



Eagle's Syndrome; Unilateral Elongated Styloid Process: A rare case report

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ABSTRACT

The styloid process is a sharp, slender, elongated pointed bone just below the ear. It extends downwards, forwards and medially. Normal length of styloid process is 2-3 cm. If it exceeds more than 3 cm it is called as elongated styloid process. Several variations in the length of styloid process have been reported. Sign and symptoms produce by elongated styloid process is known as Eagle's syndrome which is characterized by severe pain in the throat, side of the face and ear. This may be classified as an entrapment syndrome which deserves the attention of the neurosurgeons, ENT surgeons, orofacial surgeons and physicians. The aim of this case report was to familiarize neurosurgeons, ENT surgeons with Eagle's syndrome, its diagnosis, workup and management. In this case, we found an unusual case of long styloid process with ossification of stylohyoid ligament on the right side. Total length of the process from the base to the apex was 5.6 cm on the right side while on the left side it was normal in length i.e. 2.5 cm.

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Introduction

The styloid process is a sharp, slender, elongated pointed bone just below the ear. It projects downwards and forwards from the inferior surface of the temporal bone just in front of the stylo mastoid foramen. It extends downwards, forwards and medially. From its tip stylohyoid ligament passes downwards and forwards to the lesser cornu of the hyoid bone. It serves as an anchoring point for several muscles associated with the tongue and larynx. Its proximal part (tympanohyal) is ensheathed by the vaginal process of the tympanic portion. Its distal part (stylohyal) gives attachment to the two ligaments (stylohyoid and stylo mandibular) and three muscles (styloglossus, stylohyoid and stylopharyngeus). Internal jugular vein lies deep to the styloid process which is separated by stylopharyngeus muscle [1]. Normal length of styloid process is 2-3 cm [2]. If it exceeds more than 3 cm it is called as elongated styloid process. It might cause severe pain in the oropharynx, neck, and ear. It also causes difficulty in swallowing, foreign body sensation and other symptoms due to compression of carotid artery. Pietro Marchetti was the first person who described it in 1652. Later in 1937 Watt W. Eagle described the symptom complex of elongated styloid process which he called as Eagle's syndrome [3]. This syndrome is well known to both ENT surgeons [4,5&6] and oral surgeons [6,7] but the neurosurgical literature is surprisingly lacking information about it.

Case report

During routine dissection of a 55 year old male cadaver in the dissection hall of Jawaharlal Nehru Medical College Belgaum, we came across a long styloid process on the right side of the neck. The cadaver was dissected carefully on both side of the neck up to the tympanic part of temporal bone. The measurements were made with the help of a conventional measuring scale. The length of the right styloid process was 5.6 cm as inceptive point i.e. the inferior border of the tympanic bone to the tip. On the left side the styloid process was normal in length that was 2.5 cm. On the right side a flexure was present in the middle which could be presumably represents the site of

calcification between apex and ossified section of the stylohyoid ligament (Fig.1).

Discussion

Skler and Sandev proposed that the normal length of styloid process ranges from 0.5 cm to 5 cm [2]. Other authors concluded that the normal length of it ranges from 2.5 cm to 4.8 cm [8]. Similar studies done by Skozat et al [9] concluded that a length of more than 3 cm is considered to be elongated. According to the Gray's Anatomy the normal length of styloid process is 2 cm to 3 cm [1]. In our present case, we found the right sided styloid process is 5.4 cm long while that of left side is only 2.5 cm. Some rare cases exist with the length of 7.3 cm [1]. The rate of incidence of long styloid process seems to range from 1.4% to 84.4% of population which may be due to great variations in routine radiological investigation, surgical and anatomical study (cadaveric dissection and dry skull specimens study) [10]. Variation in the length and the predominance of unilateral to bilateral occurrence also depends on the ethnic group [11]. Sometimes a distinct border between apex and stylohyoid ligament is not clearly identifiable. Styloid process elongation can occur unilaterally or bilaterally. Some authors claimed that the unilateral phenomenon most common [12]. Others are contrary to these claims [13]. Clinical diagnosis of the eagle syndrome can be done by bimanual palpation of tonsillar fossa (normal styloid process is not normally palpable). Other diagnostic methods are panoramic radiography and CT scan. The latter provides additional information to plain radiographs.

An ossified stylohyoid ligament or long styloid process or both are not always produce symptoms. The symptomatic cases are known as Eagle's syndrome. The signs and symptoms varies from pain in the throat, dysphagia, otalgia, foreign body sensation, headache, pain in the neck during rotation of head, pain during tongue extension, facial and carotid pain [14,15]. Moreover it seems that there is co-existence of ossified stylohyoid ligament with other conditions such as cervical osteophytes, cervical spondylosis etc [14], anomalies in the part of the vertebral artery [16] and fracture of the ossified ligament.

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Cervical spondylosis yet very common in elderly population may have similar clinical signs with Eagle's syndrome, but could be differentiated by palpation of tonsillar fossa. Arterial anomalies should be cleared up because it is likely to co-exist with stylohyoid ossification. Very few cases have been reported with fracture of ossified ligaments in literature. Fracture of the elongated styloid process or ossified stylohyoid ligament can be caused spontaneously or traumatic or any other reasons. Violent manipulations by medical, paramedical or manual therapists and rehabilitation personnel around the neck may lead to fracture especially in asymptomatic person which may produce serious clinical consequences for the patient due to damage of surrounding structures. In present case we observed a flexure at the junction of middle and inferior 1/3 (arrow in the figure) could presumably represent the site of the union between the apex of the process and the ossified section of the stylohyoid ligament.



Figure 1: Left Styloid Process;
1. Normal left styloid process, 2. Internal carotid artery



Figure 2: Elongated Right Styloid Process; marking shows the elongated styloid Process having the length of 5.6 cm

Rate of bone growth is increased during childhood and adolescence period. After the age of 20 there is a rapid decrease in the process of ossification [17]. However, other worker support that an inconsistent trend exists toward greater ossification of the stylohyoid ligament with age advanced [18]. It has been suspected that an elongated styloid process could be due to congenital elongation of the styloid process and persistence of the cartilaginous analogue of the styloid, calcification of the stylohyoid ligament by unknown mechanism and growth of osseous tissue at the region of insertion of the stylohyoid ligament [8]. Sign and symptom of the Eagle's syndrome has various origins. Some worker has claimed that inflammation of styloid process can lead to granular tissue formation which releases pressure to nearby structures causing pain. Cranial

nerves such as glossopharyngeal, vagus and 3rd branch of trigeminal or chorda tympani can also be directly irritated by the styloid process and induce severe pain. More reasons for the symptoms include inflammation of tendons, pharyngeal mucosa excitation and impact of carotid bulb [18]. Appropriate choice of therapy like anti-inflammatory and corticosteroid drugs are advisable which depends on the intensity of pain or dysphagia and it can be conservative or invasive. If the sign and symptoms persist for longer time then excision of the styloid process could be helpful [18], but other researchers are not advised from for the surgical treatment of styloid syndrome [15].



Figure 3: Elongated right styloid process in Situ;
Arrow shows the junction between the styloid process proper and the ossified stylohyoid ligament 1. Styloid process, 2. Internal carotid artery 3. Glossopharyngeal Nerve, 4. Tympanic part of temporal bone

Conclusion

Sign and symptoms occurring in the region of ear, throat and neck could be due to elongated styloid process. It is very important to diagnose the asymptomatic case of elongated styloid process or ossification of stylohyoid ligament because fracture of it leads to more serious consequences by damaging nearby structures. So any undiagnosed and persistent pain in neck, face and ear could be due to elongated styloid process syndrome or of Eagle's Syndrome. This knowledge might be helpful for the physicians or/and surgeons during the diagnosis of persistent intermittent pain in the facial and neck region.

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