describe a case of intractable trigeminal neuralgia that resulted from a large, benign, brainstem epidermoid tumor. This presentation underscores the fact that imaging of intracranial structures, including the brainstem and posterior fossa, is warranted. MRI is the radiologic study of choice. The patient

was pain free after surgical excision of the tumor. The case provides an opportunity to review the pathogenesis, diagnosis and treatment of trigeminal neuralgia.

Author Affiliation:

Bayram Cirak, MD
103 Willow Bend Dr. 4-D
Owings Mills, MD 21117
Email: bayramcirak@yahoo.com

Nejmi Kiyamz, MD
Department of Neurosurgery
School of Medicine
Yuzuncu Yil University
Van, Turkey

Atila Arslanoglu MD
Department of Neuroradiology
Johns Hopkins Medical Institutions
600 Caroline Suite # 50738
Baltimore, MD 21287

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