



## EAGLE'S SYNDROME – A Case Report and Review.

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### ABSTRACT:

Eagle's syndrome is a condition associated with the elongation of the styloid process or calcification of the stylohyoid ligament. Eagle's syndrome can occur unilaterally or bilaterally and most frequently results in symptoms of dysphagia, headache, pain on rotation of the neck, pain on extension of the tongue, change in voice, and a sensation of hypersalivation. Differential diagnosis includes glossopharyngeal and trigeminal neuralgia, temporal arteritis, migraine, myofascial pain dysfunction and cervical arthritis. In this report, we describe the case of a 47-year-old female who presented with dysphagia, tinnitus, cervical pain and intra oral manifestations of Eagle's syndrome.

**KEYWORDS:** Eagle's syndrome, styloid process, stylohyoid ligament, dysphagia, unilateral or bilateral

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### I. INTRODUCTION

Eagle's syndrome is a combination of symptoms which was first described by Watt Weems Eagle in 1937 [1]. Later, he defined two different forms: Classic styloid syndrome, showing uncharacteristic pain and foreign body sensation, and Stylo-carotid-artery syndrome, which is responsible for TIA/stroke in some individuals. [2,3] The first case of elongated styloid process and ossification of the styloid ligament was described by Marchetti in 1652. [4]

The prevalence of Eagle's syndrome is approximately 4% of the total population, and most cases are asymptomatic. [2,4] It is more common in females than males (2:1 ratio). [3] It is characterised by elongation of the styloid processes and/or stylohyoid ligament calcification, classically unilateral or less commonly bilaterally.[5] Women in their 4 – 6<sup>th</sup> decade of life are most likely to be affected and express symptoms. [6] A higher occurrence is seen in the African race. [7]

The styloid process is a slender outgrowth at the base of the skull in the temporal bone, immediately posteromedial to the mastoid apex, arising from the inferior surface of the temporal bone at the junction of its petrous and tympanic portions; lies posterior to the tonsillar and lateral to the pharyngeal wall. It is in close proximity to glossopharyngeal, hypoglossal and facial nerves and to the internal and external carotid artery. [8,9] The styloid process with its five attachments is called the styloid apparatus. Stylohyoid and stylohyoid ligament, both are derived from Reichert's cartilage of 2nd arch, stylomandibular ligament from part of deep fascia, styloglossus-from occipital myotomes and stylopharyngeus-from third arch. [9]

Normal length of the styloid process is 25 mm and is considered to be elongated if longer than 30 mm. Etiopathogenesis is ascribed to elongation of the styloid process or calcification of the stylohyoid ligament. Several theories have been proposed for the etiology of the elongation: 1) Congenital elongation of the styloid process due to persistence of a cartilaginous analog of the stylohyal (one of the embryologic precursors of the styloid), 2) Calcification of the stylohyoid ligament by an unknown process, and 3) Growth of osseous tissue at the insertion of the stylohyoid ligament. [10,11]

The pathophysiological mechanism of symptoms is debated as well. Theories include the following: 1) Traumatic fracture of the styloid process causing proliferation of granulation tissue, which places pressure on the surrounding structures [9,11]; 2) Compression of adjacent nerves, the glossopharyngeal, lower branch of the trigeminal, or chorda tympani; 3) Degenerative and inflammatory changes in the tendonous portion of the stylohyoid insertion, called insertion tendonitis; 4) Irritation of the pharyngeal mucosa by direct compression or

post-tonsillectomy scarring (involves cranial nerves V, VII, IX, and X); and 5) Impingement of the carotid vessels, producing irritation of the sympathetic nerves in the arterial sheath. [9,10,11]

The symptoms of Eagle's syndrome vary and establishing a causative relationship between the styloid process/stylohyoid ligament can be challenging. Classically, the pain develops following tonsillectomy, presumably due to distortion of the local anatomy following surgery. [12]

Eagle's syndrome symptoms can be divided into two main subtypes. [4,11,12]

- Due to compression of cranial nerves
- Due to compression of the carotid artery

*Cranial nerve impingement:*

- Patients can have symptoms related to compression and irritation of cranial nerves in the region (cranial nerves V, VII, IX and X) such as: [1,3,11]
- Facial pain when turning the head
- Difficulty swallowing (dysphagia)
- Foreign body sensation
- Pain on extending tongue
- Change in voice
- Sensation of hypersalivation
- Tinnitus or ear pain (otalgia)

On palpation of the styloid process tip, symptoms should ideally be exacerbated.

*Arterial impingement:*

Compression of the carotid artery may produce vascular/ischemic symptoms as well as pain along the artery to the supplied territory (thought to be mediated by the sympathetic plexus), including. [9,11]

- Mechanical compression
- Visual symptoms
- Fainting (syncope)
- Carotid dissection
- Sympathetic plexus irritation (carotidynia)
- Eye pain
- Parietal pain

The diagnosis of Eagle's Syndrome is based on a good medical history and physical examination (Pharyngeal palpation within the tonsillar fossa). The relief of symptoms after injection of 2% lignocaine into the anterior pillar and tonsillar fossa can be used as a diagnostic tool. Imaging techniques includes Orthopantomogram, Lateral view radiographs of skull and 3D CT (Cone beam CT) is considered as gold standard to evaluate the exact morphology and its relationship with adjacent anatomical structures. [10,11,13] Blood analysis is required to exclude possible systemic diseases.

Eagle's syndrome can be treated surgically and nonsurgically. [14] A pharmacological approach by transpharyngeal infiltration of steroids or anaesthetics in the tonsillar fossa has been implemented, but styloidectomy is the treatment of choice.[15] Styloidectomy can be performed by an intraoral or an extraoral approach. [16] The intraoral approach may result in a restricted operative field, and the risk of deep cervical infections; while the external surgical approach results in cutaneous scars, longer hospitalization, and risks of facial nerve injuries. The overall prognosis (medical or surgical) for Eagle's syndrome is fairly good.

This article presents a case of classical Eagle's syndrome in a 47-year-old female patient with symptomatic bilateral elongation of the styloid process where the length of the right and left styloid processes were 51.00 mm and 42.00 mm respectively.

## **II. CASE REPORT**

A 47-year-old female patient reported to the Department of Periodontology, Sirte Dental school with the complaint of pain in relation to the throat region, below both the ears, upper neck region and mobility of the lower front teeth since four months.

History revealed that the pain is spontaneous in onset, continuous, dull aching type that radiates to the neck, aggravates on turning the neck to either side and on swallowing; does not relieve on taking medications. In addition, the patient had difficulty in swallowing, sensation of a foreign body stuck in her throat and ringing in the ears. The patient had previously consulted a physician and otolaryngologist for the same in the past three months; bilateral ear examination, pure tone, and impedance audiometry was performed and revealed no abnormalities was on medications that gave no relief; hence was referred for further evaluation.

On examination, there was full range of movement of the neck, with just some minor pulling in the

posterior neck muscles, there was no facial asymmetry or abnormality in the temporomandibular joint or in the muscles of mastication. There was no lymphadenopathy. No history of neck trauma.

Intra oral examination revealed multiple missing teeth (17,18,28,36,46 missing). History of extraction was due to mobility. Fixed prosthesis in relation to upper anteriors (11,12,13,21,22,23 - History of Root canal therapy). Grade II mobility was seen in relation to the lower anteriors (31,32,33,41,42,43), Generalised loss of attachment and fremitus test was positive with deep bite. Patient's oral hygiene was relatively good. The oropharynx was normal with no other significant mucosal changes. Tenderness was elicited bilaterally on palpation of the tonsillar fossa.

A provisional diagnosis of Stygia was made correlating the patients' chief complaint, history and clinical examination. Atypical facial pain was considered as the differential diagnosis.

An Orthopantomogram was advised, which showed generalized bone loss and an abnormally elongated styloid process, bilaterally measuring approximately 51mm on the right side and 42 mm on the left side (Figure 1). The length of the calcified styloid complex was measured from the cleft between the inferior margin of the tympanic plate (temporal bone) in the skull and the tip of the styloid process, as described by Jung et al. [8.9] Langlais et al. [10] type II calcification pattern (segmented type) was noted on either side with three and two segments present on the right and left side, respectively. The styloid processes had a knobby or scalloped outline, partial or complete calcification seen in some areas, with varying degrees of central radiolucency suggestive of a nodular complex pattern of calcification bilaterally. No other radiographic differential diagnosis was considered, owing to the confirmatory finding of an abnormally elongated styloid process. The patient was also diagnosed with Generalised Aggressive Periodontitis taking into consideration the amount of plaque, the bone loss, mobility and loss of attachment. The patient was examined using ultrasound and echoes due to the ossification of the elongated styloid processes were detected in the analysis.



Fig. 1. Panoramic imaging of the patient: the arrows point to the bilateral calcified styloid process lateral to the ramus measuring 51 mm on the right side and 42 mm on the left side.

Patient was educated & oral hygiene instructions were given. Phase I periodontal therapy was carried out after which orthodontic stabilization and occlusal therapy was done. Patient was put on maintenance phase and referred for surgical protocol.

The surgeon agreed with the diagnosis and transoral styloidectomy was planned under general anaesthesia; both styloid processes were resected in two procedures, left side followed by right. There were no complications from the procedure. She was discharged with excellent recovery from her pain and hearing symptoms.

### III. DISCUSSION

In the present case, Eagle's syndrome was diagnosed using panoramic imaging, ultrasound and analysis of the symptoms. In this manner, radiation dosage can be reduced and cost affective diagnostic techniques can be used. Eagle's syndrome is uncommon, and a difficult condition to diagnose, involving multidisciplinary approach. In the case presented, diagnosis was made after several months of chronic pain. It is the need of the hour that the

specialists have awareness of this syndrome (Pain, ENT, Vascular, Dental) to allow for effective diagnosis and management due to the wide range of symptom presentation.

The differential diagnosis of Eagle's syndrome should include all conditions possibly causing cervicofacial pain, such as temporomandibular joint diseases; trigeminal, sphenopalatine, or glossopharyngeal neuralgias; temporal arteritis; chronic pharyngotonsillitis; otitis media; external otitis; mastoiditis; dental pain; improperly fitting dental prostheses; submandibular sialadenitis or sialolithiasis; true pharyngeal foreign bodies and tumours of the pharynx or tongue base. [7] In differential diagnosis, we also should consider oesophageal diverticula, cervical vertebral arthritis, hyoid bursitis, Sluder's syndrome, histamine cephalgia, and cluster and migraine type headache. [15] It is vital to exclude cardiovascular and neurological disorders, such as aphasia, syncope, weakness, visual symptoms, or ischemia, associated with interruption of blood flow within the affected vessel in styloid process-carotid artery syndrome.

#### IV. CONCLUSION

In conclusion, by using easily accessible panoramic imaging and ultrasound, Eagle's syndrome can be diagnosed; and surgical resection of the elongated styloid process done to achieve a normal life for the patient.

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