Aesculapius Journal (Health Sciences & Medicine)

Volume 5 Article 6

April 2024

Bilateral Eagle Syndrome: A Surprising Cause of Neck Pain

Kailyn M. Mutsch
University of South Dakota, Kailyn.Mutsch@coyotes.usd.edu

Alla Zamulko
University of South Dakota, alla.zamulko@usd.edu

Introduction

Eagle syndrome was first described by Watt W. Eagle in 1937 where a correlation between pain and abnormalities in stylohyoid complexes was noted [1,2]. Eagle syndrome is a rare condition caused by elongation or deformity of the styloid process, which can disrupt neighboring structures leading to a constellation of symptoms [2]. Eagle syndrome may be referred to as stylohyoid syndrome, styloid syndrome, or styloid-carotid artery syndrome in some instances [3,4,5].

The normal styloid process is ~25mm in length, any process longer than 25mm may be considered elongated (Figure 1) [3,6]. Styloid process elongation can occur bilaterally or unilaterally [7,8]. An elongated styloid process is an incidental finding in approximately 4% of the population, but the incidence of Eagle syndrome is rare at around .16% [2,9]. Symptomatic patients are primarily greater than 30 years of age, with a female-to-male predominance of 3:1 [2].

We present a case that highlights the rarity of Eagle syndrome. The complex nature of diagnosis and management demonstrates the imperative need for providers to be familiar with Eagle syndrome and utilize multidisciplinary teams. Although rare, Eagle syndrome should be considered in the differential diagnosis in patients with chronic refractory orofacial and cervical pain.

Case Report

A 45-year-old female presented to an internal medicine clinic complaining of primarily left-sided chronic neck and thoracic back pain. She reported persistent dull pain exacerbated by movement of head and neck region present for greater than 2 years. Additionally, she experienced a globus sensation in the throat accompanied by dysphagia and intermittent choking which was unrelated to the consistency of food. Patient has a medical history of hypothyroidism and neurofibromatosis type II.

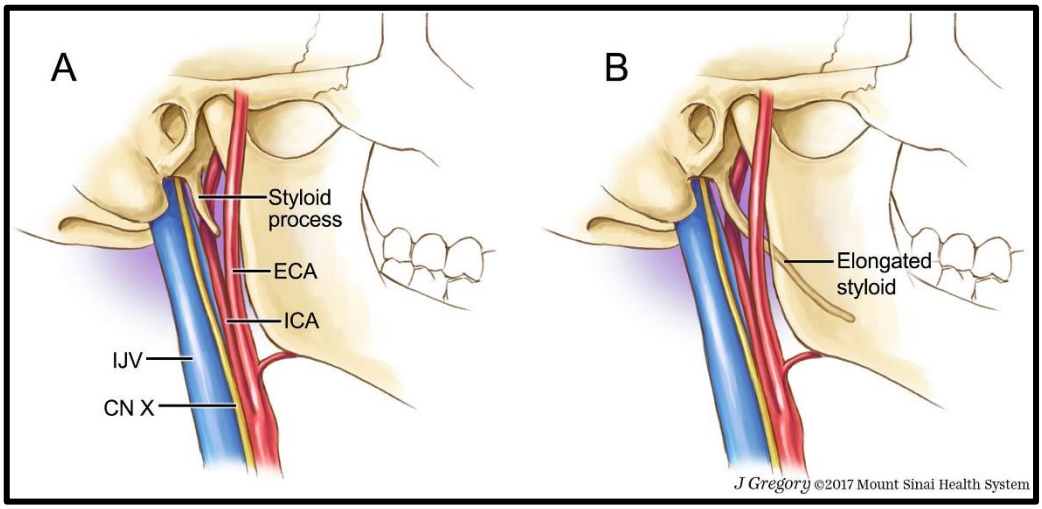

Figure 1- A. demonstrates the anatomic location of the styloid process B. illustration of an elongated styloid process.
Used with permission from © Mount Sinai Health System
Illustration by Jill K. Gregory

Her pain had been present for several years, but had increased in severity in recent months, prompting her to be seen. Approximately one month prior to this visit, she had been seen in the ER for severe left-sided neck pain as well as a migraine. A computerized tomography (CT) obtained at that time, from an outside facility, demonstrated symmetrical ossification of the bilateral styloid ligaments, with the styloid process extending, at least 50 mm in length, to the level of the hyoid bone. We were unable to obtain the imaging but received the official report.

Clinical examination showed intact cranial nerves. There was mild tenderness to level 2 region of the neck as well as multiple tender points in joint and axilla consistent with fibromyalgia. The patient was referred to Ear Nose and Throat (ENT) Surgery for further evaluation as well as referred to physical therapy for additional pain management.

Subsequent flexible fiberoptic examination of the nasopharynx and larynx performed by ENT demonstrated normal soft palate motion, symmetric base of tongue, no arytenoid edema, and normal true vocal cord motion with no pooling of secretions. Based on the clinical findings and the prior radiographic evidence of elongated styloid processes, the patient was diagnosed with bilateral Eagle syndrome.

Surgical treatment was recommended through a cervical approach opposed to a transoral approach due to the length of styloid processes. The plan was to surgically remove the left styloid process and if complete symptom resolution was achieved, removal of the right styloid process would be completed at an additional visit. The patient has not undergone surgical treatment to date.

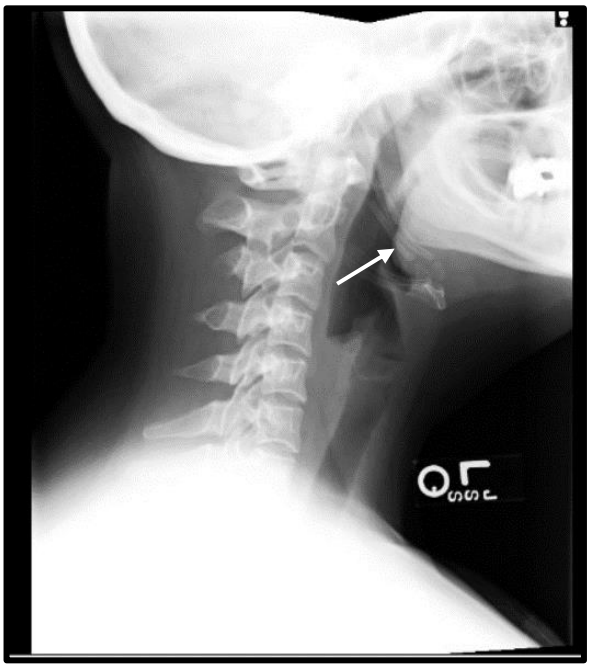

Figure 3- Xray of the soft tissue of the neck completed in 2012 showing both calcified stylohyoid ligaments come down to level of the hyoid bone (arrow).

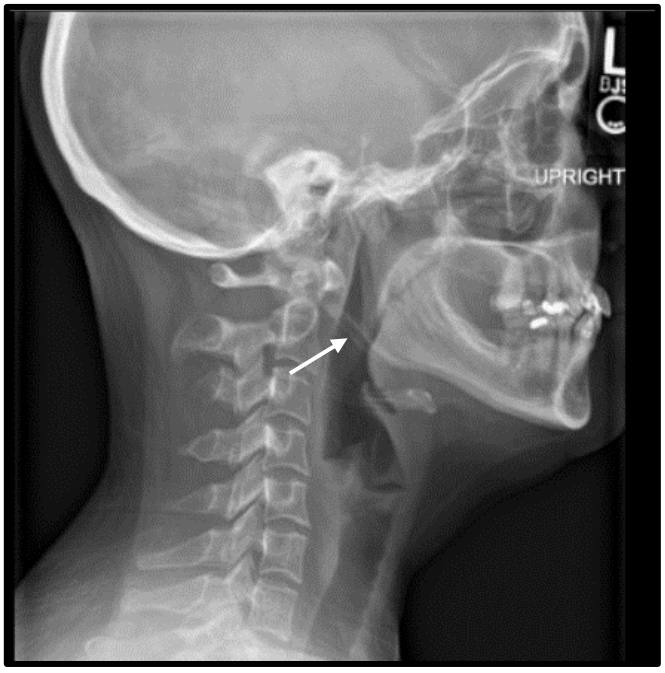

Figure 4- Xray of the cervical neck completed in 2022 demonstrating further documentation of the elongated styloid processes bilaterally (arrow).

Discussion

Embryologically, the development of stylohyoid complex begins from Reichert's cartilage of the second branchial arch [6,10,11]. As embryogenesis progresses, the second branchial arch separates into four segments: the tympano-hyal, stylo-hyal, cerato-hyal, and hypo-hyal. The tympano-hyal and stylo-hyal segments develop the proximal and distal portions of the styloid process respectively. The cerato-hyal portion deteriorates in utero becoming the stylohyoid ligament, while the hypohyal gives rise to a portion of the hyoid bone [3,10,11]. Many important anatomic structures are closely located to the styloid process and the stylohyoid ligament including a variety of nerves, arteries, and veins.

Eagle syndrome was first thought to be caused by reactive hyperplasia or ossification of the stylohyoid ligament complex occurring after tonsillectomy; however, it has been found to occur without surgical trauma [2,3,6]. Other proposed mechanisms include retained embryologic cartilage tissue, congenital elongation of the stylohyoid complex through calcification or osseous tissue, an anatomic variant, or changes associated with aging leading to shortening of the cervical spine and alteration of the styloid process angulation [3,6,11,12]. Although the underlying etiology of Eagle syndrome remains unknown, there is better understanding for the pathogenesis of symptoms.

The constellation of symptoms referred to as "classic Eagle syndrome," most often occur in patients with pharyngeal trauma or tonsillectomies. It is a consequence of stretching or compression of the nerves or nerve sheath in the location of the fifth, seventh, eighth, ninth, and tenth cranial nerves. Most commonly the ninth cranial nerve, glossopharyngeal nerve, is affected correlating to the symptoms of throat and neck pain aggravated by rotation of the head. Other symptoms noted are globus sensation of the throat, dysphagia, otalgia, voice changes, and tinnitus [2,3,9,13]. Another constellation of symptoms can be attributed to vascular Eagle syndrome, or stylocarotid syndrome. This is a consequence of compression of the internal or external carotid arteries or the sympathetic nerves travelling with the arteries on rotation of the head. This can lead to pain in the artery distribution, headaches, transient ischemic attacks, vertigo, and syncope [2,3,9,13]. Due to the fact that these symptoms may mimic many orofacial and cervical disorders, a thorough clinical history, examination, and radiological assessment are necessary to obtain a diagnosis.

The diagnosis of Eagle syndrome is made through correlation of clinical findings with confirmatory imaging. Physical examination can often lead to identification of the calcified or elongated styloid process or reproduction of symptoms. The diagnosis of the elongated styloid process is then confirmed by imaging. Several imaging techniques have been employed, but the gold standard for diagnosis is 3-D CT reconstruction [2,3,10]. This imaging modality allows for precise measurement of the length of the styloid process and demonstrates optimal anatomical involvement. Orthopantomogram (OPG) and plain radiographs can also both be used to assess the styloid process [2,6]. In case of ischemic symptoms, angiographic examination should be obtained [2,6].

Langalis classification Criteria	
Type one	An uninterrupted, elongated styloid process
Type two	A styloid process apparently joined to the stylohyoid ligament by a single pseudoarticulation
Type three	Consists of interrupted segments of mineralized ligament creating the appearance of multiple pseudoarticulations within the ligament
O'Carroll and Jackson classification criteria	
Type one	Higher than the mandibular foramen bilaterally
Type two	On the same level as mandibular foramen bilaterally
Type three	Lower than mandibular foramen bilaterally
Type four	Unilateral or having asymmetric lengths on either side

Table 1- Table demonstrating common radiographic classification criteria for Eagle syndrome.

Eagle syndrome may be classified into three subcategories depending on radiographic appearance per the Langalis classification criteria [14]. An additional classification criterion, proposed by O'Carroll and Jackson, categorizes the location of ossification of styloid processes into four subcategories [10]. (Table 1)

Eagle syndrome may be managed conservatively or with a surgical approach. Conservative treatments include analgesics, antidepressants, anticonvulsants, transpharyngeal steroid injections, nonsteroidal anti-inflammatory drugs, and the application of topical heat [6,11]. Surgical management may be considered if a patient fails conservative management or if the benefits outweigh the risks of surgical intervention. Surgical management has been found to be most effective and may be done through a variety of approaches [2,13]. A transoral approach to shortening of the styloid process may be done, which poses the advantages of reduced operation time, use of local anesthesia, and the absence of a visible external scar. This approach may not be advantageous if the length of styloid process cannot allow for proper access or visualization of surgical field. In this case, a cervical approach was opted for. This approach allows for enhanced exposure of the styloid process and adjacent structures but poses a risk of injury to the facial nerve and its branches [2,3].

Conclusion

Eagle syndrome is a rare condition that may be complex to diagnose due to the nonspecific nature of the symptoms. It is best managed with involvement from multiple specialists with optimal team relations. The imperative need for physicians to be aware of this condition to allow for effective diagnosis and management due to the wide range of symptom presentation is clear. Diagnosis of Eagle syndrome is made through a combination of clinical findings and confirmatory imaging. 3D CT scan is the gold standard imaging modality for diagnosis.

References

1. Eagle WW. Elongated styloid processes: report of two cases. *Archives of Otolaryngology-head & Neck Surgery*. 1937;25(5):584-587. doi:https://doi.org/10.1001/archotol.1937.00650010656008
2. Bokhari MR, Graham C, Mohseni M. *Eagle Syndrome*. StatPearls. 2021. https://www.ncbi.nlm.nih.gov/books/NBK430789/
3. Abuhaimed AK, Alvarez R, Menezes RG. *Anatomy, Head and Neck, Styloid Process*. StatPearls. 2023. https://www.ncbi.nlm.nih.gov/books/NBK540975
4. Zhang FL, Zhou H, Guo Z, Yang Y. Eagle syndrome as a cause of cerebral venous sinus thrombosis. *Canadian Journal of Neurological Sciences*. 2019;46(3):344-345. doi:https://doi.org/10.1017/cjn.2019.17
5. Qureshi S, Farooq MU, Gorelick PB. Ischemic stroke secondary to stylocarotid variant of eagle syndrome. *The Neurohospitalist.* 2019;9(2):105-108. doi:https://doi.org/10.1177/1941874418797763
6. Badhey A, Jategaonkar A, Anglin Kovacs AJ, et al. Eagle syndrome: a comprehensive review. *Clinical Neurology and Neurosurgery*. 2017;159:34-38. doi:https://doi.org/10.1016/j.clineuro.2017.04.021
7. Péus D, Kollias SS, Huber AM, Huber GF. Recurrent unilateral peripheral facial palsy in a patient with an enlarged styloid process. *Head & Neck*. 2018;41(3):E34-E37. doi:https://doi.org/10.1002/hed.25384
8. Montevecchi F, Caranti A, Cammaroto G, Meccariello G, Vicini C. Transoral robotic surgery (TORS) for bilateral eagle syndrome. *ORL*. 2019;81(9): 36-40. doi:https://doi.org/10.1159/000493736
9. Johnson GM, Rosdy NM, Horton SJ. Manual therapy assessment findings in patients diagnosed with eagle's syndrome: A case series. *Manual Therapy*. 2011;16(2):199-202. doi:https://doi.org/10.1016/j.math.2010.09.007
10. Scavone G, Caltabiano D, Raciti M, et al. Eagle's syndrome: a case report and CT pictorial review. *Radiology Case Reports.* 2019; 14(2):141–145. doi:https://doi.org/10.1016/j.radcr.2018.10.008.
11. Piagkou M, Anagnostopoulou S, Kouladouros K, Piagkos G. Eagle's syndrome: A review of the literature. *Clinical Anatomy*. 2009;22(5):545-558. doi:https://doi.org/10.1002/ca.20804
12. D Boedts. Styloid process syndrome or stylohyoid syndrome? *Acta Otorhinolaryngological Belgica.* 1978;32(3):273-278.
13. Politi M, Toro C, Giulia Tenani. A rare cause for cervical pain: eagle's syndrome. *International Journal of Dentistry*. 2009;2009:1-3. doi:https://doi.org/10.1155/2009/781297
14. Langlais RP, Miles DA, Van Dis ML. Elongated and mineralized stylohyoid ligament complex: A proposed classification and report of a case of Eagle's syndrome. *Oral Surgery, Oral Medicine, Oral Pathology*. 1986;61(5):527-532. doi:https://doi.org/10.1016/0030-4220(86)90400-7