

Eagle syndrome – An overview



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Eagle syndrome represents symptoms brought about by compression of vital neurovascular and muscular elements adjoining the styloid process because of the elongation of styloid process or ossification of the stylohyoid or stylomandibular ligament. It is crucial for dentists, otolaryngologists and neurologists to be aware of the elongation of the styloid process and associated signs and symptoms. This article reviews the aetiopathogenesis, classification, investigative procedures and treatment modalities associated with Eagle syndrome.

Introduction

Eagle syndrome (ES) or stylohyoid syndrome is a rare condition that occurs because of the elongation of the styloid process or calcification of the stylohyoid ligament, characterised by painful sensation in the head and neck region.¹ This condition was first elucidated by the American Otorhinolaryngologist Watt Weems Eagle in 1937.²

Most of the patients with styloid elongation or calcified stylohyoid ligament may be asymptomatic. Only 4% of affected individuals are known to experience symptoms.³

Symptoms may arise only when these ossified structures exert pressure on the various vital structures in the cervico-facial region. Symptomatic patients may experience a wide spectrum of symptoms, including pain in cervico-facial region, pharyngeal discomfort, painful neck movements, change in voice, painful tongue movements, increased secretion of saliva, otalgia and headache. The normal styloid process measures approximately 2.5 cm – 3.0 cm in length.²

Aetiopathogenesis

The aetiology of ES is not clearly known. Although few suggest that dystrophic and degenerative changes in the hyoid complex of the styloid process might be the cause for ES, others suggest that cervico-facial inflammations, tumours, tonsillectomies and trauma could play a major role in causing ES.⁴

The suggested pathophysiological mechanisms for the pain in Eagle syndrome⁵

The following are the different aspects that could lead to pain because of the Eagle syndrome:

- Compression of the neural elements, the glossopharyngeal nerve, lower branch of the trigeminal nerve and/or the chorda tympani by the elongated styloid process;
- Fracture of the ossified stylohyoid ligament, followed by proliferation of granulation tissue that causes pressure on surrounding structures, resulting in pain;
- Impingement on the carotid vessels by the styloid process, producing irritation of the sympathetic nerves in the arterial sheath (Figure 1);
- Degenerative and inflammatory changes in the tendinous portion of the stylohyoid insertion, a condition called insertion tendinosis;
- Irritation of the pharyngeal mucosa through direct compression by the styloid process; and
- Stretching and fibrosis involving the fifth, seventh, ninth and tenth cranial nerves in the post-tonsillectomy period.

Classification systems

Eagle hypothesised that the syndrome has two types: the classic type and the carotid artery type. These types were also elucidated in the studies of Breault⁶ and Lorman and Biggs.⁷ The classic type is often noticed in patients with a history of tonsillectomy and arises secondary to the stimulation of the trigeminal (fifth), facial (seventh), glossopharyngeal (ninth) and vagus

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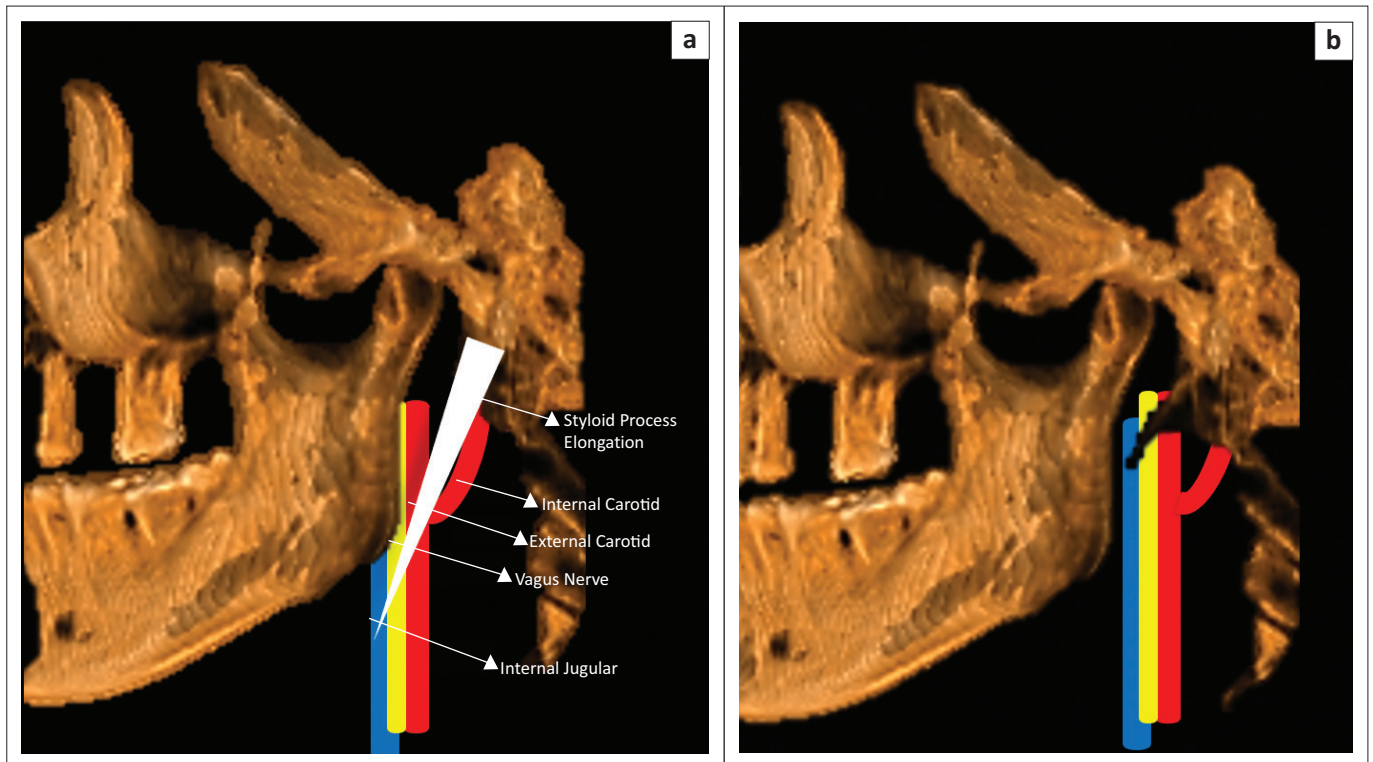
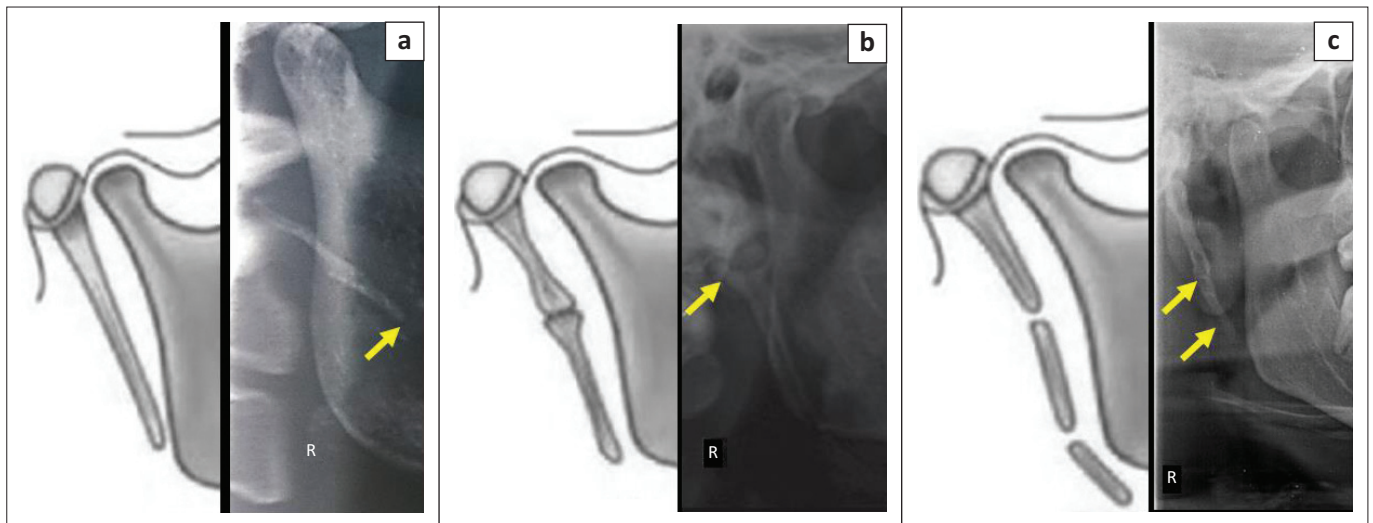


FIGURE 1: Schematic representation of (a) elongated styloid process causing impingement on vital structures versus (b) normal styloid process.



Sources: Adapted from Langlais RP, Miles DA, Van Dis ML. Elongated and mineralized stylohyoid ligament complex: A proposed classification and report of a case of Eagle's syndrome. *Oral Surg Oral Med Oral Pathol.* 1986;61:527–532. [https://doi.org/10.1016/0030-4220\(86\)90400-7](https://doi.org/10.1016/0030-4220(86)90400-7)

FIGURE 2: Radiographic classification system of styloid elongation (a) Type I (b) Type II (c) Type III, according to Langlais et al.⁸

(tenth) cranial nerves or their associated branches. Eagle speculated that after tonsillectomy, these individuals develop scarring near the styloid apex which subsequently compresses or stretches nerve structures in the space surrounding the styloid process, causing pain. In the carotid artery type, the styloid process gets associated with the carotid nerve plexus and causes a foreign body sensation in the pharynx and cervical pain on rotation of the head.

The radiographic classification system according to Langlais et al.⁸ includes the following three types of appearances (Figure 2):

- Type I – Represents an uninterrupted, elongated styloid process.
- Type II – The styloid process apparently being joined to the stylohyoid ligament by a single pseudo-articulation, which gives the appearance of an articulated elongated styloid process.
- Type III – Consists of interrupted segments of the mineralised ligament, creating the appearance of multiple pseudo-articulations within the ligament.

Classification of elongation of styloid process based on type of calcification:

- Type I – Elongated
- Type II – Pseudo articulated
- Type III – Segmented

Classification of elongation of styloid process based on pattern of calcification:

- A – Calcified outline
- B – Partially calcified
- C – Nodular
- D – Completely calcified

Classification based on angulation:

- Narrow
- Normal
- Wide

Diagnosis

The preliminary diagnosis of ES is based on a proper medical history and extraoral cum intraoral examination. The elongated styloid process can be felt intraorally by digital palpation. A gentle pressure is exerted using the index finger over the tonsillar fossa; if pain is reproduced or referred to face, head, neck or ear, the presumptive diagnosis of an elongated styloid process is very likely to be present. A styloid process of normal length is usually not palpable. Injection of local anaesthetic into the tonsillar fossa relieves the pain and can be used as a diagnostic tool.

Plain film radiographs are the commonest modality chosen. Radiographic imaging includes a panoramic radiograph (Figure 3), lateral-oblique mandible, lateral head and neck radiograph, Towne plain film radiograph and other facial projections. A threshold length of greater than 3 cm is accepted as abnormal by current conventions. Lateral views are the best to show the length of the styloid process but anteroposterior views are also required to determine bilateral involvement and lateral deviation.

CT scans have been used in difficult cases to support the diagnosis. Three-dimensional reconstruction has made it possible to envision the exact spatial orientation of the styloid processes (Figures 4 and 5). Ossification of the stylohyoid ligament can be excluded at radiographic imaging. The



FIGURE 3: Digital orthopantomogram showing the presence of bilateral elongated styloid processes.

added advantage of 3-D over 2-D radiographic imaging (conventional radiography) is the elimination of superimposition of anatomic structures and appreciation of soft-tissue changes, which is seldom visible in 2-D radiographic imaging. Barium swallow studies can show the impression of the elongated styloid process as a filling defect.³

Treatment

Both medical and surgical management options are available for treating styloid elongation, but medical management is known to provide only short-term relief. Conservative treatment modalities included transpharyngeal injection of local anaesthetics (lignocaine) and corticosteroids, non-steroidal anti-inflammatory agents (NSAIDs), diazepam, and the application of heat therapy and traditional Chinese herbs. Transpharyngeal manipulation with manual fracturing of the styloid process does not usually relieve symptoms but also poses the risk of damaging the nearby neurovascular structures. The most satisfactory and effective treatment is surgical shortening of the styloid process through either an intraoral or external approach.¹¹ Prognosis of ES is guarded by surgical failures (up to 20% of patients). This may be attributed to intraoperative injury, consequent fibrous entrapment syndrome or inappropriate shortening of the styloid process, assuming that the diagnosis was correct in the first place.¹²

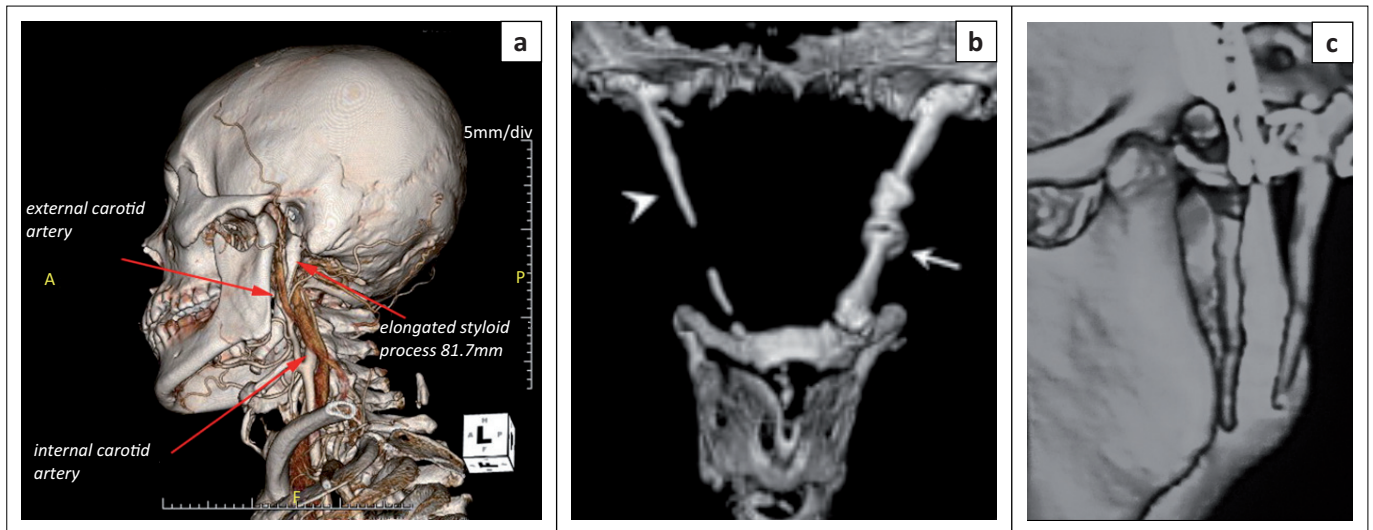
Differential diagnosis

A myriad of differential diagnoses has to be ruled out before arriving at a conclusion. Some of the differential diagnoses includes laryngopharyngeal dysesthesia, third molar impaction or dental-related pain, neuralgia of the sphenopalatine ganglia, glossopharyngeal and trigeminal nerve, temporomandibular joint disorders (TMDs), chronic tonsillo-pharyngitis, hyoid bursitis, Sluder's syndrome, cluster-type headache, migraine, atypical facial pain, oesophageal diverticula, temporal arteritis, cervical vertebral arthritis and benign or malignant neoplasms.¹³

Discussion

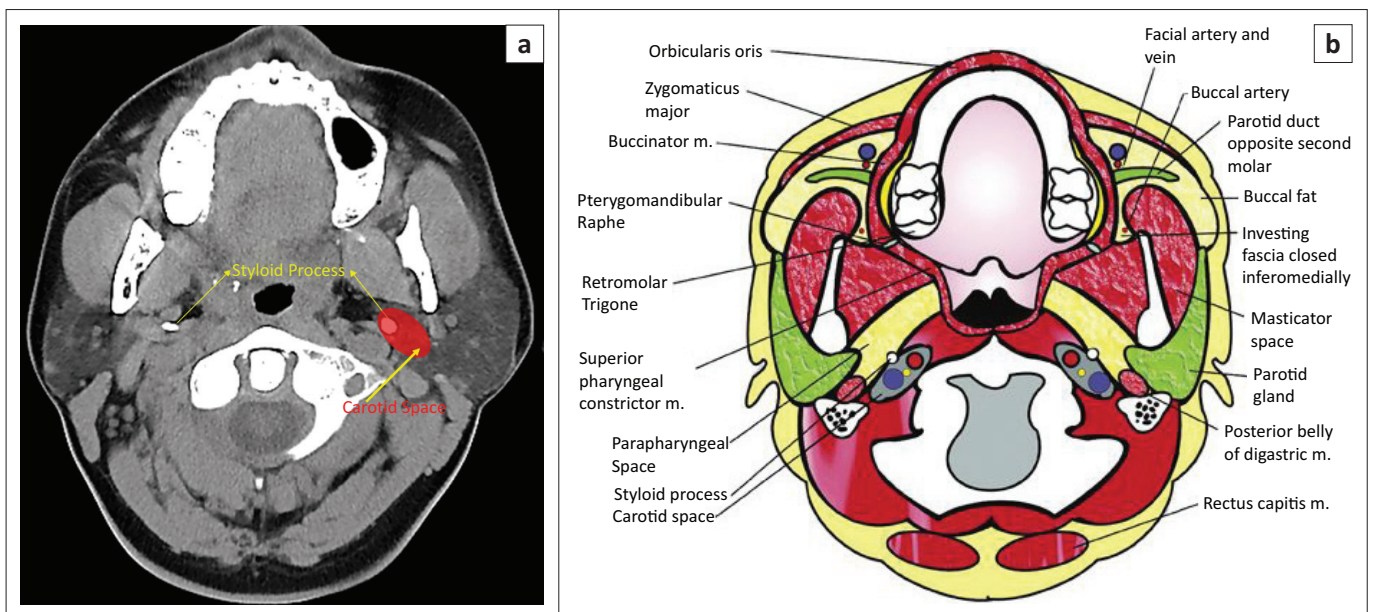
Styloid elongation was first described in 1652 by an Italian surgeon, Pietro Marchetti, who credited it to an ossifying process. In 1937, Watt W. Eagle coined the term stylalgia to describe the pain associated with this abnormality. Eagle reported over 200 cases in a 20-year study process and explained that the normal styloid process is approximately 2.5 cm – 3.0 cm in length. He observed that a slight medial deviation of the styloid process could result in severe symptoms of atypical facial pain.^{2,6}

The diagnosis of ES rests on proper medical history and prompt physical examination. An elongated styloid process can be palpated along the tonsillar fossa with the index finger, eliciting or reproducing pain, and injecting a local anaesthetic solution can relieve pain. Radiographic imaging plays a key role in providing a confirmatory diagnosis. Among all the radiographic imaging techniques, 3-D CT



Sources: (a) Dong Z, Bao H, Zhang L, Hua Z. Eagle's syndrome associated with lingual nerve paresthesia: A case report. *J Oral Maxillofac Surg.* 2014;72:886.e1–886.e4. <https://doi.org/10.1016/j.joms.2014.02.011>; (b) Raina D, Gothi R, Rajan S. Eagle syndrome. *Indian J Radiol Imaging.* 2009;19:107–108. <https://doi.org/10.4103/0971-3026.50826>; (c) Provided by author, Kavita Nedunchezian.

FIGURE 4: Various three-dimensional CT reconstruction images depicting elongation of the styloid process.^{9,10} (a) 3DCT showing elongated styloid in association with vital structures, (b) 3DCT antero-posterior view, showing complete and partial calcification of stylohyoid ligament and (c) 3DCT depicting elongated styloid process.



Sources: (a) Provided by author, Kavita Nedunchezian; (b) Seelagan D. Noujaim SE. A pictorial review of the anatomy and common pathology of the buccal space: "The overlooked space". *Appl Radiol.* 2017; Jan, 17. Available from: <http://appliedradiology.com/articles/a-pictorial-review-of-the-anatomy-and-common-pathology-of-the-buccal-space-the-overlooked-space>

FIGURE 5: (a) CT-axial view representing styloid process and associated carotid space, and (b) schematic representation.

scans seem to provide the best visualisation of the elongation and associated injuries to the adjoining neurovascular and muscular elements.

Both surgical and non-surgical means of treating ES are available. Non-surgical means provide only temporary relief, whereas surgical reduction in styloid elongation appears to have a better outcome. However, the prognosis in surgical failures should also be considered.

Compression of the internal carotid artery because of styloid elongation can lead to a transient ischaemic attack which is quite intimidating. Vagus-mediated cardiac

inhibition because of ES causing sudden death has been recorded in literature, and it was only at autopsy that it was revealed that the elongated styloid process had been compressing both the carotid sinuses; radiographic films had failed to show the anatomical changes.^{14,15}

There is a plethora of disease imitating the symptoms associated with ES. It is easy to misinterpret ES. For instance, ES can be atypically present as an exertional headache, beginning in the right ear and radiating to the neck and to the vertex.¹⁶ Another reported case was with diffuse bilateral stylohyoid chain ossification which mimicked symptoms of ES but was diagnosed as TMD.¹⁷

Conclusion

ES can be confused or mistaken for many other conditions, which can be excluded by proper and detailed history, physical examination and radiological investigations. Resection of the elongated styloid process is the treatment of choice for long-term benefit. Owing to the simple fact that a styloid process with normal length is not normally palpable, simple digital palpation over the tonsillar fossa can help us arrive at a presumptive diagnosis. A definitive diagnosis can then be arrived at after conducting appropriate radiographic investigations. Radiologists and dentists need to be cautious when viewing orthopantomograms to ensure that they assess all the structures that can be seen and not just the teeth, alveolar bone and temporomandibular joints.

Ethical consideration

Ethical consideration is not applicable for this publication.

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Competing interests

The author declares that she has no financial or personal relationship that may have inappropriately influenced her in writing this article.

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