

Surgical Management of Long-standing Eagle's Syndrome

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Abstract

Introduction: Eagle's syndrome is a rare condition that refers to chronic recurrent pain in the oropharynx, face, and neck due to elongation of the styloid process or calcification of the stylohyoid ligament. It can be treated medically or surgically through a styloidectomy. In this paper, we review our experience with the two surgical approaches for the management of Eagle's syndrome. **Materials and Methods:** We conducted a retrospective chart review, which covered the period between 1997 and 2008. The review included seven patients with a long-standing diagnosis of Eagle's syndrome. Six patients underwent surgical intervention and one patient elected to observe her condition. **Ethics Approval:** The retrospective design of the study was approved by Boston Medical Center Institutional Review Board with no need for another consent other than the one obtained before surgical interventions. **Results:** Out of the seven patients identified, there were three men and four women. The median age and mean age at diagnosis were 44 years and 26.2 years, respectively. Neck pain and odynophagia were the most common symptoms reported. Three patients underwent styloidectomy through transoral approach and the three through transcervical approach. The average time to resolution of symptoms was 26.5 days. **Conclusion:** The review suggests a favorable role for surgery in the management of Eagle's syndrome with all the patients undergoing styloidectomy experiencing complete resolution of symptoms. Both surgical approaches provide the desired outcome; however, the choice of the surgical approach depends on the patient's wishes and the surgeon's experience.

Keywords: Eagle's, pain, surgery

INTRODUCTION

Eagle's syndrome is named after W. W. Eagle, who first described the symptomatology of dysphagia, referred pain in the ear, and foreign body sensation due to an elongated styloid process.^[1] It has since been referred to in many reports and described by a wide variety of terms.^[2-5]

Similar symptoms can be caused by mineralization/calcification of the stylohyoid and stylomandibular ligaments following traumatic or spontaneous fracture of the styloid process.^[6] The length of a normal styloid process is estimated to be 2.5 cm.^[7] However, anatomic studies have shown a wide range of variation. The styloid process is considered elongated when it is <2.5 cm or the combined length of the styloid process and ossified stylohyoid ligament is <3 cm.^[1] The presenting symptoms typically include dysphagia, headache, pain on rotation of the neck, pain on extension of the tongue, change in voice, and a sensation of hypersalivation.^[8,9] The incidence of elongated styloid process has been reported between 1.4% and 30%.^[1,10,11] Of the patients with the anatomic anomaly, only an estimated 7.8%–10.3% present with symptoms.^[3,4]

Women are found to be affected more often than men, at a ratio of 3:1.^[12]

Although the precise pathophysiology has yet to be determined, new embryological evidence, epidemiological studies, and radiographical modalities are helping us understand why only some people with this anatomic anomaly develop symptoms. In this report, we present our experience with managing Eagle's syndrome through both transoral and transcervical approaches over a period of 12 years at a tertiary care referral center.

MATERIALS AND METHODS

This retrospective chart review, which covered the period between 1997 and 2008, included seven patients with a long-standing diagnosis of Eagle's syndrome. Six patients

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underwent surgical intervention and one patient elected to only observe her condition and did not undergo surgery. Each patient underwent a thorough history taking and physical examination. The data recorded included gender, age, symptoms, duration of symptoms, surgical approach, and complications. The presenting clinical features in these patients included otalgia and neck pain. In addition, computed tomography (CT) scans and/or panoramic films were obtained that showed the elongated styloid process in all of the cases [Figure 1]. The surgical approaches used for the removal of the styloid process were transoral or transcervical. All procedures were done under general anesthesia.

In the transoral approach, the styloid process can be palpated in the superior lateral pole of the tonsillar fossa. Once identified, the overlying mucosa is incised, the styloid ligament is dissected free from the calcified styloid process, and an incision is made through the periosteum of the styloid process, which is then excised as close to its base as possible. For the transcervical approach, the subplatysmal flaps are raised and the superficial layer of the deep cervical fascia is incised along the anterior border of the sternocleidomastoid muscle, which is followed superiorly to the posterior belly of the digastric. The styloid process can be palpated deep to the digastric muscle. An incision is then made on the styloid tip facilitating reflection of the periosteum and muscle attachments. Then, rongeur forceps are used to excise the styloid process. The criteria for success were partial or complete resolution of preoperative symptoms. A description of a typical case is given below.

Case 1

A 37-year-old woman who presented at the Department of Otolaryngology with a 3-month history of left-sided odynophagia that began immediately after a motor vehicle accident in which she was a belted driver. Treatment before presentation had consisted of nonsteroidal anti-inflammatory medications. The past medical history was significant for left temporomandibular joint (TMJ) surgery. On physical examination, the left anterior tonsillar pillar had a bony protuberance that elicited a sharp pain on digital palpation similar to the presenting complaint [Figure 2].

The radiographs suggested bilaterally elongated styloid processes, with the left being more prominent than the right [Figure 3], measuring approximately 3 cm. A diagnosis of traumatic Eagle's syndrome was made. The patient underwent a transoral approach to remove the left styloid process. She had an uneventful surgery and postoperatively had transient lingual paresthesia, which resolved after 2 months. She is currently asymptomatic.

RESULTS

Seven patients were identified with long-standing Eagle's syndrome through a diagnosis of exclusion over a 12-year period. Other conditions considered were TMJ pain, otalgia, and neoplastic lesion, especially in patients with odynophagia such as paragangliomas including carotid body tumors. Of

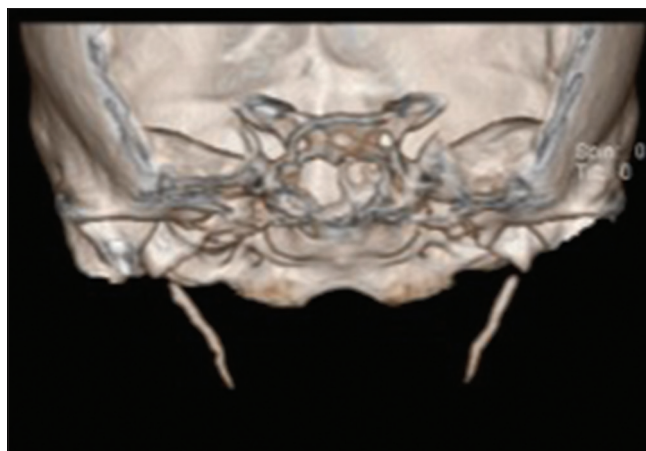


Figure 1: Elongated styloid process. Three-dimensional reconstruction of computed tomography scans showing an elongated styloid process

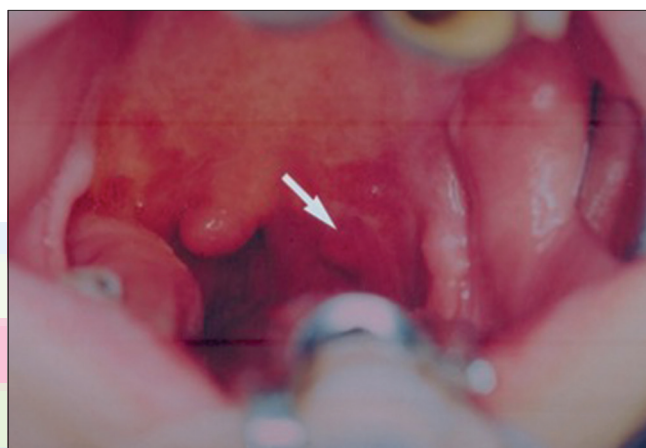


Figure 2: Palpation of styloid process intraorally. Bony protuberance of styloid process at the superior lateral pole of the tonsillar fossa

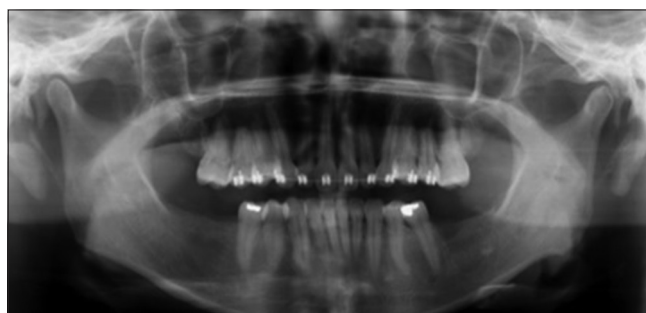


Figure 3: Case 1 panoramic film findings. Preoperative panoramic radiograph showing the elongated styloid process

the seven patients identified, there were three male and four female patients. The median age and mean age at diagnosis were 44 years and 26.2 years, respectively. Six patients had neck pain and odynophagia as their main symptom (85.7%), with an average pain score being 9 out of a severity score of 10 [Table 1]. One patient did not elect to undergo surgery and continues to have pain after having been diagnosed 3 years prior.

Three (42.8%) patients were diagnosed as having an elongated styloid process on panoramic imaging. Four (57%) patients were diagnosed through a neck CT scan with three-dimensional (3D) reconstruction. The average duration of symptoms before surgical treatment was 14.8 months. Half the surgical candidates underwent transoral resection and half underwent transcervical resection. The average length of the styloid process resected via transoral approach was 1 cm, while the average length of the styloid process resected through external approach was 2 cm. The average time to resolution of symptoms was 26.5 days. All surgically treated patients had symptom resolution. All patients resumed their diet within 24 h of surgery. Table 2 summarizes the patient demographics and response to treatment.

DISCUSSION

The first report of an ossified stylohyoid ligament was made by Marchetis in 1652.^[5] Balbuena *et al.*,^[13] in 1870, described the pain syndrome associated with an elongated styloid process. The first surgical excision was performed by Weinlecher in 1872,^[1] and Fritz^[14] published a study of 43 patients with elongated styloid processes, followed by Eagle, who published his study of 211 patients in 1948.^[7] The incidence of elongated styloid processes is unclear. Eagle estimated at 4%,^[1] Kaufman *et al.*^[10] at 28%, and Gossman and Tarsitano^[11] at 1.4%, but all patients in these series were asymptomatic.

The styloid process resides in an anatomically complex area, which helps understand how a pathologic change in size can affect neighboring structures. It extends anteromedially from the mastoid process and lies between the external and internal carotid arteries. It is the point of attachment of three muscles and two ligaments: The styloglossus (innervated by cranial nerve [CN] XII), stylohyoid (innervated by CN VII), and stylopharyngeus (innervated by CN IX) muscles and the stylohyoid and stylomandibular ligaments. Posterior to the

styloid process is the stylo mastoid foramen through which the facial nerve and stylo mastoid artery exit. Lateral to the stylo mastoid foramen is the tympanomastoid suture, which houses the auricular branch of the vagus nerve. Medial to the styloid process is the internal carotid artery, internal jugular vein, spinal accessory, and hypoglossal and glossopharyngeal nerves. Lateral to the styloid tip is the external carotid artery and medial to it is the superior constrictor muscle and pharyngobasilar fascia.^[13,15,16]

Many etiologies have been proposed to explain the anatomic disturbance in Eagle's syndrome, ranging from traumatic stimulus following a tonsillectomy or other pharyngeal surgery, to a degenerative process, to a genetically transmitted trait. However, the exact cause of Eagle's syndrome remains unclear.^[13,15,17] Likewise, there is also no consensus on the cause of the resultant symptoms due to this anomaly. Common theories include (1) traumatic fracture of the styloid process that causes proliferation of granulation tissue, which places pressure on the surrounding structures,^[18] (2) compression of adjacent nerves; the glossopharyngeal, lower branch of the trigeminal, or chorda tympani,^[19] (3) insertion tendonitis resulting from degenerative and inflammatory changes to the tendinous portion of the stylohyoid insertion,^[3] (4) irritation of the pharyngeal mucosa by direct compression or posttonsillectomy scarring (involving CNs V, VII, IX, and X),^[9] and (5) impingement of the carotid vessels, producing irritation of the sympathetic nerves in the arterial sheath.^[6,11] A study by Siéssere *et al.*^[20] quantified masseter and temporalis muscle hyperactivity resulting from the interference of the elongated styloid process. This correlates with the craniofacial signs and symptoms observed in this disease process. Less commonly, there have been reports of reversible transient ischemic attacks, consisting of ophthalmic symptoms and episodes of presyncope with extensive head turn due to an elongated styloid process compressing the carotid artery, and decreasing blood flow in the medial cerebral artery.^[21,22]

Evidence suggests that having an elongated process or calcification of the stylohyoid ligament does not necessitate symptoms of Eagle's syndrome. Studies have shown that mineralization of the stylohyoid complex is not an uncommon finding.^[11,17] Conversely, it has been noted that craniofacial pain often exists in the same region of distribution without any abnormality of the styloid process or stylohyoid ligament.^[23] A study by Krennmair and Piehslinger^[24] discovered that only

Table 1: Patients' symptoms

Symptom	Patients (%)
Neck pain	6 (85.7)
Otalgia	4 (57)
Dysphagia	3 (42.8)
Odynophagia	6 (85.7)
Pharyngeal foreign body sensation	2 (28.6)

Table 2: Patients characteristics

Patient	Age (year)	Gender	Duration of Symptoms	Surgical approach	Resolution of Sx	Complications
1	61	Female	2 years	Transoral	Complete	None
2	44	Female	8 months	Transcervical	Complete	None
3	37	Female	3 months	Transoral	Partial	Transient lingual nerve hypoesthesia, 2 months
4	45	Female	3 years	No surgery	-	-
5	34	Male	3 months	Transcervical	Partial	None
6	50	Male	10 years	Transcervical	Complete	None
7	63	Male	2.5 years	Transoral	Partial	None

136/765 patients with TMJ disorders had a radiographically identifiable abnormality in the styloid-stylohyoid chain. Thus, they concluded that stylohyoid involvement in TMJ disorders, in general, is rare.

Frommer^[25] had postulated that the direction and curvature of the styloid process were more important than length in contributing to symptoms. Likewise, they found that the variability and form of this segment result in varying sizes of stylohyoid. Furthermore, they noted that the inferior end of this segment can present with varying degrees of angulation, which may be significant enough to allow the tip of the angulated part to reach the oropharyngeal wall, which can lead to the symptoms of dysphagia and globus sensation associated with Eagle's syndrome.

The diagnosis of Eagle's syndrome requires knowledge of the anatomy, suspicion of the syndrome, and is still a diagnosis of exclusion. Other possible etiologies that can cause similar pain include glossopharyngeal neuralgia, trigeminal neuralgia, impacted third molars, cervical arthritis, otitis media, and tumors of the pharynx and base of tongue. A panoramic radiograph can diagnose an elongated styloid process as easily as a CT scan, but many practices do not have an orthopantomogram available; a CT scan may be more readily available, with 3D-CT scans being considered the gold standard in radiographic diagnosis of Eagle's syndrome.^[26] Our results show that the patients can get complete resolution of symptoms after surgery provided the correct diagnosis has been made.

Treatment of traumatic Eagle's syndrome can be medical or surgical. Medical treatment revolves around symptomatic management with the use of nonsteroidal anti-inflammatory agents, cortisone, and local anesthetic injections in the lower pole of the tonsillar fossa.^[3,18] Other methods include mandible rest, soft diet, antibiotics, and even maxillary mandibular fixation for 3–4 weeks.^[27]

The surgical approach can be transoral or transcervical (extraoral). The transoral approach was advocated by Glogoff *et al.*^[28] and requires either a local or general anesthetic. The styloid is palpated and the overlying mucosa is incised. Blunt dissection is carried down to the styloid and muscles inserting onto the styloid process are removed. Using bone rongeurs, the styloid process is shortened. The wound is then irrigated and closed with absorbable sutures.^[9,28] The advantages of this approach include shorter operating time, no cervical incision, and possible use of local anesthesia alone. The disadvantages are the lack of control of the major vessels of the neck, poor surgical exposure, and possibility of bacterial contamination of the deep neck spaces.^[9]

The transcervical or extraoral approach requires general anesthesia. An incision is made in a cervical crease and the subplatysmal flaps are elevated. The anterior border of the sternocleidomastoid muscle is delineated. The cervical fascia overlying the submandibular gland is incised low so as to expose the digastric muscle. Staying deep to the posterior

belly of the digastric muscle, dissection is carried into the parapharyngeal space and the great vessels of the neck and styloid process exposed. The styloid process is reduced with bone rongeurs. The advantages of this approach include control of the great vessels of the neck, better exposure, and lower risk of bacterial contamination of the deep neck spaces. As noted in our results, the external approach allows for a longer length of the styloid to be removed than possible through a transoral approach. The disadvantages are a longer operating time, an external incision, and possible transient weakness of the marginal mandibular nerve.^[29] Ultimately, the decision for transoral or transcervical approach depends on the surgeon's experience and the patient's wishes. In our series, both approaches led to symptom resolution. One of the patients who received a transoral incision developed transient lingual nerve hypoesthesia that was resolved within 2 months; otherwise, the surgical interventions did not result in any other complications.

CONCLUSION

Eagle's syndrome is a rare entity that should be considered in the differential diagnosis of a patient who has vague cervical pain or odynophagia. 3D-CT reconstruction and panoramic radiographs can provide much useful information for diagnosis and preparation for surgery although an isolated elongated styloid process does not necessarily equate to a diagnosis of Eagle's syndrome. The choice of the surgical approach depends on the patient's wishes and the surgeon's experience. Most of these patients experience complete resolution of symptoms after surgical intervention.

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Conflicts of interest

There are no conflicts of interest.

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