Case Report

Trigeminal neuralgia secondary to posterior fossa tumor

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ABSTRACT

Trigeminal neuralgia (TN) is by no means an uncommon entity presenting as typical or atypical pain syndrome with a standard treatment protocol consisting of medical and surgical therapies. The diagnosis of TN is mainly dependent on the characteristics of symptoms conveyed by the patient and the clinical presentation. Careful history taking, proper interpretation of the signs and symptoms and cranial nerve assessment are necessary for proper diagnosis. Here, we report a case of TN, treated for dental problems and then for neuralgia with only short-term relief. Subsequently, the patient underwent neuroimaging and was found to be having an uncommon space-occupying lesion in the posterior cranial fossa.

Key words: Posterior cranial fossa tumor, trigeminal neuralgia, cerebello-pontine angle tumors

Introduction

The International Association for the Study of Pain defines trigeminal neuralgia (TN) as a 'sudden, usually unilateral, severe, brief, stabbing, recurrent pain in the distribution of one or more branches of the fifth cranial nerve.'[1]

The clinical presentation of TN is complex; it has been classified into various subgroups depending on how 'pure' the pain is. Typical TN is a pain syndrome manifesting without any sensory deficit, the attack is usually for a brief period, for a few seconds to minutes, but may recur repeatedly. The pain may be triggered by gentle stimulation over localized areas on face, the 'trigger zones,' via activities such as chewing, speaking, swallowing, touching the face or brushing the teeth.

But in atypical TN, pain is described as throbbing, burning or aching without definite trigger zone and is more continuous. TN, whether painful or not, is associated with a structural lesion or systemic disease. It has also been found that in many cases pain slowly evolves from one category to another.[2]

The etiology in 80–90% of patients of TN is compression of the trigeminal nerve at or near the nerve root by an overlying vessel resulting in demyelination of trigeminal sensory fibers.[3] Other causes of TN include multiple sclerosis, space-occupying masses in the posterior fossa or small infarcts in the pons or medulla.[4]

TN, due to its localization in the dentition region, usually mimics pulpal pain. Misinterpretation of odontogenic symptoms of TN has led to endodontic therapy, apicoectomy, extraction and other unnecessary treatment procedures. As in this case, poor oral hygiene and carious broken teeth with periapical pathology were masking the neuralgic pain. The patient was treated initially for her obvious dental problems and then at a later stage was diagnosed as having TN.

The aim of this presentation is to emphasize the importance of thorough neurological examination and neuroimaging of all patients with TN as the presence of any underlying structural lesion might otherwise go unnoticed.

Case Report

A 37-year-old female reported to our outpatient department with the chief complaints of pain in left upper teeth region since the last 5 months and almost persistent pain on left cheek region, along with episodic sharp and unbearable pain triggered by speaking, eating or touching the face.

She gave a history of previous extraction of a few carious and broken maxillary teeth. The patient's medical history was positive for hypertension for which she was on medication.

On examination, she revealed extreme tenderness in left upper lip and labial mucosal region. The oral hygiene was very poor. Neurological examination was unremarkable.

The clinical diagnosis of TN was made and antineuralgic medication (carbamazepine sustained release 200 mg BD and gabapantine 300 mg BD) was advised.

After a brief period of relief, the patient was again in agony and insisted on immediate extraction of offending teeth.

The medication for neuralgia was changed (carbamazepine sustained release 200 mg BD and pregablin 75 mg BD), and Magnetic Resonance Imaging (MRI) head was advised, which revealed a well-marginated, extraaxial, homogenously enhancing, space-occupying lesion (3.2 × 2.6 cm) in the posterior fossa in left cerebello-pontine (CP) angle region. A differential diagnosis of meningioma/schwannoma was made.

The patient was referred to neurosurgery unit where after neurlophysiological analysis and neuroimaging, she subsequently underwent surgery. Postoperatively, the patient became asymptomatic. Figures 1 and 2 show the preoperative and Figure 3 shows the postoperative MRI scans.

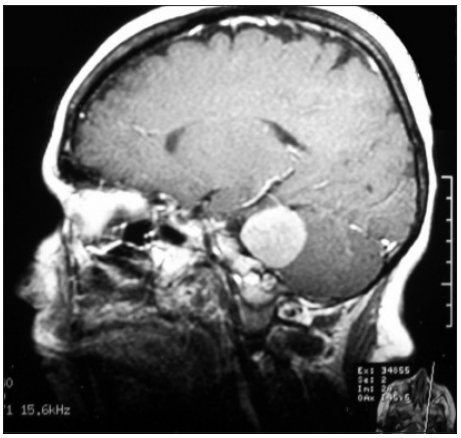

Figure 1: MRI scan (sagittal view) post contrast image demonstrating CP angle tumor

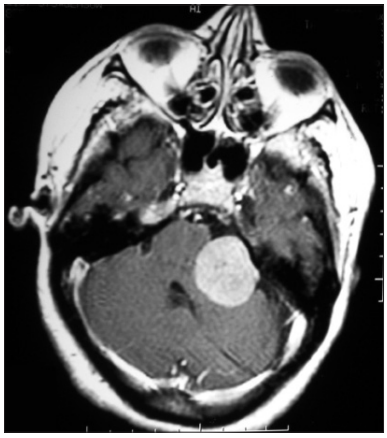

Figure 2: MRI scan (axial view) demonstrating intense enhancement of tumor

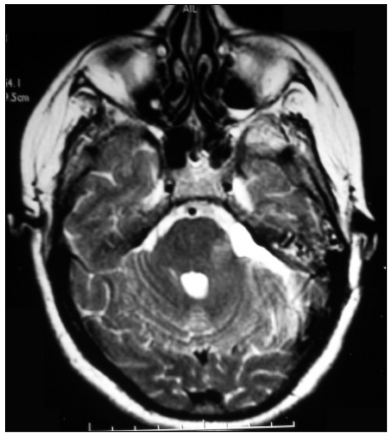

Figure 3: Postoperative MRI scan showing total tumor removal

Discussion

TN is a common and potentially disabling pain syndrome with an incidence of 4.5 per 100,000. It is more prevalent among females and affects primarily older people; the average age of onset is between the fifth and seventh decades of life.[4]

TN is termed 'idiopathic' when investigation identifies no cause other than a neurovascular contact, or 'symptomatic' when secondary to a major neurological disease such as multiple sclerosis or structural lesions.[2] Tumors, usually those involving posterior fossa, cause TN. These are slow-growing lesions. Acoustic neuromas, epidermoids, meningiomas are more common and some rare tumors found are lipomas, osteomas and tuberculomas.[5]

The most common cause for TN is compression and demyelination of the trigeminal sensory fibers at its root entry zone by a vascular loop which was first reported by Janetta.[3] Compression is now thought to account for 80–90% of cases, and all other causes including multiple sclerosis, tumors, vascular structures or aging are thought to act through a common process of demyelination of the nerve root resulting in ectopic generation of spontaneous nerve impulses

and their ephaptic conduction to adjacent fibers.[6] Fibers subserving light touch and those involved in the generation of pain are in closest proximity in this region so that ephaptic cross-talk between the two pathways is most likely to occur when the demyelination is in this region. In course of time, excessive afferent input of nerve impulses may produce central sensitization, resulting in atypical TN features such as constant pain.

No specific test can be used to clearly diagnose all cases of TN. Diagnosis is established on the basis of symptoms and thorough clinical examination including assessment of cranial nerve function and cerebral imaging.

Treatment consists of medical and surgical management. The goal of the medical management is the reduction of neuronal hyperexcitability in the peripheral and the central nervous systems. Most patients respond well to medical management. Anticonvulsant medications constitute the first line of treatment. Carbamazepine is the drug of choice, doses start with 100–200 mg two or three times daily which should be increased progressively and titrated to the severity of the patient's pain. If paroxysms of pain still occur with therapeutic blood levels, other drugs as baclofen, clonazepam and gabapentin may be added or monotherapy with phenytoin, oxcarbazepine, sodium valproate may be prescribed.[7]

With time, side effects of drugs or loss of their efficacy may result in recurrence.

The surgical treatment for TN aims at either destroying part of the nerve fiber or decompression of the nerve root as decompression produces rapid relief of symptoms in most patients with vessel-associated TN, probably because the resulting separation of demyelinated axons and their release from focal distortion reduces the spontaneous generation of impulses.[3,8]

The surgical options for TN include percutaneous procedures such as differential thermal rhizolysis, retrogasserian glycerol rhizolysis, trigeminal ganglion compression, stereotactic radiosurgery (Gamma Knife)[9] and microvascular decompression (MVD).[10,11] MVD is a widely used procedure and is reported to provide a pain free status in 70–80% cases of typical TN.

Although patients with secondary TN due to the presence of a space-occupying lesion may respond to medical treatment, definite treatment requires tumor removal and is usually followed by complete remission of TN[12] as was found in our case.

The case discussed here had been treated for dental problems and then medically for neuralgia for some time before proper neuroradiological assessment was done. The emphasis here is on thorough neurological evaluation and neuroradiological examination to explore the root cause of neuralgic pain and to rule out the possibility of any underlying structural lesion in the primary stage of treatment planning, as it will have a significant impact on the prognosis.

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