Eagle syndrome – from symptom to diagnosis Case report

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Abstract

Introduction: Eagle syndrome is a rare condition caused by elongation of the styloid process or ossification of the stylohyoid ligament. It's named after Watt W. Eagle an otolaryngologist at Duke university, who described the first case in 1937. In the clinical presentation, we found different signs of syndrome – dysphagia, odynophagia, otalgia, foreign body sensation, facial pain, trismus, headache, increased salivation, and/or voice changes. The diagnosis of Eagle's syndrome is based on an optimal medical history and physical examination. The most accurate imaging technique is the CT- scan. 3-D CT reconstruction of the neck specify the size of length of the styloid process (> 3 mm) and the ossified stylohyoid ligament. The treatment includes conservative and surgical approach.

Material and methods: We present a 65- years old female, in a good health condition, with the following complains: difficulty swallowing, foreign body sensation in the throat. The patient states that she had palpated cartilage in the right side in the area of the tonsillar fossa.

The physical examination revealed no abnormal findings, but palpable right styloid process. We performed CT and 3-D CT reconstruction, which showed an elongation of styloid process on right side.

Results: The patient refused the surgical treatment. The conservative therapy includes the nonsteroidal anti- inflammatory medications.

Conclusion: Eagle's syndrome is a rare condition with vast differential diagnosis, vague symptomatology and ambiguous incidence and etiology. When we suggest the syndrome, the CT scan is the imaging method of choice for diagnosis. It shows the measurement of styloid process, the position in the neurovascular complex and the prepositions of damages. The treatment can be surgical or non-surgical depend on the size, position and surgical risk of the approach.

Key words: Eagle' s syndrome, elongation of styloid process, dysphagia

Introduction

Styloid complex consists of styloid process, styloid ligament and stylomandibular ligament. Styloid process and ligament are derived from first and second branchial arches and Reichert's cartilage. During fetal development, Reichert's cartilage links to styloid bone to the hyoid bone. Styloid process is derived from the temporal bone just before the stylomastoid foramen. Apex of styloid process is clinically important as it is placed between external and internal carotid arteries.

Abnormalities in the stylohyoid complex were first identified in animals by Vesalius in 1543. The first description in humans was published by Marchetti in 1656. 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. have abnormalities with their stylohyoid complex have pain. He described two different presentations the classic type and the carotid artery syndrome. 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.

Normal length of styloid process in adults varies between 20–30 millimeters (fig. 1). Styloid process

longer than 3mm are named elongated. Incidence of elongation of styloid process is around 4-7% and only 4% show the symptoms. In the event that elongation of styloid process caused neck and cervicofacial pain, dysphagia, odynophagia, foreign body sensation, it is described as Eagle syndrome. Elongation of styloid process is diagnosed via physical examination and CT scan and/ or 3D- CT reconstruction

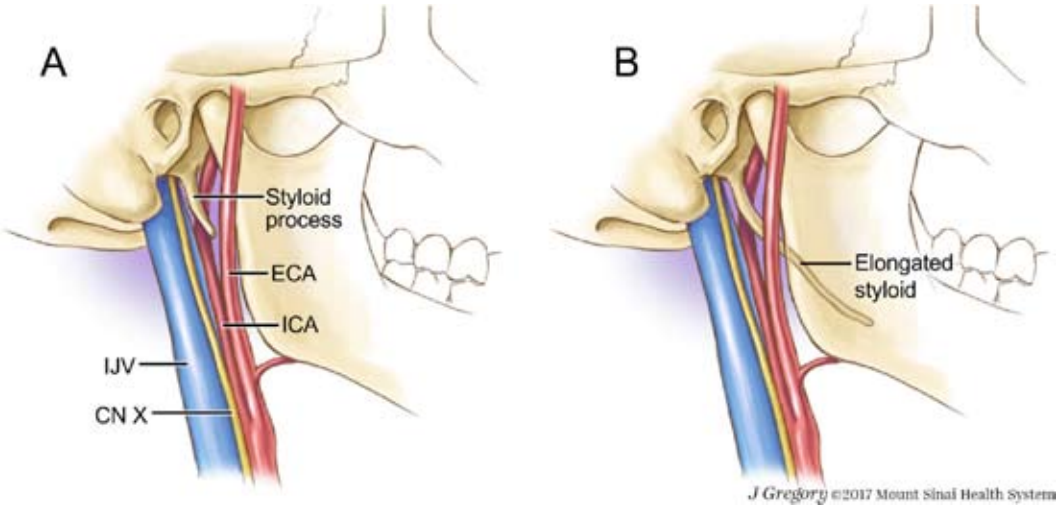


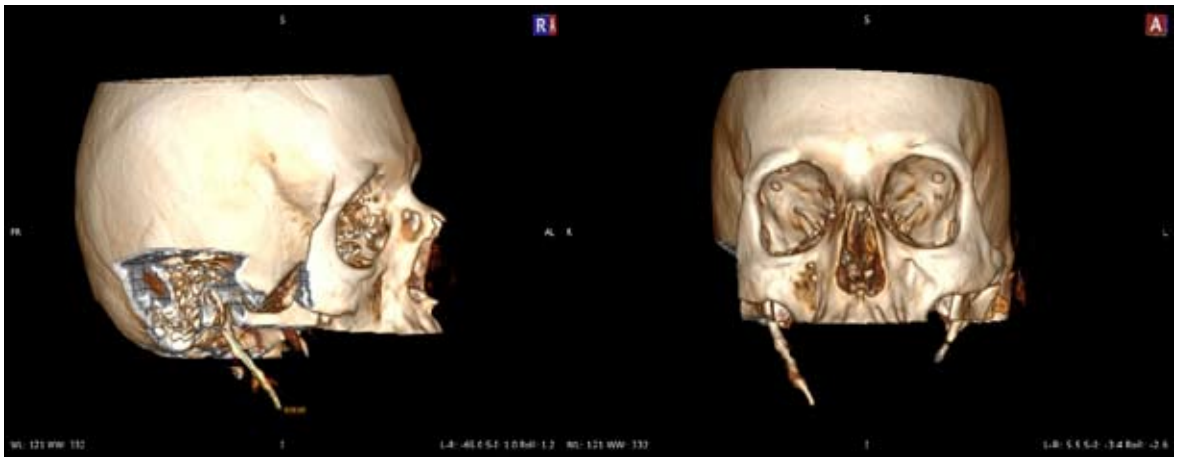
Fig. 1

Eagle syndrome is an important clinical condition for the otolaryngologist to recognize. This is due to the variety of presentations and potentially neurovascular complications.

This article aims to present the clinical and radiological performance of our case of Eagle' syndrome and the review of literature.

Material and methods

A 65 years female presented with complains of the difficulty swallowing and foreign body sensation in the area of right tonsillar fossa. She has a Hashimoto's disease, clinically controlled. Physical examination revealed no abnormal finding exclude the palpable styloid process via right tonsillar fossa. We performed CT scan and 3D CT reconstruction which show an elongated right styloid process without damage in the neurovascular boundaries (fig. 2).



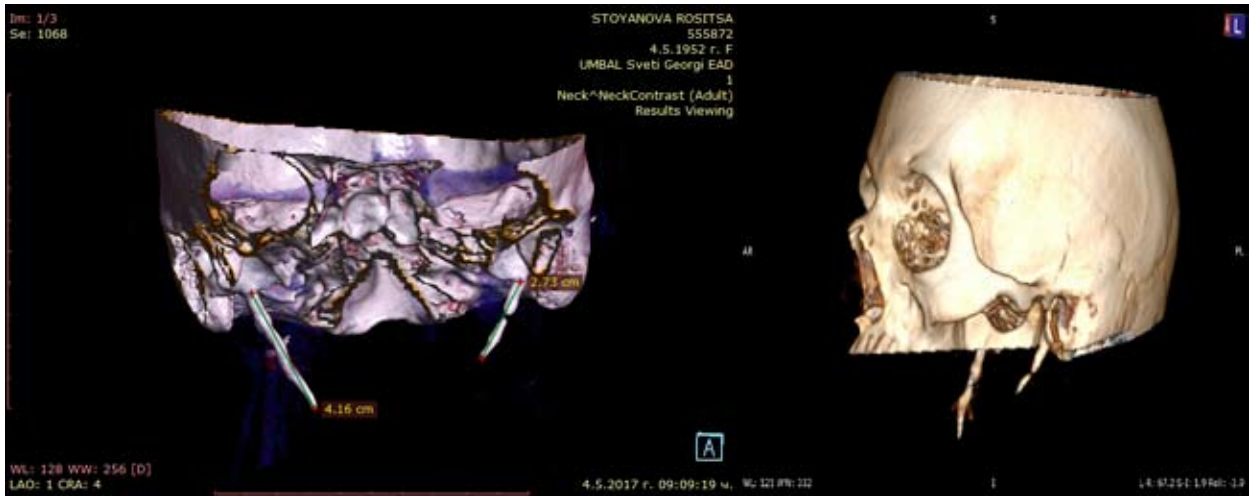


Fig. 2

Patient was asked for pain during neck movement or neck pain in rest, but she stated only painful swallowing. The 3D- CT reconstruction showed a styloid process 43,5 mm in length. We discussed with the patient the opportunities of the treatment and decided to treat it conservatively.

Discussion

Eagle's syndrome is a rare condition characterize with elongated styloid process or ossification of stylohyoid ligament. It is named after Watt Eagle an otolaryngologist at Duke University, who described first the syndrome in 1937. Anatomically, styloid process is situated to the stylomastoid foramen, lateral to the pharyngeal wall and between external and internal carotid artery. The length varies between 2-3mm. The topographic characteristic of styloid process is connected to clinical complications when it is elongated. Etiology of the syndrome is very debated and ambiguous. Watt Eagle suggested that surgical trauma like tonsillectomy or local chronic irritation could be cause of osteitis, periostitis, or tendinitis of the styloid process and stylohyoid ligament which resulted in reactive, ossifying hyperplasia. Later Lentini proposed that persistent mesenchymal elements known as Reichert's cartilage residues, could undergo osseous metaplasia. The ossification of styloid process was also related to endocrine disorders in women in menopause (Epifianio 1962). Gokce C et al. reported that the end-stage renal disease having abnormal calcium, phosphorus and Vit D could cause elongated styloid process or/and ossified stylohyoid ligament. The radiotherapy of the head and neck could be cause of osteonecrosis of styloid process and development of Eagle syndrome.

In 1949 Eagle primarily described two categories for the Eagle syndrome (styloid syndrome). Classic styloid syndrome is followed by tonsillectomy and is characterized by pain in the tonsillar fossa and sometimes attended by dysphagia, odynophagia, sore throat, foreign body sensation in the pharynx and voice changes rarely. The stylo-carotid syndrome is not correlated with tonsillectomy. In this condition, the stylohyoid apparatus compresses the internal and/or the external carotid arteries and especially their perivascular sympathetic structures, resulting in a persistent pain irradiating in the carotid space. This impingement can decrease blood flow and cause carotidynia, headache, facial pain. The presentation of syndrome can be with myriad of symptoms and with no relation to tonsillectomy. The patients can complain of unilateral pain in the neck or face , troubled swallowing-dysphagia, odynophagia, tinnitus, vertigo. Eagle's syndrome could be associated with very serious clinical complications- ischemic attacks, stroke, carotid artery dissection. The literature also contains reports of persistent mouth ulceration, toothache and glosssodynia.

The diagnose of Eagle' syndrome is based on optimal medical history and physical examination. Pharyngeal palpation, especially in tonsillar fossa can be diagnostic. Non- elongated styloid process usually is impalpable. The protrusion of styloid process into tonsillar fossa can be classified into 3 grades : 1st- in the upper pole, 2nd – in the middle pole and 3rd – in the lower pole of the tonsillar pole. The imaging methods confirm the diagnose. The

radiographs of the scull in lateral view may be useful in showing the elongated styloid process The CT scan is the most accurate tool and with 3D-CT reconstruction is gold standard for diagnosis of Eagle's syndrome.

It is more precise in determining the length and angulation of styloid process. Other important sign is the position of elongated styloid process. Kent et al. suggest that the proximity of the styloid process to the tonsillar fossa is a more appropriate diagnostic component. In their 2005 study, they concluded that the styloid process was significantly closer to the tonsillar fossa in the symptomatic group compared to the control group. This finding supports a hypothesis of glossopharyngeal nerve irritation and the consequences of it. In cases of the vascular injury, it is recommended CT angiography which can provide information regarding carotid flow especially if stroke or dissection is suspected. Other imaging modalities have been explored such as transoral ultrasound and bone scintigraphy but their efficacy remains unknown at this time. A lidocaine test can establish the diagnosis of Eagle syndrome. This test contains injection of 1ml of 2% lidocaine into the tonsillar fossa in an awake patient. If the patient' symptoms diminished or disapper within 5minutes, the test is positive and confirms the diagnosis of Eagle' syndrome.

The management of Eagle syndrome includes conservative and surgical approach. The conservative treatment is often choice of patient, because of high risk of operative approach. The conservative medications including analgesics, nonsteroidal anti-inflammatory drugs, anticonvulsants, antidepressants, transpharyngeal application of steroid. If the treatment is free of effect, the choice of treatment is surgical approach. The shortening of styloid process either via intraoral or extraoral approach, produces long- term results. If the patient with huge size of styloid process is without the Eagle's symptom disease, no conservative or surgical treatments were applied, only routine follow up.

Conclusion

Eagle Syndrome has a vague variety of clinical presentations as evidenced by the multitude of non-specific symptoms. It is important to understand the diagnostic workup, relevant imaging, and ultimate treatment options. With advances in medical management and surgical approaches to the styloid process, Eagle syndrome has become recognized more readily and has an established group of treatment options.

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