Case Report

Trigeminal neuralgia secondary to cerebellopontine angle tumor: A case report and brief overview

ABSTRACT

Trigeminal neuralgia (TN) is a paroxysmal shock-like pain restricted to innervations of the areas of one or more branches of the trigeminal nerve, often set off by light stimuli in a trigger zone. Pain attacks occur spontaneously and can also be triggered by a nonpainful sensory stimulus to the skin, intraoral mucosa surrounding the teeth, or tongue. The pathogenesis of TN is uncertain and typically is idiopathic, but it may be due to a structural lesion. Some pathologies include traumatic compression of the trigeminal nerve by neoplastic or vascular anomalies and intracranial tumors or demyelinating conditions such as multiple sclerosis. This case report describes an epidermoid cyst at the cerebellopontine angle in a 25-year-old young man with otherwise classical unilateral TN. The case highlights the difficulties of diagnosis and the importance of a multidisciplinary approach in making the correct diagnosis in symptomatic as well as classical TN.

Keywords: Cerebellopontine angle, epidermoid cyst, neurosurgery, trigeminal neuralgia

INTRODUCTION

Trigeminal neuralgia (TN) has been referred to as one of the most painful afflictions of humanity.[1] It is characterized by paroxysms of unilateral facial pain, most commonly in the second and third trigeminal divisions, that are severe and lancinating.[2,3] Pain is commonly evoked by trivial stimuli, including washing, shaving, smoking, talking, or brushing teeth (trigger factors), and frequently occurs spontaneously. The condition is usually treated with carbamazepine or other anticonvulsant medications although surgical management has also been used in recalcitrant cases. Onset is usually in the middle or old age and is rare in children and young adults.[4] The other subdivisions of TN include the symptomatic form, which is considered to be pain indistinguishable from classic TN but caused by a demonstrable structural lesion other than vascular compression. Structural lesions include intracranial tumors and cysts such as acoustic neuroma, meningioma, epidermoid cysts, schwannomas, and pituitary adenoma, and demyelination disorders such as multiple sclerosis.[4] Here, we report a case of TN secondary to epidermoid cyst at the cerebellopontine angle (CPA).

Access this article online	
Website: www.njms.in	Quick Response Code
DOI: 10.4103/njms.NJMS_2_19	

CASE REPORT

A 25-year-old gentleman with no significant medical history had reported to our dental center with a 2-month-old history of left-sided facial pain. The patient had sudden episodes of shooting electric type of pain occurring on the left side of the face near the angle of the mouth, exacerbated by touching, not associated with any facial asymmetry, or numbness. The patient had previously sought treatment from a medical specialist who was unable to provide the diagnosis and had advised him to get a secondary opinion from a maxillofacial surgeon/dental surgeon to rule out any tooth pain or impacted wisdom tooth. A number of pharmacological remedies were

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Received: 01-01-2019, **Revised:** 17-02-2019,
Accepted: 26-03-2019, **Published:** 12-11-2019

How to cite this article: Prakash C, Tanwar N. Trigeminal neuralgia secondary to cerebellopontine angle tumor: A case report and brief overview. Natl J Maxillofac Surg 2019;10:249-52.

prescribed, including codeine and ketorolac Non-Steroidal Anti-inflammatory Drug (NSAID), but the patient continued to have episodic pains. With the abovementioned complaints and history, the patient described his pain as a dull-aching pain in the area of tooth 37 and38 that was exacerbated with chewing. The clinical evaluation demonstrated no sensitivity or pain on percussion in association with tooth 37 and 38 was partially erupted with no associated pericoronitis or swelling. The tooth responded normally to temperature. The patient stated that the pain occurred spontaneously at times for short duration and radiates toward the left side of the face. The results of the clinical and panoramic radiological (Orthopantomogram) 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 radiological findings, a provisional diagnosis of neuropathic pain was made and was sent for the magnetic resonance imaging (MRI) brain to rule out any vascular cause considering his young age. MRI at MRI specifications T2 sequence was done and MRI findings revealed lobulated, poorly marginated, extra-axial mass lesion expanding the left CPA cistern, extending to the left lateral pontine cistern, left lateral medullary, and premedullary cistern [Figure 1a and b].

MR findings likely represent an epidermoid cyst in Figure 2. The mass lesion posteriorly displaces the left 7th–8th cranial nerve complex and abutting as well as laterally displacing the left 5th cranial nerve in the CPA cistern. These findings may cause the symptoms similar to those the patient described as above. The mass was also found to insinuate into the prepontine and premedullary cisterns displacing the basilar artery anteriorly and the left vertebral artery medially. As a result of his MR findings, the patient was referred to the department of neurosurgery.

The patient was treated surgically, and the lesion was excised with a left retromastoid suboccipital craniotomy, and excision of space-occupying lesion (SOL) was done. The lower cranial nerves, 7th–8th complex, and 5th cranial nerve were found to be engulfed by the lesion. Gross total excision of the mass was achieved preserving the associated nerves and vessels. Histopathological evaluation of the tissue confirmed the diagnosis of epidermoid cyst. The patient well tolerated the procedure and recovered without any complications or postoperative neurological deficits, and most importantly, with no further reports of facial pain after the surgery.

DISCUSSION

A clinical diagnosis of TN in a patient with orofacial pain may be quite obvious, but treatment is yet challenging. The coexisting pathological conditions involving the dentition especially impacted third molars, hot and cold sensitivity, sinuses, and orbit may mimic symptoms that may confuse a clinician. These patients need adequate time for history taking and thorough evaluation of the regions. Neurological conditions, such as multiple sclerosis and atypical facial pain, may always be considered before a final diagnosis of classic TN. MRI and MR angiography or computed tomography scan is done to exclude the SOLs which may compress the nerves. MRI can identify intracranial conditions and identification of vascular structures encroaching on the trigeminal nerve root entry zone.[5] Trigeminal nerve neuropathy can underlie symptomatic TN 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 TN.[6-9] Rarely, it is attributed to an intracranial tumor (incidence $<0.8\%$), which is one of the factors that make a diagnosis of the disorder so difficult.[6]

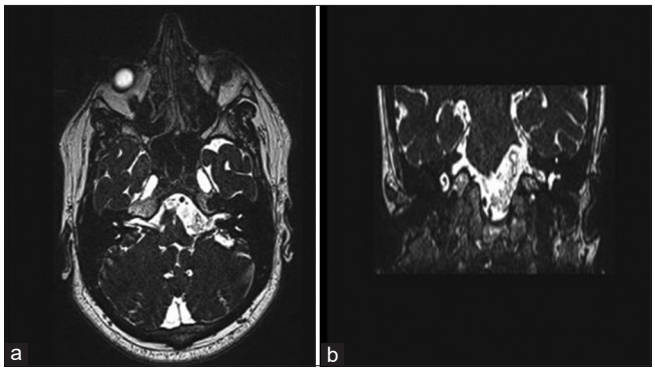

Figure 1: (a and b) T2-weighted image (axial and coronal images). T2-weighted image in axial and coronal section showing a lobulated, poorly marginated, extra-axial mass lesion in left cerebellopontine angle cistern, and extending to the left lateral pontine, medullary, and premedullary cistern. The lesion is slightly hyperintense to cerebrospinal fluid with small internal soft-tissue component with heterogeneous signal intensity, likely to be epidermoid

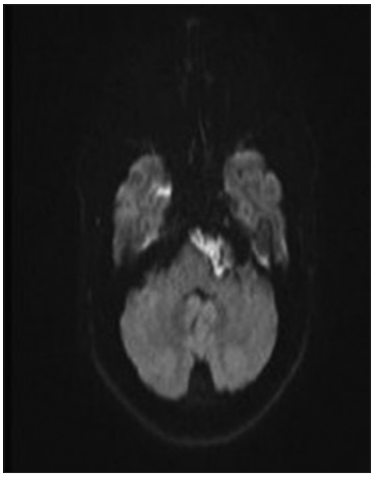

Figure 2: Diffusion-weighted image. Diffusion restricted image of same patient showing hyperintense signal which is likely to be a combination of restricted diffusion and T2 shine through as is seen in epidermoids

Epidermoid cysts, also known as primary cholesteatomas or pearly tumors, are the third most common tumors in the CPA region,[5] and they represent approximately 1% of all primary intracranial tumors.[6] They are generally located at the pontocerebellar angle but may occur in the fourth ventricle and suprasellar region also.[6] Although these tumors are present congenitally, they represent clinical symptoms in the early or middle adulthood because of their slow rate of growth.[6,10] They may be located anterolateral or posterolateral to the brainstem and tend to expand where physical resistance is low and burrow into crevices on the surface of the brain.[10] They usually surround vital structures such as cranial nerves, brainstem, and vascular structures. The tumors are covered with stratified squamous epithelium and contain keratin, cell debris, and cholesterol. Selective indications for imaging, specifically MRI, for patients with TN include young patients, patients failing to respond to medical therapy, and those with an atypical history or clinical signs such as involvement of ≥ one division of the trigeminal nerve, bilateral involvement, and presence of other cranial nerve abnormalities.[11] TN type pain was the only presenting feature in our case. There was no facial weakness or hearing loss, and the patient was younger than usual to be experiencing a classic TN. Therefore, further imaging in the form of MRI was done to rule out any vascular cause. It has also been noted that the occurrence of TN at a younger age is characteristic of TN patients with epidermoids, in contrast to patients with TN due to a vascular cause. Hyperactive dysfunction of the cranial nerves, especially TN, may be the initial as well as only symptom in patients with CPA epidermoids.[12] It has also been noted that the occurrence of TN at a younger age is characteristic of TN patients with epidermoids, in contrast to patients with TN due to a vascular cause.[12] This was a significant feature in this patient, who was in her second decade of life and manifested only with TN. Epidermoid cysts may also lead to multiple irritative syndromes such as the combination of TN, hemifacial spasms, and tinnitus.[13] Patients having a definite cause such as intracranial tumors as seen in this case are well suited for surgical intervention. In this case, the chief complaints followed by the confirmatory diagnosis by MRI have clearly explained the symptoms which were due to mass lesion posteriorly displacing the left 7–8th cranial nerve complex as well as laterally displacing 5th cranial nerve in the CPA cistern.

TN may be an initial presenting symptom indicating a CPA tumor, especially in young patients. We can assume from different sources that from 1% to 9.9% of cases of TN are caused by CPA tumors.[14] Because clinical findings do not differentiate classical (basically idiopathic) TN from TN that might be caused by neoplastic disease, therefore, the diagnostic brain imaging studies should be a part of the initial evaluation of any patient with symptoms of TN.

This case experience underscores the importance of a multidisciplinary and transdisciplinary approach to the diagnosis and management of orofacial pain, because several forms of the dentoalveolar disease, including temporomandibular joint and other dental pain, 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.

CONCLUSION

The TN-like symptoms in young age if reported to a dental specialist for any intermittent or constant pain must go for thorough radiological examination to rule out any dental pathology-like impacted third molars. One must take a secondary opinion with neurologist/neurosurgeon or general physician for other possible etiologies in case of absence of any dental pathologies.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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