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Trigeminal Neuralgia: A Review

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Trigeminal Neuralgia (TN) is a well-recognized syndrome characterized by lancinating attacks of severe facial pain. The diagnosis of TN is based on a history of characteristic pain attacks that are consistent with specific widely accepted criteria for the diagnosis. TN pain attacks may result from physiologic changes induced by a chronic partial injury to the brainstem trigeminal nerve root from a variety of causes. An early and accurate diagnosis of TN is important, because therapeutic interventions can reduce or eliminate pain attacks in the large majority of TN patients.

KEYWORDS: Trigeminal Neuralgia, Orofacial pain, Tic Douloureux.

INTRODUCTION

Trigeminal neuralgia (TN), tic douloureux (also known as Prosopalgia, the Suicide disease or Fothergill's disease) is a neuropathic disorder characterized by episodes of intense pain in the face, originating from the trigeminal nerve.¹ One, two or all three branches of the nerve may be affected. It is "one of the most painful conditions known to humans, yet remains an enigma to many health professionals."²

HISTORICAL BACKGROUND

Aretaeus of Cappadocia is credited with the first clinical description of TN. At the end of first century he described a condition (heterocrania) where "spasm and distortion of the countenance take place." Jujani, an 11th century Middle Eastern physician, also discussed a problem of unilateral facial pain with associated spasm and anxiety. He even suggested the pain resulted from "the proximity of the artery to the nerve."³ In 1677 John Locke, a noted American physician and philosopher, accurately identified the major clinical features of TN in the Countess of North UMBER land. He correctly recognized that the facial pain was not caused by dental pathology but rather by a neuralgia of the trigeminal nerve.³ In 1756, the French physician Nicolaus Andre recognized the unique nature of syndrome, commenting that it was exclusive and distinctive

from all the diseases. He gave the name tic Douloureux to the condition. The English physician John Fothergill in 1773 also outlined the major clinical features of TN, clearly establishing the disorder as a discrete syndrome. In the early 20th century, Oppenheim alluded to an association between multiple sclerosis (MS) and TGN and Patrick commented on its familial incidence.¹

DEFINITIONS OF TGN⁴

TN is defined as "a chronic pain condition that causes extreme, sporadic, sudden burning or shock-like face pain". (National Institute of Neurological Disorders and Stroke)

TN can also be defined as "sudden unilateral, severe brief stabbing recurrent pains in the distribution of one or more branches of the Vth cranial nerve". (International Association for the Study of Pain)

"Painful unilateral affliction of the face, characterized by brief electric shock like pain limited to the distribution of one or more divisions of the trigeminal nerve. Pain is commonly evoked by trivial stimuli including washing, shaving, smoking, talking and brushing the teeth, but may also occur spontaneously. The pain is abrupt in

onset and termination and may remit for varying Periods". (International Headache Society)

EPIDEMIOLOGY AND DEMOGRAPHICS

TN has an incidence of approximately 1.9 per 100,000. Although familial examples of TN are reported, the large majority of cases occur spontaneously, and TN does not appear to be more common in any particular ethnic group, geographic region, or climate.³

PATHOPHYSIOLOGY, CAUSES AND DIFFERENTIAL DIAGNOSIS

The symptoms of TN are often falsely attributed to a pathology of dental origin. Extractions do not help. The pain is originating in the trigeminal nerve itself – often in its roots – and not in an individual nerve of a tooth but real tooth pain may be referred to the same areas of the face as that of TN. Because of this difficulty, many patients go untreated unless a correct diagnosis is made. Several hypothesis exist to explain the possible causes of this pain syndrome. It was once believed that the nerve was compressed in the opening from the inside to the outside of the skull; but newer leading research indicates it is an enlarged blood vessel – possibly the superior cerebellar artery – compressing or throbbing against the microvasculature of the trigeminal nerve near its connection with the pons. Such a compression can injure the nerve's protective myelin sheath and cause erratic and hyperactive functioning of the nerve.⁵ This can lead to pain attacks at the slightest stimulation of any area served by the nerve as well as hinder the nerve's ability to shut off the pain signals after the stimulation ends. This type of injury may rarely be caused by an aneurysm (an out pouching of a blood vessel); by a tumor; by an arachnoid cyst in the cerebellopontine angle;⁶ or by atraumatic event such as car accident or even a tongue piercing.⁷ Another hypothesis states that TN arises secondary to a vascular loop that cross-compresses the trigeminal nerve a few millimeters proximal to the pons. This vulnerable area, known as the nerve REZ, marks the transition from central to peripheral myelin. Analogous cranial nerve neuralgias, such as hemifacial spasm and glossopharyngeal neuralgia, often involve similar lesions in their nerve REZ. Post herpetic Neuralgia (PHN), which occurs after shingles, may cause similar symptoms if the trigeminal nerve is

damaged. When there is no structural cause, the syndrome is called idiopathic.

Diagnosis is made from a well taken history. The classic clinical pattern will lead towards the diagnosis. Sometimes, symptoms may be less classic and may mimic toothache, sinusitis, stomatitis or other inflammatory condition. The neuralgic symptoms in younger group of patients should alert the clinicians to a possible intracranial space occupying lesion or intracranial arteriovenous anomalies. Other differential diagnosis should include acoustic neurilemoma, multiple sclerosis, post-herpetic neuroma or post-traumatic neuralgia. All patients should ideally have MRI scanning or at least a CT scan.

CLINICAL MANIFESTATIONS

The disorder is characterized by episodes of intense facial pain that usually last from a few seconds to several minutes or hours. The episodes of intense pain may occur paroxysmally. To describe the pain sensation, patients may describe a trigger area on the face, so sensitive that touching or even air currents can trigger an episode. It affects the lifestyle as it can be triggered by common activities such as eating, talking, shaving and tooth brushing.⁸ The attacks are said by those affected to feel like stabbing, electric shocks, burning, pressing, crushing, exploding or shooting pain that becomes intractable.

Individual attacks usually affect one side of the face at a time, lasting from several seconds to a few minutes and repeat up to hundreds of times throughout the day. The pain also tends to occur in cycles with remissions lasting months or years. 10-12% of cases are bilateral, or occurring on both sides. This normally indicates problems with both trigeminal nerves since one serves strictly the left side of the face and the other serves the right side. Pain attacks typically worsen in frequency or severity over time.⁸ Many patients develop the pain in one branch, then over years the pain will travel through the other nerve branches.

Outwardly visible signs of TN can sometimes be seen in males who may deliberately miss an area of their face when shaving, in order to avoid triggering an episode.⁹ Successive recurrences are incapacitating and the dread of provoking an

attack may make patients unable to engage in normal daily activities.

DIAGNOSIS

Sweet diagnostic criteria for TN:-

1. The pain is paroxysmal.
2. The pain may be provoked by light touch to the face (trigger zones).
3. The pain is confined to the trigeminal distribution.
4. The pain is unilateral.
5. The clinical sensory examination is normal.

DIAGNOSTIC ALGORITHM FOR TN³

Refer Figure 1.

WORKUP AND MANAGEMENT OF TRIGEMINAL NEURALGIA¹⁰

Refer Figure 2. The surgical management of TN is depicted in figure 3.

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LEGENDS

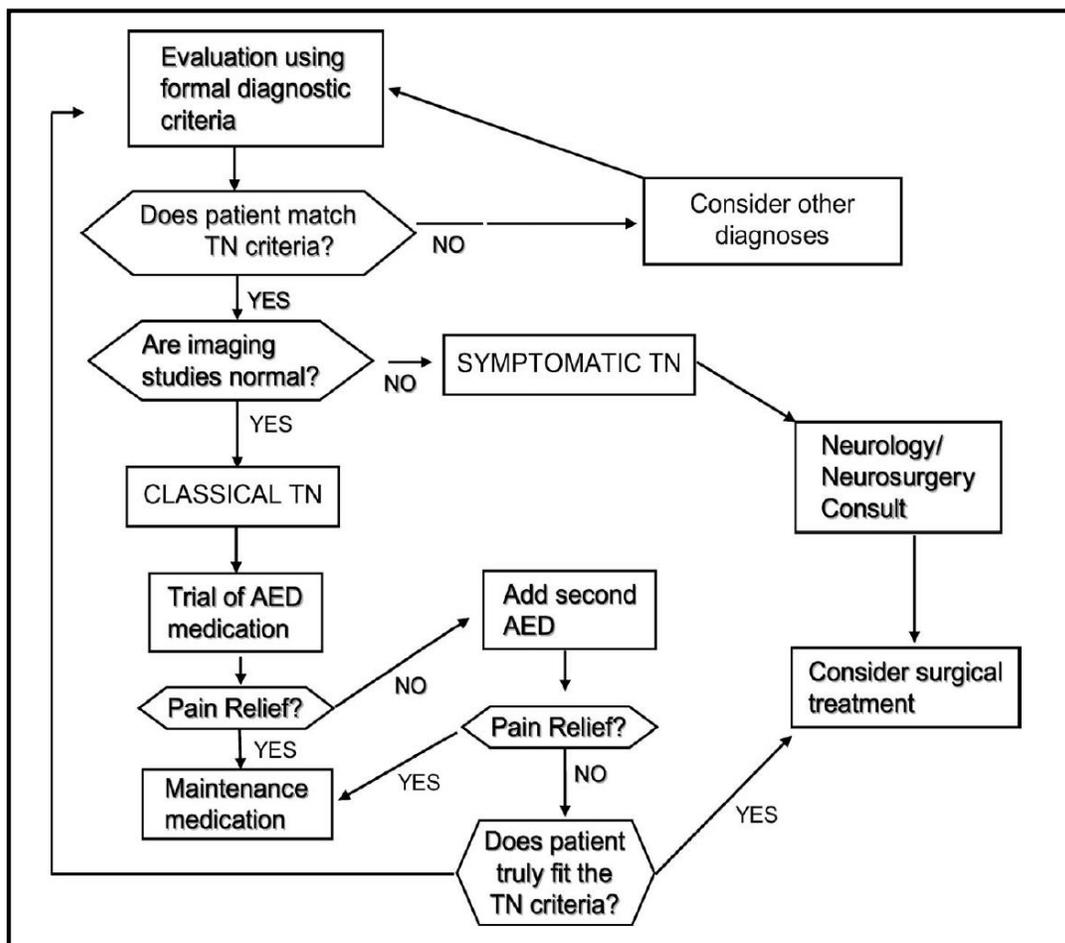


Figure 1. Diagnostic Algorithm for Trigeminal Neuralgia³

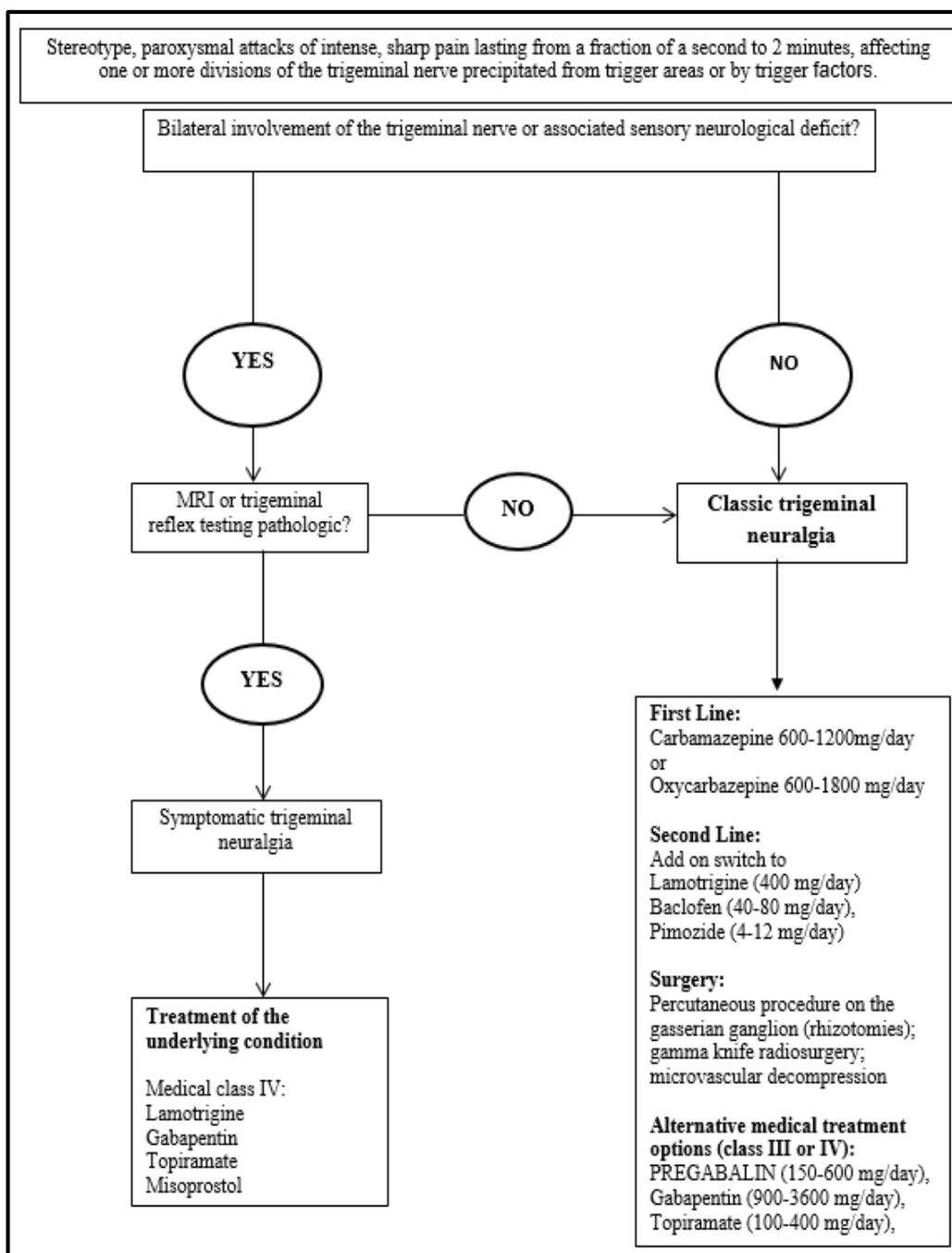


Figure 2. Workup and management of Trigeminal Neuralgia¹⁰

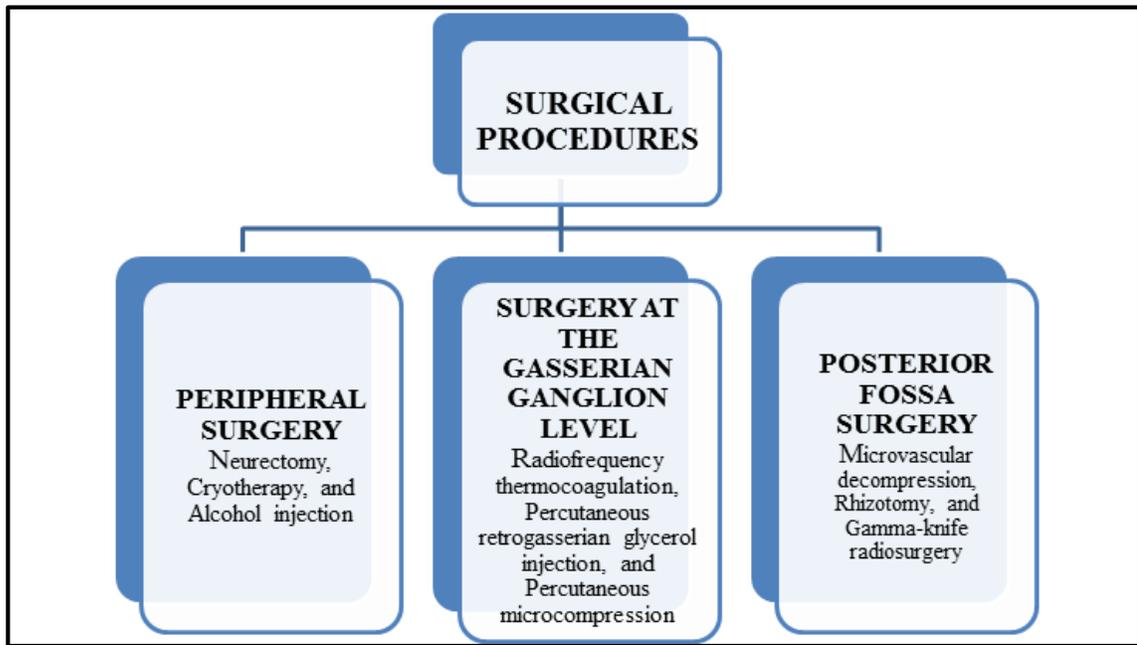


Figure 3. Surgical management of Trigeminal Neuralgia¹⁰