

**EAGLE SYNDROME: A CHALLENGE FOR ORAL AND MAXILLOFACIAL DOCTORS
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ABSTRACT

Eagle syndrome is a complication of elongated styloid process. The most common symptoms are: Unilateral Throat pain, dysphagia, facial pain, pain at the back of the tongue of the affected side, feeling of something struck in the throat, facial pain, difficulty in speech with voice changes. Eagle syndrome decreases the quality of life. Sometimes the pain of eagle syndrome can involve head, ear and TMJ region of affected site. Proper knowledge of anatomy of styloid process and its attachment is necessary for the early diagnosis on the basis of involvement of site, sign and symptoms and for planning the treatment. As health care professionals it's our duty to diagnose the case early and provide better treatment so that compromised quality of life of the patient is improved. OPG can be helpful in the radio graphical evaluation of elongated styloid process but CT image can be used as advanced tool for the radiographical evaluation of elongated styloid process. Local application of anaesthesia (lidocaine) can be used for the diagnosis.

KEYWORDS: Eagle syndrome; Styloid process; Sore throat; Stylohyoid ligament and Stylomandibular Ligament.**INTRODUCTION**

Eagle's syndrome, defined by Eagle in 1949, is characterised by morphological abnormality/ossification of the styloid process 1. The average length of the styloid process is 20-30 mm in the adult Caucasians and 15.4-18.8 mm in the Asian population. An elongated styloid process is defined by being at least 30 mm long.^[1,2,3]

Basic anatomy of styloid process:**Vessels and nerves**

Styloid process is surrounded by significant vessels and nerves

Medially:

- Internal jugular Vein
- Internal carotid artery
- Glossopharyngeal nerve(CNIX)
- Vagus nerve (CNX)
- Accessory nerve (CNXI)

Laterally:

- Occipital artery and Hypoglossal nerve (CNXII)

Ligaments:**Styloid process apex is the origin of two ligaments:**

1. The stylohyoid ligament (Attaches to the lesser cornu of the hyoid)
2. The stylomandibular ligament (Attaches to the ramus of mandible) both the ligaments facilitate the movement of the tongue, pharynx, larynx, hyoid bone and mandible

Muscles:

The distal component of the styloid process consists of the shaft and is the origin of 3 muscles:

1. The stylohyoid
2. Stylopharyngeous
3. Styloglossus

Table 1: Nerves And Its Supply: any hindrance/compression to these cranial nerves will directly hamper with the areas they supply and their functioning.

Glossopharyngeal nerve (IX)	Sensation /taste to posterior one third of the tongue, posterior pharynx, stylopharyngeus- swallowing, parotid gland- salivation
Vagus nerve (X)	Parasympathetic supply to eye, heart, gut, lungs, larynx (Sensation to airway, motor to vocal cords)
Accessory nerve (XI)	Supplies sternocleidomustoid (Rotates head) and trapezius (lifts shoulder)
Hypoglossal nerve (XII)	Supplies tongue muscles

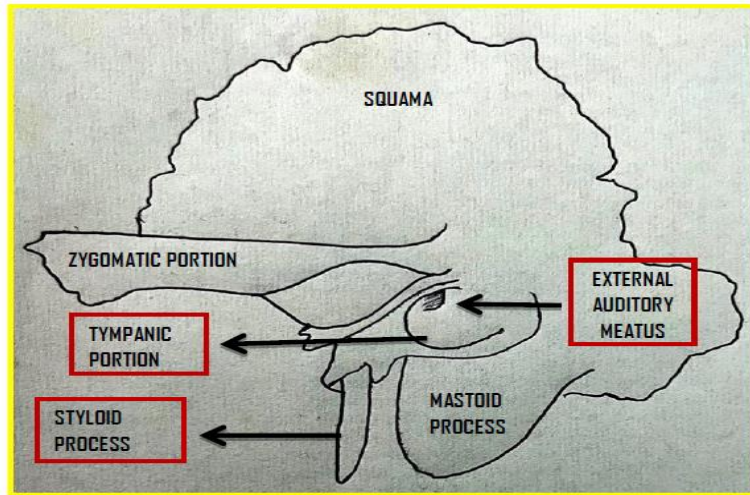


Fig. 1:- Line diagram showing styloid Process and Adjacent structures.

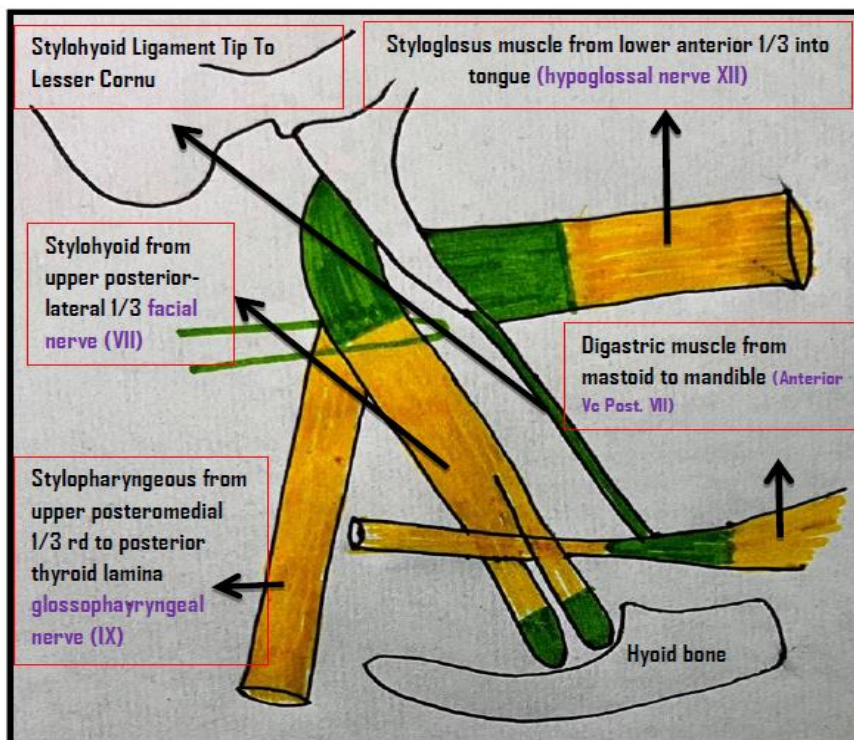


Fig. 2:- Right styloid process Muscle and Ligament attachments lateral view.

Epidemiology:

Approximately 4% of the general population has an elongated styloid process, and of these about 4% gives rise to the symptoms of Eagle syndrome.^[4] Therefore, the incidence of stylohyoid syndrome may be about

0.16%.^[4] More recent studies have reported the incidence of styloid elongation to be as high as 54%. The condition was first described by American otorhinolaryngologist Watt Weems Eagle in 1937.^[4]

Types:- Two Types

- a. Classical Eagle Syndrome or Stylohyoid Syndrome
- b. Stylocarotid Artery Syndrome

A. Classical eagle syndrome or stylohyoid syndrome

Presents as a sharp pain in the neck or the ear that extends to the maxilla, Face and Oral cavity.

Predisposing factors of pain:

- Rotation of Head
- Chewing
- Swallowing
- Extending the Tongue
- Yawning

It might also be associated with a foreign body sensation in the pharynx, tinnitus or vertigo. Additionally a mass might be palpable in the tonsillar fossa. Usually unilateral but could rarely present bilaterally. These symptoms occur due to irritation or possible entrapment of the nearby cranial nerves (CN:- V, VI, IX, X). It has been commonly observed that classic eagle syndrome presents post tonsillectomy or other pharyngeal surgery. The irritation or entrapment that occurs may be secondary to the formation of local granular cells.

B. Stylocarotid artery syndrome

Occurs when the styloid process impinges upon the internal or external carotid Artery and The nerve plexus accompanying those presents as:

- Pharyngeal Pain
- Eye pain
- Parietal cephalgia
- Resembling as a migraine or cluster headache

Compression of the internal carotid artery might present with symptoms of internal carotid vascular insufficiency such as:

- Weakness
- Visual changes
- Syncope

- Exacerbates with head movement
- The elongated styloid process might also pose the risk of carotid artery dissection leading to a transient ischemic attack or stroke

1. **Etiology-** Eagle syndrome occurs due to elongation of the styloid process or calcification of the stylohyoid ligament, potentially compressing the nearby carotid artery or glossopharyngeal nerve.^[4] However, the cause of the elongation hasn't been known clearly. It could occur spontaneously or could arise since birth. Usually normal stylohyoid process is 2.5–3 cm in length, if the length is longer than 3 cm, it is classified as an elongated stylohyoid process.^[5] There are reports of eagles syndrome being elicited after wisdom tooth removal.^[6]

Etiopathogenesis:

There is debate regarding the etiology of Eagle syndrome. Dr. Watt Eagle proposed that surgical trauma (tonsillectomy) or local chronic irritation causes osteitis, periostitis, or tendonitis of the styloid process and the stylohyoid ligaments which resulted in reactive, ossifying hyperplasia. Later Lentini (1975) suggested the hypothesis that persistent mesenchymal elements, also known as Reichert cartilage residues, could undergo osseous metaplasia in the setting of an appropriate traumatic or stressful event. Epifanio in 1962 considered that the ossification of the styloid process was also corresponding to endocrine disorders in women at menopause, who also had ossification of other ligaments in the body. Gokce C et al. (2008) reported that patients with the end-stage renal disease having abnormal calcium, phosphorus, and vitamin D metabolism had heterotopic calcification which caused elongation of the styloid process and thus the presentation of Eagle Syndrome. Finally, a retrospective study by Sekerci in 2015 indicated that a relationship exists between the presence of an arcuate foramen and an elongated styloid process. Results were derived from data from 542 patients employing three-dimensional CT scans.^[7,8]

Styloid process deviates laterally	Styloid process deviates posteriorly	Styloid process deviates medially and anteriorly
It will likely impinge upon the external carotid artery and its branches	It may impinge upon CN (IX, X, XI & XII) between it & the transverse process of atlas	It will irritate the tonsillar fossa and the important structures continues within it

Previously it was hypothesized that the formation of scar tissue around the styloid apex after tonsillectomy caused compression and straining of the neurovascular structures present in the retro styloid compartment. However, Eagle syndrome also presents in patients who have never been operated on for tonsillectomy. Several possible mechanisms for the pathogenesis of pain in Eagle syndrome have been proposed. The first considers that the elongated styloid process causes compression of cranial nerves, most commonly the glossopharyngeal nerve, with subsequent throat and neck pain.

Alternatively, there is the possibility of compression of the internal carotid artery by the styloid process, which can cause transient ischemic attacks or compression of the sympathetic nerves running along the artery, leading to an array of symptoms. The pain in Eagle syndrome often resembles glossopharyngeal neuralgia but is typically more dull and constant; however, cases with sharp intermittent pain along the path of the glossopharyngeal nerve have also been reported. Furthermore, theories of reactive hyperplasia and reactive metaplasia exist which associate the elongation

with either overgrowth of the styloid process itself or ossification of the stylohyoid ligament complex as a consequence of trauma. This phenomenon may explain the incidence of Eagle syndrome in patients after tonsillectomy, as it was originally described by Eagle. Other possible considered causes are the abnormal angulation associated with abnormally lengthy styloid process causing irritation of adjacent musculature or mucosa. Stretching and fibrosis involving the fifth, seventh, ninth, and tenth cranial nerves in the post-tonsillectomy period could also be a possible etiology. Finally, the symptoms may be a result of the normal process of aging. As normal aging is associated with a decrease in elasticity of soft tissues, degenerative and inflammatory changes in the tendinous portion of the stylohyoid insertion, a condition called insertion tendinosis, may cause pain in the distribution of glossopharyngeal nerve resembling Eagle syndrome. To avoid confusion, this manifestation better is called a pseudo-stylohyoid syndrome.^[9,10,11]

Clinical features

Age: Eagle's syndrome often observed in the third and fourth decades of life and in women more frequently than in men. Bilateral involvement is quite common but does not always involve bilateral symptoms.^[12]

Gender: Eagle's syndrome often observed in the third and fourth decades of life and in women more frequently than in men.^[12]

Sign and Symptoms: The main symptom of Eagle syndrome is pain usually on one side of your neck or face, especially near your jaw. The pain may come and go or be constant. It's often worse when you yawn or move or turn your head. You may also feel the pain radiate towards your ear.

Other symptoms of Eagle syndrome include:

- Headaches
- Dizziness
- Difficulty swallowing
- Feeling like something's stuck in your throat
- Ringing in your ears

Possible symptoms include:

- Sharp, shooting pain in the jaw, back of the throat, base of the tongue,^[1] ears, neck, and/or face^[5]
- Difficulty swallowing^[5]
- Sensation of having a foreign object in throat^[5]
- Pain from chewing, swallowing, turning the neck, or touching the back of the throat^[6]
- Ringing or buzzing in the ears

Classic Eagle syndrome is present on only one side; however, it may rarely be present on both sides. In vascular Eagle syndrome, the elongated styloid process comes in contact with the internal carotid artery below the skull. In these cases, turning the head can cause compression of the artery or a tear inside the blood

vessel, which restricts blood flow and can potentially lead to a transient ischemic attack or stroke. Sometimes, compression of the internal jugular vein can also occur and might lead to increased intracranial pressure.^[13,14]

Investigations:

1. Blood work is required to exclude possible systemic diseases. A complete blood count (CBC) is obtained if infection is suspected.
2. In the lidocaine infiltration test, 1 mL of 2% lidocaine is infiltrated into the anterior pillar and deep into the tonsillar fossa; after a few minutes, if the patient's symptoms are relieved temporarily, the test is regarded as positive, confirming the diagnosis of Eagle syndrome.
3. Diagnosis is suspected when a patient presents with the symptoms of the classic form of "Eagle syndrome" e.g. unilateral neck pain, sore throat or tinnitus. Sometimes the tip of the styloid process is palpable in the back of the throat. The diagnosis of the vascular type is more difficult and requires an expert opinion. One should have a high level of suspicion when neurological symptoms occur upon head rotation. Symptoms tend to be worsened on bimanual palpation of the styloid through the tonsillar bed. They may be relieved by infiltration of lidocaine into the tonsillar bed. Because of the proximity of several large vascular structures in this area this procedure should not be considered to be risk free.
4. Imaging is important and is diagnostic. Visualizing the styloid process on a CT scan with 3D reconstruction is the suggested imaging technique.^[12]
5. The enlarged styloid may be visible on an orthopantomogram (OPG) or a lateral soft tissue X ray of the neck.

Differential diagnosis:^[11]

- Cervical arthritis
- Cervical mass
- Faulty dental prostheses
- Migraine-type headaches
- Esophageal diverticula
- Otitis
- Possible tumors
- Salivary gland disease
- Temporal arteritis
- Trigeminal neuralgia
- cluster

Management

Conservative management

The management of Eagle syndrome is commonly divided into conservative methods of medical management or more definitive surgical treatment. Basic medical therapy can be further divided into first-line analgesics such as NSAIDs and alternative management consisting of a combination of anticonvulsants, antidepressants, local injections.^[15] For patients who

refuse surgery, Han et al report on a viable multi-drug approach with gabapentin, tianeptine, tramadol, and acetaminophen, coupled with local injections of 1 mg triamcinolone/mepivacaine. A weekly stellate ganglion block added to this regimen can result in near complete resolution of symptoms.^[16] Case reports have shown additional success utilizing a dual therapy with pregabalin and anticonvulsants such as carbamazepine.^[17] Taheri et al. demonstrated an 80% decrease in symptoms after 6 months of treatment with pregabalin (75 mg daily) and amitriptyline (10 mg daily).^[18] Local injection of anesthetic and dexamethasone has been proven effective as a final conservative option. However the effects of these injections diminish with time.^[19] While there have been reports of physical manipulation and manual transpharyngeal fracture, recommendations have come out against these techniques due to the risk to adjacent structures such as the carotid artery.^[20]

Surgical treatment

The literature tends to support that surgical treatment results in more definitive treatment and long lasting symptomatic relief.^[21] Surgical management is typically divided into the intraoral and cervical approaches.^[22] The traditional intraoral approach begins with a tonsillectomy followed by the removal of styloid process tip.

CONCLUSION

Eagle's syndrome should always be suspected when idiopathic unilateral pain occurs during swallowing, yawning and crying. As oral and maxillofacial specialist we face many such cases in our clinics. Proper diagnosis and treatment planning help the patients more. The elongation of the styloid process is a relatively common condition, although not all the affected patients complain of symptoms. Idiopathic unilateral throat and neck pain, mild dysphagia and foreign body sensation in the pharynx can help in the initial diagnosis. Investigations such as OPG and other advanced imaging modalities help in final diagnosis.

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