



Eagle Syndrome: A Case Report

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Abstract

Introduction: Eagle's syndrome is a pathology characterized by signs and symptoms affecting the pharyngeal and cervical regions, in relation to the elongated styloid process or the calcification of the ligaments that originate from the same. The styloid process is a bony structure originated from the tympanic part of the temporal bone projecting inferiorly and anteriorly at the base of the skull. The styloid apophysis, or styloid process, has a thin, cylindrical and pointed aspect located in close proximity to noble structures of the cervical region. The styloid process is considered normal when its dimensions do not exceed 30 mm. Eagle Syndrome is characterized by the elongation of the styloid process, and either calcification of the stylomandibular or stylohyoid ligaments, with painful symptomatology in the head and neck region. This symptomatology is varied, including: recurrent or swallowing pharyngeal pain, neck pain, foreign body sensation in the throat and pain when moving the head. Differential diagnosis should be made due to the symptomatology similar to other pathologies of neurological, otorhinolaryngological or orthopedic origin.

Objective: This study aims to report a clinical case of Eagle Syndrome, as well as to review the literature in order to broaden the information regarding clinical diagnosis and treatment.

Case Description: patient, ASA I, submitted to surgical treatment of unilateral styloidectomy, with regression of symptoms after surgery.

Conclusion: There is still a lack of information regarding the pathology, which requires a differential diagnosis with several pathologies with similar signs and symptoms.

Keywords: Cervicalgia; Heterotopic ossification; Buccomaxillofacial surgeons

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Introduction

Eagle Syndrome (ES) is characterized by signs and symptoms that affect the cervical/pharyngeal region in relation to the elongated Styloid Process (SP) or the calcification of the ligaments that originate from it [1].

The SP is a bony structure originated from the tympanic part of the temporal bone projecting inferior and anterior to the base of the skull, antero-medial to the mastoid process [2]. It has a thin, cylindrical and pointed aspect; it is related to the internal carotid arteries and external, auriculotemporal and facial nerves, stylo-mastoid artery and posterior portion of the parotid gland [2]. It serves as an insertion for the muscles: styloglossus, stylohyoid and stylopharyngus [1]. The mandibular and stylohyoid ligaments originate in PE and insert in the angle of the mandible a smaller portion of the hyoid bone, respectively [2,3]. The SP can measure between 25 and 30 mm within the normality patterns and is considered elongated when it exceeds this average [1,3].

The symptomatology of ES is varied, including recurrent pharyngeal pain or during swallowing, which can radiate to the ear ipsilaterally, intense salivation, headache, earache, toothache, neck pain, moving the head, facial pain, tongue and along the internal and external carotid arteries, and during episodes of severe pain, visual disturbance, dizziness and syncope may occur. In addition, there are varying degrees of dysphagia, odynophagia, dysphonia, sialorrhea, hearing loss, restriction of cervical movements, sensation of a foreign body in the throat and trismus [1-3]. ES has similar symptoms to pharyngeal, otorhinolaryngological, neurological, and other pathologies, being extremely important the differential diagnosis [2]. There are cases of stroke, cited in the literature, as a result of the compression of the carotid arteries by the elongated SP, preventing normal blood flow in the region. These evidences reinforce the importance of normal anatomical knowledge and its variations, as well as the careful clinical and radiographic examination of the stylohyoid complex [3].



Figure 1: Clinical evaluation of the patient.



Figure 2: A bilateral elongated styloid process can be observed in the panoramic radiograph performed to confirm diagnostic suspicion. Arrows indicate the position of the elongated styloid process.

The etiology of ES has not yet been clarified. There are several hypotheses for the nature of ossification of abnormal SP, such as heredity, metaplasia, after tonsillectomy or trauma in the region of the hyoid complex [1-4]. The recommended treatment and proven efficacy in the remission of signs and symptoms and improvement in patients' quality of life is surgical treatment, which can be performed inside the mouth or out of the mouth [1-4]. In the absence of a better understanding of Eagle Syndrome, this paper aims to report a clinical case of a patient, ASA I, submitted to the surgical treatment of unilateral styloidectomy. It is hoped that this clinical case and its correlations with the literature may contribute to increase knowledge about the Syndrome in order to broaden information regarding clinical diagnosis and treatment, as there is still a lack of information in the literature, and it can easily be confused with other pathologies that present similar symptomatology.

Case Presentation

A 23-year-old female patient, ASA 1, attended the Oral and Maxillofacial Surgery and Traumatology Service of the Hospital Santo Antônio OSID/UFBA (FEDERAL UNIVERSITY OF BAHIA), presenting painful complaints in the left cervical region and intense headache for approximately 4 months, with episodes of fainting. The patient reported a sensation of a foreign body in the oropharynx on the left side with slight discomfort when swallowing (Figure 1), denying a painful sensation when moving the head.

Treatment plan established: Multidisciplinary follow-up with Orthodontics, Speech Therapy and Neurology; Surgical intervention, unilateral styloidectomy, to remove the left side styloid process, stimulating exacerbated pain sensation, keeping intact the elongated styloid process on the right side. The surgery approach was

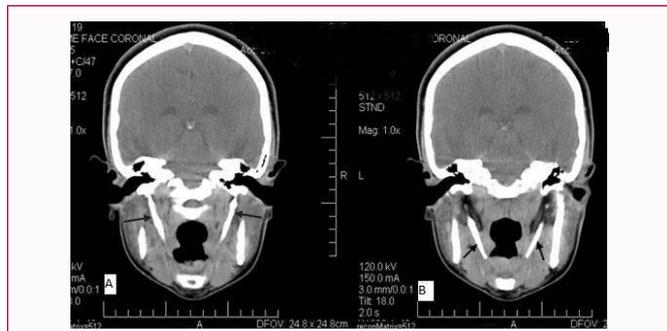


Figure 3: Elongated styloid process indicated by the arrows A side: posterior coronal section B side: anterior coronal section.



Figure 4: Surgical access evidencing noble structures.



Figure 5: Styloid process removed.

performed by out of the mouth access, in the region of the carotid triangle, in order to preserve nearby noble structures such as facial, glossopharyngeal, hypoglossal and vagus nerves, as well as external and internal carotid arteries (Figure 4). The left elongated styloid process was dissected and removed during surgery having about 40 mm length (Figure 5). Surgery performed under general anesthesia resulted in a small cutaneous scar in the incision region after healing (Figure 6). The patient evolves well, with total absence of symptoms, returning to her work activities without further problems or pain.

Discussion

The SE was first described in 1937 by the American otorhinolaryngologist W. Eagle, reporting 200 cases of elongated styloid process accompanied by varied symptoms in the cervical and pharyngeal region [1-4]. The prevalence of SP elongation is higher in females, according to Guzzo et al. [1], since the third decade of life; there is no predisposition for either uni or bilaterality [1]. Others, researchers argue that SE has no gender preference, such as Bagga et al. [2] when assessing 2706 adult panoramic radiographs in their study where there was no correlation between SE and gender, but an



Figure 6: Clinical evaluation of the patient after surgery.

increase in the number of cases with advancing age.

The etiology of ES has not yet been clarified. Studies by Steinmman culminated in the postulation of three theories [1-5,9]. The first one refers to reactive hyperplasia, in which SP is stimulated to ossify in its terminal portion, due to a cervical trauma pharyngeal ligament, for example, with subsequent ossification of the stylohyoid ligament. The second theory is that of reactive metaplasia, in which trauma in the region of the stylohyoid complex can induce metaplastic changes in the stylohyoid ligament, causing the segmented ossification of this ligament. The theory of anatomical variation is the third postulated by Steinmman, who attempts to explain the calcification of the styloid ligaments in children and young people with no history of trauma in the cervical pharyngeal region [2-4,9]. The first and second theories, which correlate the occurrence of SE to a previous trauma, can be classified as classic SE [1-3]. However, there is no report of cervical/pharyngeal trauma prior to the appearance of the symptomatology by the patient of the present case. The ES may still be related to tonsillectomy, and the resulting fibrous tissue would involve the elongate SP, causing a compromise of the fibers of the cranial nerves that innervate the inserted muscles. The affected nerves would be the glossopharyngeal, facial, lower trigeminal branch and hypoglossal, causing pharyngeal pain with ipsilateral ear irradiation, dysphagia, odynophagia, foreign body sensation in the throat, trismus and facial or cervical pain [1,9,12,13]. In the present case, the patient does not report having undergone the tonsillectomy procedure.

Another form of presentation of the ES would be the Syndrome of the carotid artery or stylocarotid. Mechanical irritation causes stimulation of the sympathetic plexus of the carotid artery wall (internal or external), which generates cervical pain with irradiation to the respective areas of vascularization. The worsening of painful symptomatology occurs when the patient rotates the head, regardless of which carotid artery is the focus of sympathetic stimulation [1,3]. This form of ES can be diagnosed as vascular, but this form of ES presentation can cause transient ischemia and stroke by compression of the internal carotid artery [1,14]. There is still no consensus in the literature regarding the etiological factor triggering ES, which evidences the need for further studies to elucidate the origin of this pathology [1-10,14]. In trying to explain the mechanism of ES, some researchers defend the hypothesis that it is a consequence of a mechanical stress of the structures of the stylohyoid complex [14].

The structures of the stylohyoid complex have embryological origin from the second branchial arch, Reichert's cartilage [2]. The ES hypothesis as the sequel to a mechanical stress of the stylohyoid

ligament is based on a histochemical study performed in styloid processes removed after styloidectomy, in which the presence of osteoblasts, osteogenic proteins and chondrogenic cells, usually present in response to tensile strength and mechanical stress in the fibrous ligament tissues, promoting ossification of the stylohyoid ligament and stretching of SP [14].

The hypothesis of mechanical stress as a triggering factor for ES requires more relevant studies with more sample data for its confirmation, since it may be an attempt to clarify the cases in older patients [14]. However, it is not a factor that triggers patient study in this work, since it belongs to an age group of young adults. Diagnosis is made based on clinical examination and imaging tests such as panoramic radiography, lateral cervical radiography and computed tomography [3,6,10]. In the present case, the diagnostic exams were panoramic radiography and multislice computed tomography. During the clinical examination, the palpation of the SP in the tonsillar fossa represents a strong indication of its extension, and there may be exacerbation of the symptomatology, since SP with normal proportions (between 25 mm and 30 mm) is not palpable in this region [1]. Image exams represent an important tool in the determination of the diagnosis of ES, with emphasis on multislice computed tomography, as it plays an important role in obtaining information necessary for surgical planning, such as: extension of SP elongation, angular deviation and anatomical relationships with cervical structures [1,3]. Several degrees of calcification and presentation of SP, observed by some authors, allowed for a radiographic classification. According to this classification, SP can be described as elongated, pseudoarticulate or segmented [1,3].

The differential diagnosis of ES is essential for the accurate determination of the diagnosis and effective treatment, where surgical excision of the elongate SP is usually chosen. Temporomandibular dysfunction, cervical arthritis, otitis, mastoiditis, sialadenitis, sialolithiasis, esophageal diverticulosis, temporal arteritis, myofascial pain, chronic pharyngotonsillitis, impacted or non-erupted third molars, migraine, pharyngeal or tongue base tumors, and trigeminal, glossopharyngeal, upper laryngeal and sphenopalatine neuralgias are involved in the differential diagnosis of ES, which makes its diagnosis to be performed by exclusion [1].

Surgical treatment, which can be performed by inside or out of the mouth access, is satisfactory and effective, and there is no recurrence in the researched literature. Inside the mouth access surgery can be performed, but it is not the most indicated, it has the advantage of not leaving visible scar, however, the risk of injuring noble areas of the region should be analyzed in case of emergency or carotid ligation, as well as increase the risk of infection of the deep facial spaces and reduce the visibility of the surgical field [1-10,18]. Out of the mouth access has advantages for providing good visualization, reducing the possibility of infection or intracervical hemorrhage, reducing the risk of lessening facial, glossopharyngeal, hypoglossal and vagus nerves, in addition to the external carotid artery, however, requires general anesthesia, and results in external cutaneous scarring [1-10,18].

The surgery out of the mouth requires general anesthesia. The incision is made in the region of the carotid triangle, followed by the division of the platysma and sternocleidomastoid muscle. Low incision performed at the level of the cervical fascia that covers the submandibular gland to expose the digastric muscle, dissection extends into the parapharyngeal space with the objective of exposing the posterior belly of the digastric muscle and the elongated SP to

remove it [10,18,19]. The muscles inserted in the SP can be removed or only deflated, which can atrophy or the body finds a new insertion without sequelae to the patient [18]. The styloidectomy, that is, the surgical removal of the styloid process, was chosen the best option of treatment for this case, where we proceeded with the unilateral removal (left side, origin of the symptomatology) by extraoral access. Conservative treatment consists of the use of non-hormonal anti-inflammatories and the local infiltration of corticosteroids or anesthetics, which shows little effectiveness, since it does not act in the cause of the pathology [1,18,19].

Final Considerations

ES is pathology of unknown etiology with difficult diagnosis in view of the various diseases affecting the cervical and facial region, which present similar symptoms. The most effective treatment is the surgical approach to remove the styloid process and provide remission of symptoms, improvement of the patient's quality of life and favorable prognosis since the pathology is not recurrent.

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