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Case Report

Eagle's syndrome with neck discomfort: A report of three cases ☆

Poppy Kristina Sasmita, PhD[a,b,*], *Herlina Uinarni, PhD*[a,c], *Liliana Sugiharto*[a]

[a] *Department of Anatomy, School of Medicine and Health Sciences, Atma Jaya Catholic University of Indonesia, Jl. Pluit Raya No. 2, Jakarta, 14440 Indonesia*
[b] *Neurologist at Mitra Keluarga Kelapa Gading Hospital, Jakarta, Indonesia*
[c] *Radiologist at Pantai Indah Kapuk Hospital, Jakarta, Indonesia*

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ABSTRACT

Eagle's syndrome, or ES, is associated with the elongation of the styloid process and partial or complete mineralization of the stylohyoid ligament. Clinically, the symptoms of ES are characterized by sore throat, neck pain radiating to the ear, dysphagia, and a foreign body sensation when swallowing, resulting from disruption of the neck or pharyngeal region. This report describes 3 male patients (40, 60, and 43 years old) with neck discomfort. These patients were inadvertently diagnosed with ES using multidetector computer tomography and 3-dimensional volumetric computed tomography (MDCT-3D CT). The length of the left styloid process in the first case was 42 mm. In the second case, the size of the right styloid process was 53 mm. In the last case, the length of the right styloid process was 41 mm, while the left side was 43 mm. This syndrome should always be suspected when pain is unilateral and unresponsive to analgesics, mainly in women. Diagnosis requires appropriate examination through radiological examination, special techniques, and experiences. We aim to present and re-emphasize the consideration of a differential diagnosis of ES for diagnosticians.

Introduction

In 1937, Eagle first discovered an elongated styloid [1]. Eagle's syndrome (ES) or styloid or stylohyoid syndromes are disorders caused by lengthening of the styloid process due to ossification of the styloid ligament. The symptoms depend on the structure surrounding the process, which is disturbed by the increased length of the process. As the symptoms are not specific, patients can visit an otolaryngologist, neurologist, dentist, or psychiatrist. Diagnosing ES is difficult, and ES is often overlooked by doctors and accidentally discovered. Conventional radiology studies, such as panoramic photographs, can help establish a diagnosis, but are sometimes unclear due to

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* Corresponding author.
E-mail address: poppy.kristina@atmajaya.ac.id (P.K. Sasmita).

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the overlap between anatomical structures. This case report describes the findings of ES using multidetector computer tomography and 3-dimensional volumetric computed tomography (MDCT-3D CT) with multiplanar reconstruction to assess the length and angulation of the styloid process. Moreover, we aim to present and re-emphasize for diagnosticians the consideration of a differential diagnosis of ES.

Case reports

Three cases with different complaints came to the surgery, neurology, and ENT clinics and were diagnosed with Eagle's syndrome after the MDCT-3D CT examination.

Case 1

A 40-year-old man complaining of hoarseness visited our surgical clinic for the first time. He also experienced pain when moving his head to the left and swallowing, with a mild-to-moderate pain scale and neck discomfort for the last year. The patient often visited a general practitioner for pain relief medication. The pain disappeared when the patient took medication. Within 2 weeks, the patient complained of hoarseness. On extraoral examination, pain was felt in the left posterior auricle and mastoid area. The surgeon recommended an assessment using a CT scan of the neck. A CT scan of the neck revealed linear calcifications originating from the left styloid process towards the hyoid bone below the angle of the mandible with a size of about 42 mm (Fig. 1). This structure clinically describes mineralization of the left styloid ligament. The size of the right side of the styloid process was approximately 23 mm, and there was no mass in the neck. The patient was administered nonsteroidal anti-inflammatory drugs (NSAIDs) to relieve pain, and no surgery was performed.

Case 2

A 60-year-old man visited a neurologist with frequent headaches and neck discomfort. The patient had a history of a cerebral hypervascular space-occupying lesion (SOL) on previous MRI. A neurologist suggested a CT brain angiography. On CT angiography, the SOL was not found, but there was an elongation of the styloid process on the right side with a size of 53 mm (Fig. 2). The neurologist prescribed only nonsteroidal anti-inflammatory drugs (NSAIDs) to treat headaches.

Case 3

A 43-year-old man visited the ENT clinic complaining of a cold, headache, and neck discomfort. The working diagnosis was sinusitis, and the doctor advised the patient to undergo a head CT Scan. On head CT, maxillary sinusitis was seen on the left side and an ossified styloid process, with a right-side length of 41 mm and a left-side length of 43 mm (Fig. 3). The ENT doctor prescribed antibiotics and analgesics. No surgery was performed on this patient.

Discussion

The styloid process is a cylindrical bony projection of the temporal bone from Reichert's cartilage of the second pharyngeal arch. The Reichert cartilage is divided into 4 parts: tympanohyal, stylohyal, ceratohyal, and hypohyal. The tympanohyal part, which develops antenatally, attaches to the petrous part of the temporal bone and forms the base of the styloid process. The stylohyal portion develops after birth, forming the trunk to the tip of the styloid process and the proximal portion of the stylohyoid ligament. The hypohyal and stylohyal regions

Fig. 1 – CT scan volume rendering (3D) of the neck showed an ossified styloid process extending from the base of the skull anterolaterally to the hyoid bone on the left side.

Fig. 2 – CT scan view of the neck showed the ossified styloid process extending from the base of the skull anterolaterally and caudally to the hyoid bone on the right side.

may not fuse or fuse after puberty. If these 2 parts unite, a long styloid process develops. If the stylohyal part does not undergo ossification, a short styloid process develops. The ceratohyal part and its fibrous sheath form the stylohyoid ligament, which extends to the lesser cornu of the hyoid bone. The hypohyal region forms the cornu minus the hyoid bone [2,3]. The styloid process is located anterior and inferior to the external auditory canal, anterior and medial to the mastoid process, and anterior to the stylomastoid foramen. Several vital structures such as the internal carotid artery, internal jugular vein, and cranial nerves X, XI, and XII are located medially around the styloid process.

The styloid process consists of proximal (tympanohyal) and distal (stylohyal) segments. The distal end is the origin of the stylohyoid, stylopharyngeus, and styloglossus muscles, and the base of the stylohyoid and stylomandibular ligaments. These 5 structures play roles in the movement of the tongue, pharynx, larynx, hyoid, and mandible [3,4]. The cartilage of the styloid process becomes ossified and elongated between the ages of 5 and 8 years. By the age of 30 years, elongation of the styloid process slows. The structure and length of the styloid process vary among individuals. The styloid process of the temporal bone varies in length from 2 to 3 cm. It is considered elongated if it exceeds 30 mm [1]. Several theories have explained the occurrence of stylohyoid and stylomandibular ligament ossification, including reactive metaplasia, dysplasia, trauma-induced ligament loss of elasticity, and tendon degeneration [5].

ES accounts for approximately 4% of the population and is more common in women [6,7]. According to Mathews' study of 100 subjects aged 20-80 years, the prevalence of styloid process elongation was 35% and was higher in males than in females, that is, 68.6%. Elongation was more in the 41-60-year-old age group with prevalence of 54.4% [8]. In a study by Vieira et al. [9] in the Brazilian population, the prevalence of styloid process elongation was 43.89% and was significantly higher in women (25.54 %) than in men (18.35 %).

The symptoms in patients with an elongated styloid process are atypical. Most symptoms are pain associated with restricted movement of the head or neck; pain in the temporomandibular joint, difficulty in swallowing, pain while swallowing or as a foreign body in the throat area, or pain while swallowing with or without radiation to the affected ear or mastoid. Other symptoms include tinnitus, ear pain, and stroke, secondary to carotid artery compression [10–13]. Differential diagnoses of ES include malocclusion, unerupted molars, denture implants, sphenopalatine neuralgia, temporomandibular arthritis, trigeminal neuralgia, chronic tonsillopharyngitis, cluster headache, cervical vertebral arthritis, migraine, pharyngitis, and mastoiditis [1,14,15].

The styloid process elongation rate in the East Aegean population was 7.01%, of which only 3.5% were symptomatic, and the most serious clinical complaint was headache, account-

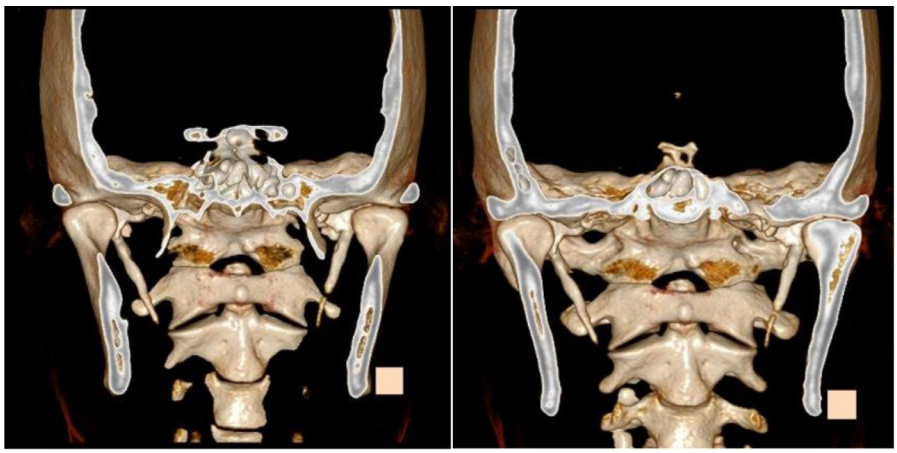

Fig. 3 – CT scans of the neck (coronal) showed the ossified styloid process extending from the base of the skull anterolaterally and caudally to the hyoid bone on both sides (left dominant).

ing for 18.7% [16]. According to Ceylan, several mechanisms of pain experienced by individuals with ES are as follows: 1) compression of neural structures such as the N. IX, inferior ramus of N. V, and chorda tympani; 2) fracture of the stylohyoid ligament ossification followed by proliferation of granulation tissue, which causes compression of the surrounding structures; 3) irritation of the sympathetic nervous system due to impingement of the carotid artery; 4) degenerative changes and inflammation of the tendon portion of the styloid process; 5) irritation of the pharyngeal mucosa due to compression; and 6) stretching and fibrosis involving the CN V, VII, IX, and X periods post-tonsillectomy [7].

The Langlais category was used to assess styloid system elongation in 1986. There are 3 types based on the degree of elongation: type 0 is a normal styloid process, type 1 is an elongation greater than 30 mm, type 2 is a pseudo-segmented styloid process, and type 3 is segmented, with a length of more than 30 mm. According to Langlais, there are 4 types of calcification patterns: 1) external calcification and not using a signal of calcification within the styloid process, 2) partial calcification with a radiopaque calcified segment within the styloid process, 3) nodular calcification with nodular radiopaque calcification inside the styloid process, and 4) complete homogenous calcification [8,13]. In a study by Mathew et al. [8], external calcification was the most common type of calcification (67%). In Ilgüy's study, type I (elongated) was the most common type, occurring on both sides (42/59 patients) [13]. A study conducted by Bagga et al. [5] on 2706 adults in Mathura, India, using panoramic radiographs showed that the highest bilateral elongation was 79.5% with a prevalence of type 1 and partial calcification >55%.

Three cases of Eagle's syndrome have been reported, 2 on one side and 1 bilaterally. According to the Langlais category, the degree of elongation of the styloid process in all cases reported in this study was type 1. In the first case, the length of the styloid process on the left side was 42 mm, and in the second case, the length of the styloid process on the right side was 53 mm. The third patient had bilateral styloid process elongation, with lengths of 41 mm on the right side and 43 mm on the left side. ES often occurs in women; however, all reported cases were found in men. These conditions were diagnosed accidentally. All 3 patients were treated with symptomatic therapy.

Conclusion

Eagle's syndrome is a rare syndrome with non-specific symptoms. The elongated styloid process should be considered when the clinician faces neck discomfort, headache, dysphagia, or temporomandibular joint pain. It can be challenging to diagnose in patients with head, neck, and facial pain. MDCT-3D CT is a valuable tool in the diagnosis of ES.

Patient consent

Patients had been informed that their cases would be published and had provided informed consent.

REFERENCES

[1] Chickooree D, Ram V. Eagle's syndrome-view from the general practitioners perspective. Clin Med Diagn 2014;4:9–13.
[2] Patil S, Ghosh S, Vasudeva N. Morphometric study of the styloid process of temporal bone. J Clin Diagn Res 2014;8(9):AC04–6.
[3] Piagkou M, Anagnostopoulou S, Kouladouros K, Piagkos G. Eagle's syndrome: a review of the literature. Clin Anat 2009;22(5):545–58.
[4] Vadgaonkar R, Murlimanju BV, Prabhu LV, Rai R, Pai MM, Tonse M, et al. Morphological study of styloid process of temporal bone and its clinical implications. Anat Cell Biol 2015;48(3):195–200.
[5] Bagga MB, Kumar CA, Yeluri G. Clinicoradiologic evaluation of styloid process calcification. Imaging Sci Dent 2012;42:155–61.
[6] Prasad KC, Kamath MP, Reddy KJ, Raju K, Agarwal S. Elongated styloid process (Eagle's syndrome): a clinical study. J Oral Maxillofac Surg 2002;60:171–5.
[7] Ceylan A, Köybasioglu A, Çelenk F, Yilmaz O, AU Uslu. Surgical treatment of elongated styloid process: experience of 61 cases. Skull Base 2008;18:289–95.
[8] Mathew AL, Cherian SA, BB Joseph. Syloid Process elongation and calcification pattern in a South Kerala population-radiographic aspects. J Ora Med 2017;1(1):11.
[9] Vieira EMM, Guides OA, De Morais S, De Musis CR, De Albuquerque PAA, Borges AH. Prevalence of elongated styloid process in a Central Brazilian Population. J Clin Diagn Res 2015;9(9):ZC90–2.
[10] Feldman VB. Eagle's syndrome: a case of symptomatic calcification of the stylohyoid ligaments. J Can Chiropr Assoc 2003;47:21–7.
[11] Galletta K, Granata F, Longo M, Alafaci C, De Ponte FS, Squillaci D, et al. An unusual internal carotid artery compression as a possible cause of Eagle syndrome – a novel hypothesis and an innovative surgical technique. Surg Neurol Int 2019;10:174.
[12] Gallaway E, Bayoumi S, Hammond D, Halsnad M. Case report: an atypical presentation of Eagle syndrome. JSCR 2017;8:1–3.
[13] Ilgüy M, Ilgüy D, Güler N, Bayirli G. Incidence of the type and calcification pattern in patients with elongated styloid process. J Int Med Res 2005;33:96–102.
[14] Savranlar A, Uzun L, Ugur MB, Özer T. Three-dimensional CT of Eagle's syndrome. Diagn Interv Radiol 2005;11:206–9.
[15] Yildiz A, Akmansoy BP, Pekiner FN. Eagle Syndrome: report of a case and a review of literature. J Dent Fac Atatürk Uni 2018;28(3):391–5.
[16] Asutay F, Erdem NF, Atalay Y, Acar AH, Asutay H. Prevalence of elongated styloid process and Eagle Syndrome in East Eagean population. Bezmialem Sci 2019;7(1):28–32.