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Case Report

Eagle's syndrome –A case report

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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, clinically characterised by throat and neck pain, radiating into the ear. 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. In this report, we describe the case of a 71-year-old man who presented with long standing orofacial pain due to unilateral elongated styloid process on the right side.

1. Introduction

Eagle's syndrome, defined by Eagle in 1949, is characterised by elongation or ossification of the styloid process. The styloid process is a slender bone projecting from the lower surface of the petrous portion of the temporal bone. When the styloid process is longer than 30 mm, it is referred to as an elongated styloid process. This syndrome had been divided into two groups; "the classical styloid process syndrome" and "the stylo-carotid artery syndrome".[1] According to Watt Eagle, the typical presentation includes a foreign body sensation in the throat, dysphagia, otalgia, and throat pain.[2] The radiological findings are sometimes detected incidentally.[3]

2. Case Report

A 71 year old, male patient reported to the department of oral medicine and radiology with the complaint of pain below the angle of mandible on the right side and a feeling of discomfort while swallowing for the last 10 years. History of presenting illness revealed that the pain increased in intensity in the last 6 months and it radiated to the right side of the head and ear and aggravated on eating and swallowing. The patient had visited multiple hospitals in the past for the same but did not get any permanent solution. On extraoral examination, the patient had pain on the right side while opening the mouth widely and during side to side (lateral) movements of the neck. The patient had discomfort while swallowing and had a sensation of foreign body stuck in his throat. After thorough clinical examination, the patient was advised orthopantomogram along with computed tomography. The orthopantomogram revealed an elongated styloid process on the right side as compared to that on the left side and it was almost close to the angle of the mandible. Langlais et al. Type 1 elongation and completely calcified styloid process was noted on the right side.

The 3D CT revealed unilateral elongated styloid process on the right side measuring 35.9mm and it is slightly deviated towards the medial aspect.

Patient was further advised surgical management.

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Fig. 1: The OPG reveals the styloid process is elongated on the right side as compared to the left side and is almost close to the angle of the mandible.

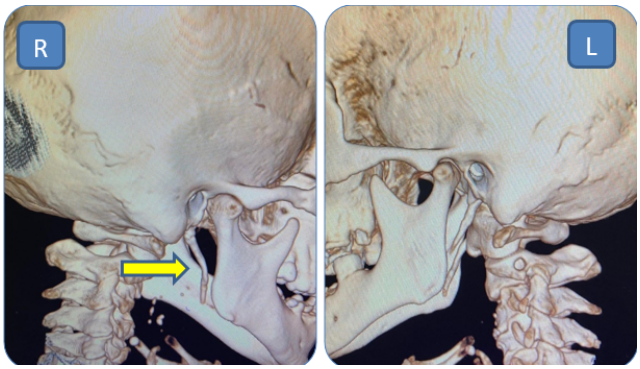

Fig. 2: 3D CT scan revealing elongated and medially deviating styloid process on the right side.

3. Discussion

The styloid process is a slender bone which projects immediately below the ear. It extends downward and forward from the inferior aspect of the temporal bone posterior to the mastoid apex. The stylohyoid ligament connects the tip of the styloid process to the ipsilateral lesser cornu of the hyoid bone.[4]

Eagle's Syndrome was first reported by otorhinolaryngology's, Watt W. Eagle in 1937 and is characterized by the simultaneous presence of elongated styloid process and/or face pain. It frequently occurs as an incidental finding when asymptomatic. The prevalence is about four percent of the population with most of them being asymptomatic and only four to ten percent of the patients experience the symptoms with mostly over 40 years of age. Eagle's syndrome is more commonly seen in females.[5]

Eagle's syndrome refers to a constellation of neuropathic and vascular occlusive symptoms caused by pathologic elongation or angulation of the styloid process and styloid chain, clinically characterized by throat and neck pain, radiating into the ear.[6] In most of the individuals, the normal length of the styloid process ranges between 20mm to 30 mm; when it exceeds 30mm, it is considered as

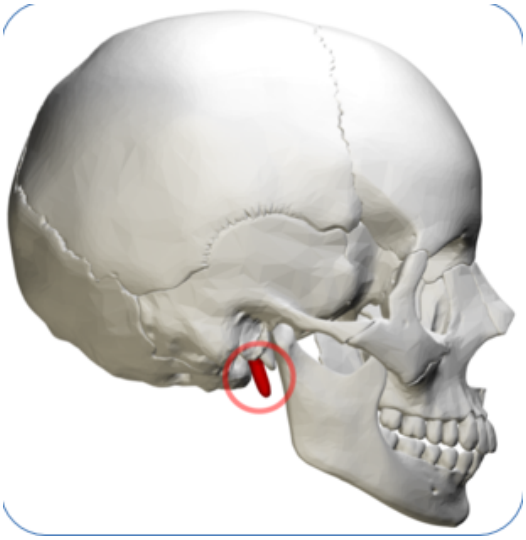

Fig. 3: The styloid process of the temporal bone projecting below the ear.

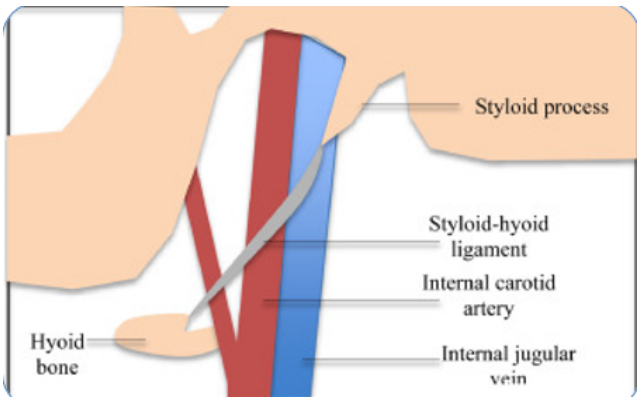

Fig. 4: The stylohyoid complex and its anatomic relation to neurovascular structures.

elongated.[5] Panwar et al (2022) reported a case of 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 60.5mm and 70.74mm,respectively, which is one of the longest ever reported.[4]

The etiopathogenesis of Eagle's Syndrome is still believed to be uncertain. But, various theories have been proposed, including congenitally elongated styloid process due to perseverance of the stylohyoid cartilage, idiopathic calcification of the stylohyoid ligament and ossification at the origin of the stylohyoid ligament.[4] It was suggested that surgical trauma, chronic irritations at a localized site, endocrine disorders and trauma during development of the styloid process could lead to ossified hyperplasia of the styloid process.[7]

Eagle's Syndrome has been found to be associated with both unilateral and bilateral elongated styloid process. The 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.

Eagle reported that medial deviation of the SP leads to increase in the severity of the symptoms.[4] The carotid artery subtype may be considered when the posteroanterior radiographs show extreme medial deviation.[8]

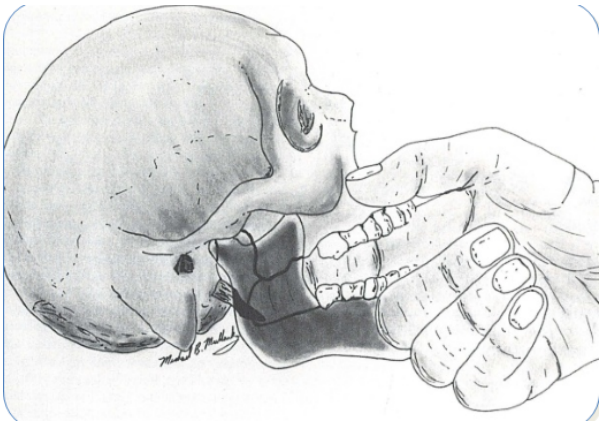

Fig. 5: Intraoral palpation of the elongated styloid process using index finger.

For a precise diagnosis, a thorough history and clinical examination are required. The elongated styloid process can be palpated intraorally and can provoke pain. A gentle pressure is exerted using the index finger over the tonsillar fossa to palpate the elongated styloid process.[8]

Eagle first identified two syndromes associated with elongation/ aberrant ossification of the stylo-hyoid apparatus. They are the classic Eagle syndrome and stylocarotid Eagle syndrome. "The classical styloid process syndrome" is most commonly caused due to post tonsillectomy fibrosis. It is characterized by spastic and nagging pain in the pharynx, radiating into the mastoid region. The pain is attributed due to stretching of the sensory nerve endings of the fifth, seventh, ninth, and tenth cranial nerves by elongated styloid process.[1] Stylocarotid artery syndrome is a rare condition which is caused by the compression of the internal or external carotid arteries by the elongated styloid process. This vascular variation of Eagle syndrome can cause neurological symptoms such as transient ischemic attack (TIA), syncope, and even stroke. The 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.[4]

There have been numerous pathophysiological mechanisms explaining the symptoms of Eagle's Syndrome - proliferation of granulation tissue secondary to traumatic fracture of the styloid process creating pressure on the surrounding structures, compression of the adjacent lying glossopharyngeal nerve or chorda tympani nerve, degeneration and inflammation in the tendonous segment of the styloid process, called insertion tendonitis or 'pseudostylohyoid syndrome', irritation to the pharyngeal mucosa either through compression or by post-tonsil surgery scarring and irritation of the sympathetic nerve plexus in the arterial sheath due to impingement of the arterial vessels.[4]

Any suspicion of Eagle's syndrome is further followed by radiographic examination. The basic imaging modalities frequently used are panoramic radiography, lateral head and neck, antero-posterior view, and lateral-oblique view.[8] Plain film radiograph may be a disadvantage due to superimposition of adjacent structures and inability to demonstrate the orientation and the dimension of the styloid process. Advanced imaging techniques are preferred. Three dimensional computed tomography represents the gold standard. It provides necessary information on the dimension, direction and anatomical relation between the styloid process and the adjacent neurological and vascular structures.[7]

A radiographic classification was proposed by Langlais et al. of the elongated and mineralized stylohyoid ligament complex. This classification included three types of abnormal radiographic appearances and four patterns of calcification/mineralization of the styloid process or the stylohyoid ligament.[8,9]

Table 1: Three types of elongation.

Characteristics	
Type I – elongated	Characterized by uninterrupted elongation (>25-28mm)
Type II – pseudoarticulated	Less frequent than elongation. The styloid process is joined to the mineralized stylomandibular or stylohyoid ligament by a pseudoarticulation, usually located superior to the inferior border of the mandible.
Type III – segmented	Consists of short or long non continuous portions of the styloid process or interrupted segments of mineralized ligament.

A variety of differential diagnoses can be considered for Eagle's syndrome such as atypical cervicofacial pain, TMD's, trigeminal or glossopharyngeal neuralgias, pharyngotonsillitis, otitis media, true pharyngeal foreign bodies, external otitis, mastoiditis, temporal arteritis, and unerupted or impacted teeth.[4]

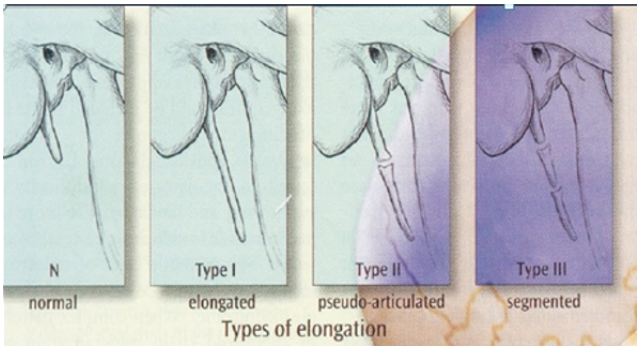

Fig. 6: Various types of elongation commonly observed in the stylohyoid complex.

Table 2: Patterns of calcification

Characteristics	
Calcified outline	Reminiscent of the radiographic appearance of a long bone with a thin radiopaque cortex and a central lucency that constitutes most of the process.
Partially calcified	Thicker radiopaque outline, with almost complete opacification as well as a small and occasionally discontinuous radiolucent core.
Nodular complex	This pattern has a scalloped outline and maybe partially or completely calcified with varying degrees of central lucency.
Completely calcified	This pattern is totally radiopaque with no evidence of a radiolucent inner core.

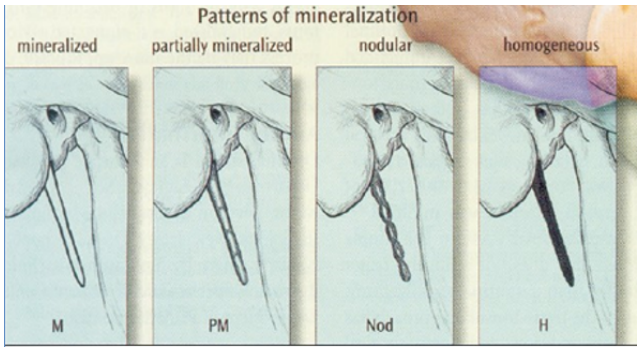

Fig. 7: Patterns of mineralization/calcification that are commonly observed in the stylohoid complex.

The complications of Eagle's syndrome includes sudden death, transient ischemic attack and syncope.[10] Eagle jugular syndrome which involves the internal jugular vein (IJV) compression caused by an elongated styloid process has been proposed as a venous variant of the Eagle syndrome. It has the potential to induce venous congestion due to the compression the IJV.[1] Eagle syndrome is a rare cause of stroke and can cause carotid dissection and consequent ischemic stroke.[2] The diagnosis of sudden death due to mechanical irritation of the carotid sinus by elongated styloid process was made as the sign of acute cardiovascular failure was present and upon exclusion of other causes of death.[11]

Successful and permanent management of Eagle's Syndrome is achieved only by surgical management which includes transoral styloid fracture or surgical shortening, carried out transorally or extraorally. However, some non surgical treatments are suggested if the patient is not willing for surgery. These include transpharyngeal infiltration of steroids and local anesthesia in the tonsillar fossa. Although, the transoral surgical approach has cosmetic benefits, it has disadvantages of deep cervical infection, poor visualization of surgical field which may increase the risk of neurovascular injury and post operative edema.To get over these drawbacks, endoscopic assisted transoral resection has been used recently.[7]

Early diagnosis of both symptomatic and asymptomatic Eagle's syndrome is very important, as any compression on the carotid artery due to elongated styloid process may be fatal. For dentists, identifying Eagle's syndrome in patients complaining of atypical facial pain presents a diagnostic challenge, therefore, adequate knowledge of clinical signs and symptoms of Eagle's Syndrome will help to diagnose such atypical oro-facial pain.[4]

4. Source of Funding

None.

5. Conflict of Interest

None.

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