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Eagle syndrome: Case report and literature review on a rare pathology treated at the cancer hospital of Muriaé

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ABSTRACT

The styloid process consists of a thin bony projection originating from the temporal bone, in its inferior face, where muscles and ligaments of the head and neck are inserted. Eagle Syndrome was first described in 1937 by Watt Eagle, and is configured by the increase in the styloid process associated with symptoms of pain in the cervical region and carotid compression. It is a rare pathology and still little widespread, but of great relevance as a differential diagnosis for causes of temporo-mandibular dysfunctions, odynophagia and cervicalgia. With varied etiology, the elongation of the styloid process causes symptoms to the patient according to its size and compression of adjacent structures, being the most effective surgical treatment in the resolution of the complaints of the affected patient.

Keywords: Eagle syndrome, Head and Neck Surgery, Temporomandibular disorders.

1 INTRODUCTION

The styloid process consists of a bone prolongation with an average length of 25mm, which is located on the lower surface of the temporal bone, being between the internal and external carotid arteries and posterior to the pharynx, where the stylohyoid, styloglossal and stylopharyngeal muscles originate, innervated respectively by the glossopharyngeal nerves (CN IX), hypoglossal nerve (CN XII) and facial nerve (CN VII), and also the stylohyoid and stylomandibular ligaments. This bone process is considered elongated when its length exceeds the average normal values of 25 mm to 30 mm, being, abnormally, between 30 mm and 105 mm, being more common a length of 40 mm, consisting of a pathognomonic sign of Eagle Syndrome. According to studies, it is estimated that Eagle Syndrome affects about 4% of the world's population, where 50% occurs bilaterally. The highest incidence predominates among 30 to 50 years of age, especially in females.

The stylohyoid apparatus, affected by this syndrome, is composed of the stylohyoid ligament, the minor horn of the hyoid bone and the styloid process of the temporal bone. This begins its formation in the fourth week of embryonic development, since in it the branchial arches are formed, being the second branchial arch that, when divided into 4 segments, will form this apparatus. Anomalies in this complex have been described in humans since 1656, but it was in 1937 that the German otolaryngologist Watt W. Eagle first described Eagle Syndrome as a pathology associated with the abnormal elongation of the styloid process or the aberrant ossification of the apparatus to which this process is part, which will cause symptoms according to the neurovascular structures affected by this abnormality. It is pertinent to list that several structures are closely related to the style-hyoid complex, such as the facial nerve, the auriculotemporal nerve, the lingual nerve, the chorda tympani nerve, the glossopharyngeal nerve, the hypoglossal nerve and the bifurcation of the common carotid artery into the internal and external carotid artery, all subject to compression in a patient with Eagle syndrome.

From this, Watt W. Eagle divided this syndrome into 2 categories: the first called classic, related to pharyngeal involvement with pain radiating to the ear, associated with dysphagia and odynophagia, usually associated with tonsillectomy surgery and the local healing process developed in the postoperative period. The second category, called Carotid artery-apostoid syndrome, occurs when the enlarged styloid apophysis or calcification of the hyoid ligament compresses the internal carotid artery or the external carotid artery, generating pain in the course of these vessels due to sympathetic stimulation and, in more severe cases, there may be dissection of the vessels involved and even a transient ischemic attack or a stroke.

2 MATERIALS AND METHODS

The information contained in this study was obtained through review of the medical record, interview with the patient, photographic record of the diagnostic and treatment methods to which the patient was submitted and literature review.

3 GOAL

To present a case of a patient who received the diagnosis of Eagle syndrome and its clinical manifestations and treatment.

4 CASE REPORT

CFS patient, 48 years old, male, was referred to the Head and Neck Surgery outpatient clinic with the main complaint of "pain in the thick nerve of the neck". Patient reports that for about 3 months presents discomfort in the pharyngeal region associated with pain in the cervical region, especially on the right, of moderate intensity and spontaneous onset, aggravated when opening and closing the mouth

and when moving the neck to the sides. He also had significant odynophagia and foreign body sensation when swallowing. He denies weight loss, adenopathies, or other symptoms associated with upper digestive airways. He denies previous comorbidities, allergies, smoking and alcoholism.

On examination, the patient presented no palpable adenopathies in the neck and no lesions visible at oroscopy, based on the tongue free on palpation. Cervical resonance was requested, which showed no significant alterations.

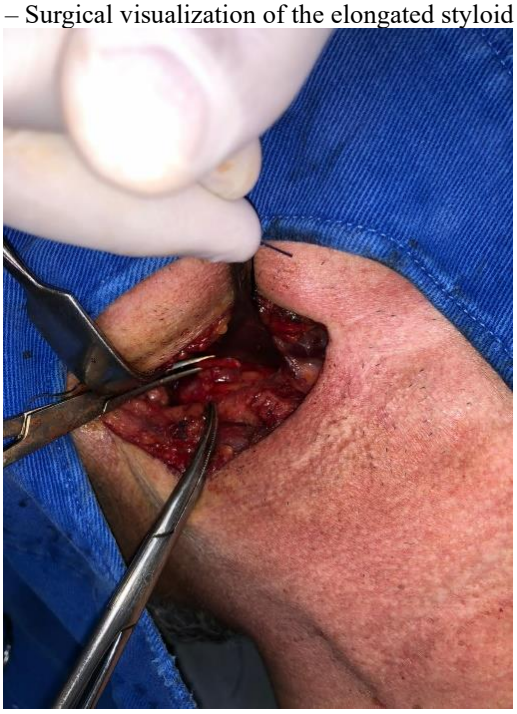
Based on these data, the main diagnostic hypothesis was Eagle Syndrome and, in order to confirm the use of complementary tests, computed tomography of the neck and endoscopic examinations were requested.

Endoscopic examinations without relevant presentations for the diagnostic elucidation of the patient. Computed Tomography of the neck showed bilateral elongation of the styloid processes of the mastoid bone, the right being of greater length and presenting intimate contact with the spine of the pharynx, so the diagnosis of Eagle Syndrome was confirmed and surgical treatment was indicated for the patient through styloidectomy on the right by cervicotomy.

For surgical treatment of Eagle Syndrome in the patient in question, resection of the styloid process was performed on the right, under general anesthesia and without intercurrences.

On the first postoperative day, the patient presented significant improvement of the odynophagia, with pain and edema at the incision site. The patient was discharged from the hospital and was instructed to return in 6 months.

Figure 1 – Surgical visualization of the elongated styloid process.



Source: author.

Figure 2 – Removed styloid process measuring 37mm.

Source: author.

5 DISCUSSION

The most common clinical presentation for patients with this syndrome is pain in the cervicofacial region, which can be unilateral or bilateral, with irradiation to the mandible and ear. Otalgia, odontalgia, cervicalgia, glossalgia, pain in the temporo-mandibular joint and in the course of the carotid arteries are examples of painful manifestations associated with this pathology. There are also other manifestations such as dysphagia, odynophagia, foreign body sensation, pain when yawning, pain when moving the head, trismus, sialorrhea, hypoacusis, tinnitus and syncope, mainly associated with compression of the neurovascular structures near the anomalous style-hyoid complex. Many patients may be asymptomatic.

From the case presented, we observed that the patient presented classic painful symptoms of Eagle Syndrome, with significant neck pain neglecting organic functions such as swallowing and movement of the temporomandibular joint, especially on the right side, in which he presented the classic pathognomonic sign of Eagle Syndrome: elongated styloid process with 40mm in length, of the elongated type, being a continuous bone structure, as we can see in Image X, of the styloid process of the patient after surgical resection.

Although there is still no consensus, there are several etiologies proposed for the development of this abnormality, such as embryological retention of cartilaginous tissue of Reichert's cartilage (second branchial arch) during embryological development, development of fibrosis in tonsillectomized patients, calcification of the stylohyoid ligament, calcification of the stylomandibular ligament or the expansion of bone tissue during its formation, post-traumatic hyperplasia or metaplasia

inducing ossification of the stylohyoid apparatus and also a possible anatomical variation in the size of the styloid process.

To reach the diagnosis of Eagle Syndrome one must have a clinical history, digital palpation and imaging tests. Therefore, a data collection of the main complaints, signs and symptoms of the patient should be made through the anamnesis. Then, the physical examination, which consists of digital palpation, the tonsillar fossa should be palpated with the index finger in order to identify the styloid process, a firm structure found in this region indicates a stretching of the styloid process, otherwise it cannot be felt by this maneuver. Thus, the exacerbation of pain in the performance of this clinical examination, followed by relief of symptoms after anesthetic injection into the tonsillar fossa, shows a positive sign for stretching. And BP, lateral mandible or panoramic radiographs should be requested and observed, which showed the presence of the elongated styloid process, confirming the diagnosis. In order to rule out malignant causes, computed tomography with cranial and neck contrast, cervical resonance and endoscopy should be ordered.

In the differential diagnosis of the syndrome are some dysfunctions of the temporomandibular joint, such as: missing tumors and dental prostheses, tongue base tumors, trigeminal and glossopharyngeal neuralgias; of migraine and histamine headache, non-erupted or impacted third molars, myofascial pain, cervical arthritis and temporal arteritis.

The treatment of this syndrome can be done in two different ways, which will be chosen according to the degree of discomfort and the symptomatology of the patient. The first and most satisfactory form is the surgical treatment, chosen for the reported patient, which consists of the removal of the styloid process intra-orally or by cervicotomy, the second most used being due to the close relationship with noble anatomical structures (carotid artery and facial nerve). Despite having the styloid process elongated on both sides, as the patient's symptoms were only on the right side, the treatment of choice was resection only on the side of the symptoms, with no need to remove the elongated styloid process on the left. The second form consists of pharmacological treatment, which includes analgesics, anti-inflammatories, psychotropic drugs and measures to control the symptomatology, however, if there is no significant improvement, surgical treatment should be done.

6 CONCLUSION

Eagle syndrome is a rare pathology, difficult to diagnose, for which it is important to know the history of the patient through the anamnesis and identify signs and symptoms in this and in the physical examination that can guide the clinical reasoning for it, and thus seek confirmation or exclusion of the diagnosis through complementary tests. It is a disease that has a great importance and relevance in relation to temporomandibular disorders, contributing to their diagnosis and etiology, since it requires an anatomical and physiological knowledge of the joints and all adjacent areas of this region.

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