Bilateral elongated styloid process (Eagle’s syndrome) - a case report and short review

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ABSTRACT
Eagle’s syndrome refers to unilateral or bilateral elongation of the styloid process or calcified stylohyoid ligament, along with other symptoms, such as dysphagia, otalgia, tinnitus, and globus pharyngeus. A styloid process is considered elongated or abnormal when it exceeds a normal length of 25-27 mm. However, elongation of the ossified styloid ligament is insufficient for diagnosing Eagle’s syndrome. Instead, a constellation of symptoms in history, as mentioned above, and physical and radiographic examinations contribute to diagnosis. This disorder has long been a diagnostic challenge for clinicians. Hence, it is essential for dentists to have a superior knowledge of this relatively rare entity and its anatomic base. This report presents a case of classical Eagle’s syndrome in a 47-year-old woman with symptomatic bilateral elongation of the styloid process. The lengths of the right and left styloid processes were 60.5 mm and 70.74 mm, respectively, which is one of the longest ever reported.

Introduction
The styloid process (SP) is a slender bone projecting immediately below the ear. It extends downward and forward from the inferior aspect of the temporal bone posterior to the mastoid apex. The tip of the SP is attached to the ipsilateral lesser cornu of the hyoid bone via stylohyoid ligament. All of the aforementioned features constitute the stylohyoid chain. The entire chain is embryologically derived from four cartilages: hypohyale, tympanohyale, stylohyale, and ceratothale. The SP originates from the second branchial arch (1). The SP can sometimes elongate enough to cause symptoms owing to compression of surrounding the vital anatomical structures. In 1949, Eagle described homonymous syndrome, which is characterized by an ossified stylohyoid ligament or elongated SP (2).

The first case of elongated SP and ossification of the styloid ligament was described by Marchetti in 1652 (3). In early 1872, Wein Lechner reported the first surgically treated case of a symptomatic elongated SP (4). Later, in 1937, Eagle, an otorhinolaryngologist first described a syndrome characterized by the simultaneous occurrence of elongated SP and orofacial pain. He described two cases of pharyngeal irritation associated with SP (5).

The prevalence of elongated SP is approximately 4% of the total population (6,7), and most cases are asymptomatic. Women around 40-years of age experience more symptoms of stygalgia than men and women in other age groups (8). It presents a diagnostic challenge to the clinicians and often eludes for a long time. Hence, superior knowledge of this relatively rare entity is essential for all dentists.

This case report 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 60.5 mm and 70.74 mm, respectively, which is one of the longest ever reported.

Case report
A 47-year-old female patient reported to the outpatient department of oral medicine and radiology with chief complaints of pain in the throat region and below both ears for six months. History revealed that...
the pain is spontaneous in onset, continuous, dull aching type that radiates to the neck, aggravates on turning her neck to either side and swallowing, and does not relieve on taking medications. In addition, the patient had difficulty in swallowing, sensation of a foreign body stuck in her throat, ringing in the ears and noise on turning the neck to either side. The patient had previously consulted a physician and otorhinolaryngologist for the same complaint in the past three months, who advised her medications that gave no relief; hence, she was referred for further evaluation. Patients’ medical and dental histories were not significant.

A previous bilateral ear examination, pure tone, and impedance audiometry performed by an otorhinolaryngologist revealed no abnormalities. There was no facial asymmetry or abnormality in the temporomandibular joint or in the muscles of mastication on extraoral examination. Examinations of the nose, nasal cavity, and neck were normal, except that the patient had pain in the postauricular and submandibular regions and side of the neck on turning the neck to either side with tinnitus and popping sound. The oropharynx was normal with no other significant mucosal changes on intraoral examination. However, tenderness was observed bilaterally on palpation of the tonsillar fossa. Intraoral hard-tissue examination revealed no contributory findings. The present case was provisionally diagnosed to be a case of styalgia on correlating the patients’ chief complaint, history and clinical examination. Atypical facial pain was considered in the differential diagnosis. An orthopantomogram followed by cone beam computed tomography (CBCT) of the right and left styloid processes was advised. An Orthopantomogram showed generalized horizontal bone loss, a partially edentulous area in relation to 36, a radiopaque restoration in 46, and an abnormally elongated styloid process, bilaterally measuring approximately 53 mm on the right side and 72.1 mm on the left side (Figure 1). Whereas, it measured about 60.50 mm on the right side and 70.74 mm on the left side in the CBCT (Figure 2 and 3). 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. (9). Langlais et al. (10) type III calcification pattern (segmented type) was noted on either side with five and three 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. Routine blood investigations such as complete hemogram, serum calcium, total serum cholesterol and liver and kidney function tests, were within normal limits. Transoral styloidectomy was planned under general anaesthesia, and the same procedure was performed. The transoral approach was chosen because it avoids external incision and the resulting neck scarring. The styloid was palpated and an incision was made along the ascending border of the ramus, cutting through the mucosa and sub mucosa. Blunt dissection was performed using a curved hemostat down to the styloid, dissecting...
posteriorly, medially, and laterally to the medial pterygoid and the superior constrictor muscles. The styloid apparatus was exposed and the styloid process was resected with a bone rongeur. The muscles and mucosa were irrigated and closed with absorbable sutures. The advantages of this approach include shorter operating time and no cervical incision. The disadvantage of this procedure is poor surgical

Figure 2. (a) 3-D reconstructed view of the right styloid process, (b) Right styloid process measuring 60.50 mm.

Figure 3. (a) 3-D reconstructed view of the left styloid process, (b) Left styloid process measuring 70.74 mm.
exposure. The patient was prescribed antibiotics and analgesics for one week post-surgery. The patient was symptom free with a better quality of life during the 3-month follow-up and was advised to report back if any of the previous symptoms recurred.

**Discussion**

Eagle’s syndrome is a combination of symptoms caused by an elongated ossified SP. An SP longer than 25 mm is considered elongated, but the elongated SP is not only diagnostic for Eagle’s syndrome, because most patients with incidental findings of an elongated SP are asymptomatic (6,7). Eagle observed that mesial deviation of the SP leads to more severe symptoms. In addition, Eagle’s syndrome has a female predilection and is mostly diagnosed during the 3rd and 4th decade of life. Eagle’s syndrome has rarely been observed in young individuals. Eagle’s Syndrome has been found to be associated with both unilateral and bilateral elongated SP. Symptoms may include globus pharyngeus, sensation of a foreign body in the throat, hoarseness of voice, pain radiating to the ear, throat pain on swallowing or side movements of the neck and hypersalivation (11).

The etiopathogenesis of Eagle’s Syndrome is still believed to be uncertain. Various theories have been proposed, including (a) congenitally elongated SP due to perseverance of the stylohyoid cartilage, (b) idiopathic calcification of the stylohyoid ligament, and (c) ossification at the origin of the stylohyoid ligament (11).

At the prenatal stage, the stylohyoid complex has four segments (superior portion of the hyoid corpus, SP, lesser cornua of the hyoid, and stylohyoid ligament). These are all derivatives of Reichert’s cartilage (2nd branchial arch), which can be further divided into four parts based on the consequent development of the stylohyoid complex. Tympanohyal, being the first and most proximal segment, gives origin to the tympanic (proximal) segment of the SP, as well as the stapes. The second segment is called the stylohyal segment and gives rise to the distal portion of the SP. The third segment is ceratohyal and degenerates in utero, forming the stylohyoid ligament. The fourth and most distal segment is called the hypohyal segment and forms the lesser cornua of the hyoid. The stylohyoid process arises from the temporal bone immediately medial and anterior to the stylomastoid foramen, extends anteromedially, rarely shows any anatomical variations in its course, and is encircled on both sides by the internal carotid artery (ICA) and external carotid artery (ECA). The stylopharyngeus, styloglossus, and stylohyoid along with the two ligaments being stylohyoid ligament and stylomandibular ligament originate from the SP (6,7).

Stylocarotid artery syndrome is a rare condition caused by the compression of the internal or external carotid arteries by the styloid process. This vascular variation of Eagle syndrome can cause neurological symptoms, such as transient ischemic attack (TIA), syncope, and even stroke. Other rare symptoms include aphasia, visual abnormalities caused by the mechanical compression of the carotid artery, and irritation of the sympathetic plexus adjacent to the carotid artery. Symptoms may be triggered or aggravated by head and neck motion or rotation, respectively (12).

It was suggested that trauma due to surgery, chronic irritations at a localized site, endocrine disorders and trauma during development of the SP could lead to ossified hyperplasia of the SP. Okabe et al. (7) established a significant correlation between serum calcium concentration and SP length in 80-year-old subjects.

Various pathophysiological mechanisms proposed to explain the symptoms of Eagle’s syndrome are: Due to (a) Proliferation of granulation tissue secondary to traumatic fracture of the SP creating pressure on the surrounding structures (13); (b) compressing adjacent lying glossopharyngeal nerve or chorda tympani nerve; (c) Degeneration and inflammation in the tendinous segment of the SP, called insertion tendonitis or ‘pseudostylohyoid syndrome’; (d) irritation to the pharyngeal mucosa either through compression or by post-tonsill surgery scarring and (e) irritation of the sympathetic nerve plexus in the arterial sheath due to impingement of the arterial vessels (11).

Various types of SP mineralization result in symptomatic pathological conditions; in the literature, there are different classification systems used to describe SP mineralization (14). Langlais et al. classified SP into three types based on radiographic ossification patterns (I) elongated – uninterrupted mineralization of the SP; (II) pseudoarticulated – SP joined to the mineralized stylomandibular or stylohyoid ligament; and (III) segmented – segmented SP has multiple pseudo-articulations as a segmented mineralized stylohyoid complex. This pathological condition is described according to these authors by a SP length more than 32 mm (15).

O’Carroll and Jackson classified SP ossification into four types on the basis of the location of the ossified SP: (a) both sides higher than the mandibular foramen; (b) both sides on the same level as the
mandibular foramen; (c) both sides lower than the mandibular foramen; (d) unilateral or different lengths on the two different sides (16). In addition, Hardy et al. (14) found an unusual unilateral complete ossification of the stylohyoid apparatus that was always associated with laryngeal and vertebral calcifications.

Chuang et al. (14) published a rare case report of a patient who developed left hemispheric ischemia within 15 s of turning his head to the left, which completely vanished upon returning the head to the central position, with no long-term sequelae evident clinically or radiographically.

Camrada et al. (17) found that symptoms similar to stylohyoid syndrome can occur in non-elongated SP due to tendinosis at the junction of the lesser cornua of the hyoid bone and stylohyoid ligament; they termed this condition as ‘pseudostylohyoid syndrome’ (18). Bareiss et al. (19) reported a case of Eagle’s syndrome resulting from osteoradionecrosis of the SP following head and neck radiation therapy.

Usually, the initial diagnosis of an elongated SP is made radiographically and confirmed by palpation of the tonsillar fossa because non-elongated SP is not palpable (4). Moreover, palpation of the tip of the elongated SP exacerbates associated orofacial symptoms (5). After diagnostic imaging of Eagle’s syndrome and palpation of the tonsillar fossa, infiltration of a local anaesthetic agent allows for a final diagnosis.

Different radiological diagnostic criteria have been described in literature. The majority of authors describe a stylohyoid length of more than 25 mm as abnormal (8), whereas a few suggest a length of 40 mm (20). It is important to remember that the syndrome is not only dependent on the elongation of the SP, but may also be due to SP of normal length with a deviation of the tip (6,8). Only a combination of symptoms and abnormal elongation of the SP confirmed the syndrome. As Eagle’s syndrome is rarely suspected in clinical practice, the visualization of an elongated SP is often an incidental finding. Radiographic studies are usually advised to visualize other more frequent diseases.

Various imaging modalities are used for the diagnosis of Eagle’s syndrome, including Towne’s view or panoramic radiography, lateral head and neck or anteroposterior view, and lateral-oblique view. These conventional extraoral radiographs have various possible disadvantages, such as superimposition of osseous structures and inaccuracy in measuring the definitive length of the SP. These drawbacks are eliminated in CBCT imaging.

Clinically diagnosed cases of Eagle’s Syndrome should be directly advised with CBCT scans, without conventional radiographs as to decrease the time to confirm the diagnosis. CBCT should be performed using axial, coronal, and sagittal reconstruction images and volume-rendering reconstruction. CBCT allows the measurement and evaluation of SP length and angulation, as well as its relationship with nearby vital anatomical structures. Detailed information obtained from CBCT also helps in planning extra-oral or intraoral surgery for elongated SP.

The differential diagnoses which can be considered for Eagle’s syndrome are atypical cervicofacial pain, TMD’s, trigeminal or glossopharyngeal neuralgias, pharyngotonsillitis, otitis media, true pharyngeal foreign bodies, external otitis, mastoiditis, temporal arteritis, submandibular sialadenitis or sialolithiasis, and tumours of the floor of the mouth or pharynx (18,21). In the differential diagnosis, we can also consider histamine cephalgia, oesophageal diverticula, Sluder’s syndrome, cervical vertebral arthritis, hyoid bursitis, and cluster and migraine type headache (22).

**Conclusion**

Early diagnosis of both symptomatic and asymptomatic Eagle’s syndrome is very important, as any pressure due to a sharp elongated SP may be fatal due to its close proximity to the carotid artery. Eagle’s syndrome is a diagnostic dilemma for dentists in patients with complaint of atypical facial pain, therefore, adequate knowledge of eagle syndrome’s clinical signs and symptoms will help to diagnose such oro-facial pain in elderly patients.

**Informed consent**

The authors certify that we have obtained all appropriate patient consent forms from the patient for imaging and other clinical information to be reported in the journal.

**Disclosure statement**

No potential conflict of interest was reported by the author(s).

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