Clinical Neurology and Neurosurgery 159 (2017) 34–38

Contents lists available at ScienceDirect

Clinical Neurology and Neurosurgery

journal homepage: www.elsevier.com/locate/clineuro

Review Article

Eagle syndrome: A comprehensive review

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ARTICLE INFO

Keywords:
Eagle syndrome
Management
Styloid process

ABSTRACT

The objective of this report is to summarize the symptoms, diagnostic workup, necessary imaging, and management of Eagle syndrome. A comprehensive literature review was conducted on peer-reviewed publications of Eagle syndrome across multiple disciplines in order to gain a thorough understanding of the presentation, diagnosis, and management of this disorder. Diagnoses of Eagle Syndrome have increased, in part due to the awareness of physicians to patient symptomatology. While cervical pain and dysphagia are among the typical symptoms, patients can present with a wide spectrum of benign and dangerous symptoms. CT scan is the gold standard for diagnosis and can be aided by both 3D reconstructive imaging and Angiography. Treatment strategies include medical management (analgesics, corticosteroids, antidepressants, and anticonvulsants) and varied surgical approaches (extraoral, transoral, endoscopic assisted). Increased understanding by providers treating patients with Eagle Syndrome allows for a more comprehensive treatment plan. With a variety of medical regimens and more definitive surgical approaches, Eagle Syndrome can be treated safely and effectively.

1. Introduction

Eagle Syndrome is a rare and poorly understood clinical condition that presents with a myriad of symptoms that typically include pain in the anterolateral neck. These symptoms are associated with an abnormal styloid process. Fig. 1 shows a schematic of normal anatomy along with an elongated styloid process. Eagle initially described a pain syndrome associated with an elongated styloid process in 1937 as "stylalgia" [1]. Eagle subsequently expanded on his initial descriptions [2–6]. Historically stylohyoid pain syndromes have been delineated based upon their etiology, i.e. acquired versus congenital. Eagle syndrome proper has been described as a pain syndrome associated with an elongated styloid. The congenital variant, often described as stylohyoid syndrome has been described as a syndrome with pain and symptoms of carotid compression (presyncope, syncope, and even transient ischemic events) caused by an ossified stylohyoid ligament. Subsequent studies have expanded Eagle syndrome to include a myriad of symptoms beyond pain. Furthermore the literature has also blurred the demarcation between the acquired and congenital syndromes.

Eagle syndrome is an important clinical condition for the otolaryngologist to recognize. This is due to the variety of presentations, potentially serious complications, and the fact that Eagle syndrome is often amenable to treatment. In this paper we present an updated review of the literature regarding Eagle/stylohyoid syndrome and discuss current/emerging treatment modalities.

2. Methods

For this study we aimed to identify all full-text, peer-reviewed publications pertaining to Eagle Syndrome in otolaryngology or head and neck surgery. The searches were conducted in the Ovid MEDLINE, Google Scholar, PubMed, and NCBI databases there were no regional restrictions. However only English language results were reviewed. The following search terms were used: "Eagle Syndrome", "styloid syndrome", "stylocarotid syndrome", "stylohyoid syndrome", "styloid-carotid artery syndrome", and "elongated styloid syndrome". The results were combined with the terms "otolaryngology" or "head and neck" to retrieve articles. All such articles were reviewed. Finally the works cited by each of these papers were reviewed In order to obtain a more complete and expansive collection of relevant literature.

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http://dx.doi.org/10.1016/j.clineuro.2017.04.021
Received 31 January 2017; Received in revised form 26 April 2017; Accepted 27 April 2017
Available online 06 May 2017
0303-8467/

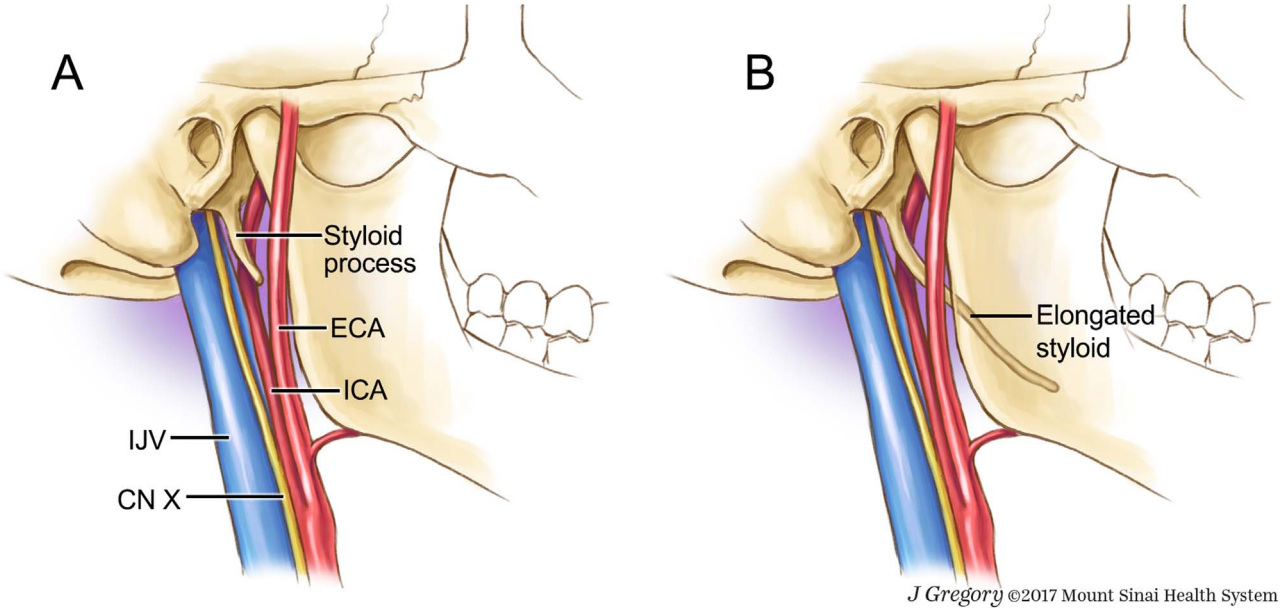

Fig. 1. Normal anatomy of the styloid process (Fig. 1a). Elongated styloid process as found in patients with Eagle Syndrome (Fig. 1b). (We would like to acknowledge Jill Gregory, senior illustrator, from the Mount Sinai Health System for providing this artwork. This artwork has been used with permission from Mount Sinai Health System).

3. Background

Abnormalities in the stylohyoid complex were first identified in animals by Vesalius in 1543 [7]. The first description in humans was published by Marchetti in 1656 [8]. Watt Eagle first described the combination of pain associated with an abnormal stylohyoid complex in 1937 and later reported a case series of over 200 patients [1,5]. Eagle found that about 4% of patients that have abnormalities with their stylohyoid complex have pain [5]. He described two different presentations the classic type and the carotid artery syndrome. At the end of his studies he concluded that tonsillectomy was a potential cause. Further studies have examined the relationship between tonsillectomy and Eagle syndrome and have not found the same relationship [9]. Prior to beginning a detailed discussion of Eagle Syndrome, a brief illustrative case will be presented to provide context for the ensuing report.

4. Example case illustration

A 56-year old female presented to the clinic with a 2-year history of right-sided throat pain and globus sensation. The patient denied dysphagia, weight loss, or other associated symptoms. The patient had tried several courses of anti-inflammatory medication and antibiotics without sufficient relief. A complete head and neck examination including fiberoptic laryngoscopy was performed, and the only pertinent finding was pain in the right neck with deep palpation. Intraoral palpation of the right retromolar trigone and right peritonsillar region also elicited pain. A computed tomography scan of the neck was obtained as displayed in Figs. 2 and 3. Based on the radiographic analysis, the diagnosis of Eagle Syndrome was made. As the patient had failed medical management, surgical intervention was offered and the patient was keen on pursuing this option. The patient underwent a tonsillectomy sparing transoral resection of the styloid process with primary closure, and had immediate resolution of symptoms following awakening in the recovery unit. She was discharged the same day and had an uneventful post-operative course with only minimal pain medication requirements and quick resolution of regular oral intake.

5. Epidemiology

There have been many studies since the identification of Eagle Syndrome that have sought to determine the incidence and prevalence

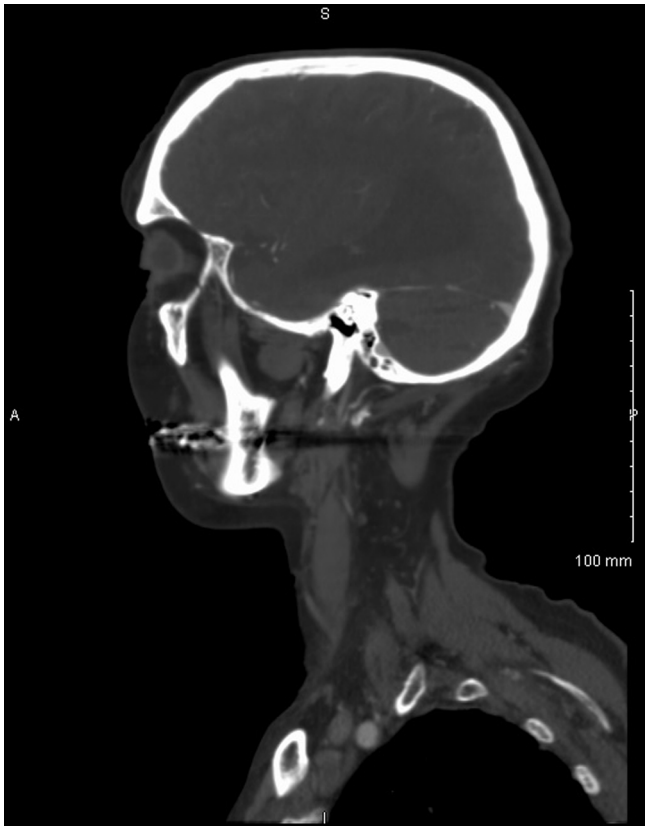

Fig. 2. A sagittal CT scan of a patient with an elongated styloid hyoid complex (We would like to acknowledge Dr. Roy Holliday from Mount Sinai Health System for providing these images).

of this condition. The variability in determining the epidemiology of this condition is likely due to differences in the diagnostic criteria in radiologic imaging. Some have suggested that the accepted length of the normal styloid process is approximately 2.5 cm, with 3 cm regarded as the upper limit of normal [10,11]. Radiologically, some studies have used stylohyoid length greater than 2.5 cm as abnormal [10]. Other studies have defined length greater than 4.0 cm as abnormal as this