

Eagle's syndrome: The external surgical approach.

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Abstract: Eagle's syndrome is a condition where the styloid process is elongated or the stylohyoid ligament is ossified. In ES the result of such deformity is the compression of the V, VII, IX, and X of the carinal nerves. Patient presentation varies and they can present with one or more of the following symptoms otalgia, dysphagia, foreign body sensation, facial pain, headache, tinnitus, odynophagia, increased salivation, or voice changes.

Keywords: Eagle syndrome, Elongated styloid process, Styloidectomy



1. Introduction

Eagle's syndrome develops due to an elongation or deformation of the styloid process and "ossification" of the stylohyoid ligament.[1]It is known to occur when either the total length of the styloid process is longer than 25 mm or when stylohyoid or stylomandibular ligaments are ossified. Eagle's syndrome may be the source of craniofacial and cervical pain. [2-4]

The first case reported in scientific literature described a calcified elongation of the stylohyoid ligament. This description was undertaken by Pietro Marchetti (1652), an anatomist.[5]Weinlecher (1872) is credited with first reported surgical treatment of symptoms related to an elongated styloid process.[5]Watt Eagle (1937)[6], at Duke's University was first to provide a comprehensive description of symptoms associated with elongation of styloid process and/or calcification of styloid ligament.[7-9] He described the disease clinically in two syndromes the classical syndrome (after tonsillectomy) and the stylocarotid syndrome (not associated to tonsillectomy).[10, 11]

Reichert's cartilage is an important 2nd arch component. It gives rise to the stapes, styloid process, stylohyoid ligament, lesser horns and upper rim of the hyoid. This Reichert's cartilage consists of four developing components; the tympanohyal, which arises from the periotic capsule of the temporal bone and is a process attached to the inferior surface of the petrous part of the temporal bone. The second part is the stylohyale, which usually forms the greater part of the styloid process proper. The tympanohyal and stylohyale segments usually fuse at puberty. The third part is the ceratohyal, which forms the stylohyoid ligament; and the lastly the hypohyal (Epihyal), which forms the lesser cornu of the hyoid bone.[12]

Although approximately 4 % of the population is thought to have an elongated styloid process, only a small percentage (between 4 and 10.3 %) of this group is thought to actually be symptomatic.[13]The cause is considered to be a mystery. Authors have proposed different theories to explain such abnormal elongation and ossification of the styloid process.[14, 15] Treatment options are conservative medical treatment and the

surgical excision. There are two surgical approaches, intraoral and extraoral.[16]

Presented here is a case study of a patient with elongated styloid process that was symptomatic and required a surgical excision.

1. Case Report

A 25 years old male without previous medical history presented to the ENT clinic complaining of right-sided neck tenderness, right otalgia, andodynophagia. Since adulthood the patient has been complaining of recurrent right-sided neck pain, and teeth pain. He underwent several teeth extractions and dental caries management due to bad oral hygiene. However, the pain still persisted. He was referred in 2008 to dental clinic complaining of right-sided pain while chewing and treated as a temporomandibular joint arthritis. In 2011 he was referred to the endocrinology clinic due to his right neck pain. The patient laboratory works were within normal limits, and his ultrasonography showed enlarged thyroid gland and he was suspected to have thyroiditis. He received medical treatment and the follow up ultrasonography of the thyroid was performed and it was normal. During the period 2012- 2017 he regularly visited the primary healthcare clinic complaining of neck pain, swelling, and headache and kept receiving analgesics and anti-inflammatory medications about without any actual improvement in the clinical condition. Finally he was referred to the otolaryngology clinic where the clinical presentation of the patient coupled with his long journey seeking for relief raised the suspicion of the presence of an elongated styloid process. His neurological examination

was normal and he only complain was tenderness during palpation of right retroauricular region. Computed tomography (CT) scanning revealed styloid process elongation that goes with Eagle syndrome **fig 1**

Management plan

The patient after going through the different conservative medications was not willing to precede more with the drugs and he was offered the surgical option. Neck CT scan was performed and revealed bilateral elongated styloid processes more on the right side. (4.8 cm). The two surgical approaches were discussed with the patient and He was booked for external styloidectomy.

2. Method

Operation was done under general anesthesia. The patient is positioned in the supine position with the head extended and tilted to the other side. Identifying the external landmarks and marking the site of the planned incision 2 cm below the angle of the mandible. **fig 2a**

The skin is incised and subplatysmal flaps were elevated with special care to avoid injury of the marginal mandibular nerve. The submandibular gland was dissected and retracted anteriorly. The posterior belly of the digastric muscle was dissected and identified, then was retracted laterally. Figure palpation of the surgical field identified the location of the elongated styloid process. The styloid process and the stylohyoid ligament were dissected and identified. **fig 2b**

The stylohyoid ligament was transected at the tip of the styloid process as it was not calcified. A bone

cutting scissor was used to cut a 2.5 segment of the elongated styloid process. **fig 2c& 2d**The surgical field was inspected carefully of hemostasis and the wound was closed in layers and a compressive dressing was applied.

3. Discussion

Eagle syndrome is one of the benign causes of cervical dysphagia and chronic orofacial pain. [17]Eagle syndrome is primarily an otorhinolaryngologic condition characterized by odynophagia and cervicofacial pain due to an elongated or ossified stylohyoid ligament, in its severest form ipsilateral head turn or relative systemic hypoperfusion may precipitate focal cerebral hypoperfusion and ischemia.[18]

Different values were reported as concerns the length of the styloid process in the literature.[19]Reviews on the literature and radiological studies suggest that the length of the styloid process should not be more than 25 mm and its considered to be accepted as 3 cm in general. [1, 20-22]Although the incidence of elongated styloid processes varies between 1% and 30% (mean 24%), most elongated styloid process cases are usually asymptomatic and only about 4% are symptomatic.[19]It has been reported that the eagle 'syndrome is generally more common in women than men and occurs at the age of 30.[23], the patient presented here at the age of 25 and was a male.

In most cases the styloid process is elongated bilaterally yet the symptoms appear to be associated with one side only.[24-26]The patient presented here also had a bilaterally elongated styloid process, as stated in the

literature.[24-26] However, the symptoms and findings were observed to be associated with the right side only.

Clinically, the patient had pain in the tonsillar region and there was tenderness in the posterior tonsillar pillar on manual palpation of the right tonsil. Palpating the styloid process in the tonsillar region can facilitate Eagle syndrome diagnosis. However, objective findings are revealed by radiographic imaging.[27] Computed tomography was of help in establishing the diagnosis and also in measuring the length of the styloid process in this patient and it was 48.34 mm. fig 1b Lateral head-neck radiography, anteroposterior radiography, townes radiography, and panoramic mandible radiography are another imaging methods that can be used for diagnosing the elongated styloid process.[26, 28]

The occurrence of symptoms in the Classic Stylohyoid Syndrome causes direct compression on the pharyngeal mucosa and cranial nerves that result from a traumatic fracture of the elongated styloid process.[2, 7]. However, it can happen in those with no trauma or surgical history as a result from the congenital length of styloid processes or the embryonic ossification of the stylohyoid ligament.[19]

Pain can be observed along the course of ICA. Patients can be misdiagnosed with migraine or cluster-type headache.[29-31] Under-eye pain caused by the external carotid artery involvement. In addition, and more rarely, direct compression on the carotid artery may cause symptoms. Cases in which vascular compression is more evident and with clinical findings, such as aphasia due to interruption of blood flow in the affected artery, visual

symptoms, transient ischemic attack, vertigo and syncope, can also be there.[32]

The patient was offered a number of treatment options as an alternative to surgical treatment (use of non-steroidal anti-inflammatory and the use of carbamazepine) however he was not satisfied by the results of this conservative treatment and agreed to proceed with the surgical option. Surgical shortening of the elongated styloid process is the only effective treatment.[19] However, there are other conservative options that could be offered such as the use of non-steroidal anti-inflammatory drugs, injection of transpharyngeal steroids or long-acting local anaesthetics, use of oral carbamazepine. [33, 34]

4. Conclusions

Eagle 'syndrome' is manifested with a wide variety of symptoms and causes diagnostic difficulties when it is not considered in the differential diagnosis

Radiological diagnosis of the Eagle syndrome is easy in patients with specific findings. CT scan is a fast and effective examination in terms of showing the bony deformity in patients with the Eagle syndrome that is considered to be abnormal and causes vascular compression.

Surgical shortening of the elongated styloid process is the only effective treatment. However, there are other conservative options that could be offered to the patient yet the responses are individual.

5. Compliance with Ethical Standards

- i. **Disclosure of potential conflicts of interest:** Author **Maram M.A. Alghumlas, MBBS** declares that she has

no conflict of interest. Author **Hisham E.M. Elbadan, M.D** declares that he has no conflict of interest. Author **Walid Farouk Sobhy Afifi, M.Sc.** declares that he has no conflict of interest. Author **Abdullah O.A. Aljaafari, MBBS** declares that he has no conflict of interest. Author **Hind N.M. Alotaibi, MBBS** declares that she has no conflict of interest.

- ii. All procedures performed in this study involving human participants were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki declaration and its later amendments.
- iii. **Informed consent:** Informed consent was obtained from all individual participants included in the study.

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Figure Legends

Fig. 1. CT scan of the patient showing an elongated styloid process. [a] Coronal view showing bilateral elongated styloid processes. [b] Sagittal view of the left side showing a 48.34 mm long styloid process.

Fig 2. External surgical styloidectomy. [a] Identifying the external landmarks and marking the site of the planned incision 2 cm below the angle of the mandible. [b] The styloid process and the stylohyoid ligament were dissected and identified. [c] A bone cutting

scissor was used to cut a 2.5 segment of the elongated styloid process. [d] The excised portion of the styloid process.

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