The present investigation summarizes relevant symptoms, differential diagnosis, imaging, and treatment options of Eagle Syndrome. A comprehensive literature review of peer-reviewed literature was employed utilizing most relevant databases. The diagnoses of Eagle Syndrome have recently increased because of increased awareness of physicians of Eagle Syndrome and the earlier identification of the cardinal symptoms of the disease. The most important symptoms are dysphagia in the setting of cervical neck pain, but there is a wide variety of symptomatology that make Eagle Syndrome a challenge to recognize and diagnose clinically. CT scan continues to be the standard of care for diagnosing Eagle Syndrome and CT Angiography has an important role in aiding diagnosis as well. Medical treatment options include steroids, antidepressants, and anticonvulsants however not all cases of Eagle Syndrome can be managed medically. Surgical approaches are varied but typically are either extraoral or transoral. This report aims to update providers on the important diagnostic criteria of Eagle Syndrome and how physicians can develop a treatment plan that addresses all the symptoms of patients with Eagle Syndrome because it can be treated safely and appropriately.

INTRODUCTION

BACKGROUND

Eagle Syndrome can be classified by an elongated styloid process or by the ossification of the styloid ligament. It can occur unilaterally or bilaterally and often co-occurs with headaches, dysphagia, and hypersalivation.1 The most common symptoms of an elongated styloid process are associated with the pharynx and the ear. Other irregular symptoms can be related to the hypopharynx and the upper esophagus.2 An elongated styloid process may irritate nerves and blood vessels and thus result in pain.3 This may include glossopharyngeal nerve compression, irritation of pharyngeal mucosa due to post-tonsillectomy scarring, and callus formation from a prior styloid fracture.4 Eagle Syndrome may commonly be mistaken for glossopharyngeal neuralgia, a severe pain that is caused by damage to the glossopharyngeal nerve. The oversights presented in this disease are often due to the lack of information and awareness on Eagle Syndrome in the medical community.4

The exact etiology of Eagle Syndrome is unknown. Dr. Eagle believed that one reason for the styloid elongation may be acquired secondary to traumatic or spontaneous fracture of the styloid process and hyperplasia from previous tonsillectomies.3,5 Bone development and homeostasis appear to contribute to an elongated styloid process as well as the presence of two ossification centers.5 Congenital elongation may also be due to the persistence of a cartilaginous analog of the stylohyal, which is an embryologic precursor of the styloid.1 Diseases affecting calcium phosphate
It is important to note, however, that the mere presence of an elongated or calcified styloid process does not necessarily point to a diagnosis of Eagle Syndrome since many patients with such similar findings are also asymptomatic. For instance, of the patients that present with such an anatomic anomaly, it has been found that only an estimated 7.8-10.3% are symptomatic. Dr. Eagle also noted there to be two different forms of the disease: classic styloid syndrome and stylo carotid-artery syndrome. Classic styloid syndrome presents with uncharacteristic pains as well as a foreign body sensation. In contrast, stylo carotid-artery syndrome is primarily responsible for transient ischemic attack or stroke. The threat for a TIA may be related to the enlarged styloid process compressing the internal carotid artery.

Upon initial examination, it is important to note that normal styloid processes are not palpable. However, the palpation of the styloid process in the tonsillar fossa may be indicative of elongation. Eagle Syndrome may be diagnosed using panoramic radiography of the mouth, computed tomography of the skull base and neck, or skull posterior-anterior conventional x-ray. Studies report that the average styloid process measures anywhere from 2.5 to 3 centimeters. Elongation of the process may be diagnosed with radiography. In one case report conducted by Khandelwal, Singh Hada, and Harsh, a 56-year-old male presented with complaints of upper neck pain for six months. Upon radiologic examination, it was found that his left side was noted to be approximately 6 cm and right side was 5.5 cm, both severely long. Following the radiographic findings, the patient was reexamined and found to have a hard bony mass which was palpated in the left tonsillar fossa. The patient was given a primary diagnosis of Eagle Syndrome.

Epidemiology and Risk Factors

Elongated styloid processes have often been overlooked as a cause of pharyngeal or ear pains. It has been found that only 4% of the population present with elongated styloid processes. Of those abnormal elongations, only 4-10% of them reveal symptomatic issues such as pharyngeal pains, ear pains, or headaches. One radiologic study noted that elongation and calcification had no significant correlation to age. However, there may be some correlation with symptomatology. In a study conducted by Monsour and Young, it was found that subjects that had elongated styloid processes (>40mm) exhibited dysphagia while those with ossification of the stylohyoid ligament had a much higher incidence of pain during side to side head turning. Additionally, as noted by Morris et al., ossification of the styloid process may also be hereditary and is most likely due to an autosomal dominant method of inheritance.

As noted earlier, the exact etiology of the disease is unknown. However, elongation and calcification of the styloid process diagnosed during radiologic findings is an important finding. In one case, a 59-year-old woman presented with "shooting pains" to her left ear and was subsequently diagnosed with an elongated styloid process after a diagnostic radiograph and CT. Her original lateral radiograph was noted to have a heavily ossified structure that extended from the base of her skull anterolaterally and caudally to the hyoid bone. Neck CT confirmed the presence of a calcified styloid process and has become a standard for diagnosis. The discomfort caused by an elongated or calcified styloid process is quite significant and ought to be made more aware by the medical community. Thus, the goal of this review will be to bring awareness and comprehensively discuss the literature related to Eagle Syndrome.

Methods

The present investigation aimed to summarize related literature for treatment and management of Eagle Syndrome. Ovid MEDLINE, PubMed, NCBI databases, and Google scholar were searched for all publications related to Eagle Syndrome that also involved otolaryngology or head and neck surgery. Eagle Syndrome is also called stylo carotid artery syndrome, although there are subtle differences. All these terms were used as MeSH terms to search for literature relating to Eagle Syndrome. The present investigation was filtered according to relevance to otolaryngology and the references referenced in this manuscript were identified and then reviewed to provide a more comprehensive understanding of Eagle Syndrome.

Clinical Presentation and Diagnosis

Clinical Signs and Symptoms

In 1957, Watt W. Eagle began publishing his experiences with patients who presented with a classic syndrome of unilateral, sometimes bilateral, pharyngeal pain, pain referred to the ear by vagal irritation, excessive salivation, abnormal gag reflex, and foreign body sensation; through physical exam and radiography, he came to associate these symptoms with an elongated styloid process (SP). A palpable styloid in the tonsillar fossa was diagnostic, and confirmed then with lateral and anteroposterior x-ray. Elongation of the SP causes abnormal anatomical relationships, frequently leading to palsies of cranial nerves IX, V, VII, and X, in order of decreasing incidence, with oropharyngeal or cervical muscular movement often associated with symptomatic exacerbation.

The common clinical picture to keep in mind is that of the post-tonsillectomy patient who presents with cervicofacial pain, odynophagia, and painful head movements. These cranial nerve palsies can present as additional symptoms of altered taste, glossodynia, and esophageal and/or pharyngeal spasm. Eagle's atypical, or stylocarotid syndrome concerns symptoms related to compression and irritation of the branches of the carotid artery by the styloid process, which can result in impaired vascular function and compression of sympathetic nerve fibers that innervate arterial walls, producing pain. These symptoms can include tinnitus, cervical pain, headache, and even signs of cerebral ischemia. It is important to note that grossly abnormal elongation is not necessary for the stylocarotid variant since lateral or medial deviation is the underlying cause;
therefore, palpation of the SP may not be possible as a clinical sign.\textsuperscript{15} In all, these presentations fall under what is known, but frequently forgotten in the differential, as Eagle Syndrome.

With better imaging and greater access to healthcare, the description of Eagle Syndrome has greatly expanded, and the presenting signs and symptoms caused by an abnormal styloid process are now better-defined. Based on recent literature, cervicofacial pain, foreign body sensation, pharyngeal pain, odynophagia, painful head movements, and otalgia are shown to be the most common presenting complaints in order of decreasing frequency.\textsuperscript{16} When Eagle Syndrome is in the differential diagnosis, palpation of the styloid process in the tonsillar fossa is one of the most important signs in clinically for its propensity to agitate symptoms, especially those caused by nerve irritation.\textsuperscript{12}

**COMPLICATIONS FOR EAGLE SYNDROME**

The location of the styloid process in a compact region with complex neurovasculature, as well as being a site of muscular attachment, gives a wide range of complications that could arise with either variant of Eagle Syndrome. There is an array of complaints that are often typically more serious than the usual symptoms of Eagle Syndrome described previously in this review, and they can make diagnosis more difficult. Although there are definitive diagnostic tests and criteria, the rarity of Eagle Syndrome means it is frequently dubbed a diagnosis of exclusion and finds no place in an initial differential; this is compounded by the vague and varied symptoms that send patients to a variety of specialists.\textsuperscript{17} Thus, one of the major complications for patients who have Eagle Syndrome is that of misdiagnosis, which frequently leads to mistreatment and delay of definitive treatment.\textsuperscript{18} Many patients present with a history of numerous visits prior to diagnosis, having received diagnosis and treatment of temporomandibular disorders, which includes those disorders causing pain in the same area but attributed to masticatory muscles and the temporomandibular joint.\textsuperscript{3,19}

Cerebrovascular complications, although rare, are a serious manifestation of the stylocarotid syndrome. Compression of the large vessels of the neck, frequently the internal carotid artery, can result from rotational movement of the neck, which can give a presentation of transient ischemic attacks in ES patients.\textsuperscript{20,21} Likewise, frank cerebrovascular accidents, can arise from position-independent vascular compression by the styloid, which can cause atheromatous plaque formation, sinus thrombosis due to jugular compression, and dissection of the cervical carotid arteries.\textsuperscript{14,22,25} Related to misdiagnosis, mistreatment, and neurological complications is the fracture of carotid artery stents placed after presentation of TIA or CVA, but the patient had yet to be diagnosed with ES as the cause of the event. There are cases reported in the literature of endovascular stenting being performed at the site of carotid stenosis, and on follow-up or presentation of a new cerebrovascular event, the stent had been fractured by an elongated SP that was present yet overlooked on initial imaging.\textsuperscript{24,25}

While cranial nerve palsies are common in Eagle Syndrome, involvement of the vagus nerve is one of the less frequently involved nerves. This leads to cases in which patients complain of hoarseness that can be subacute to insidious onset and can easily steer diagnosis in another direction.\textsuperscript{26,27} Horner syndrome is rare in its own right, but the classic triad of a ptosis, miosis, and anhidrosis have been reported related to the even rarer cause of an elongated SP compressing the cervical sympathetic plexus.\textsuperscript{12,28}

**DIAGNOSIS: CT AND CT ANGIOGRAPHY**

The historical means of diagnosing Eagle Syndrome, e.g., palpation and x-ray, are still at the forefront of the clinical evaluation today. As mentioned, palpation of the tonsillar fossa is reliable and a very simple first step when suspecting Eagle Syndrome, normally producing local tenderness and aggravation of other symptoms in in the vast majority of patients with a symptomatic elongated SP.\textsuperscript{16,27,29} Lidocaine or xylocaine infiltration of the tonsillar fossa is not only a compelling part of diagnosis when it decreases symptoms of pain and other symptoms caused by cranial nerve irritation, but infiltration has some predictive value for response to surgical management as well.\textsuperscript{16,30,31} Simple radiographs (lateral, anteroposterior, and modified Towne’s view) and orthopantomogram are still mainstays in the Eagle Syndrome diagnostic workup; while they are accessible in most settings and are good indicators of elongation and devia-

Computed tomography (CT) is now considered the gold standard for diagnostic confirmation and has been shown as an effective means to relate symptomology and the corresponding aberrancies in cervicocranial anatomy.\textsuperscript{1,29,32} In addition to giving a more complete anatomical picture, CT has been shown to be significantly more accurate than x-ray and lessens the burden of unnecessary treatment of some elongated styloid processes.\textsuperscript{12,36} In atypical or stylo-carotid syndrome, use of cervical CT angiography is particularly useful for showing the consequences of SP proximity to the carotid arteries and jugular veins; these include reduced patency, transient occlusion, and dissection.\textsuperscript{12,14,21}

Other emerging imaging modalities that have been reported as used in settings suspicious for Eagle Syndrome include transoral ultrasonography and bone scintigraphy. The promise of transoral ultrasonography applies mostly to
observing the effects of compression, as well as visualizing dissection, on the carotid artery branches in stylocarotid syndrome and is to be used in conjunction with other imaging, such as CTA. Bone scintigraphy shows areas of abnormal bony growth, thus its use is likely one of excluding other pathological processes that could result in new bone growth or degeneration in the cervicocranial region, though reports of scintigraphic evaluation are still limited.37

TREATMENTS OF EAGLE SYNDROME

Treatment for Eagle Syndrome is based on the severity of the case. Typically, it has been found that conservative management consisting of physiotherapy, long-acting anesthetics in the tonsillar fossa and treatment with anti-inflammatory drugs are all ineffective at relieving pain.3 Thus, treatment involves an intraoral or extraoral surgical approach to shortening the elongated styloid process.3 During surgery, it is important to note that there is a risk of harming parapharyngeal structures such as the carotid artery or vagal nerve. This is related to difficulty in full visualization of the area. Thus, CT-based navigation and endoscopy-assisted surgery are used to precisely identify the surrounding anatomical areas in this operation.5 In a retrospective chart review study, seven patients with symptoms of neck pain and odynophagia were treated. Six of the seven patients underwent the surgery. The average time for resolution of symptoms was found to be 26.5 days.5 The seventh patient who did not undergo the surgery was found to still have existing pain after being diagnosed three years prior. Thus, surgery has been found to be an important treatment option for Eagle Syndrome.

CONSERVATIVE TREATMENTS OF EAGLE SYNDROME

The pathogenesis and severity of the presenting case factors into the selection of a management plan/treatment for patients afflicted with Eagle syndrome. Eagle syndrome treatments are commonly divided into conservative management or surgical excision.58 Though surgery is the more definitive treatment, conservative management of Eagle Syndrome should be used first.59 Conservative methods of medical management can further be divided into first-line analgesics such as non-steroidal anti-inflammatory drugs (NSAIDs) and a multi-drug approach consisting of anticonvulsants, antidepressants, local injections, and manipulation.58–43 NSAIDs provide analgesia in Eagle Syndrome and will likely provide the most benefit to patients suffering symptoms that are inflammatory in nature.41 Targeting of the styloid ligament and surrounding inflammation has been shown to decrease painful symptoms associated with Eagle Syndrome. Taheri et al. demonstrated an 80% reduction in severity of pain with pain-free intervals 6-months post-treatment report of treating a patient with only oral medications (amitriptyline and pregabalin). Similar cases, with similar results, used oral medications in conjunction with local injections and/or a stellate ganglion block.42

Conservative treatment of Eagle Syndrome also includes ultrasound guided injection of steroids or long-lasting anesthetics.38,39,45 Ultrasound guided injections begin with identification of important neurovascular structures adjacent to the styloid process. Once the neurovascular structures are identified, a 1.5 inch, 25-gauge needle is directed into tonsillar fossa to reach the styloid process. At the styloid process, physician chosen analgesics (i.e., bupivacaine or ropivacaine and dexamethasone) can be injected. Patients receiving local injections should be monitored following the procedure and assessed for any measurable relief from symptoms. Local injections can serve as temporary relief for patients awaiting surgery or as an alternative to patients refusing surgical excision of the ossified styloid process.43

Stellate ganglion blocks (SGBs) provide a therapeutic benefit to those suffering with pain from head and neck syndromes like Eagle Syndrome. SGBs can be used alone or in combination with a multi-drug approach. The latter has shown to result in a near complete resolution of symptoms related to Eagle Syndrome.44

Han et al., used an oral regimen of gabapentin 300 mg, d, tianeptine 1.5 mg/d, and tramadol hydrochloride 37.5 mg/acetaminophen 325 mg 3 tablets/d. A local injection of 1 ml of triamcinolone 10 mg combined with 0.3% mepivacaine 3 ml was injected once into tender areas surrounding the tonsils. In addition to oral medications and the local injections, a stellate ganglion block was performed in the problematic area 1/well for a period of 4 weeks. Treatments administered by Han et al., were only expected to be temporary; however, the patient showed no signs of worsening pain during a 3-month monitoring period following conservative treatment.44

Physical manipulation with physical therapy aims to reduce overall pain by relaxing previously constricted, stretched, or in-spasms muscles surrounding the calcified styloid ligament.45 Trans pharyngeal manipulation with manual fracture while under general anesthesia has been proposed as a treatment modality, however the maneuver risks damage to adjacent neurovascular structures and provides little relief for symptoms experienced by patients.45

Treatment of stylocarotid artery syndrome, a variant of Eagle Syndrome, centers around preserving the integrity of the internal and external carotid arteries. Patients with stylocarotid artery syndrome present with symptoms associated with the stimulation of the sympathetic nerve plexus surrounding the blood vessels.44 Presenting symptoms include syncope, impaired vision, facial pain, headaches, migraines, and other cerebrovascular symptoms. In such cases, anticoagulant or antiplatelet drugs in monotherapy or combination can be used to lower the risk of stroke or dissection of the carotid arteries.46–48 Depending on the severity of involvement of the surrounding vascular and nervous structures, it may be in the best interest of the patient to undergo surgical excision.49 Only after conservative measures fail, should surgical excision of the elongated styloid process be considered.

SURGICAL TREATMENT FOR EAGLE SYNDROME

The most definitive treatment of Eagle Syndrome is surgical amputation or excision of the elongated styloid process.50

Health Psychology Research
Two different approaches have been proposed for the surgical treatment of Eagle syndrome: 1) the intraoral or transpharyngeal and 2) the extraoral or transcervical approach. Each surgical approach has its advantages and disadvantages; however, the selected surgical approach is largely based on the surgeon’s preference.\textsuperscript{51}

Traditionally the intraoral approach follows a tonsillectomy, however, a tonsil-sparing transoral approach has been developed.\textsuperscript{52,53} Whether following tonsillectomy, or not, the intraoral approach involves the surgeon locating the styloid process via digital palpation within the tonsillar fossa. Once the styloid process is identified, the surgeon then makes a surgical excision from which the stylohyoid and its ligamentous attachments are excised.\textsuperscript{54} Advantages of the intraoral approach include its simplicity, shorter operation time, ability to be performed under local anesthetic, decreased risk of injury to the facial nerve, and a lack of external scar. The disadvantages of the intraoral approach are poor visualization, risk of iatrogenic injury to major neurovascular structures (i.e., carotid arteries), and other transient postoperative complications (dysphagia, trismus, and airway edema).\textsuperscript{54–56}

The extraoral approach begins with an oblique incision made at the angle of the mandible and carried out to the sternocleidomastoid muscle. With the sternocleidomastoid retracted posteriorly, visualization of the styloid process in made from the space between the parotid gland and the posterior belly of the digastric muscle.\textsuperscript{56} Advantages of the extraoral approach include the markedly enhanced exposure of the styloid process and its adjacent structures. The disadvantages of the extraoral approach include longer duration ang greater risk of injury to the facial nerve and its branches, a disfiguring scar, and significantly longer recovery period.\textsuperscript{51,54,55}

CONCLUSIONS

The original description of Eagle Syndrome included one sided or sometimes bilateral pharyngeal and/or cervicofacial pain that is referred to the ear in the setting of dysphagia. Dr. Watt Eagle came to associate these symptoms with an elongated styloid process and this anatomical finding can be confirmed with X-ray showing this anatomical abnormality, but CT scan is needed to confirm the diagnosis and to definitively measure the styloid process which typically measures around 4 cm in patients with Eagle Syndrome. Transoral ultrasound and bone scintigraphy are emerging diagnostic tools in this clinical picture. Treatment of Eagle Syndrome is based on the severity of the case and it can be managed conservatively with physiotherapy and/or long-acting anesthetics. The surgical approach involves amputation or excision of the elongated the styloid process via an intra or extraoral approach.

If left untreated, the most serious complications of Eagle Syndrome are cerebrovascular in nature and frequently involve the internal carotid artery, which can present with a transient ischemic attack and eventual stroke. Cranial nerve palsies have also been seen with Eagle Syndrome. However, the most frequent complication is misdiagnosis and delay of either conservative or definitive treatment to alleviate symptomatology. It is therefore critical that physicians are increasingly aware of how-to diagnosis and treat Eagle Syndrome. The challenge for the modern Otolaryngologist and physician in general is that there is a wide variety of symptomatology that make Eagle Syndrome a challenge to recognize and diagnose clinically.

DISCLAIMER

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