Overview of Eagle's syndrome, consequences, and treatment

Abdulaziz Fahad Altowairqi , Saud Mohammed Saud Aleisa , Raad Hameed Altowairqi , Rayan Darwish Habib

Abstract— The aim of this study was to identify the symptoms and evaluate the causes behind of Eagle's Syndrome, as well as to review the evidence in treatment options that are available. The searches were conducted through MEDLINE, Google Scholar, and PubMed databases there were no regional restrictions but English lanaguge restriction with human subject for studies published up to October, 2017. Along with a medical background and physical examination, radiological assessment is likewise essential for diagnosis. Patients with Eagle's syndrome typically have a chronic pain. Appropriate diagnosis is needed for treatment ,usually surgical treatment is required. . When a patient is presenting long-term pain of the head and neck region, dentists and physicians should consider Eagle's syndrome as a possible cause.

Index Terms— Eagle's syndrome, Eagle's disease, prolongation of the styloid apophysis.

INTRODUCTION

Eagle's syndrome is defined by cervicopharyngeal symptoms and signs related to prolongation of the styloid apophysis [1]. The styloid apophysis is composed of a bone projection coming from the tympanic section of the temporal bone, which is roughly 25 mm in size [1]. Prolongation of the styloid apophysis might happen via ossification of the stylohyoid ligament, or via the progression of the apophysis because of osteogenesis caused by an aspect such as injury [2]. ES is an unusual entity which is not generally presumed in medical practice [3] and also just a little percent of the populace is thought to have actually an extended styloid process and a calcified stylohyoid ligament shows up the signs [4]. Watt Eagle explained it for the first time in 1937, separating it right into 2 subtypes: the" classic syndrome" and also the" stylo-carotid artery syndrome." Several concepts have actually been presented concerning its pathogenesis.

Eagle syndrome is believed to be an unusual incident; nevertheless, the reported occurrence of an extended styloid process differs. Eagle reported a clinical occurrence of 4%, while Kaufman discovered an occurrence of 28% radiographically [5]. While the occurrence of a lengthened styloid process in the basic populace is debatable, just 4%-10% of patients with this structural variant record symptoms [6].

Prolongation of the styloid apophysis or calcification of the stylohyoid ligament could trigger serious face pain, migraine, dysphagia or odynophagia, otalgia, ringing experiences, and also trismus [1], [2]. Specific medical diagnosis of the syndrome is difficult. It is normally confused by temporomandibular joint disorder, chronic tonsillitis, headache, collection headache, glossopharyngeal as well as trigeminal neuralgia, myofascial pain dysfunction syndrome, pain secondary to unerupted or affected 3rd molars, neck joint inflammation, as well as tumors of the base of the tongue. Finger palpation of the palatine tonsil raises the discomfort related to Eagle's syndrome and also supports its medical diagnosis.

The aim of this study was to identify the symptoms and evaluate the causes behind of Eagle's Syndrome, as well as to review the evidence in treatment options that are available.

METHODOLOGY

The searches were conducted through MEDLINE, Google Scholar, and PubMed databases there were no regional restrictions but English lanaguge restriction with human subject for studies published up to October, 2017. The following search terms were used: "Eagle Syndrome", "styloid syndrome", "stylocarotid syndrome", "stylohyoid syndrome", "styloidcarotid artery syndrome", and "elongated styloid syndrome". References list of identified artciles were scaned for more relvant articles.

DISCUSSION

Background:

Eagle syndrome was first explained by Watt Eagle in 1937 when he presented 2 situations entailing prolongation of the styloid procedure causing a constellation of signs and symptoms that were specific from primary glossopharyngeal neuralgia [4]. The styloid process (SP) is a cylindrical, lengthened cartilaginous bone situated on the temporal bone and also re-

mains in a significant area of the neck, as it is bordered by the exterior carotid artery, the interior carotid artery including the sympathetic chain, the occipital artery, the inner jugular vein, the accessory nerve, the hypoglossal nerve, the vagus nerve, and also the glossopharyngeal nerve [7]. Any type of deviation of the styloid process could disrupt this essential anatomy as well as trigger signs and symptoms of Eagle syndrome. The muscles and also ligaments which take a function in chewing and also swallowing are attached to the SP. There are lots of nerves as well as vessels such as carotid arteries beside the SP. The SP and also the stylohyoid ligament create from the Reichert's cartilage (2nd pharyngeal arch) [8]. There is no certain gender predisposition for prolongation of the styloid process, yet women are recognized to be much more symptomatic as compared to the male equivalents [4]. The usual SP length is roughly 20-- 30 mm [8], [9]. The styloid process prolongation (SPE) can be presumed if either the SP or the surrounding stylohyoid ligament ossification reveals a general size over of 30 mm (Figure 1) [9].

Etiology:

The etiology is still being debated Watt Eagle [5] offered that surgical injury (tonsillectomy) or regional chronic irritation triggers osteitis, periostitis, or tendonitis of the styloid process and also the stylohyoid ligaments which led to responsive, hardening hyperplasia. Later On Lentini (1975) proposed the theory that consistent mesenchymal components, additionally called Reichert cartilage material deposits, can undertake bony metaplasia in the setup of a proper traumatic or difficult occasion. Epifanio in 1962 taken into consideration that the ossification of the styloid process was likewise relating endocrine disorders in females at menopause, that likewise had ossification of various other ligaments in the body. Gokce C [10] (2008) reported that in patients with the end-stage kidney illness having unusual calcium, phosphorus, as well as vitamin D metabolic process had heterotopic calcification which triggered prolongation of styloid process and also therefore the discussion of Eagle Syndrome.

Relation of ectopic calcification and ES

In a research, Okabe et alia [11] discovered an important relationship in between the serum calcium (Ca) level and also the SP length amongst 80-year-old patients. The lengthier the SP was, the elevated the serum Ca level remained in this example.

Extraskeletal (ectopic) calcification (deposition calcium phosphate crystals) or ossification (real bone development) could have a function for the prolongation of SP. Ectopic calcification (EC) in nonosseous soft tissue could be because of 3 systems; metastatic calcification because of problems triggering unnatural serum Ca and also P amounts, dystrophic calcification because of mineral deposition right into metabolically damaged or dead tissue in spite of typical serum degrees of Ca and also P, as well as ectopic ossification. In patients with end-stage kidney illness (ESRD) have dangers for EC or ossification because of disorders (kidney failing, dialysis, secondary hyperparathyroidism) triggering metastatic calcification [12].

Unusual Ca, P and also vitamin D metabolic process is typical in patients with end-stage kidney illness ESRD [13]. Disruptions in this metabolic process in these patients triggers extended provocation of parathyroid glands. This causes raised synthesis and also production of parathyroid hormone (PTH). For that reason, it creates parathyroid hyperplasia-secondary hyperparathyroidism (SHPT). SHPT triggers skeletal disruption that defines kidney osteodystrophy. It is related to vascular and also various other soft tissue calcifications [14]. In addition, high PTH amounts, a raised hyperphosphatemia as well as calcium X phosphate item are related to enhanced death rate in patients with ESRD [15].

EC-metastatic calcification- in nonosseous soft tissue because of irregular serum Ca and also P amounts (problem in Ca as well as P metabolic process) is typical in patients with ESRD [12]. As a result, this illness might be a great version for the examination of the EC in the prolongation of SP. Nevertheless, a detailed differential medical diagnosis must be provided for this problem. For that reason, the ESRD patients had co-disorders creating dystrophic calcification (scleroderma, dermatomyositis, and also systemic lupus erythematosis, trauma-induced) and also ectopic ossification (post-surgical operation, burns, neurological injury, myositis ossificans) need to be removed. The level of EC boosts with the dialysis duration and also age [16]. EC because of the problem in Ca and also P metabolic rate which relates to the period of dialysis is likewise crucial for this calcification in these patients [12].

Pathophysiology:

It was taken into consideration before that development of scar tissue around the styloid apex after tonsillectomy, triggered compression and also straining of the neurovascular constructs existing in the retro styloid area. Nevertheless, Eagle syndrome [13] likewise shows in patients that have actually never ever been operated for tonsillectomy. A number of potential mechanisms for the pathogenesis of discomfort in Eagle syndrome have actually been suggested. The very first one regards that the extended styloid process triggers compression of cranial nerves, the majority of generally the glossopharyngeal nerve, with succeeding throat and also neck pain. Next off one is based upon the opportunity of compression of the inner carotid artery by the styloid procedure, which could trigger short-term ischemic attacks or compression of the sympathetic nerves running along the artery, causing a variety of signs. The ache in Eagle syndrome typically appears like glossopharyngeal neuralgia however is usually much more blunt and also continuous, nevertheless, situations with sharp periodic pain along the path of the glossopharyngeal nerve are additionally reported. More ideas of responsive hyperplasia and also responsive metaplasia exist which relate the prolongation with either overgrowth of the styloid process itself or ossification of the stylohyoid ligament complicated consequently of injury. This sensation might clarify the occurrence of Eagle syndrome in patients after tonsillectomy, as it was initially explained by Eagle. Several of the various other feasible thought about reasons are the irregular angulation related to extraordinarily extensive styloid procedure creating irritability of surrounding musculature or mucosa. Stretching and also fibrosis entailing the 5th, 7th, nine, as well as tenth cranial nerves in the post-tonsillectomy duration might likewise be a feasible etiology [13].

Osteoradionecrosis, one of the serious complications of radiotherapy, arises from hypovascular-hypoxic-hypocellular problems that trigger failure of tissue that causes bone direct exposure. Furthermore, any kind of inflammation or trauma to the location more rises the energy/oxygen requirements of the

regional tissues, intensifying the danger of osteoradionecrosis[13]. The probability of osteoradionecrosis growth is dose-dependent and also greatest at a dosage of > 60 Gy. While intensity-modulated radiation treatment has actually numerous advantages as compared to standard radiation treatment, just how this therapy influences the possibility of establishing osteoradionecrosis is unclear [14]. Therefore, it appears sensibly essential to think about the growth of osteoradionecrosis of the styloid procedure creating Eagle syndrome adhering to radiation therapies of the head and also neck.

Table1. Summary of the pathophysiological mechanisms for the pain of Eagle's syndrome

Compression of the neural elements, the glossopharyngeal nerve, the lower branch of the trigeminal nerve, and/or the chorda tympani nerve by the elongated styloid process
Fracture of the ossified stylohyoid ligament, followed by proliferation of granulation tissue that causes pressure on surrounding structures and results in pain
Impingement on the carotid vessels by the styloid process producing irritation of the sympathetic nerves in the arterial sheath
Degenerative and inflammatory changes in the tendinous portion of the stylohyoid insertion, a condition called an insertion tendinosis
Irritation of the pharyngeal mucosa by direct compression by the styloid process
Stretching and fibrosis involving the 5^{th}, 7^{th}, 9^{th}, and 10^{th} cranial nerves in the post-tonsillectomy period.

Arterial impingement:

Compression of the carotid artery may produce vascular/ischaemic symptoms as well as pain along the artery to the supplied territory (thought to be mediated by the sympathetic plexus), including:

- mechanical compression
  - visual symptoms
  - syncope
  - carotid dissection has also been described [20]
- sympathetic plexus irritation (carotidynia)
  - eye pain
  - parietal pain

Eventually, the symptoms could be an outcome of the regular procedure of aging. As typical aging is related to a reduction in flexibility of soft cells, degenerative and also inflammatory modifications in the tendinous part of the stylohyoid insertion, a problem called insertion tendinosis which could trigger discomfort in the distribution of glossopharyngeal nerve, appearing like Eagle syndrome. To prevent confusion, this indication much better is called pseudo stylohyoid syndrome. Eagle syndrome has actually been identified in patients with a normal-length styloid process. In this circumstance, the pathology is associateded with inflammatory and also degenerative adjustments in the location, particularly insertion tendonitis [21].

Complications:

In few situations, the lengthy styloid process could put pressure on the inner carotid arteries [21] on both side of your neck. This stress might trigger a stroke. Immediate emergency care is required if instantly experience any one of these signs and symptoms: headache, weak point, vertigo, adjustments in vision, comp.

Diagnosis:

The medical diagnosis of Eagle's syndrome is based upon an ideal case history and also physical examination. A lengthened styloid process could be examined by intraoral palpation, putting the index finger in the tonsillar fossa as well as using mild pressure. If the pain is duplicated by palpation and also either described the ear, face or head, the medical diagnosis of an extended styloid process is most likely. A styloid procedure of typical length is normally impalpable.

The medical diagnosis of the extended styloid process is after that verified by imaging. Numerous imaging methods have actually been used, however CT is one of the most precise. 3-D CT [22] remodeling of the neck allows exact measurement of the size of the styloid process as well as the ossified stylohyoid ligament.The typical size of the grown-up styloid is roughly 2.5 centimeters while higher than 3 centimeters is regarded extended. If this requirement is employed, practically 4% of the populace have actually a lengthened procedure. Nevertheless, just a tiny percentage is symptomatic. Orthopantomogram (OPG) as well as CT could both be applied to evaluate the styloid process/stylohyoid ligament complicated. In situations where mechanical vascular compression is possibly the source of ischaemic signs, angiographic exam (CT angiography or catheter angiography) is gotten with the patient's head suitably placed to duplicate signs and symptoms which could show vital for showing mechanical constriction of the carotid artery.

Much more typically; a panoramic radiography (PR) is applied to identify whether the SP is lengthened. Panoramic pictures are most helpful medically for detecting problems associated with face structures consisting of maxillary and also mandibular bones and also their sustaining structures [23]. Although PRs have an essential function for showing the variants of SP, they are unable to reveal the alignment as well as measurements of this bone.

On the other hand, multislice calculated tomography (MSCT) gives a trusted visualization of this features [24]. Nevertheless, lots of records have actually revealed that SPE was not the only factor of the signs and indicators, and also they recommended that elements such as mediolateral angling (MLA), anteroposterior angling (APA) as well as the flexing of the SP head were additionally essential [25]. As a result, radiographic imaging might suffice for the patients with Eagle's syndrome that has regular bone structure. Yet, if there is any type of issue concerning this bone like tumor, osteomyelitis or fracture, this problem could be simply identified by bone scintigraphy as a result of the high level of sensitivity of this imaging technique [25].

A lidocaine infiltration examination could develop the medical diagnosis of Eagle syndrome. This examination entails injection of 1 mL of 2% lidocaine right into the tonsillar fossa in a conscious patient. If the patient's signs decrease or go away within 5 mins of shot, the examination is thought about favorable and also sustains the medical diagnosis of Eagle syn-

drome [26].

Treatment:

The lengthened styloid process syndrome may be taken care of either cautiously or surgically. Traditional therapies [27] consisting of normal anesthetics, antidepressant drugs, anticonvulsant, trans pharyngeal shot of steroids as well as lidocaine, diazepam, nonsteroidal anti-inflammatory medicines, as well as the application of topical warmth all have actually been utilized for the objective. Transpharyngeal control with manual fracture the extended styloid procedure does not typically eliminate signs as well as dangers damages to nearby neurovascular structures. Patients that cannot react to several drugs could need medical manipulations.

One of the most efficient treatment is the medical shortening of the styloid procedure either using an intraoral or outside technique as it generates far better lasting outcomes. The benefits of an intraoral strategy [28] are the simpleness of the method, decreased procedure time, achievability under regional anesthetic, as well as lack of any kind of noticeable outside mark. Nonetheless, the major negative aspects are an absence of accessibility, especially if there are a subsequent hemorrhage as well as deep neck infections, inadequate visualization of the medical area specifically in patients with considerably decreased jaw opening, the danger of iatrogenic injury to significant neurovascular structures, changes of speech as well as ingesting from postoperative edema.

One of the most substantial benefit of an outside technique [28] is boosted direct exposure of the styloid procedure as well as the nearby structures which outweigh all various other advantages. It likewise helps the elimination of a partly ossified stylohyoid tendon. Significant drawbacks consist of even more time-consumption, the threat of injury to facial nerve and also its branches, damaging neck mark and also longer healing duration. For the most parts, nonetheless, this selection is normally based upon the medical specialized of the carrying out surgeon.

In 2015, Kiralj et al offered that the length of the styloid process or the calcified ligament need to determined by the surgical approach[28]. In the existance of ossification of the stylohyoid structure or a styloid process extending to the hyoid bone, the extraoral approach is safer. But, if the styloid process is shorter or not ossified, the intraoral approach should suffice.

CONCLUSION

Along with a medical background and physical examination, radiological assessment is likewise essential for diagnosis. Patients with Eagle's syndrome typically have a chronic pain. Appropriate diagnosis is needed for treatment ,usually surgical treatment is required. . When a patient is presenting long-term pain of the head and neck region, dentists and physicians should consider Eagle's syndrome as a possible cause.

FIGURES

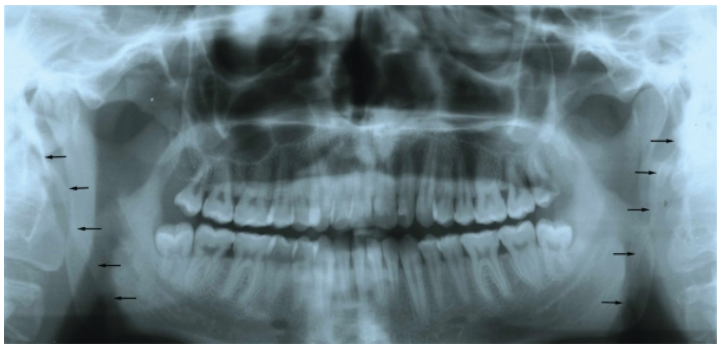

Figure 1: Bileteral styloid process elongation in a subject on a panoramic radiography [9].

REFERENCES

(1) Fini G, Gasparini G, Filippini F, Becelli R, Marcotullio D. The long styloid process syndrome or Eagle's syndrome. J Craniomaxillofac Surg. 2000;28:123–7.
(2) Prasad K C, Kamath M P, Reddy J M, Raju K, Agarwal S. Elongated styloid process (Eagle's syndrome): a clinical study. J Oral Maxillofac Surg. 2002;60(2):171–5.
(3) Winkler S., Sammartino F.J., Sr., Sammartino F.J., Jr., Monari J.H. Stylohyoid syndrome. Report of a case. Oral. Surg. Oral. Med. Oral. Pathol. 1981;51:215–217.
(4) Fini G., Gasparini G., Filippini F., Becelli R., Marcotullio D. The long styloid process syndrome or Eagle's syndrome. J. Craniomaxillofac. Surg. 2000;28(2):123–127.
(5) Eagle WW. Symptomatic elongated styloid process: Report of two cases of styloid process-carotid artery syndrome with operation. Arch Otolaryngol. 1949. May; 49 5: 490- 503.
(6) Kaufman SM, Elzay RP, Irish EF. Styloid process variation: radiologic and clinical study. Arch Otolaryngol. 1970. May; 91 5: 460- 463.
(7) Bafaqeeh SA. Eagle syndrome: classic and carotid artery types. J Otolaryngol. 2000. April; 29 2: 88- 94.
(8) Carter L. Soft tissue calcification and ossification. In: White SC, Pharoah MJ, editors. Oral radiology, principles and interpretation.Missouri: Mosby; 2004. pp. 597–614.
(9) Gokce C, Sisman Y, Tarim Ertas E, Akgunlu F, Ozturk A. Prevalence of styloid process elongation on panoramic radiography in the Turkey population from Cappodocia region. Eur J Dent. 2008;2:18–22.
(10) Sisman Y, Gokce C, Sipahioglu M, Tarim Ertas E, Oymak O, Utas C. Bilateral elongated styloid process in an end-stage renal disease patient with peritoneal dialysis: is there any role for ectopic calcification? Eur J Dent. 2008.
(11) Okabe S, Morimoto Y, Ansai T, et al. Clinical significance and variation of the advanced calcified stylohyoid complex detected by panoramic radiographs among 80-year-old subjects. Dentomaxillofac Radiol. 2006;35:191–199.
(12) Favus MJ, Vokes TJ. Paget disease and other dysplasias of bone. In: Jameson JL, editor. Harrison's endocrinology. USA: McGraw-Hill Companies; 2006. pp. 485–498.
(13) Hruska KA, Teitelbaum SL. Renal osteodystrophy. N Engl J Med. 1995;333:166–174.
(14) Slatopolsky E. The role of calcium, phosphorus and vitamin D metabolism in the development of secondary hyperparathyroidism. Nephrol Dial Transplant. 1998;13:3–8.
(15) Locatelli F, Cannata-Andía JB, Drüeke TB, et al. Management of disturbances of calcium and phosphate metabolism in chronic renal insufficiency, with emphasis on the control of hyperphosphataemia. Nephrol Dial Transplant. 2002;17:723–731.
(16) Guérin AP, London GM, Marchais SJ, Metivier F. Arterial stiffening and vascular calcifications in end-stage renal disease. Nephrol Dial Transplant. 2000;15:1014–1021.
(17) Ceylan A, Köybasioglu A, Celenk F, Yilmaz O, Uslu S. Surgical treatment of elongated styloid process: Experience of 61 cases. Skull Base. 2008;18:289–95.
(18) Murtagh RD, Caracciolo JT, Fernandez G. CT findings associated with Eagle syndrome. AJNR Am J Neuroradiol. 2001;22 (7): 1401-2.
(19) Chuang WC, Short JH, McKinney AM et-al. Reversible left hemispheric ischemia secondary to carotid compression in Eagle syndrome: surgical and CT angiographic correlation. AJNR Am J Neuroradiol. 2007;28 (1): 143-5.
(20) Faivre A, Abdelfettah Z, Rodriguez S et-al. Neurological picture. Bilateral internal carotid artery dissection due to elongated styloid processes and shaking dancing. J. Neurol. Neurosurg. Psychiatr. 2009;80 (10): 1154-5.
(21) Steinmann EP. Styloid syndrome in absence of an elongated process. Acta Otolaryngol. 1968. October; 66 4: 347- 356.
(22) Shah K, Miller DJ. Three-dimensional modeling of Eagle syndrome. Neurology. 2016 Nov 22;87(21):2279-2280.
(23) Monsour PA, Young WG. Variability of the styloid process and stylohyoid ligament in panoramic radiographs. Oral Surg Oral Med Oral Pathol. 1986;61:522–526.
(24) Murtagh RD, Caracciolo JT, Fernandez G. CT findings associated with Eagle syndrome. Am J Neuroradiol. 2001;22:1401–1402.
(25) Onbas O, Kantarci M, Murat Karasen R, et al. Angulation, length, and morphology of the styloid process o the temporal bone analyzed by multidetector computed tomography. Acta Radiol. 2005;46:881–886.
(26) Prasad KC, Kamath MP, Reddy KJ, Raju K, Agarwal S.Elongated styloid process (Eagle's syndrome): a clinical study. J Oral Maxillofac Surg. 2002. February; 60 2: 171- 175.
(27) Ceylan A, Köybaşioğlu A, Celenk F et-al. Surgical treatment of elongated styloid process: experience of 61 cases. Skull Base. 2008;18 (05): 289-95.
(28) Kiralj A, Illić M, Pejaković B, Markov B, Mijatov S, Mijatov I.Eagle's syndrome: a report of two cases. Vojnosanit Pregl. 2015. May; 72 5: 458- 462.