CASE REPORT

Dual findings: A clinically symptomatic case of trigeminal neuralgia with incidental radiographic finding of elongated styloid process: A rare case report and review of literature

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Abstract

Trigeminal neuralgia (TN) is a severe, short-lasting, unilateral facial pain. The most common is the classical type which is unrelated to pathology and most commonly caused by neurovascular compression of the trigeminal nerve, i.e., V cranial nerve root. Most (>85%) of TN cases are diagnosed as classical TN. In adults, the length of styloid process is about 25 mm with a tip located between the external and internal carotid arteries and seen lateral to the pharyngeal wall and the tonsillar fossa. Ossification of the stylohyoid and stylomandibular ligaments leads to elongation of the styloid process and causes clinical symptoms. This article describes a treated case of 55-year-old female patient with clinical symptoms of TN and an incidental radiographic finding of elongated styloid process.

Keywords:
Styloid process, tonsillar fossa, trigeminal nerve

Introduction

The IASP defined trigeminal neuralgia (TN) or Tic douloureux is sudden, usually unilateral, severe, brief, excruciating, recurrent episodes of pain in one or more divisions of the trigeminal nerve, i.e., V cranial nerve.[1]

The styloid process is a bony prominence, located immediately anterior to the stylomastoid foramen, varying from 20 to 25 mm in length. It is of cylindrical form and extends downward from the infratemporal surface toward the front, downward and medial side which narrows toward the tip of process.

Stylopharyngeus muscle gets its attachment medially and from the posterior side next to styloid process base, stylohyoid muscle from the posterior side and laterally on the central part of the process. Styloglossus muscle which begins from the anterior part of the process directly next to the tip. These muscles are innervated by glossopharyngeal, hypoglossal, and facial nerve.

The ligament of stylohyoid projects from styloid process tip to the lesser horn of the hyoid bone and the stylomandibular ligament which begins beneath the attachment of styloglossus muscle and ends on the angle of mandible. Some of the studies showed that a length of over 30 mm is considered elongated and few others consider 40 mm as elongated. Some rare cases exist with length of 73 mm.[2,3]

Case Report

A female patient aged 55 years old came to the Department of Oral Medicine and Maxillofacial Radiology with a chief complaint of pain and burning sensation in her right facial region since 3 months. Pain was sudden in onset, sharp, shooting, intermittent, radiating to the lower lip, aggravated on exposure to cold and chewing food and she was unable to chew on that side. Pain lasted for time duration of few seconds to 2 minutes and relieved on its own. History of 20-30 attacks per day. Pain was present on the right facial region and it was seems to extending from preauricular region to the lower lip and also in the vestibular region. History of disturbed sleep was noted. There was no relief of pain even after taking analgesic medications.
Medical history disclosed patient was known asthmatic since 10 years and was under medication for the same.
No history of drug allergy and had no adverse habits.
Past dental history disclosed history of extraction 3 months back in the upper and lower right posterior teeth region due to pain. The patient also gave history of aggravation of pain after extraction of teeth.
Extraoral examination revealed the presence of scar in the infraorbital region as shown in Figure 1.
The patient gave history of scar formation due to post-treatment for pain in her right upper back teeth region in her village and that area was burnt due to medications placed extraorally in that region 45 years back.
Intraoral examination of hard tissue revealed suspected presence of root stumps irt 18.

\[
\text{Missing teeth} \quad 876|678 \\
\text{876543}
\]

As shown in Figures 2 and 3
Dental caries was seen irt 37 and generalized attrition was present.

As an investigatory measure, a maxillary nerve block was given to the patient and the patient was symptom free for sometime after the nerve block.
With the above findings of unilateral occurrence of pain, age, triggering factors, sex and clinical history, we gave a provisional diagnosis of the TN.
Root stumps were not seen in intraoral periapical radiograph irt 18 as shown in Figure 3.

\[
\text{Orthopentamogram revealed missing teeth} \\
\text{irt Missing teeth} \quad 876|678 \\
\text{876543}
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Presence of significant lengthening of styloid process as shown in Figure 4.
The patient was referred to neurologist to rule out any pathology and for physician for her overall health status including her asthmatic condition and ENT consultation was taken.
After considering neurologist, physician, ENT surgeon’s opinion and no pathology (e.g., vascular compressions, tumors) was noticed related to it, the patient was advised to take tab Tegretol with a dose of 400 mg thrice daily for 1 week and recalled her again.

**Figure 1:** Extraoral examination showing presence of scar in the infraorbital region

![Figure 1](image1)

**Figure 2:** (a and b) Intraoral Figure of missing teeth irt 16, 17, 18, 26, 27, 28, 43, 44, 45, 46, 47, 48

![Figure 2](image2)

**Figure 3:** Intraoral periapical radiograph irt 18

![Figure 3](image3)

**Figure 4:** Panoramic radiograph showing missing teeth irt 16, 17, 18, 26, 27, 28, 43, 44, 45, 46, 47, 48 and presence of significant elongation of styloid process

![Figure 4](image4)
After 1 week, she reported us with complete reduction of pain and slight reduction of burning sensation after taking medication.

The patient was advised to take gabapentin 400 mg and nortriptyline hydrochloride 100 mg (Pentanerve nt) along with tegretol. Patient reported with complete reduction in pain and burning sensation after 1 month. Patient is being monitored.

Discussion

The trigeminal nerve is the \( V^6 \) cranial nerve and it is a mixed nerve with a large sensory root and small motor root. The sensory root carries information from the unilateral hemiside. The nerve is divided into three branches, i.e., \( V1 \), \( V2 \) and \( V3 \) which correspond to three different dermatomes. The unilateral muscles of mastication get its innervation from the motor root.\(^4\)

\( V1 \) gives nerve supply to scalp, forehead, and around eye. The area around cheek gets its nerve innervation from \( V2 \). \( V3 \) innervates area around jaw as shown in Figure 5.\(^5\)

The sensitive (sensory) portion takes to the central nervous system, from the skin and mucous membrane of greater portion of the face, which leads to “TN.”\(^6\)

TN occurs more frequently in the \( V2 \) and \( V3 \) divisions of the trigeminal nerve, so deep and limited to the trigeminal nerve distribution. The attacks may be accompanied by salivation, lacrimation, rhinorrhea, congestion of nasal mucosa, erythematous skin, facial edema, or clonic contraction, ipsilateral salivation, lacrimation, rhinorrhea, congestion of nasal mucosa, earache. Other features are vertigo, tinnitus, pain on turning the head, reduced opening of mandible, dysphonia, carotidynia and change in voice, hypersalivation, and even altered taste sensation. Although 4% of people are thought to have a lengthened styloid process among them only 4-10% is symptomatic.\(^8\)

History of TN

The 1st known description of TN, or a similar condition, was written in the 2nd century AD by Aretaeus of Cappadocia, a contemporary of Galen.\(^9\)

Jujani, an 11th century Arab physician in his writings, mentioned that unilateral facial pain causing spasms and anxiety. Interestingly, he suggested that the cause of the pain was due to the proximity of the artery to the nerve.\(^10\)

In 1773, the first full account of TN was published when John Fothergill presented a paper to the Medical Society of London. He gave the detailed elaboration of the typical features of the condition, including paroxysms of unilateral facial pain, evoked by speaking or eating or touch, starting and ending abruptly, and associated with anxiety.\(^9\)

Sometime earlier, Nicolaus Andrea had used the term tic douloureux to describe TN. However, it had been suggested, that no more than two of the patients he described, in fact, had TN.

Later in the 18th and 19th century, Sporadic observations by Pujol, Chapman and Tiffany helped to complete the clinical Figure and differentiate TN from common facial pain conditions like toothache.

In the early 20th century, Oppenheim mentioned a significant relation between a demyelinating disease like multiple sclerosis (MS) and TN and Patrick suggested on its familial incidence.

History of stylohyoid process elongation

In 1652, elongation of the stylohyoid ligament was first reported by Marchetti. Later in 1937, Eagle reported the first two symptomatic cases of ossified stylohyoid process.\(^11\)

Epidemiology

TN is a rare disease with few reported studies on its prevalence. The available literature suggests that the prevalence might be between 0.01% and 0.3% in the general individuals, even though few other studies suggested that it may be around 12%/1 lakh persons per annum. This, however, could be due to misdiagnosis. The females are twice commonly affected than males. TN can first occur at any age, but in greater than 90% of affected individuals onset occurs in the 4-5th decades, and maximum incidence is between 5th and 6th decades.\(^12\)
Incidence of ossified styloid process

Approximately, 4-10.3% of symptomatic cases of ossified styloid process occur in 4% of the general people. It is common in the third and fourth decade of life. Women are thrice commonly affected than males. Bilateral occurrence is frequent but does not always cause bilateral symptoms.[13]

Etiology of TN

The pathophysiological mechanisms of TN are not fully understood, mostly being related to trigeminal nerve compression by intracranial peripheral vessels, categorized as idiopathic TN. The mechanism responsible for triggering neuralgia would be the cooccurrence of the degenerative processes of aging associated with long-standing vascular compression on the trigeminal nerve posterior root.[14]

TN has an enormous psychological impact, few scholarly papers highlighted the ways how it can affect the quality of life of affected individuals.

In 2010, Luna et al. stated that 80-90% of TN may be idiopathic or triggered by trauma, emotional factors, external stimuli, and neoplasms.

Siqueira, Nóbrega, Valle et al. (2004)[15] in their study mentioned that TN may occur as a cause of brain lesions, arachnoid adhesions, vascular loops or plaques of demyelination situated at the entrance of the nerve root, which will create potential ectopic for TN.

Other possible phenomena related to the TN are autosomal dominant, autosomal recessive, possibly X-linked, as well as related to senility. The symptomatic causes of TN include MS, viral infections, tumors, aneurysms, and impaired post-dental extractions. The TN symptom may also be related to maxillofacial trauma, such as facial bone fractures, chiefly fractures of the zygomaticomaxillary complex, mainly when it involves the orbital floor causing injury to the infraorbital nerve.

Etiology of elongation of styloid process

1. Gokce et al. reported that ectopic calcification might play a role for the etiology of it, especially in the individuals with abnormal vitamin D, calcium and phosphorus metabolism. [7]
2. The calcification of muscles of tendons causes irritation of the adjacent structures thereby leading to elongation.
3. An association of the abnormal angulation with length of the styloid process.

Clinical features

Location, radiation: V2 and V3 divisions are the most commonly affected in TN. The pain is mostly unilateral, with only 3% of bilateral occurrence, and it is of little radiating in nature outside the distribution of trigeminal nerve.

Character: It is of sharp, shooting, Electric shock-like or stabbing in intensity.

Severity: Mostly moderate to severe but can be milder at times.

Classification

The revised International classification of headache disorders -3 suggest three variants of TN:[16]
1. Classical TN (CTN)
2. Symptomatic TN and
3. TN with concomitant persistent facial pain.

Langlis classification of styloid process ossification as:
1. Type I: Which is uninterrupted
2. Type II: Which is a pseudo-articulation between process of styloid and ligament of stylohyoid
3. Type III: In which there are interrupted segments of the mineralized ligament, creating the appearance of multiple pseudo-articulations.[17]

Classification is shown in Figure 6.

Figure 6: Langlis radiological classification of elongated styloid process
Diagnostic criteria for classic TN
• Each attack of pain can range from a time duration of few seconds to 2 minutes
• Pain should have at least one of the following features such as intense, superficial, sharp or stabbing in character and should provoke from trigger zones or by trigger factors
• Attacks of pain are similar in individual patients
• Clinically no evident neurological deficit
• Another disorder should not attribute to it.
  Olesen (1997) criteria are similar to above mentioned diagnostic criteria for CTN. TN is mainly diagnosed based on clinical signs and symptoms mentioned by the patient as typical paroxysms, refractory periods and trigger zones.[14]

Diagnosis of elongated process of styloid
It must be diagnosed on thorough medical history, and proper physical examination which includes careful intraoral palpation, i.e., by inserting the index finger in the tonsillar fossa region and gentle pressure is applied. By doing the clinician should be able to feel an elongated process of styloid. The diagnosis is very likely if pain occurred by palpation and referred to the ear, face, or head. It is usually not palpable if it is of normal length. One of the useful diagnostic tools is relief of pain after injecting local anesthetic solution into tonsillar fossa.[17]
• Radiological diagnosis
• Panoramic radiographs
• Soft tissue lateral radiograph of the neck
• Towne’s view of the skull
• CT scan.[18]

Differential diagnosis of TN
Dental infection/cracked tooth Well localized to tooth; local swelling and erythema; appropriate finding on dental examinations or X-ray. Temporomandibular joint pain often bilateral and may radiate around ear, neck and temples; jaw opening may be limited and “click” can be heard.

Persistant idiopathic facial pain (odontalgia)
Often bilateral and may extend out of trigeminal territory; pain often continuous, mild to moderate in severity, aching or throbbing in nature.

Temporal arteritis
Common in elderly; pain constant, a/w jaw claudication, fever. Temporal arteritis may be firm, tender, non-pulsatile on palpation. Sinusitis facial tenderness. Aching increasing with position. Acute ear infections A/w inflammatory changes in the ear Migraine Aura; severe unilateral a/w nausea, photophobia, phonophobia, and neck stiffness.[1]

Differential diagnosis of lengthened process of styloid
Laryngopharyngeal dysesthesia as well as dental malocclusion, glossopharyngeal, sphenopalatine, and TN’s, temporomandibular arthritis, temporal arteritis, diverticulum of esophagus, cervical vertebral arthritis, malignant or benign tumors, and cluster type headache and migraine has to be considered.

Investigations
• No specific investigations exist for the diagnosis of TN
• Dental X-rays can be done to clarify differential diagnosis
• Visual analog scale to rate pain score.
  MRI and Angiography is the best option to image trigeminal nerve and associated vascular malformations and diagnose the presence of tumors or cysts, plaques of MS and trigeminal nerve compressions.

Treatment
Medical treatment is mainly based on the use of antiepileptic medications. Based on present evidence-based guidelines, the first-line of choice is carbamazepine (200-1200 mg/day) and oxcarbazepine (600-1800 mg/day). The second-line therapy includes add-on therapy with lamotrigine (400 mg/day) or a switch to lamotrigine or baclofen (40-80 mg/day). Other antiepileptic drugs - such as gabapentin, phenytoin, valproate, and pregabalin - are also been suggested to be useful in the treatment.
  In the case of emergency, an IV infusion of fosphenytoin, as well as injections of lidocaine locally into trigger points can be helpful.[19]

Surgical treatment
Surgery should be considered as the treatment of choice if successful outcome is not obtained with medical therapy or if it results in evident deterioration of day-day activities. Surgical procedures include decompression of the affected nerve/vessel or destruction of the Gasserian ganglion. This modality provides the best pain relief, with results showing initial pain relief in 90% of affected individuals, >80% were pain free after 1 year and 75% were pain free after 3 years.
  Up to 4% of affected individuals showed important side effects such as cerebrospinal fluid leakage, aseptic meningitis, infarcts or hematomas.[12]
  Radiofrequency thermocoagulation, balloon compression, and percutaneous glycerol rhizolysis are the gasserian ganglion percutaneous techniques. 90% of patients reported relief of pain following these procedures. The follow-up results were 1 year after radiofrequency thermocoagulation technique, 68-85% were free from pain, after 3 years 54-64% were pain free, and after 5 years only 50% were pain free. Gasserian ganglion level surgery needs short-acting anesthetics and mainly includes minor overnight procedures with a very low rate of mortality.
  In gamma knife surgery in which a focused beam of radiation is passed at the trigeminal root located in the posterior fossa. It is an optional treatment for patients who are unfit for open surgery or those patients who are on coagulants (e.g., patients on warfarin).
Treatment of ossified process of styloid

Conservative or surgical approaches are used for the treatment of ossified process of styloid. Conservative approaches consist of transpharyngeal injection of corticosteroids and lignocaine, diazepam, NSAIDS, heat fomentation. The surgical approach includes manual fracturing of the ossified styloid process.[20]

Surgical shortening of the ossified styloid process either through intraoral or external approaches is considered as most satisfactory treatment outcome. Among intraoral or external approach, the later has the significant advantage of enhanced exposure of the process of styloid and the adjacent vital structures, also helps in performing the surgical resection of a partially elongated stylohyoid ligament. It. Transoral resection does not lead to obvious outside scars, but it has the disadvantage of the risk of deep cervical infection and associated neurovascular injury.[8,21]

References
