



Eagle's syndrome and associated pain in head and neck region: A case report and review literature

Rachatawan Leelarangsun¹, Surakit Visutiwatanakorn²

¹ DDS, Post-grad Dip (Oral and Maxillofacial Surgery) Department of Hospital Dentistry, Faculty of Dentistry, Mahidol University.

² DDS, Department of Oral and Maxillofacial Surgery, Faculty of Dentistry, Mahidol University.

Abstract

Orofacial pain is an unpleasant symptom that can originate from several etiologies, including inflammation of the eustachian tube, parotid gland infection, impacted teeth, trauma, carcinoma of pharynx, and trigeminal neuralgia. The most common diagnoses of orofacial pain are cluster headache, migraine, and temporomandibular joint disorders (TMD). On the other hand, elongation of styloid process, also known as Eagle's syndrome, is often neglected because of its rare occurrence. Orofacial pain related to an elongated process can be relieved by its surgical removal. Therefore, it is important to gather all necessary information by history taking and clinical and radiographic investigation to make the correct diagnosis so that unnecessary treatment and surgeries can be avoided.

Key words: Eagle's syndrome, styloid process, orofacial pain, stylalgia, neck pain, langlais classification

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Correspondence author:

Surakit Visutiwatanakorn
Department of Oral and Maxillofacial
Surgery, Faculty of Dentistry,
Mahidol University,
6 Yothi Road, Rajthevi, Phyathai,
Bangkok, Thailand
email address: surakit.vis@mahidol.ac.th

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Introduction

Eagle's syndrome is a rare condition which was first described in 1652 by Marchetti^{1,2}. In 1870, Luke published the clinical presentation of an elongated styloid process^{3,4}. Thereafter, in 1872, Weinlechner reported a case of elongated styloid process and its surgical removal which eradicated the symptoms^{3,2}. In 1937, Watt W. Eagle started his work on the clinical and radiological presentations, diagnosis, and treatment of elongated styloid process. He named it as "Stylalgia" and defined it as an abnormal elongation of styloid process (more than 30 mm) which irritates the adjacent structures, especially glossopharyngeal nerve and causes recurrent throat and neck pain that radiates to the auricular and mastoid region without any previous history of trauma⁵. Although many cases have been reported in the literature, its etiology is unknown^{6,7}.

Several symptoms has been reported with Eagle's syndrome, including cervicofacial pain, odynophagia, dysphagia, foreign body sensation, and headache. These symptoms are provoked by mastication, swallowing, tongue movement, speech, and changes in the head posture. Voice changes also have also been mentioned, lasting for a few minutes^{7,8,9}. Limited mouth opening following pain on orofacial region is another commonly associated complaint which can be significantly resolved by injection of local anesthetic agents and corticosteroid⁶. Epidermologically, there is a 4% incidence of elongated styloid process in the population but only 4% of these patients have related symptoms^{9,10}. It is mostly found in individuals aged 30-50 year-old and there is also a female predominance. Although it can be found more bilaterally than unilaterally, most patients often complain of pain at only one side^{8,11}.

Eagle's syndrome presents as an atypical pain at the craniofacial and neck region which

are often misdiagnosed as eustachian tube infection, parotid gland infection, pain due to impacted teeth, carcinoma of pharynx, trigeminal neuralgia, and faulty dental prosthesis. In most cases cluster headache, migraine, and temporomandibular joint disorder (TMD) are the most commonly diagnoses. This reflects that history taking alone may not sufficient. Physical examination by the digital palpation of the styloid process in the tonsillar fossa is also required^{10,12,13}. If there is a presence of pain during palpation at the region, it should be thoroughly examined for signs of an elongated styloid process^{6,9,14}. Radiographic imaging has an important role for establishing the definitive diagnosis. Panoramic radiographs is one of the best modalities which is recommended by many authors, as it shows the styloid processes bilaterally and easy to perform. Computed tomography (CT) scan, magnetic resonance image (MRI) and lateral cephalometric technique have also been used. The normal length of the styloid process is 20-30 mm^{1,12,15}. If the length is longer than 30 mm, it is suggestive of an elongated styloid process^{1,15,16,17}.

According to pathogenic mechanism as purposed by Watt W. Eagle in 1937, the elongation of styloid process or ossification of stylohyoid ligament is classified into 2 types-Classic styloid syndrome and Stylocarotid artery syndrome. The classic type presents as an atypical pain at oropharynx and neck region that radiates to mastoid and temporal areas. Tonsillitis is a common diagnosis in these situations¹. Consequently, the patient may have a history of tonsillectomy, without any relief of the symptoms⁹. Imaging techniques may reveal an abnormally elongated styloid process, bilaterally or unilaterally. On the other hand, some authors believe that the classic type occurs secondary to the tonsillectomy due to the fibrosis during the healing process^{18,19}. Stylo-carotid artery syndrome presents with

headache, dizziness, and syncope. Some patients also have reported of transient ischemic attacks with numbness and muscle weakness at one side of face and body. It is caused by compression on internal carotid artery and nerve plexus such as glossopharyngeal nerve, accessory nerve, and vagus nerve from elongated styloid process. Moreover, sympathetic plexus which lies on a surface of internal carotid artery is also affected. However, in a clinical report by Chuang et al, sclerosis of the internal carotid artery was not observed on computed tomography angiogram²⁰.

Based on the radiographic presentations, Langlais et al divided Eagle's syndrome into 3 groups: Elongated type, pseudo-articulated type, and segmented type (Figure 1) (ref). Elongated

type illustrates the continuity between styloid process and stylohyoid ligament. On the other hand, pseudo-articulated type shows a discontinuous line between them. Segmented type is described as multiple ossification of stylohyoid ligament^{1,7,10}. This clinical report explains a rare condition of craniofacial pain in a patient, which was caused due to elongated styloid processes (classic styloid syndrome) and its surgical management.

Clinical report

A 31 year-old Thai female came to the Faculty of Dentistry, Mahidol University in 2011 with a chief complaint of pain at the right ear, which she had been having for the past 3 years. The pain occasionally radiated to the right

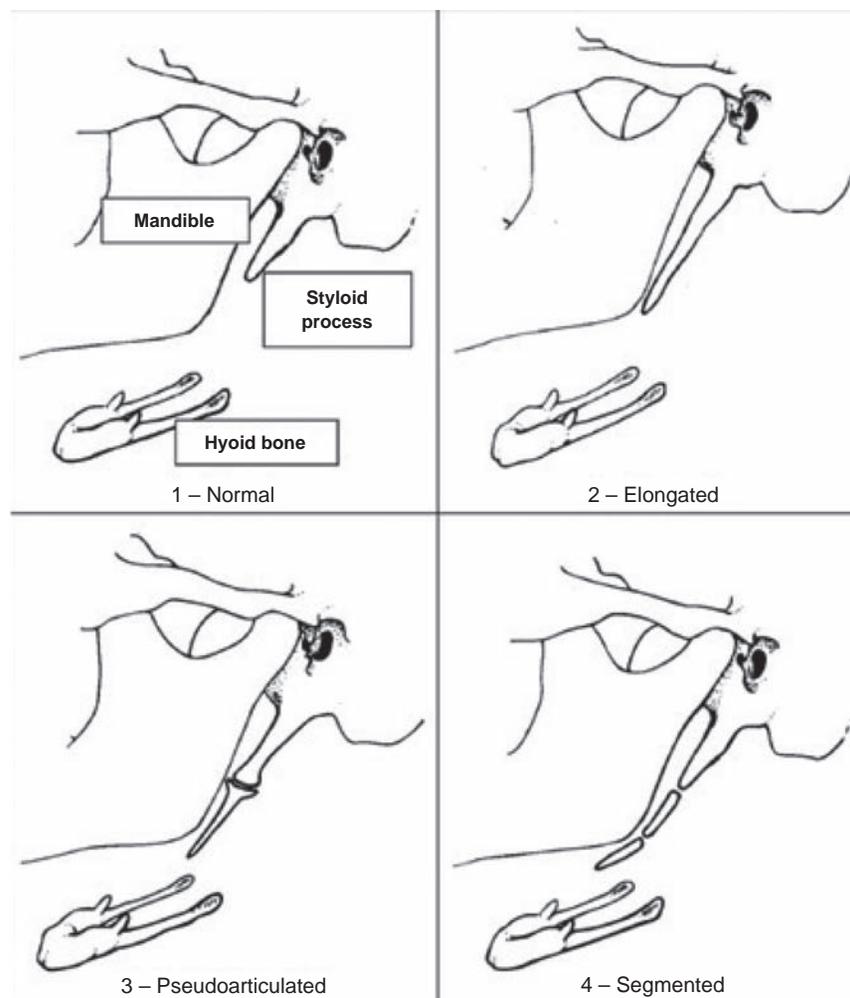


Figure 1 Langlais classification of elongated styloid process; (1) Normal styloid process, (2) Type 1 Elongated, (3) Type 2 Pseudoarticulated, (4) Type 3 Segmented.

temporal, mastoid, and cervical regions and exacerbated during talking, chewing, and changing of the head posture. There were no relevant systemic diseases or drugs allergies. On external examination, facial asymmetry along with deviation of the chin to the left side was observed. There was also a canting of maxillary arch to the left but no traumatic occlusion was noted (Figure 2).

The patient had been referred to an occlusion specialist because the initial diagnosis of the orofacial pain was made as TMD. After continuing physical and occlusal splint therapy for a year, the symptoms had not been relieved. Following which she was referred to an orthodontist for occlusal realignment and correction of the facial asymmetry. During the treatment with fixed appliance, the patient continued to have pain without any relief. As a part of the continuing orthodontic treatment, she was accepted to the Department of Oral and Maxillofacial Surgery for orthognathic surgery to correct the facial asymmetry. After history taking and radiographic imaging, the cause of orofacial pain unfolded. Panoramic radiograph clearly showed elongated styloid processes bilaterally, which were approximately

5 cms in length (Figure 3). Lateral cephalometric radiograph also confirmed the findings (Figure 4). Finally, the definitive diagnosis of Eagle's syndrome was made for the origin of the orofacial pain. Amputation of right elongated styloid process and Le Fort I orthognathic surgery with bilateral sagittal split ramus



Figure 2 Frontal view of the patient exhibiting profound facial asymmetry.



Figure 3 Panoramic radiograph shows bilateral abnormal elongated styloid process (arrows).

osteotomy (BSSO) to correct the facial asymmetry were planned to be carried out simultaneously. After thoroughly explanation of the the treatment plan, patient's consent was obtained.

Surgical procedure

The patient was placed in supine position and general anaesthesia was obtained via nasoendotracheal tube (NETT). One gram of Cefazolin was injected parenterally prior to the surgery. After styloid process was identified by localized palpation at tonsillar fossa, a 1 cm incision was made along the lateral edge of soft palate to the retromolar area (Figure 5). Oral mucosa, submucosa, and superior constrictor muscle were resected by blade number 15 and electrocauterized, following by blunt dissection through loose connective tissue was made by a small hemostat into parapharyngeal space. When the tip of styloid process was directly

visualized, stylohyoid, styloglossous, and stylopharygeous muscles were detached. A freer was used to catch the styloid process. This technique can also protect important neurovascular structures around that region such as, internal and external carotid artery, internal jugular vein, and ix, x, xi, xii cranial nerves (Figure 6). Rongeur forceps was used to resect the right elongated styloid process (Figure 7). The resected styloid process was 3.5 cm in length (Figure 8). Normal saline was used to irrigated the wound, which was then closed in two layers (superficial constrictor muscle and mucosa) before by resorbable suture. Following which Le Fort I osteotomy and BSSO were performed to correct the facial asymmetry as planned.

After the surgical procedure, the patient was admitted in a hospital for 4 days for observation of post-op complications such as upper airway obstruction, bleeding, pain, and infection and for supportive treatment. In the first follow-up visit (7 days post-op), pain at the right ear had disappeared. Mastication, talking, and changing a head posture did not exacerbate any discomfort. However, during the one and a half month follow-up visit, the patient complained of pain at the left ear region which was similar in nature to the pain she had at the



Figure 4 Lateral cephalometric image shows abnormal elongated styloid process (arrow).

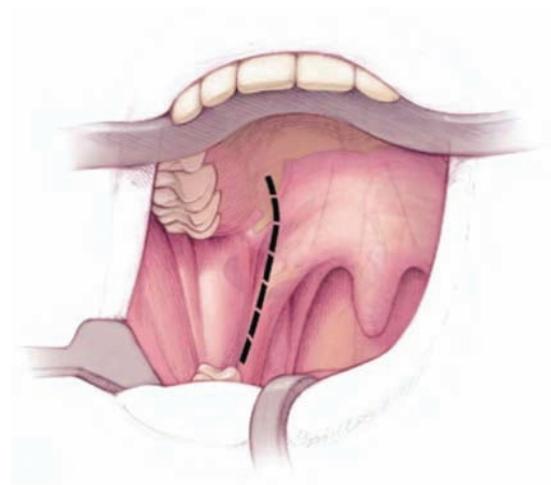


Figure 5 Incision outline for transoral incision.

right ear. Further treatment has been planned for the surgical resection of the left elongated styloid process.



Figure 6 Freer was used to hold the styloid process and it also protected the neurovascular bundles in pharyngeal space.

Discussion

Eagle's syndrome is a rare condition associated with elongated styloid process. Radiographic imaging technique best helps to confirm the diagnosis but it is of equal importance not to neglect any information during history taking.

In the literature, various surgical and non-surgical managements of Eagle's syndrome have been described. Severity of the symptoms, degree of elongation, and location are considered as important factors in establishing the most suitable treatment¹¹. A conservative treatment composes of administration of oral antidepressants, anticonvulsant, opioids, and non-steroidal anti-inflammatory drugs. Transpharyngeal injection of steroid or local anesthetic agents has been used. If pain disappears after injection of steroid or local anesthetic agents into tonsillar fossa, it is suggestive of an elongation of styloid process. Therefore transpharyngeal injection can also be used as diagnostic tools^{6,7,9,12}. Physical therapy and warm compressions are considered to relax muscle spasm that is caused from the irritation due to the elongated styloid process¹¹. A



Figure 7 Rongeur forceps was used to cut the elongated styloid process and an artery forceps was used to remove the elongated styloid process after resection.

conservative management is less invasive and is considered to be a good treatment option. However, it alone may not relieve persistent pain and a surgical management may be required^{11,14,19}.

Both intraoral and extraoral approaches can be used to gain access to the styloid process. Intraoral or transoral method was first described by Watt W. Eagle^{5,18,22} and the extraoral procedure was explained by Loeser and Cardwell²³. Intraoral approach is simple, easy and less time consuming. External scars can also be avoided but the visualization is limited with this approach and there are high risks associated with injury to the greater vessels, deep neck infections, and post-operative airway obstruction, especially when both sides are operated simultaneously. On the other hand, extraoral approach provides more access and visualization but external scars are difficult to avoid. Moreover, facial nerve tends to be injured and the operating time is longer. Rongeur forceps, rotary instruments and piezosurgery(R) can be used to shorten the styloid process¹⁸. In this case, the patient had medicated for pain relief and physiotherapy had been tried for 2 years but the symptoms were not relieved, therefore surgical management was indicated. An intraoral

approach was used to access the styloid process because of its ease and precision and the avoidance of external scars. According to Chrcanovic et al, intraoral approach is a safe procedure but should not be performed bilaterally to avoid upper airway obstruction. Therefore unilateral styloidectomy via intraoral approach is recommended²⁴. Many studies on transoral approach have given good results. Postoperatively, the patient have had a long pain-free period without recurrence of the symptoms. However, the surgical approach to styloid process should be considered as per individual but the surgeon's experience also plays a vital role in the treatment outcome. Although surgical management is the first choice of treatment, a failure rate of 20% has been reported due to entrapment of fibrous tissue and adjacent nerves or inadequate shortening of the styloid process^{13,22}.

In our patient, after resection of the right styloid process, pain at the right ear had disappeared. One and a half month later, the patient complained of pain on the left ear which may have been due to increased pressure and irritation during the rotation of the mandible to the right. Therefore, styloidectomy of the left styloid process has been planned for the future.



Figure 8 The resected styloid process specimen measured at 3.5 cm in length.

In conclusion, orofacial pain is a discomfort with many potential etiologies. Although rare, Eagle's syndrome can be one of the possible etiologies; therefore importance should be given on clinical and radiographic investigations as well as a thorough history of the illness so that a timely and correct diagnosis can be made such that the management can be initiated to treat the condition and the associated symptoms.

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