

TRIGEMINAL NEURALGIA

Etiopathogenesis

Compression of nerve root in the posterior fossa as it exits the Pons before reaching the gasserian ganglion.

Sources of compression:

- Aberrant loop of superior Cerebellar artery lying on nerve root
- Space occupying lesions – Cerebello pontine angle tumor, posterior fossa tumors or basilar artery aneurysms, Meningioma of Meckel's cavity.
- Systemic condition : Multiple sclerosis

Pathophysiology

- Demyelination causing ectopic impulses and ephaptic cross firing between nerve fibres leading to pain hypersensitivity.

Salient features

- Unilateral
- **Electric shock like pain**
- Paroxysmal few seconds to few minutes
- Trigger zones
- Distribution of nerve areas
- Absence of neurological deficit
- Absence of night pain
- Inconspicuous stimuli

Pre Trigeminal syndrome

Some patients may present with this syndrome for a period of weeks or even before developing the customary symptoms of Trigeminal neuralgia. They complain of an unrelenting sinus pain or tooth ache lasting for hours triggered by moving the jaw or drinking fluids. They benefit from Carbamazepine or baclofen

Natural history and Prognosis

After an initial attack the disorder may remit for months or even years. Thereafter the attacks may become more frequent more easily triggered more disabling and may require long term medication. Dull ache persist between episodes of stabbing pain. Patients may find immediate and satisfying relief with one medication typically carbamazepine. However over years they may require a second or third drug to control breakthrough episodes and finally may need surgical intervention. With more invasive operations, the risk of disabling adverse effect of anaesthesia dolorosa increases. The long term prognosis of this disorder varies.

Differential diagnosis

1. Atypical facial pain: Usually extends beyond the distribution of 5th cranial nerve (main location – face, neck, ear). It is rarely triggered. It presents with a steady unrelenting discomfort lasting hours to days and it can be associated with sensory abnormalities.
2. Glossopharyngeal neuralgia: It causes pain in the tonsillar fossa, posterior pharynx and ear. It may be initiated by coughing, yawning or swallowing cold liquids. During acute attack of this disease which frequently is associated with an underlying pathology, the patient is unable to speak and tries to avoid moving the lips or tongue.
3. Occipital neuralgia: It causes pain in the posterior head region.
4. Paroxysmal hemicranias syndrome: The pain typically lasts only seconds similar to trigeminal neuralgia. But occur in and around one eye. It does not respond to carbamazepine.
5. Migraine and Cluster headache: They produce severe unilateral pain not triggered by movement or contact with the face. They don't respond to carbamazepine.
6. Secondary trigeminal neuralgia: Pain is associated with hyperesthesia along the course of the 5th nerve or is observed with other cranial neuropathies. Investigations may reveal multiple sclerosis, tumor in posterior fossa, tumor on trigeminal nerve. Acoustic neuromas, cerebral aneurysms, trigeminal neuromas and meningiomas can produce syndromes similar to idiopathic trigeminal neuralgia. Age <40 years, predominant forehead and or orbit pain ie, first division of trigeminal nerve or those with bilateral facial pain. Bilateral sensory loss or weakness of facial muscles or jaw.
7. Tic convulsif: Prominent hemifacial spasm. Associated with dilated and ecstatic basilar artery or other vascular malformation compressing the 5th nerve.

8. Trigeminal neuropathy: It presents as a constant, unilateral often mild facial pain with prominent sensory loss. It is non triggerable and unremitting. It may be symptomatic or idiopathic.

9. Atypical Trigeminal neuralgia: A syndrome that overlaps trigeminal neuralgia and trigeminal neuropathy. It is associated with constant pain that episodically intensifies. Pain character is both lancinating triggered pain and a constant dull and throbbing discomfort. The atypical form may occur in upto 5% of people after facial surgery or significant trauma and in 1-5 % after removal of impacted teeth.

Investigations

No laboratory, electrophysiologic or radiologic testing routinely is indicated for diagnosis.

The initial response to carbamazepine therapy typically is diagnostic and successful.

Imaging studies: Brain MRI with and without contrast helps to distinguish secondary causes of Trigeminal neuralgia from idiopathic form .

Therapeutic diagnosis: A clear relief of pain with carbamazepine or another anti convulsant confirms the diagnosis of idiopathic trigeminal neuralgia.

Treatment

Medical management

Carbamazepine is the drug of choice. 100 mg tablet may produce significant and complete relief within 2 hours. If the above dosage does not provide relief a higher dose is administered or other drugs may be tried.

Medications:

Anti convulsants: They reduce the excitability of gasserian ganglion neurons, preventing anomalous discharges and related lancinating volleys of pain. Few investigators suggest that the medications work by interrupting the temporal summation of afferent impulses that precipitate the attack.

1. Carbamazepine (tegretol) : 100 mg twice daily initially

Maximum dose not >1200 mg; Maintenance dose as low as 200mg per day to prevent recurrences

2. Gabapentin (Neurontin) : 900 -2700mg per day - It is given in patients refractory to carbamazepine
3. Lamotrigine(Lamictal) : 100 – 400 mg per day - Newer anti epileptic
4. Phenytoin(dilantin): 200 – 400 mg four times daily - Less effective

5. Oxcarbazepine(trileptal) : 300 – 2400 mg twice daily - Not yet approved by FDA for Trigeminal neuralgia

Experimental drug: Topiramate (topamax) : 150 – 300 mg four times daily

In patients where carbamazepine and Gabapentin have failed.

Anti spasmotic drugs: Baclofen (Lioresal): 60-80 mg per oral in divided doses

When it is added to an anti convulsant the dosage of anti convulsant often is reduced.

Surgical management:

1. Microvascular decompression
2. Radiofrequency thermorhizotomy
3. Per cutaneous procedures
 - a. Per cutaneous retrogasserian glycerol rhizotomy
 - b. Per cutaneous radiofrequency trigeminal gangliolysis
 - c. Per cutaneous balloon microcompression
4. Gamma knife surgery

Miscellaneous:

Anti convulsants: Sodium valproate

Anti depressants : Tricyclic anti depressants – amitriptyline, nortriptyline

Dextromethorphan

L – baclofen