



Eagles Syndrome: A Current Update

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Abstract

Eagle's syndrome is a relatively rare disorder whereby a calcified stylohyoid ligament or elongated styloid process gives rise to a multitude of otolaryngological symptoms. It is also a source of craniofacial and cervical pain. Eagle's syndrome is one of the glaring examples where the exact etiology eludes the treating doctor for a long time. It is important for a dental practitioner to be aware of this anomaly and its anatomic basis. Diagnosis can be made both radiographically as well as by physical examination of the patient. Most frequently, a panoramic radiograph is used to determine the appearance of stylohyoid ligament complex.

Keywords: Eagle Syndrome; Elongated Styloid Process; Orofacial Pain; Panoramic Radiography

Introduction

The styloid process and ligament are embryological remnants of the second pharyngeal arch. The styloid process is a long, slender and pointed bony protuberance that project downwards, forwards and slightly medially from the temporal bone. It descends between the external and internal carotid arteries to reach the side of the pharynx [1]. In adults, the styloid process is usually 2.0 to 3.0 cm long [2]. The length of an elongated styloid process is greater than 3 cm, and presents with varied clinical manifestations such as throat pain, difficulty in deglutition, feeling of trapped foreign body, and carotid artery compression syndrome. The first description of the elongation was given by Marchetti, an Italian scientist as early as late 16th century. Eagle, in 1937 defined the pain originating from elongated styloid process as Stylalgia [3]. Eagle's syndrome is a rare disease entity characterized by the symptomatic elongation of the styloid process or mineralization of the stylohyoid ligament complex [4]. Eagle's syndrome presents with a plethora of varied clinical manifestations and may mimic neuralgic pain [5], or oral, dental and temporomandibular joint pain [6]. An accurate diagnosis is established by thorough physical examination and radiographic evaluation. The condition may be managed by pharmacological therapy (oral carbamazepine, steroids/local anaesthetic solution injection) or surgical intervention by an intraoral or extraoral resection of styloid process [7]. In this paper, a general overview of Eagle's syndrome is presented.

Epidemiology

Epidemiological studies suggest that an elongated styloid process is seen in approximately 4% of the population [7], and only 4 - 10% of the affected individuals present with clinical signs and symptoms [8]. The condition has a slight predilection for females [9] and individuals of older age group [10]. The severity of symptoms and the severity of the ossification do not show a significant association. Even though ossification occurs in individuals under the age of 31, very few become symptomatic (1 - 5%) [11]. Patients exhibiting symptoms are typically over the age of 40 [12].

Etio-Pathogenesis

Murtagh et al. put forward three causative theories to describe the progression of Eagle's syndrome. The first theory is congenital elongation of the styloid process due to the persistence of the cartilaginous precursor, the second is stylohyoid ligament calcification by an unexplained process, and the third is the bony tissue growth at the site of stylohyoid ligament insertion [13].

Steinmann further proposed three theories to explain the ossification [14].

The "Theory of Reactive Hyperplasia" states that trauma can cause ossification at the end of the styloid process down the length of the stylohyoid ligament. Since the stylohyoid ligament contains remnants of its connective tissue and fibrocartilagenous origins,

the potential for ossification remains. The post traumatic symptoms are due to impingement of the carotid arterial system especially upon rotation or flexion of the head.

The second theory “Theory of Reactive Metaplasia” also involves an aberrant post traumatic healing response. Here, the presence of ossifying centres within the four segments initiates the calcification of the ligament. Thus, symptoms arising from the stiffened ligaments are primarily of a pharyngeal character. The symptoms resembles the “object stuck in the throat” complaint and pain upon swallowing.

The third theory “Theory of Anatomic variance” states that the early elongation of the styloid process and ossification of the stylohyoid ligament are anatomic variations that occur without recognizable trauma.

According to the “Theory of Aging developmental anomaly”, the ageing process is associated with loss of soft tissue suppleness and a localized inflammatory response. This inflammatory reaction results in inflammation of the tendon at the junction of ligament and the lesser cornu. This would not be a true calcified stylohyoid syndrome, Carmada, *et al.* termed this as “pseudostylohyoid ligament” [15].

Clinical types and features

Eagle’s syndrome manifest with a plethora of bizarre clinical signs and symptoms. Throat and neck pain, and the feeling of a trapped foreign body in the throat are the most frequently encountered manifestations. Referred ear pain, headache, carotidynia, dizziness, and dysphagia are the less common features. Turning the head and/or neck flexion may aggravate the pain. Few cases may also present with episodes of transient ischemia and syncope [16,17]. Cases where styloid process shows bilateral elongation may be fatal due to due to compression of both carotid arteries [18].

Eagle described the syndrome into two distinct types. The classic type of Eagle’s syndrome usually occurs following tonsillectomy. The resultant scar tissue causes compression and stretching of cranial nerves supplying the region (V, VII, IX and X nerves). The patient usually present with feeling of foreign body in throat, pain referred to the ear, and difficulty in swallowing.

The carotid artery type of Eagle’s syndrome causes compression of sympathetic nerve plexus. The affected individuals usually present with migrainous and neurological manifestations. Compression of internal carotid artery causes ipsilateral headaches and external carotid artery compression results in pain in the temporal and maxillary branch areas [19,20].

Radiographic features

A radiographic classification of the elongated and mineralized stylohyoid ligament complex was given by Langlais, *et al* [22]. There classification included three types of abnormal radiographic appearances and four patterns of calcification/mineralization.

Types of elongation

Elongated: An uninterrupted elongation of the styloid process is seen radiographically (> 25 - 28 mm) (Figure 1).

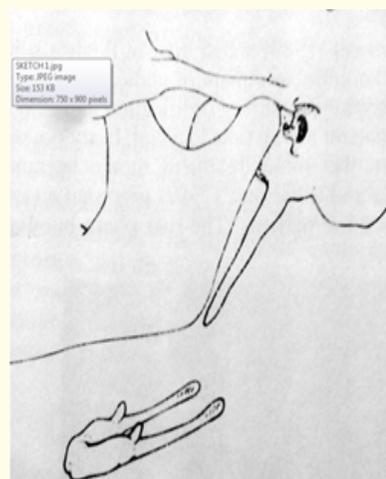


Figure 1

Pseudo-Articulated: Less frequent than the elongated type, the two mineralized segments are joined by a single pseudo-articulation that is usually located at the level superior to lower border of the mandible (Figure 2).

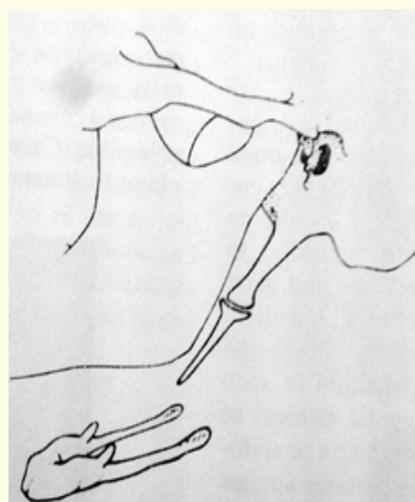


Figure 2

Segmented: Consists of long or short non-continuous portions of the styloid process or uninterrupted segments of mineralized ligament (Figure 3).



Figure 3

Patterns of calcification

- Calcified outline:** Reminiscent of the radiographic appearance of a long bone with a thick radio opaque cortex and a central lucency that constitutes most of the process.
- Partially calcified:** Thicker radio opaque outline, with almost complete opacification as well as a small and occasionally discontinuous radiolucent core.
- Nodular complex:** This pattern has a scalloped outline and may be partially or completely calcified with varying degree of central lucency.
- Completely calcified:** Totally radiopaque with no evidence of lucent inner core.

Diagnosis

Various parameters are taken in to account to establish a precise diagnosis of this condition. (1) Presenting manifestations (2) manual palpation of styloid process in the tonsillar fossa (3) infiltration of anesthetic solution (4) Radiologic assessment.

Thorough clinical examination is done by palpation of the styloid process in the tonsillar fossa. It is felt as a bone spicule causing pain, which is relieved by infiltrating lidocaine [23]. Temporary remission from pain after infiltration of local anaesthetic solution in tonsillar fossa is highly suspicious for a diagnosis of eagle's syndrome [24].

Various imaging techniques (panoramic, oblique lateral of skull and neck, anteroposterior skull, Towne, neck angiography, or magnetic resonance) are employed to reveal the presence of elongated and calcified stylohyoid process.



Figure 4: OPG showing bilaterally elongated styloid process.

Currently, computed axial tomography (CT) and, primarily, cone beam computed tomography (CBCT) is preferred for radiographic evaluation of eagle's syndrome because it prevents image overlap and provides low levels of distortion, larger scale contrast, precise length of the styloid process along with its angulation and vicinity to anatomical landmarks [23].

Differential diagnosis [25]

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.

Treatment

Noninvasive treatment protocol is the choice of therapy for managing the neurologic complications of Eagle's syndrome. Short term remission may be achieved by intake of oral agents (carbamazepine, amitriptyline, gabapentin, and valproate) and image-guided corticosteroid injections in the tonsillar fossa. Surgical management is reserved for cases recalcitrant for conservative treatment. Both Intra oral and extra oral approaches are employed for surgical resection of styloid process, although

the intraoral procedure is most frequently used [21]. The external approach is easily accessible and can be performed with minimal blood loss and cervical infection. However, the major drawback is the resulting cutaneous scar mark [26].

Conclusion

The clinician should be familiar with the bizarre clinical presentation of Eagle's syndrome, and an elongated styloid process should be kept as a differential diagnosis in cases of pain due to impacted or unerupted third molars. Thorough history taking, clinical examination and radiographic evaluation usually facilitate the dentist in making an accurate diagnosis of Eagle's syndrome.

Bibliography

1. BD Chaurasia. "Human Anatomy: Part III: Head, neck and Brain".
2. Eagle WW. "Elongated styloid process: Further observations and a new syndrome". *Archives of Otolaryngology* 47.5 (1948): 630-640.
3. Kim E., et al. "Eagle Syndrome: A Case report and review of literature". *Ear, Nose and Throat Journal* 87.11 (2008): 631-633.
4. Victor B Feldman. "Eagle's syndrome: A Case of symptomatic calcification of the stylohyoid ligament". *Journal of the Canadian Chiropractic Association* 47.1 (2003): 21-27.
5. Pierrakou ED. "Eagle's syndrome: Review of the literature and a case report". *Annals of Dentistry* 49 (1990): 30-33.
6. Zohar Y., et al. "Elongated styloid process syndrome masquerading as pain of dental origin". *Journal of Oral and Maxillofacial Surgery* 14.5 (1986): 294-297.
7. Diamond LH., et al. "Eagle's syndrome: A Report of 4 cases treated using a modified extraoral approach". *Journal of Oral and Maxillofacial Surgery* 59.12 (2001): 1420-1426.
8. Rechtweg JS and Wax MK. "Eagle's syndrome: A Review". *American Journal of Otolaryngology* 19.5 (1998): 316-321.
9. Woolery WA. "The diagnostic challenge of styloid elongation (Eagle's syndrome)". *Journal of the American Osteopathic Association* 90.1 (1990): 88-89.
10. Unlu Z., et al. "Elongated styloid process and cervical spondylosis". *Clinical Medicine Case Reports* 1 (2008): 57-64.
11. Gossmann JR and Tarsitano JJ. "The styloid-stylohyoid syndrome". *Journal of Oral Surgery* 35 (1997): 555-560.
12. Balasubramanian S. "The ossification of stylohyoid ligament and its relation to facial pain". *British Dental Journal* 116 (1964): 108-111.
13. Moon C-S., et al. "Eagle's syndrome: a case report". *Journal of the Korean Association of Oral and Maxillofacial Surgeons* 40.1 (2014): 43-47.
14. Steinmann EP. "Styloid syndrome in absence of an elongated process". *Acta Otolaryngologica* 66.4 (1968): 347-356.
15. Carmada AJ., et al. "Stylohyoid chain ossification A Discussion of etiology". *Oral Surgery, Oral Medicine, Oral Pathology, and Oral Radiology* 67.5 (1989): 508-514.
16. Farhat HI., et al. "Eagle syndrome as a cause of transient ischaemic attacks". *Journal of Neurosurgery* 110.1 (2009): 90-93.
17. Martin TJ., et al. "Transcervical resection of the styloid process in Eagle syndrome". *Ear Nose and Throat Journal* 87.7 (2008): 399-401.
18. Ruwanpura PR., et al. "Sudden unexpected death probably due to Eagle's syndrome: a case report". *Medicine, Science and the Law* 48.4 (2008): 350-353.
19. Colby CC and Del Gaudio JM. "Stylohyoid complex syndrome: a new diagnostic classification". *Archives of Otolaryngology - Head and Neck Surgery* 137.3 (2011): 248-252.
20. Costantinides F., et al. "Eagle's syndrome: signs and symptoms". *Cranio* 31.1 (2013): 56-60.
21. Fusco DJ., et al. "Eagle's syndrome: embryology, anatomy, and clinical management". *Acta Neurochirurgica* 154.7 (2012): 1119-1126.
22. Langlais RP., et al. "Diagnostic imaging of the jaws". Philadelphia: Lea and Febiger (1995): 617-623.
23. Arbildo H., et al. "Eagle syndrome. A narrative review". *Journal of Oral Research* 5.6 (2016): 248-254.
24. Jayachandran S and Khushboo G. "Stylocarotid syndrome- An unusual case report". *Contemporary Clinical Dentistry* 3.4 (2012): 503-507.
25. Nedunchezian K. "Eagle syndrome - An Overview". *South African Journal of Radiology* 21.1 (2017): 1247.
26. Strauss M., et al. "Elongated styloid process syndrome. Intraoral vs external approach for styloid surgery". *Laryngoscope* 95.8 (1985): 976-979.

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