

Eagle's syndrome and its various manifestations

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Abstract. Objectives: We present an updated review of the literature regarding Eagle/stylohyoid syndrome and discuss the variety of presentations, and current/emerging treatment modalities. Methods: A non-systematic qualitative review of articles pertaining to Eagle syndrome in the Google Scholar, PubMed, and Web of Science databases. The following search terms were used: Eagle syndrome, styloid or stylohyoid syndrome, styloarotid syndrome, and elongated styloid syndrome. After excluding the duplicates, we further excluded the irrelevant articles for the purpose of this revision. In the qualitative analysis, we excluded the publications already mentioned in the analyzed papers, and conference abstracts or clinical photography. Results: The literature search resulted in 606 publications (n = 507 after removal of duplicates). Ninety-seven publications were analyzed as full text, and their quality was assessed according to the SIGN recommendations. Seventeen of these publications met all eligibility criteria and were included in the qualitative review. These articles are mainly case reports, a brief clinical study, two case series, two commentaries on surgical management of patients with Eagle syndrome, and two literature reviews. Conclusion: Eagle's syndrome has a wide variety of presentations expressed by multiple nonspecific symptoms. Even that Eagle's syndrome has become recognized more readily by understanding diagnostic workup and imaging, many questions remain regarding the treatment options considering that the etiopathogenetic mechanism has not been clarified and the diagnosis is made by exclusion of other diseases.

Key Words: endoscopic sinus surgery (ESS); chronic rhinosinusitis; tissue injury; murine experimental model; osteitis- neo-osteogenesis

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Introduction

Patients often seek consultations for a sore throat or foreign body sensations in the pharynx that prompts frequent swallowing. These complaints are related to several disorders that may exhibit a variety of signs and symptoms. However, all are characterized by a sensation of pain of variable intensity in the head and neck region with a chronic and nonresponsive pattern, leading to errors in diagnosis and treatment (Constantinides et al 2013). One of these is Eagle's syndrome, a condition associated with ossification or abnormality of the stylohyoid chain, which may cause various cervico-pharyngeal symptoms such as dysphagia, odynophagia, headache, and chronic neck pain that may radiate to the ear (Badhey et al 2017). Usually, Eagle's syndrome is associated with trauma, but the symptoms rarely appear spontaneously without a specific cause. Spontaneous fracture of the styloid process produces unexplained symptoms for which appropriate treatment cannot be performed due to indefinite complaints that are not thoroughly examined (Lee & Chung 2020). Styloarotid syndrome, a less common variant syndrome, refers to external compression of the internal carotid artery by an elongated styloid process (Dewan et al 2016). In this paper, we present an updated review of the literature regarding Eagle/stylohyoid syndrome and discuss the variety of presentations, and current/emerging treatment modalities.

Methods

We aimed to identify the review articles, systematic reviews, meta-analysis articles and case reports pertaining to Eagle syndrome in the Google Scholar, PubMed, and Web of Science databases. The following search terms were used: Eagle syndrome, styloid or stylohyoid syndrome, styloarotid syndrome, and elongated styloid syndrome. Eligibility was discussed by authors by screening the title and abstracts of the retrieved articles. Whenever in doubt about the inclusion or exclusion of an abstract, the full text was accessed. After excluding the duplicates, we further excluded the irrelevant articles for the purpose of this revision (an obvious traumatic cause that does not raise diagnostic issues Figure 1). In the qualitative analysis, we excluded the publications already mentioned in the analyzes published between 1999 – 2020, and conference abstracts or clinical photography.

The review protocol was performed in accordance with the guideline Preferred Reporting Items for Systematic Reviews and Meta-Analysis (PRISMA) statement (Moher et al 2015). The criteria of a meta-analysis not being met because most articles were a case presentation, we decided to conduct a review of the literature.

Results

The literature search resulted in 606 publications (n = 507 after removal of duplicates). The study selection process is illustrated

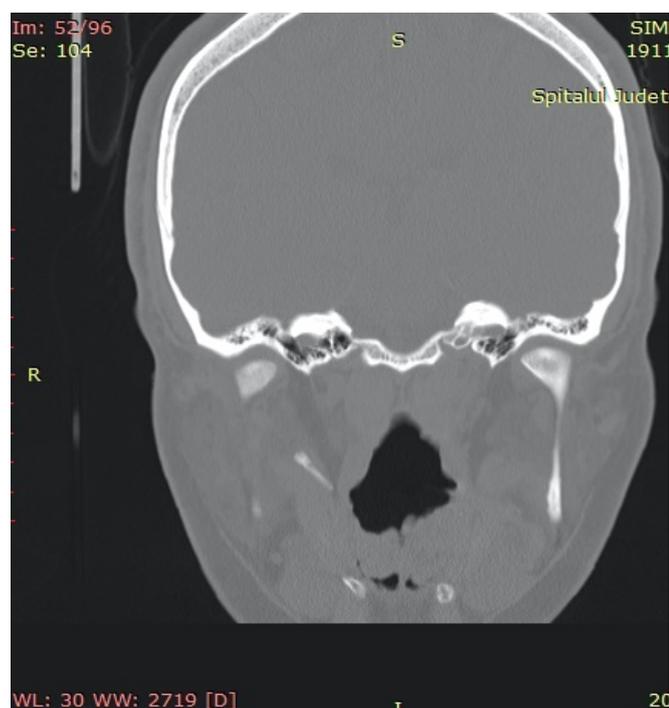


Fig. 1. Fracture of the right styloid process - the dislocated bone fragment “pushes” the pharyngeal lateral wall

in Figure 2. Ninety-seven publications were analyzed as full text, and their quality was assessed according to the SIGN recommendations. Seventeen of these publications met all eligibility criteria and were included in the qualitative review.

These articles are mainly case reports (Dewan et al 2016; Hirano et al 2020; Farhat et al 2009; Cano et al 2010; Krohn et al 2018; Galletta et al 2019; Rissardo & Caprara 2019; Slavin 2002), a brief clinical study (Bal et al 2018), six case series (Waters et al 2018; Fitzpatrick et al 2020; Singhania et al 2013; Burulday et al 2017; Kent et al 2015; Jalisi et al 2017), two commentaries on surgical management of patients with Eagle syndrome (Lou 2019; Xu 2021), and two literature reviews (Constantinides et al 2013; Badhey et al 2017).

The most commonly found syndromic form is the classic type characterized by symptoms such as headache, dysphagia, cervical pain, pharyngeal pain, chronic cough, and foreign body sensation. An elongated styloid process occurs in about 4% of the general population, while only a small percentage (between 4-10.3%) of these patients are symptomatic (Constantinides et al 2013). In most of the cases, the diagnosis of Eagle syndrome was suspected on anamnesis and objective examination. The objective exam was normal or revealed the presence of a mass or a bone protrusion due to an elongated styloid process or the reproduction of the patient's symptoms by palpation of the tonsillar fossa. Subsidence of the patient's symptoms after lidocaine infiltration of the anterior pillar and tonsillar fossa may also support the diagnosis (Singhania et al 2013). In all reviewed articles, Eagle syndrome was confirmed by a head and neck 3D computed tomography (Hirano et al 2020; Krohn et al 2018; Rissardo et al Caprara 2019; Slavin 2002). In case of the vascular form of the syndrome, the authors used angiography (Dewan et al 2016) or angiocomputed tomography (Farhat et al 2009; Cano et al 2010; Galletta et al 2019, Bal et al 2018). The treatment applied to patients with Eagle syndrome was differentiated according to the intensity of the symptoms, the

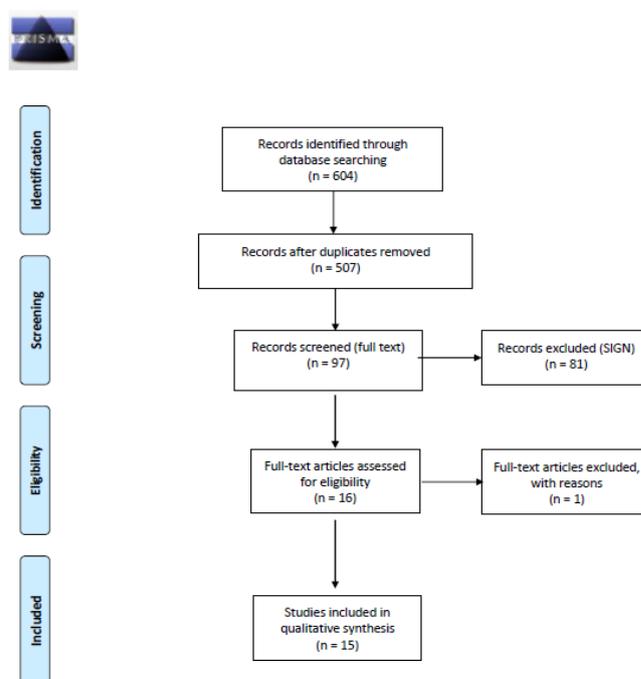


Fig. 2. PRISMA 2009 Flow Diagram with study selection process

identified etiology, and the severity of the quality-of-life impairment. The medical therapy could be divided into first-line analgesics or NSAIDs and alternative management consisting of a combination of anticonvulsants/antidepressants (carbamazepine, amitriptyline) (Constantinides et al 2013 ; Badhey et al 2017). All articles with vascular forms of Eagle syndrome described surgical treatment, either by transcervical or transoral approach (Dewan et al 2016; Farhat et al 2009; Cano et al 2010; Galletta et al 2019, Bal et al 2018). Waters and al (2018) performed transcervical styloidectomy for 41 patients identified with Eagle's syndrome. In their 17-year experience with open or transoral robotic surgery (TORS), Fitzpatrick and his co-workers performed 21 styloidectomies, 6 were performed via TORS and 15 were done transcervically (Fitzpatrick et al 2020).

Discussions

In 1937, when dr. Eagle defined the relationship between the length of the styloid process and various symptoms, he was referring to the elongation of the styloid process. Then, in 1948 and 1949 (Constantinides et al 2013; Badhey et al 2017), he differentiated two clinical conditions: the classic type that usually appears after a tonsillectomy due to scar tissue formation under the tonsillar fossa, which compresses and stretches the nervous structures around the styloid process, and the carotid artery type that is the result of the impingement of the artery by the elongated process or an ossified stylohyoid ligament. If the external carotid artery is involved, the symptoms are increased by the head's rotation towards the affected part with the appearance of odynophagia with ipsilateral reflex otalgia. If the internal carotid artery is involved, the irritation of the periarterial sympathetic nerve plexus produces pain from the occipital to the ophthalmic region (carotidynia) or even transient ischemic attacks when the head is turned to the symptomatic side (Farhat et al 2009) or even carotid dissection (Cano et al 2010).

When the clinician is faced with a patient complaining of chronic orofacial pain, the diagnostic may be challenging. The differential diagnosis of temporomandibular disorder or unerupted molar, middle ear disease, Sluder's syndrome, essential neuralgia, cluster headache, migraine, cervical vertebral disorder or fibromyalgia syndrome, chronic tonsillitis or gastro-esophageal reflux, or even a neoplasm should be excluded (Constantinides et al 2013; Krohn et al 2019). Traumatic fractures of the styloid process can appear after motor vehicle accidents or as dental treatment and tonsillectomy complications (Lee & Chung 2020). Spontaneous fractures may occur because of sudden laughter, swallowing, yawning, or coughing, the patient having no previous complaints related to Eagle's syndrome (Badhey et al 2017; Lee & Chung 2020). The normal length of the styloid process ranges from 1.52 to 4.77 cm (Constantinides et al 2013). However, if the ossified portion appearing on radiographs is longer than 30 mm, this could be considered an elongated process. Diagnosis is made both by radiological and physical examination. Palpation of the styloid process in the tonsillar fossa is indicative of an elongated styloid process, whereas processes with normal length are generally not palpable (Lee & Chung 2020).

The accurate history and clinical examination are mandatory to establish the diagnosis, and plain radiographs reveal reliable documents. Three-dimensional imaging techniques, such as 3D-CT or cone-beam-computed tomography (CBCT), are often necessary (Rissardo et al 2019; Slavin 2002; Bal et al 2018; Waters et al 2018; Fitzpatrick et al 2020; Singhania et al 2013; Burulday et al 2017; Kent et al 2015; Jalisi et al 2017). CT Angiography can provide further information regarding carotid flow, especially if stroke or dissection is suspected. Other imaging modalities have been explored, such as transoral ultrasound and bone scintigraphy, but their efficacy remains unknown at this time (Badhey et al 2017).

Management of Eagle's syndrome starts with standard medical treatment. In addition to regular analgesic, anticonvulsant, and antidepressant medications, local infiltration of the region of the styloid process with a mixture of steroids and local anesthetic agents may result in lasting pain relief (Slavin 2002). Patients with vascular type of the syndrome or those with non-vascular form which do not respond to medical treatment, require surgery. It is possible to fracture the elongated styloid process manually by transpharyngeal manipulation, however it is not recommended since it has been associated with carotid artery injury. Surgical resection of the distal styloid process yields better long-term results (Waters et al 2018; Fitzpatrick et al 2020) This resection may be performed via the transpharyngeal approach as originally described by Eagle or by using the external route. In the two reviewed case series (Waters et al 2018; Fitzpatrick et al 2020), the authors presented the benefits of styloidectomy, but their studies have several limitations (Fitzpatrick et al 2020; Xu 2021): most surgeries performed by Waters et al (2018) had as their first indication the symptomatology suggestive for Eagle's syndrome with evidence of an elongated styloid, but 31.3% of patients reported a history of tonsillectomy, a possible cause for globus sensation. The authors did not record the duration of symptoms prior to surgery and the follow-up period was less than one year. From the experience of other authors (Lou 2019), the long-term results of styloidectomy are far from satisfactory. Another limitation of Waters' study is that they only described

the length of the styloid, and the angulation of the process is more important for the symptoms than the length (Burulday et al 2017; Kent et al 2015). The direction and curvature of the styloid process are considered more important than length in contributing to symptoms. Furthermore, the inferior end of this segment can present with varying degrees of angulation, which may be significant enough to allow the tip of the angulated part to reach the oropharyngeal wall, which can lead to the symptoms of dysphagia and globus sensation associated with Eagle's syndrome (Jalisi et al 2017). Fitzpatrick et al (2020) compared the surgical outcomes of the use of TORS and transcervical approach in the treatment of patients with Eagle's syndrome. As the Waters' study limitation, Fitzpatrick's study has the same problem: the surgical indication was based only on the size of the styloid without considering its angulation. The long-term effect was not satisfying for some patients. In their study, the authors did not strictly define the surgical indication; 90% reported some degree of lasting improvement in symptoms while 55% reported significant improvement.

When the underlying causes of Eagle's syndrome are clear, the surgical treatment is indicated for individuals with a significant dislocation and angulation of an elongated styloid process or in the carotid artery type. The choice of treatment should be discussed with the patient and depends on the severity of the symptoms (Constantinides et al 2013).

Conclusions

Eagle's syndrome has a wide variety of presentations expressed by multiple nonspecific symptoms. Even that Eagle's syndrome has become recognized more readily by understanding diagnostic workup and imaging, many questions remain regarding the treatment options considering that the etiopathogenetic mechanism has not been clarified and the diagnosis is made by exclusion of other diseases.

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