

Class Room

Pain In The Chain (Eagle's Syndrome) - An Overview

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Abstract

Eagle's syndrome was first proposed by an otorhinolaryngologist Watt Weems Eagle in the year 1937. Eagle's syndrome occurs due to the ossification of the stylohyoid chain and patients with Eagle's Syndrome may present with a history of pain in the tonsillar region, dysphagia and pain on turning the head, foreign body sensation in the throat and referred otalgia. Extra oral radiographs like panoramic radiographs, lateral head and neck radiographs aid in the diagnosis of this syndrome, followed by surgical management to relieve the clinical symptoms.

Key Words: Styloid Chain, Pain in the Throat, Stylohyoid Syndrome.

Introduction

Eagle's syndrome is a rare clinical condition which is of interest to the dentists and ENT surgeons. The limitation in knowledge about this particular syndrome can be the reason for less clinical exposure to this condition. It is also considered a field of interest for dentists, as the surgical correction for this particular condition is often done by an intraoral approach. This paper gives a complete overview on Eagle's syndrome with appropriate differential diagnosis to be considered when encountered by dentists.

Normal Anatomy

Location: The styloid process is a thin bony projection of the petrous temporal bone which lies anterior to the stylomastoid foramen. Normally, it measures around 25 mm – 28 mm in length, but might vary from person to person and even from one side to the other. The styloid process projects forward, downward, and slightly medially with the tip of the process located between the internal and external carotid arteries. It lies posterior to the tonsillar fossa and lateral to the pharyngeal wall¹.

Attachments: The styloid process along with its attachments is called the Styloid Apparatus. Three muscles and two ligaments are attached to the process. The muscles are - the stylopharyngeus (from the base of the process; innervation: glossopharyngeal nerve), the stylohyoid (attached to the middle portion; innervation: facial nerve), and the styloglossus (arising from extremity of process; innervation: hypoglossal nerve). The two ligaments are - the stylomandibular ligament (from the apex of the process attaching at the angle of the mandible), and the stylohyoid ligament (from the apex of the process attaching at the lesser cornu of the hyoid bone)¹.

Embryological Considerations

Eagle's Syndrome may occur due to a stylohyoid chain ossification. This chain consists of the styloid process, the stylohyoid ligament and the lesser cornu of the hyoid bone. The stylohyoid chain is a derivative of the second branchial arch (hyoid arch) arising from the Reichert's cartilage. In many mammals, this cartilage gives rise to a series of four bony parts: the tympanohyal, stylohyal, epihyal and ceratohyal. In humans, it is believed that the tympanohyal part fuses with the stylohyal and petrous part of the temporal bone and ossifies to form the styloid process. Normally, the epihyal cartilage degenerates but its fibrous sheath remains; this sheath is the stylohyoid ligament. The ceratohyal or hypohyal cartilage calcifies and becomes the lesser cornu of the hyoid bone¹.

Synonyms of Eagle's Syndrome

Elongated styloid process syndrome, Mineralized Elongated Styloid Process (MESP), Styloid process and carotid artery syndrome, Stylohyoid syndrome, Styloid process neuralgia, Styalgia^{1,2}.

History and Epidemiology

Literature reveals that in 1652, Pietro Marchetti identified an elongated styloid process formed due to ossification of a portion of the stylohyoid ligament. However, in 1937 it was otorhinolaryngologist Watt Weems Eagle who first defined Eagle's syndrome as an arbitrary entity related to an elongated styloid process formed due to mineralization of the stylohyoid ligament complex. He divided it into two subtypes: the "classic syndrome" and the "stylo-carotid artery syndrome"³.

Types

Eagle primarily described two subtypes of the syndrome:

1. Classical styloid syndrome: This subtype occurs due to fibrosis around the tip of the elongated styloid process following tonsillectomy, causing distortion of the cranial nerve endings in the tonsillar bed and presents with sensation of foreign body in throat, pain on deglutition and pain in the throat². It is characterized by pharyngodynia localized in the tonsillar fossa and is sometimes accompanied by dysphagia, odynophagia, hyper salivation and rarely by temporary voice changes⁴.
2. The stylo-carotid syndrome: This subtypes is not correlated with tonsillectomy and is due to the compression of the internal and/or the external carotid arteries, especially their perivascular sympathetic chain in the carotid sheath by the ossified ligament or tip of the styloid process, resulting in an incessant pain irradiating in the carotid zone^{2,4}.

Clinical features and Examination

Diagnosis requires a thorough clinical examination of the head and neck and should be confirmed radiographically. Patients with Eagle's Syndrome may present with a history of pain in the tonsillar region, dysphagia, pain on turning the head, foreign body sensation in the throat, referred otalgia and persistent pain in the carotid territory. Duration of patients' symptoms generally range from 8 months to 37 months (average: 14 months)³.

Examination of the styloid region can be performed by seating the patient in a chair with firm back support. Palpation of the styloid region of the neck is done bimanually and digitally standing behind the patient. Intraoral examination method includes visualizing the tonsillar region with a tongue blade or mouth mirror¹. This is followed by digital palpation of the tonsillar fossa, which may reveal a bony formation. Palpation may exacerbate pain aggravating symptoms with local tenderness⁴. The diagnosis can be confirmed by infiltration of local anesthetics into the tonsillar bed, which alleviates the pain³. This acts as a chair-side diagnostic test for Eagle's syndrome.

Radiographic Features and classification

Radiographic evaluation helps to confirm diagnosis and subsequently precedes the treatment required. Different types of imaging have been used for the diagnosis of Eagle's syndrome. These include panoramic radiographs, lateral head and neck radiographs, Towne's view radiographs, lateral oblique radiograph of mandible, antero-posterior head radiograph and computed tomography (CT)³. Recent advancements like cone – beam computed tomography (CBCT) imaging technique are preferred for a three dimensional visualization of the elongated styloid process. Langlais et al proposed a radiographic classification of the mineralized elongated styloid process. This included three types of elongation and four patterns of calcification or mineralization as shown in Table 1 and 2¹.

Types of elongation	Characteristics
Elongated	Uninterrupted elongation (>25- 28 mm)
Pseudo articulated / Pseudo segmented	Less frequent type; the styloid process is joined to the mineralized stylohyoid ligament by a single pseudo-articulation
Segmented	Consists of short /long non-continuous portions of the styloid process or interrupted segments of mineralized ligament

Table 1 - Types of elongation

Patterns of calcification	Characteristics
Calcified outline	Similar to the radiographic appearance of a long bone with a thin radiopaque cortex and a central lucency constituting most of the process
Partially calcified	Thicker radiopaque outline, with almost complete opacification as well as a small, discontinuous radiolucent core
Nodular complex	It has a scalloped outline and may be partially or completely calcified with varying degrees of central lucency – like nodules
Completely Calcified	This pattern is totally radiopaque with no radiolucent inner core

Table 2 - Patterns of Calcification

Management

Surgical shortening is the universally accepted treatment for an elongated styloid process. Multi-disciplinary management yields optimal results; some studies have recommended an intraoral approach (i.e., trans-tonsillar fossa excision)¹. In this, an incision is made anterior to the tonsillar fossa and the tip of the styloid process is exposed via blunt dissection. This dissection is carried out as proximally as possible to the styloid process, ultimately leading to the removal of the elongated part. Complete exposure and thus complete excision of the styloid process are often not possible with this approach, although the excised component is almost always sufficient to relieve symptoms⁵.

It has been accepted that intraoral approach or trans-tonsillar fossa excision is advised only if the distal tip of the styloid process can be palpated in the tonsillar fossa. Some advantages of intraoral approach are that it is a simple procedure with less operative time, no external scar formation and does not result in greater morbidity than a routine tonsillectomy. Disadvantages are inadequate surgical shortening of the styloid process due to poor visualization of the surgical field, possible injury of nearby neurovascular structures, and risk of deep neck space infection^{1,3}.

Styloidectomy can also be performed by the extra-oral approach. This is done by making an incision that extends from the mastoid process along the

sternocleidomastoid to the level of the hyoid, then up across the neck to the midline of the chin. This approach is aesthetically less pleasing with more morbidity compared to the intra-oral route⁶. However, the extra oral cervical approach was preferred because surgical visualization was optimal and the risk of deep cervical infection was minimal.

The external approach is favoured by N. Asherson (1957) and Moffat et al. (1977) while the intraoral approach via tonsillar fossa is advocated by Eagle (1949)³. Dentists must weigh the advantages and disadvantages of each technique and select the appropriate procedure for the patient.

Differential Diagnosis

Patients with an underlying Eagle's syndrome may be examined by a surgeon, a dentist, a neurologist, and a psychiatrist, often receiving various modalities of treatments that do not relieve the symptoms and cloud the clinical picture. This is because of its vagueness of symptoms and the infrequent clinical observation which are often misleading.

Stylalgia is misdiagnosed or often overlooked as a possible diagnosis in cases of vague cervicofacial pain. A variety of head and neck conditions should be considered in the differential diagnosis of Eagle's Syndrome and cervicopharyngeal pain⁷. These include temporomandibular disorders, glossopharyngeal neuralgia, trigeminal neuralgia, migraine-type headaches, sphenopalatine neuralgia, cervical arthritis, temporal arteritis, possibilities of projected pain caused by inflammation of the eustachium and middle ear (otitis), parotid diseases, carotidynia, impeded eruption of the upper and lower impacted or retained wisdom teeth, and possible tumours. Other pathology should be eliminated by a careful medical history, clinical and radiographic examination^{2,7}.

Several syndromes should also be considered which are marked by the symptom of pain in the same region and whose symptoms are occasionally very similar to those of the styloid process syndrome. They include: a. Costen's syndrome; b. Trotter's syndrome; c. Miofacial painful syndrome².

Thus, awareness of the differential diagnosis of Eagle's syndrome helps in handling the cases in a more professional manner in order to diagnose the condition accurately.

Conclusion

Oro-facial pain is a vague condition with a number of differential diagnosis. Thorough and adequate knowledge on the subject is of utmost significance in diagnosis of orofacial pain. Eagle's syndrome is often missed by the general dental practitioners causing diagnostic dilemma and unnecessary expenses for the patient by undergoing a wide array of diagnostic radiographs. Hence, a deep insight on this particular topic is essential to consider Eagle's syndrome when pain is experienced in the submandibular region, base of the tongue or neck.

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