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Eagle Syndrome: A Comprehensive Review of Pathophysiology, Diagnosis and Management Strategies

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Abstract:

Introduction: Eagle syndrome, also known as elongated styloid process syndrome, is a rare condition resulting from excessive elongation of the styloid process or calcification of the stylohyoid ligament. These abnormalities may compress adjacent neural and vascular structures, producing a wide range of symptoms such as throat, ear or facial pain, dysphagia, or the sensation of a foreign body in the throat. Due to the proximity of critical anatomic structures and the nonspecific clinical presentation, Eagle syndrome poses a significant diagnostic challenge and should be considered in the evaluation of craniofacial and cervical pain.

Purpose of the work: The aim of this study is to analyze and present the current state of knowledge regarding Eagle syndrome, its pathophysiology, symptoms, complications, the impact on patients' quality of life, diagnostic methods, and management strategies.

Materials and methods: An analysis of scientific articles available on PubMed and Google Scholar was conducted using the following keywords: Eagle syndrome; diagnosis of Eagle syndrome; Eagle diagnosis; styloidectomy; calcification of the styloid-hamate ligament; neuralgia.

Results: A review of the literature showed that elongated styloid process occurs with a frequency of 0.4% to 84.4%, but symptomatic course is found in approximately 4% of the population. The most commonly observed symptoms were sore throat, dysphagia, ear pain and neuropathic symptoms, and in isolated cases also vascular complications such as carotid artery dissection. Computed tomography with 3D reconstruction proved to be the most effective diagnostic method. Conservative treatment (NSAIDs, neuropathic pain medication, steroid injections) resulted in only partial and temporary improvement. Styloidectomy, performed using an intraoral or extraoral technique, proved to be the most effective. Despite appropriate surgical treatment, approximately 20% of patients still reported persistent symptoms.

Keywords: Eagle's syndrome, glossopharyngeal neuralgia, elongated styloid process, styloidectomy, 3D-CT/CBCT, surgical treatment

ANATOMICAL AND CLINICAL SIGNIFICANCE OF THE ELONGATED STYLOID PROCESS IN EAGLE SYNDROME

Eagle's syndrome, or more accurately elongated styloid process syndrome, is a rare condition in which the main cause of the patient's symptoms is an excessively long styloid process or calcification of the styloid-olecranon ligament, which is much more common on one side [1]. According to medical literature, the length of the stylohyoid ligament should be 20-30 mm, with values above this considered to be an elongated ligament [2,3,4]; some consider the process to be elongated if it is one third of the length of the mandibular ramus [5]. There may also be cases where the styloid process reaches the hyoid bone (C4 level). [6] Both the styloid process (1) and the stylohyoid ligament (2) are very important for clinicians dealing with head and neck disorders, such as radiologists and maxillofacial surgeons. It is very important to familiarise oneself thoroughly with the structural anatomy of the surrounding area (1 and 2) due to a number of conditions occurring in their vicinity, important structures, including the internal and external carotid arteries, facial nerve, vagus nerve, accessory nerve, glossopharyngeal nerve [7,8], and related ailments [9]. Ailments that may occur in the course of Eagle's syndrome include sore throat, difficulty swallowing, severe pain when pressing on the tonsillar fossa, dysphagia, ear pain, facial pain, painful neck movements or a feeling of

a foreign body in the throat [6], a range of neuropathic symptoms or disorders of the mandibular-gnathic complex [10,11]. There are also known cases of ischaemia and dissection of the carotid artery [12].

PATHOPHYSIOLOGY

The first description of elongation of the styloid process was made by Weinlecher in the 19th century, with its prevalence estimated at around 4% [13]. The latest data indicate a percentage ranging from 0.4 to 84.4% [14,15,16]. According to current sources, it is estimated to be between 2 and 11.8%, with pain symptoms occurring in approximately 4% of the population [17]. Eagle's syndrome was described as a separate disease entity in the 1940s by Watt Weems [18]. It is a rare syndrome that affects approximately 0.06% of the population [19], the majority of whom are women [20,21,22]. There are also studies that show a predominance in men [23,13] or no correlation between gender and incidence [17, 24]. Ossification of the stylohyoid ligament

is the metaplasia of fibrous connective tissue into bone or cartilage tissue. This process occurs in stages: degenerative changes in collagen (mainly type I), deposition of calcium salts (calcium phosphate) in the extracellular matrix (dystrophic calcification), recruitment of osteogenic cells from surrounding vessels and periosteum, metaplasia of fibroblasts and formation of osteoblasts and chondroblasts, formation of bone trabeculae, i.e. complete or partial ossification of the ligament. This process is heterotopic ossification, i.e. ossification in tissue that physiologically should not contain bone. Eagle divided the syndrome into the classic form, which is associated with tonsillectomy, and the so-called carotid artery syndrome [25], where mechanical compression of the elongated styloid process on the sympathetic fibres of the cervical plexus occurs [26,27]. According to the current literature, a division into three main classifications is also used [28]. Another division of the styloid process is determined during radiological examination and the position relative to the mandibular foramen [29,30]. Factors contributing to ossification can be divided into physiological (1) and pathological (2). The former include, among others age - after the age of 30, the calcification of connective tissues increases - procedures in younger people are also known [17], degenerative changes – loss of collagen fibre elasticity, natural predisposition to metaplasia in structures of cartilaginous origin (such as Reicher's cartilage). Pathological factors include: chronic inflammation in the throat or neck (e.g. after tonsillitis, tonsillectomy) causing local release of pro-inflammatory cytokines (IL-1, TNF-α), which activate osteogenesis, mechanical trauma causing microhaemorrhages and healing with bone tissue deposition, calcium and phosphorus metabolic disorders (less common), genetic predisposition to excessive heterotopic ossification.

Physiological	**Pathological**
Age over 30 years old.	Chronic inflammation
Degenerative changes	Mechanical injury
Predisposition to metaplasia	Ca and P metabolic disorders
Calcification of connective tissue	Genetic predisposition

Steinmann is one of the authors of the hypothesis explaining the causes of differences in the ossification and elongation of the styloid process [31]. He presented four main theories.

The first of these, known as reactive hypertrophy theory, assumes that the elongation of the styloid process is the result of reactive hypertrophy caused by trauma to the throat.

The second, known as the theory of reactive metaplasia, suggests that metaplastic changes occur within the sphenomandibular ligament, leading to its partial ossification, which results in an abnormal ossification process.

The third theory refers to anatomical differences and assumes that ossification of the styloid process and stylo-mandibular ligament may be a physiological phenomenon reflecting individual variations in anatomical structure.

The fourth theory suggests that the elongation of the styloid process may result from the presence of embryonic tissue remnants originating from Reichert's cartilage [32].

Although each of the theories presented provides a credible explanation for the mechanisms leading to the elongation of the styloid process, no clear consensus has yet been reached among researchers on this issue.

DIAGNOSTIC

Symptoms that may occur in Eagle syndrome include chronic sore throat, difficulty swallowing, severe pain when pressing on the tonsillar fossa, dysphagia, pain in the ear and side of the neck—especially when opening the mouth, drooling, pain and sensory disturbances in the face, painful neck movements or a feeling of a foreign body in the throat, Horner's syndrome, numbness at the root of the tongue, hoarseness, pain radiating to the chest, pain in the sternocleidomastoid muscle, headaches located in the ethmoid sinus projection, lateral deviation of the tongue towards the ossifying ligament, pain in the temporomandibular joint [6,17, 32, 33], a number of neuropathic symptoms, or disorders of the mandibular-gnathic complex [10,11]. Ophthalmic symptoms such as diplopia and retro-ocular pain, eye swelling, blurred vision, and visual field disorders have also been described in the past [20,35]. Cases of ischemia and carotid artery dissection are also known [12,17]. These symptoms should prompt further diagnosis. Very often, these symptoms overlap and are therefore nonspecific, resulting in the patient being misdiagnosed. The first and simplest test for suspected Eagle syndrome is palpation of the styloid process area to assess pain intensity. in the palatine tonsil bed, a hard, sharp tip of the hypertrophied styloid process can be felt. A more invasive method to confirm

the diagnosis of Eagle syndrome is the temporary relief of symptoms after administering 1 ml of 1% lidocaine to the palatine tonsil bed. The most important tool, the so-called "gold standard" in the assessment of elongated styloid process syndrome, is CT with 3D reconstruction (3D-CT), which allows for a very accurate illustration of the structures with an assessment of the angle of the styloid process and its adjacent space [8, 24]. CBCT is also a useful examination for the diagnosis of elongated styloid process or ossification of the mandibular-zygomatic ligament, as it allows for detailed visualization of structures and very accurate linear measurements [24, 38]. Often, a basic panoramic image can be helpful, especially in showing the ossified ligament-process area [33]. For this reason, Eagle syndrome is most often diagnosed as a result of ruling out other syndromes. Failure to accurately diagnose Eagle syndrome significantly delays the initiation of appropriate treatment, resulting in patients suffering from long-term head and neck complaints. [39] According to the classification presented by Correll, elongation of the styloid process can be divided into three basic types [31].

The **first** type is characterized by continuous, uninterrupted elongation of the styloid process**.**

The **second** type includes cases in which the styloid process is connected to the styloid-olecranon ligament via a single pseudo-joint.

The **third** type describes the occurrence of numerous mineralized fragments of the ligament, which form a structure resembling many pseudo-joints along its course.

Another author who attempted to classify calcification was Langlai [31]. He divided the calcification process into four types.

Type I involves contour calcification, characterized by the deposition of calcium salts along the outline of the structure.

Type II is referred to as partial calcification, in which the mineralization process affects only parts of the appendix or ligament.

Type III is nodular calcification, characterized by the presence of irregular, nodular foci of mineralization.

Type IV describes complete calcification, in which the entire structure is fully mineralized.

TREATMENT: **CONSERVATIVE TREATMENT** The treatment of Eagle syndrome can be divided into two currently used methods, i.e., conservative and surgical treatment. The aim of conservative treatment is to reduce inflammation, muscle tension, neuropathic pain, and irritation of anatomical structures. It is carried out using measures such as pharmacotherapy, local injections, and physiotherapy. Pharmacotherapy	NSAIDs – ibuprofen, ketoprofen, diclofenac – reduction of pain and inflammation. Centrally acting analgesics – paracetamol, tramadol in the short term. Medications for neuropathic pain – gabapentin, pregabalin, amitriptyline – if there is a neuralgic component (irritation of the IX nerve). Corticosteroids (systemic or topical) – less frequently, in cases of severe inflammatory symptoms.
Local injections	Blockade of the styloid process/cervical ganglion: injection of lidocaine ± steroid (e.g., triamcinolone) under ultrasound or CT guidance. - Has diagnostic and therapeutic effects (if improvement occurs after the blockade, it confirms the diagnosis).
Physiotherapy	Manual therapy and relaxation techniques for the neck and throat muscles. Avoid tilting your head back for long periods of time (e.g., when working at a computer). Warm compresses, soft diet, postural techniques.

In addition, a single injection of 1 ml of triamcinolone combined with a 0.3% solution of mepivacaine (3 mg) into the tonsillar fossa can be used to treat the symptoms of elongated styloid process syndrome [40].
If symptoms persist after 6–12 weeks of conservative treatment, the patient is eligible for surgical treatment.

SURGICAL TREATMENT - STYLOIDECTOMY

Most authors emphasize that styloidectomy is the most effective method of treating Eagle syndrome. [41]
Styloidectomy is the resection of the elongated styloid process or removal of the calcified stylohyoid ligament. Surgeons use two types of access: through the mouth (intraoral) and from the outside (extraoral). Access through the mouth allows for shorter operations and better aesthetic results, but at the same time creates a greater risk of complications associated with limited visibility and an increased risk of infection [39, 42, 43], no visible scar, shorter procedure time, the procedure can be performed under local anesthesia, the risk of accidental exposure of the tonsillar fossa is minimal, and thanks to the flexibility of its bed, intraoperative bleeding remains low. The healing process is faster, the likelihood of infection in the cervical space is low, but more frequent than in the case of extraoral incision [44], and the duration of the operation is reduced. In addition, postoperative pain is less severe and patients recover more quickly. This technique does not require blind dissection, as preparation is limited to the tip of the styloid process [45]. The disadvantages of intraoral access include limited visibility, increased risk of infection, and unintentional excessive displacement of the facial nerve [46]. The average duration of an intraoral procedure is approximately 45 minutes. This approach is preferred in clinical practice due to the minimal risk of damage to the marginal mandibular branch of the facial nerve and the lack of need for skin incisions on the neck, which eliminates scarring [41].

The main indications for surgical treatment are: lack of improvement after conservative treatment, severe pain or vascular symptoms (carotid artery compression - dizziness, fainting), appendix length >3 cm with correlation of symptoms. Factors such as the severity of clinical symptoms, the degree of elongation, and the location of the styloid process are also important factors in the choice of surgical technique.

Intraoral access	A cut in the area of the palatine tonsil (often after a previous tonsillectomy).
Extraoral access	Incision in the neck, access to the appendix through the parapharyngeal space.

Extraoral access reduces damage to surrounding tissues and nerves that may occur during intraoral access due to limited exposure of anatomical structures. Despite a properly performed surgical excision, symptoms persist in approximately 20% of patients. This phenomenon is explained by the entrapment of the glossopharyngeal nerve in fibrous tissue or irritation of the covering placenta by a shortened styloid process, which remains abnormally covered by the placenta of the tonsillar fossa [31, 47].

Due to the prevalence of Eagle syndrome and its nonspecific symptoms, this condition is often misdiagnosed. Therefore, every physician and dentist should consider it in the differential diagnosis of pain in the neck, face, and throat.

Conclusions: Eagle syndrome remains a rare but clinically significant condition which, due to the nonspecific nature of its symptoms, is often undiagnosed or misdiagnosed. A thorough knowledge of the anatomy of the styloid process and stylohyoid ligament is crucial for the proper assessment of pain in the head, neck, and throat. Computed tomography with 3D reconstruction is currently the most accurate and recommended diagnostic method, allowing for the assessment of the length, angle, and degree of calcification of the structures responsible for the symptoms.

Conservative treatment may bring improvement, but if symptoms persist, the most effective method remains styloidectomy, performed using an intraoral or extraoral technique, selected individually according to the patient's anatomical conditions. Despite the high effectiveness of the procedure, some patients may continue to experience symptoms, which indicates the complexity of the pathophysiology of the syndrome and the potential coexistence of neuropathic disorders.

In light of the available data, Eagle syndrome should be regularly considered in the differential diagnosis of chronic facial, throat, and neck pain, as well as neurological and vascular symptoms of unclear etiology. Early diagnosis and appropriate selection of therapy significantly

improve patients' quality of life and reduce the risk of complications, including rare but potentially dangerous vascular disorders.

Disclosure

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