Eagle syndrome. A narrative review.

Abstract: Painful disorders in the maxillofacial region are common in dental practice. Most of these conditions are not properly diagnosed because of inadequate knowledge of craniofacial and cervico-pharyngeal syndromes such as Eagle Syndrome. The aim of this review is to describe the general aspects, diagnosis and treatment of Eagle syndrome. Eagle syndrome or stylohyoid syndrome was first described by Watt W. Eagle in 1937. It was defined as orofacial pain related to the elongation of the styloid process and ligament stylohyoid calcification. The condition is accompanied by symptoms such as dysphonia, dysphagia, sore throat, glossitis, earache, tonsillitis, facial pain, headache, pain in the temporomandibular joint and inability to perform lateral movements of the neck. Diagnosis and treatment of Eagle syndrome based on symptoms and radiographic examination of the patient will determine the need for surgical or nonsurgical treatment. Eagle syndrome is a complex disorder demanding a thorough knowledge of its signs and symptoms to make a correct diagnosis and provide an appropriate subsequent treatment. Disseminating information about this syndrome among medical-dental professionals is essential to provide adequate dental care to patients.

Keywords: Eagle syndrome, Styloid process, Estylohyoid ligament, Facial pain, Review.

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INTRODUCTION.

Eagle Syndrome (ES) or Stylohyoid syndrome is an unusual pathology of the head and neck, which produces orofacial pain. Pain is caused by the elongation of the styloid process and by the partial or total calcification of the stylohyoid ligament. The group consisting of the styloid process and the stylohyoid ligament is called “stylohyoid complex”, which has its embryonic origin in the cartilage of the second pharyngeal arch or hyoid arch (Reichert’s cartilage). ES is part of musculoskeletal disorders related to the temporomandibular joint; masticatory and cervical muscles as well as associated structures. Diagnosis of ES is made by clinical and radiographic methods. However, ES is frequently underdiagnosed due to its low prevalence and inadequate knowledge of craniofacial and cervico-pharyngeal syndromes. Moreover, as its symptoms are varied, health professionals and patients tend to confuse this disease with other disorders. The aim of this review is to describe the general aspects, diagnosis and treatment of Eagle syndrome.

DEFINITION AND HISTORICAL DATA

Eagle syndrome is also known as styloid process, styloid syndrome, Stylohyoid syndrome, Stylohyoid Complex Syndrome, Carotid Artery Syndrome, Carotid style Syndrome, Elongated and Ossified Stylohyoid Process Syndrome or Stylohyoid Neuralgia. It was first des-
The styloid process is usually located between the internal and external carotid arteries and allows the insertion of the stylopharyngeus, stylohyoid and styloglossus muscles and also of the stylomandibular ligaments. The stylohyoid ligament originates from the tip of the process and is inserted into the horn or lesser horn of the hyoid bone and the second originates from the medial part of the process and is directed forward and down to the inner surface of the mandibular gonial angle.

**CLASSIFICATION**

Eagle suggests that the normal length of the styloid process is approximately 25mm, therefore any length exceeding that size would be considered elongated. However, other authors, in current studies, indicate that the length of the styloid is variable. They agree that a styloid process exceeding 30mm should be considered elongated.

According to the symptoms, Eagle describes two types of syndrome: the classical or typical syndrome associated with carotid artery and the atypical one. The first type occurs in patients with previous tonsillectomy and is related to the extension of nerve endings, mainly cranial nerve IX (glossopharyngeal), causing constant or dull pain in the pharyngeal region that often radiates to the ear resulting in dysphagia, foreign body sensation in the throat, increased salivation, nausea, facial and neck pain (unilateral), trismus and limited mandibular lateral excursion. Eagle attributed the pain to the scarring process around the styloid process after a tonsillectomy.

The second type would be present in patients with or without tonsillectomy, and occurs when the styloid process contacts the external carotid artery on the affected side, resulting in carotodynia (pain caused by compression of the carotid and referred to the neck and to the area around the eyes), intermittent headache in the frontal, temporal or parietal regions, earache and dizziness, and tenderness on palpation in the area of the carotid artery. Pain would be associated with the elongation of the styloid process.
associated with pressure on sympathetic fibers of perivascular carotid artery,\textsuperscript{13,16,27,38} as well as elongation and calcification of stylohyoid complex,\textsuperscript{1,25} causing a transient ischemic event,\textsuperscript{2,22,31} a stroke,\textsuperscript{2,25,36} and even death.\textsuperscript{12,14,19,21}

Colby and Del Gaudio,\textsuperscript{16} developed a new classification based on cone beam computed tomography (CBT), dividing it into: elongated styloid process, ossified stylohyoid ligaments and elongated hyoid bone. Langlais \textit{et al.}\textsuperscript{1,36,40,41} made a classification for elongation and calcification patterns of the stylohyoid complex by radiographic appearance. Three radiographic patterns are known:

1. Type I or elongated\textsuperscript{1,23,24}: This calcified stylohyoid complex is characterized by a continuous styloid process.\textsuperscript{36,41} If the study is conducted by observing panoramic radiographs, a length of 28mm is accepted, taking into account the normal magnification affecting this type of radiographs.\textsuperscript{4,40}

2. Type II or pseudoarticulated\textsuperscript{1,23,24}: In this radiographic image, the styloid process is attached to the stylomandibular ligament or to the stylohyoid ligament by a pseudoarticulation,\textsuperscript{36} which is located on the lower edge of the mandible (tangentially). This gives the appearance of an articulated and elongated complex.\textsuperscript{4,40,41}

3. Type III or segmented\textsuperscript{1,23,24}: It consists of short or long portions of the styloid process that are discontinuous or interrupted segments of the mineralized ligament.\textsuperscript{36,40} Two or more segments are observed, with interruptions either above or below the lower border of the mandible, or both.\textsuperscript{4,41}

Also, according to the pattern of calcification, four patterns can be determined:

1. Calcified contour\textsuperscript{1,23,24,41}: A radiopaque and thin edge with a central radiolucency comprising most of the process is observed,\textsuperscript{4,40} giving the radiographic appearance of a long bone.\textsuperscript{34,40}

2. Partially calcified\textsuperscript{23,24,41}: It indicates that the process has a radiopaque and almost completely opaque contour, however, it may have discontinuous radiolucent centers.\textsuperscript{1,4,34,40}

3. Nodular complex\textsuperscript{23,24,41}: It is characterized by an undulating or scalloping contour, partially or completely calcified with varying degrees of central radiolucency.\textsuperscript{1,4,34,40}

4. Completely calcified\textsuperscript{14,23,40}: It is totally radiopaque.\textsuperscript{24,34,41}

**EPIDEMIOLOGY**

Lirios\textsuperscript{3} states that the prevalence of an elongated styloid process has great variability in the population. Eagle, in its original publication, found the elongated styloid process in 4\% of his cases.\textsuperscript{2,4,13,16,29,30,38,42}

It is estimated that between 3 and 30\% of the world population has elongated styloid process, and of them, from 4 to 10.3\% suffer from painful symptoms.\textsuperscript{1,14,36} The wide variation in the prevalence of elongated styloid process evidenced in different studies can be explained by variations in diagnostic criteria and interpretation of images, geographic location and characteristics of the local population.\textsuperscript{37}

When comparing both processes, findings suggest that bilateral elongations are more frequent,\textsuperscript{15,25,29,33,41} larger on the right side.\textsuperscript{1,38} Studies indicate that the highest prevalence of the disease is found in women (3:1)\textsuperscript{4,10,18,28} and in the age range of 30 to 50 years.\textsuperscript{12,13}

**SYMPTOMATOLOGY**

A wide variety of symptoms have been associated with the elongation of the styloid process, including an intense facial pain,\textsuperscript{3,5} nonspecific cervical pain or during rotation of the head, headaches, sore throat, ear pain, dysphagia or odynophagia,\textsuperscript{6,7,19} pain by extending the tongue, burning sensation on the tongue, pain when opening the mouth, discomfort during chewing, difficulty to speak, voice change,\textsuperscript{8,9,32} sensation of hypersalivation, foreign body sensation in the throat, tinnitus, trismus, syncope, submandibular swelling and brain vascular symptoms induced by the change of position.\textsuperscript{13,14,36} Pain is the main characteristic of this condition. It is constant and distressing, and can last from minutes to days.\textsuperscript{25,26,31} Also, most patients experience this discomfort unilaterally.\textsuperscript{18,28,41} However, most patients with ES are asymptomatic.\textsuperscript{36,43}

Absence of pain in patients with an elongated styloid process can be explained because there may be regions of
incomplete mineralization, which reduce the compression in the surrounding tissues.\textsuperscript{22-24}

**ETIOPATHOGENESIS**

Pathogenesis is caused by the compression of the glossopharyngeal nerve and associated vascular structures, because of the enlarged process.\textsuperscript{5,9,26,32} The etiology is still unknown,\textsuperscript{21,24,29} however, there are different theories that attempt to explain it.\textsuperscript{17,18,30}

Steinmann\textsuperscript{2,4,13,33} proposed three theories to explain the process of ossification:

1. Theory of reactive hyperplasia, which suggests that surgery or chronic irritation of the throat can cause tendinitis, ossifying periostitis or osteitis in the stylohyoid ligament.\textsuperscript{17}

2. Theory of reactive metaplasia (Heteromorphosis) associated with a partial ossification of fibrocartilaginous tissue of the stylohyoid ligament, usually in response to a previous trauma.\textsuperscript{17}

3. Theory of anatomical variation: it proposes that early elongation of the styloid process and ligament ossification are anatomical variations which occur without the presence of previous trauma; this would explain the presence of ossification in children.\textsuperscript{30}

In addition, there is a theory involving congenital elongation,\textsuperscript{21} which is the most accepted model, and explains that ES is the result of the persistence of embryonic mesenchymal sheet capable of producing bone tissue in adults;\textsuperscript{21,29} and ossification of stylohyoid ligament related to mechanical stress during fetal development.\textsuperscript{9,13}

In rare cases they are associated with endocrine disorders (menopause).\textsuperscript{17,30} One study reports the possibility that ES could be the result of the expression of an autosomal dominant gene.\textsuperscript{13,43}

Furthermore, it has been explained that the cause of pain symptoms is the relationship of the stylohyoid complex with anatomical structures,\textsuperscript{1,5,20} such as muscles, nerves, arteries, veins, fasciae and increased length of styloid process, as it can lead to irritation and inflammation of those structures.\textsuperscript{21,29,42} There are some mechanisms that may explain the cause of pain produced by ES, such as:

1. Secondary mechanical irritation of the pharyngeal mucosa.\textsuperscript{13,14,36}

2. Extension of nerve endings of cranial nerves V, VII, IX and X, resulting from the fibrosis that occurs after tonsillectomy.\textsuperscript{13,14,36}

3. Fracture of the calcified stylohyoid ligament,\textsuperscript{15,31,33} caused by cervical trauma or a sudden movement of the neck, with the consequent proliferation of granulation tissue that can cause pressure on the structures surrounding the stylohyoid complex.\textsuperscript{16,37,41}

4. Pressure on the carotid artery, with stimulation of the sympathetic chain involving the carotid sheath.\textsuperscript{1}

5. Degenerative changes in the insertion of the stylohyoid muscle or insertion tendinitis.\textsuperscript{17,18,30}

**DIAGNOSIS**

Diagnosis of ES is based on four parameters: clinical manifestations, digital palpation of the styloid process, lidocaine infiltration test, and radiological findings.\textsuperscript{1,25} Therefore a correct anamnesis\textsuperscript{30,39,41} is required, as well as the use of clinical\textsuperscript{13,16,33} and radiographic\textsuperscript{3,5,24,42} methods.

**Clinical examination**

Clinical examination is performed by means of palpation of the styloid process in the tonsillar fossa.\textsuperscript{3,20,24,34} It is perceived as a bone spicules that causes pain,\textsuperscript{13,35,42} which is relieved by infiltrating lidocaine.\textsuperscript{2,16,25,30}

**Radiographic examination**

Various imaging techniques (panoramic,\textsuperscript{20,30,33,34} oblique lateral of skull and neck,\textsuperscript{4,9,24} anteroposterior skull,\textsuperscript{10,13,39} Towne,\textsuperscript{18,25,29} neck angiography,\textsuperscript{8,21} or magnetic resonance\textsuperscript{16,37}) allow to demonstrate the presence of elongation\textsuperscript{12,23} and calcification of stylohyoid process.\textsuperscript{35,41} However, computed axial tomography (CT)\textsuperscript{3,7,12,36} and, particularly, computed tomography cone beam is considered the radiological examination of choice\textsuperscript{32,37} because it prevents image overlaps and provides low levels of distortion, larger scale contrast, accurate measurement of the length of the styloid process as well as its angulation and anatomical relationship.\textsuperscript{2}
DIFFERENTIAL DIAGNOSIS

It is important to establish the differential diagnosis of ES when the patient has pain in the cervicopharyngeal or cervicofacial region, considering the following conditions: sphenopalatine neuralgia, trigeminal neuralgia, occipital neuralgia, glossopharyngeal neuralgia, arthrosis and temporal tendinitis, dysphagia, migraine, atypical facial pain, sialadenitis and sialolithiasis, myofascial pain syndrome, pain related to impacted third molars, cervical arthropathy, degenerative disc disease, diverticulosis of the esophagus, defective or excessively tight dentures, chronic laryngopharyngeal reflux, tumors, otitis, mastoiditis, chronic tonsillitis, otitis, pharyngitis, benign and malignant tumors in the oropharyngeal area, internal carotid artery syndrome, carotidynia, fracture of the hyoid bone, Ernest syndrome and temporomandibular dysfunction.

TREATMENT

Treatment of ES is surgical or nonsurgical and depends on the intensity and severity of symptoms.

Nonsurgical Treatment

In cases where the symptoms are of medium intensity, treatment is not recommended, except reassuring the patient. Some authors suggest the use of: antidepressants (amitriptilina), anticonvulsants (valproic acid, gabapentin and carbamazepine), analgesics and nonsteroidal anti-inflammatory drugs, corticosteroids infiltration (hydrocortisone), infiltration of steroids and non infiltration of local anesthetics (lidocaine, mepivacaine).

Surgical treatment

In severe cases surgical resection of the styloid process is the most effective and satisfactory treatment, either extraoral or transcervical, or intraoral or transpharyngeal.

Extraoral or transcervical approach

Loeser and Caldwell described this technique: A proximal incision is made near the sternocleidomastoid muscle to the hyoid bone; parotid fascia is retracted upwards and the carotid sheath along with the sternocleidomastoid muscle are placed on the back. Muscle insertions of the stylohyoid process are dissected and to then remove the portion of the elongated styloid process.

Advantages of extraoral procedures include: better view of the work area, exhibition and preservation of vascular and nerve structures, broader styloid process resection, minimum generation of edema in the airway, and reduced risk of infection. The disadvantages are: longer surgical time and paresthesia of the cutaneous nerves; the resulting external scar of this surgical approach: is cervical, high, small and aesthetically acceptable.

Hoffmann and Spalthoff describe the use of a piezoelectric device as a safe and effective way to surgically treat this disease.

Intraoral or transpharyngeal approach

Eagle described this technique. It consists of a longitudinal incision with blunt dissection performed to locate and remove the styloid process. If the tonsils are present, a tonsillectomy is performed in the same surgical event. Despite being an easier technique, leaving no visible external scar, it does not allow adequate visualization of the structures adjacent to the styloid process, thrombosis of the internal carotid artery, subcutaneous emphysema and an increased risk of contaminating cervical spaces.

Some authors recommend an intraoral or extraoral endoscopically-assisted approach to solve the disadvantages of this procedure because it decreases the amount of bleeding, duration of surgery and hospitalization time, and improves the subjective symptoms of patients, being this an effective and minimally invasive solution to treat Eagle syndrome.

In general, it is accepted that the intraoral approach should be used only if the distal tip of the styloid process is palpable in the tonsillar fossa, and only if the surgeon is familiar with the technique and management of complications. The success rate of surgical treatment is 80% to 95.6%. For the nonsurgical treatment there is no real positive long-term effect and after 6 to 12 months, the symptoms reappear.
CONCLUSION

Eagle syndrome is a complex disorder demanding a thorough knowledge of its signs and symptoms to make a correct diagnosis and provide an appropriate subsequent treatment.

Spread information about this syndrome among health professionals is essential to provide adequate dental care to each patient.

Síndrome de Eagle. Una revisión narrativa.

Resumen: En la práctica odontológica, es frecuente encontrar alteraciones con sintomatología dolorosa en la región maxilofacial, las cuales no son apropiadamente diagnosticadas, a causa del desconocimiento de síndromes craneofaciales y cervicofaríngeos, como el Síndrome de Eagle. El objetivo de esta revisión es describir los aspectos generales, diagnóstico y tratamiento del Síndrome de Eagle. El Síndrome de Eagle o estilalgia es la entidad nosológica, descrita por Watt W. Eagle en 1937, definida como aquel dolor orofacial relacionado con la elongación de la apófisis estiloides y calcificación del ligamento estilohiodio; el cual está acompañado de síntomas como: disfonía, disfagia, dolor faríngeo, glositis, otalgia, tonsilitis, dolor facial, cefalea, odinofagia, dolor en la articulación temporomandibular e imposibilidad de realizar movimientos laterales del cuello. El diagnóstico y tratamiento del Síndrome de Eagle está basado en la sintomatología y el examen radiográfico del paciente, lo cual determinará el tratamiento quirúrgico o no quirúrgico. El Síndrome de Eagle es una patología compleja que requiere un conocimiento amplio de sus signos y síntomas, para establecer un correcto diagnóstico y posteriormente un adecuado tratamiento. Para ello, es necesario difundir la información sobre este síndrome entre los profesionales médico-odontológico y así brindar una atención adecuada a cada uno de los pacientes.

Palabras clave: Síndrome de Eagle, Apófisis estiloides, Ligamento estilohiodio, dolor facial, Revisión.

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