EAGLE’S SYNDROME: CASE REPORT


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Abstract:
Eagle's syndrome is a disease caused by an elongated styloid process or calcified stylohyoid ligament. Eagle's syndrome is usually characterized by neck, throat, or ear pain; pharyngeal foreign body sensation; dysphagia; pain upon head movement; and headache. The diagnosis of Eagle's syndrome must be made in association with data from the clinical history, physical examination, and imaging studies. Patients with increased symptom severity require surgical excision of the styloid process, which can be performed through an intraoral or an extraoral approach. Here, we report a case of stylohyoid process bilaterally elongated in a 31-year-old female. We did a surgery by extraoral approach and patient's symptom was improved.

Keywords: Eagle syndrome, Elongated styloid process, calcified stylohyoid ligament.

Introduction:
In 1652, Pietro Marchetti first described an elongated styloid process related to an ossifying process of the stylohyoid ligament. Eagle, an otolaryngologist, later defined Eagle's syndrome in 1937 [1-2]. Eagle considered any styloid process longer than 25 mm in an adult to be abnormal. He found that 4% of the population had elongated styloid processes, but only 4% of those with this trait had symptoms [3-4]. Here, we report a case of the stylohyoid ligament bilaterally elongated in a 31-year-old female. We did a surgery by extraoral approach and patient's symptom was improved.

Case Report:
A 31-year-old White female reported to the Department of Otorhinolaryngology of Casablanca. She reported a three year history of persistent left mandibular and submandibular pain which radiated to the neck and the left ear, and this pain increased whenever she turned her head or moved her mandible, especially upon yawning and swallowing. The Patient took several treatments (paracetamol, steroids and carbamazepine). She did not present a previous history of tonsillectomy or cervical trauma. At the physical manipulation, it was found a bone enlargement at the anterior pillar of the right tonsil bed, and a painful (when hand-manipulated) bone projection in

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the left high carotid-jugular area. A skull base CT (Figure 1) showed bilaterally elongated origins of the styloid processes and partial ossification of the stylohyoid ligament. Based on the clinical examination and radiographic findings, surgical treatment under general anesthesia, with an extra-oral approach was recommended (figure 2).

Figure 1: Cervical CT with 3D reconstruction [sagittal view]: ossification of ligament left stylohyoid

Figure 2: Excision of the ligament stylohyoid left externally: Ligament stylohyoid ossified, extending the styloid process
Discussion:
Eagle’s syndrome is characterized by the presence of pharyngeal-cervical symptoms combined with the elongation of styloid process [1]. It is more common in females aging from 30 to 50 years [5-7]. Diagnosis is based on clinical history and physical manipulation, and its confirmation depends on the imaging exam. The impairment is usually bilateral while symptoms are usually unilateral. The severity of these symptoms is not correlated to the size of the styloid. The major symptoms are cervical pain; foreign body sensation in the pharynx; dysphagia; odynophagia and ear pain. At the physical exam, the styloid process can be hand-manipulated as a firm resistance in the lateral wall of the oropharynx (tonsil bed). This procedure, which usually worsens pain and symptom healing after anesthesia in the tonsil bed, helps diagnosing the disease [5-7]. The imaging investigation can be performed through cervical planigraphy (body-section radiography); side and anteroposterior skull radiography; mandible radiography; CT of the base of the skull with three dimensional reconstructions [5,6]. Among all imaging exams, CT of the base of the skull with three dimensional reconstructions seems to be the most effective one, since it enables measuring styloid process length [5,6]. The adequate length (25-30mm) inferiorly reaches the level of C1 and C2 articulation [7]. Clinical therapy may be performed through application of anesthesia or steroids in the tonsil bed; surgical therapy may be performed through intra-oral approach or high cervicotomy. Surgical removal is the most effective therapy for styloid process [5, 6] and if performed through intra-oral approach does not cause scars, though it presents contamination risks of the cervical space and vascular injury due to the difficulty visualization of adjacent structures to the process [5]. High cervicotomy establishes better exposition, preserves nervous and vascular structures and enables more extensive removals of the elongated process [5, 6]. It consequently reduces morbidity rates, as seen through the approaches for the reported cases. In this case, we planned surgical excision because the patient’s symptoms were severe. After surgery, the patient's symptoms improved but a scar remained. At the 3-month postoperative follow-up, the patient's symptoms were further decreased although some discomfort remained, at two years follow up, the patient presented complete symptoms healing with no complications from surgical therapy.

Conclusion:
The syndrome of Eagle is a rare, little known entity which it is necessary to know how to think of in front of algie faciale after having eliminated other ORL and neurological pathologies. It’s diagnosis can be made with clinical examination and radiographic imaging and it’s Surgical therapy by removing part of the styloid process is related to symptom healing.

References: