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內文:

ABSTRACT :

Trigeminal neuralgia is a disorder of the fifth cranial nerve. We report an unusual case of trigeminal neuralgia affecting right side of face presenting atypical features of neuralgia and not **responding to the usual course of treatment**. A large **extra-axial mass involving right cerebellopontine angle**. The aim of this case report is to show a tumor of cerebellopontine angle, presenting clinically as atypical trigeminal neuralgia.

INTRODUCTION

• Trigeminal neuralgia (TN), also known as **prosopalgia** [1], **suicide disease** [2], or **Fothergill's disease** [3], The clinical association between TN and hemi facial spasm is the so-called **tic douloureux (i.e painful jeking)**[4].

--Described as one of the **most painful** conditions known to mankind --It is estimated that **1 in 15,000 or 20,000 people** suffers from TN

- Lesions of cerebellopontine angle (CPA) are frequent and represent 6–10% of all intracranial tumors .
- Acoustic neuromas is also called vestibular schwannomas and meningiomas, are the two most frequent lesions and account for approximately 85–90% of all cerebellopontine angle tumors [10].
- It is usually associated with pathosis along the course of the nerve. <u>Most</u> of the times the cause is not known and trigeminal neuralgia is termed as <u>idiopathic TN.</u>
- Anomaly of the superior cerebral artery causing demyelization and pressure of the aneurysms of the intrapetrous portion of the internal carotid artery
- Many lesions such as trigeminal neuromas in the middle cranial or the posterior fossa, epidermoid tumors, Meckle's cave, arteriovenous malformations, aneurysms, and vascular compression have been suggested as the causes. It is also recognized that TN occurs in about 2–4% of patients with multiple sclerosis [7].
- Pain is always unilateral and does not cross the midline. The pain is of short duration and lasts from a few seconds to several minutes.
 Refractory period is also present between the attacks. Most of these patients respond to treatment with carbamazepine and this can be used as a diagnostic indicator in TN.
- neuralgic symptoms in **younger age group** (**less than 35yrs**) should alert the clinician to a possible **intracranial space** occupying lesion or **intracranial arteriovenous anomalies**.
- Lesions of cerebellopontine angle (CPA) are frequent and represent 6– 10% of all intracranial tumors. vestibular schwannomas and meningiomas, are the two most frequent lesions and account for approximately 85–90% of all cerebellopontine angle tumors [10].

• MRI & CT show typical features of acoustic neuromas or meningiomas and are sufficient to establish the diagnosis.

CASE REPORT

- <u>GENERAL DATA</u>: A 40-year-old male patient
- <u>Chief complaint</u> : pain in the right ear and preauricular region for 2 years
- <u>P.I</u>: Patient was diagnosed as a case of **glossopharyngeal neuralgia** three months prior to his visit and was treated **with tablets carbamazepine 100 mg** thrice daily which then increased to 200 mg thrice daily but without any satisfactory relief. Severity and intensity of pain increase on the same side of face in spite of taking increased dose of carbamazepine.
- PAIN : severe, intermittent, and lancinating pain in that region.
 --Pain was paroxysmal and each episode lasted from few seconds to few minutes

--Trigger points in the <u>preauricular</u> and <u>postauricular</u> region on right side of face .

--Patient also gave history of episodes of severe awakening pain during nights but no history of pain on deglutition(吞嚥) or during movements of tongue.

- There was no history of seizures, nausea, vomiting, and vision changes. (meningiomas)
- On examination, trigger zones were present on malar area, zygomatic buttress area, and also body and ramus of mandible
- The pains are bright and stimulating and are accurately located by the patient. The behavioral characteristics of the pain are neurogenous.
- All these findings were suggestive of trigeminal neuralgia but as the patient was not responding to the usual course of treatment and the severity of pain increases, an intracranial pathology was suspected in this case

 Magnetic resonance imaging (MRI) showed well-defined intensity enhancing extra-axial mass involving right cerebral pontine angle region measuring 2.5 × 2.0 × 1.8cm in anteroposterior, transverse, and superoinferior direction, respectively, and causing moderate mass effect on trigeminal nerve and brain stem (Figures 1 and 2).



Figure 1: Axial view of T1W image showing isointense, well-defined lesion seen in right cerebellopontine angle causing rotation of brain stem and compression of contralateral CP angle.

Figure 2: Flair axial image showing hyperintense, well-defined lesion seen in right cerebellopontine angle causing rotation of brain stem and compression of contralateral CP angle. • Further MRI with contrast revealed possibility of cerebellopontine angle Schwannoma or meningioma



• Figure 3: MRI T1W axial image **with contrast** shows well-defined intensity enhancing extra-axial mass lesion involving right cerebral pontine angle region.

--MRI impression: MRI study in contrast revealed possibility of CP angle Schwannoma or Meningioma.

• The patient was referred to a neurologist for further management. The tumor was surgically removed at Bombay Hospital, Mumbai, under general anesthesia. Histopathological exami- nation showed characteristic whorling suggestive of meningioma (Figure 4). Final diagnosis of trigeminal neuralgia involving maxillary and mandibular nerve secondary to meningioma was confirmed.



FIGURE 4: Histopathology slide: histopathologically showing characteristic whorling suggestive of meningioma.

DISSCUSSION

- 15% of patients with trigeminal neuralgia have intracranial pathology --This often occurs in the cerebellopontine angle, pons, and medulla . Failure to properly assess for secondary TN is a major potential pitfall
- Examination of all the cranial nerves along with MRI of the brain is advisable in patients younger than 60 yrs.
- One should also lookout for symptoms related to compression of surrounding tissues, which can also affect other cranial nerves and blood vessels.
- The typical features of **meningiomas are seizures**, **headaches**, **nausea**, **vomiting**, **vision**, **and behavioral and cognitive changes**. Sometimes no symptoms occur and tumor is detected incidentally [12]. In this case, no typical symptoms of meningioma were seen; it rather presented as a typical case of trigeminal neuralgia.

• Medicinal therapy is usually given for the treatment of TN. Surgical intervention can be attempted only if and when the pharmacologic therapy fails .

--Carbamazepine 100 mg tablet thrice daily usually resolves pain of TN.
 -- Side effects of this drug may cause bone marrow depression, drowsiness, blurred vision, and vertigo. Therefore, monitoring the bone marrow activity by obtaining the complete blood count prior to initiating therapy and routinely thereafter is indicated.

-- Anticonvulsant medications pose risk of sedation and ataxia particularly in elderly patients, which may make driving or operating machinery hazardous

-- palliative and often are of limited and temporary value

- If the patient does not respond at least partially to carbamazepine, we should suspect for **idiopathic TN**.
- Tumor arising in the CPA or the tumor of the base of the skull or brainstem encroaching upon the CPA can produce TN like symptoms.
- The other associated symptoms of these tumors depend on the direction of growth of these tumors. In our case although the lesion arose from CPA, it did not have any feature of this tumor and rather it presented as atypical TN; therefore, diagnosis was difficult.
- As the tumor grows

 -upward into the superior aspect of the cerebellopontine angle, it
 encroaches upon the trigeminal nerve, producing a gradual decrease of the
 corneal reflex and facial analgesia and anaesthesia
 -Downward growth of these lesions results in hoarseness, numbness of
 the throat, or complaints of difficulty in swallowing.
 -As with acoustic tumors, large meningiomas can produce cerebellar
 symptoms and signs or hydrocephalus with increased intracranial pressure
- Neurosurgical interventions including anesthesia dolorosa
 -- Numbness of the face is a constant part, which has also been referred to as trigeminal deafferentation pain. It results from damage to peripheral or

central sensory pathways leading to partial or total loss of sensory nerve supply to a region of the body

-- Deafferentation pain is usually severe in intensity and highly **resistant to treatment**. It can spread to other orofacial structures by extension of the receptive field.

CONCLUSION

- It is not always possible to determine what causes trigeminal pain
- The present case shows an unusual lesion of cerebellopontine angle tumor, rare cause of atypical TN, emphasizing the importance of investigating each and every case of trigeminal neuralgia for proper clinical diagnosis
- As we cannot establish a relation between pathology in the brain or brainstem and a clinical picture in a patient, imaging technique preferably an MRI scan with a focus on the course of the trigeminal nerve can be of utmost importance for the diagnosis of such cases
- Follow-up scans are needed because meningioma can recur years or even decades after treatment

題號	題目
1	Which of the following statement of TN is true?
	 (A) Pain located most in V1 and V3 (B) Male < female (C) Pain "spasm" duration 2~4 minutes
	(D) Average onset less then 40 years old
答案 (B)	出處: oral and Maxillofacial Pathology, third edition, p.861

題號	題目
2	造成三叉神經痛(trigeminal neuralgia)最常見的原因是:
	(A) 基底動脈的小動脈瘤(aneurysm of the basilar artery)
	(B) 三叉袖經瘤(trigeminal neuroma)
	(C) 多發性硬化症(multiple sclerosis)
	(D) 原因不明(idiopathic)
答案	出處: oral and Maxillofacial Pathology, third edition, p.862
(D)	