Eagle Syndrome : A Review

Dr. Saloni Kumari
[BDS (Hons), MDS(OMR)]
Dental surgeon
GB Pant Govt Medical College & Hospital
A4/112 syamalima apt Agartala , Tripura, India

Dr. Sonali Sharma
(BDS, MDS)Lecturer
College of Dentistry, University of Dammam
Kingdom of Saudi Arabia

Keywords : Neck pain, Osteogenesis, Styloid syndrome

Objective : Dental surgeon often find eagle syndrome as extreme difficult to be diagnose clinically and its consequent treatment due to asymptomatic condition and when become symptomatic it becomes difficult to be treated and controlled

Introduction

Eagle's syndrome is characterized by cervicopharyngeal signs and symptoms associated with elongation of the styloid apophysis. The styloid apophysis consists of a bone projection originating in the tympanic portion of the temporal bone, which is approximately 25 mm in length . Elongation of the styloid apophysis may occur through ossification of the stylohyoid ligament, or through growth of the apophysis due to osteogenesis triggered by a factor such as trauma[1]. An abnormally long styloid process or stylohyoid chain ossification producing cluster of symptoms gives rise to "Eagle's syndrome" (ES) or "Stylohyoid syndrome"which is characterized by craniofacial or cervical pain. ES is a rare entity which is not commonly suspected in clinical practice, and only a small percentage of 4% of the population is believed to have an elongated styloid process and a calcified stylohyoid ligament manifests the symptoms[2].Patients with Eagle syndrome often report symptoms that include dysphagia, otalgia, throat pain, globus sensation, facial pain, headache, taste disturbances,and dental pain that worsen with chewing, head and tongue movements, and swallowing[3].

Embryology and Anatomy

The embryologic history of the styloid process, stylohyoid ligament, and hyoid bone is a subject of debate. The styloid process, stylohyoid ligament,and lesser cornu of the hyoid bone develop from endochondral ossification of Reichert's cartilage, the cartilaginous component of the second branchial arch. They assert that after 3 months of fetal life, Reichert's cartilage is disrupted and divided into five distinct components (from proximal to distal):tympanohyal, stylohyal, ceratohyal, hypohyal, and basyhyal. The tympanohyal component contributes to the tympanic bone and the base of the styloid process. The stylohyal component contributes to the majority of the styloid process, and the hypohyal and basyhyal components contribute to the hyoid bone. In many animals the ceratohyal component ultimately becomes the epihyoid bone. In humans, Revilla and Stuyt contend that it degenerates to form the stylohyoid ligament[4].

The styloid process is a thin, elongated, pointed, cylindrical bony projection that is situated anteromedially to the mastoid process[5] (and projecting downward, forward and slightly medially from the temporal bone[6] . Its length varies from 2 to 3 cm. Posterior to the styloid is the facial nerve, which emerges from the stylomastoid foramen. Medial to styloid, moving posterior to anterior are the internal jugular vein (with XI, XII, X and IX cranial nerves)[5] and descends between the external and internal carotid arteries to reach the side of pharynx[6]. Medial to the tip of the styloid process are the superior constrictor muscle and the pharyngobasilar fascia, which lie adjacent to the tonsillar fossa. Lateral to the tip of the process is the external carotid artery that bifurcates into superficial temporal and maxillary arteries. The stylohyoid ligament extends from the styloid to the lesser cornu of hyoid bone[5].

Clinical features

The styloid process varies in length in different individuals and often on the two sides of the same person[7] and normally measures 2.5–3 cm in length. When the length of the styloid process exceeds 3 cm, it is considered to be elongated and can be associated with a wide variety of symptoms that were first described by Eagle in 1937[8].

Eagle originally described two morbidity forms. The first classic styloid process syndrome form is caused by the calcification of the stylohyoid complex resulting in the scar tissue impinging on cranial nerves V, VII, IX, X or XII, all of which are situated near the styloid process. The second carotid artery Eagle syndrome form is caused by compression of the sympathetic chain in the carotid sheath. It often occurs in the absence of ossification of the stylohyoid complex. Only a slight deviation of the styloid process medially is required for the tip of the process to start impinging on an artery and cause a wide variety of symptoms[8]. The syndrome presents clinically with pain in the throat that is frequently referred to the ear. Usually there is a sensation of a foreign body in the tonsillar area producing dysphagia. It is believed to be because of physical irritation of nerves or vessels passing near the elongated styloid tip[9] .The ossified ligament usually can be detected by palpation over the tonsil as a hard, pointed structure. Very little correlation exists between the extent of ossification and the intensity of the accompanying symptoms. One symptom is vague, nagging to intense pain in the pharynx on swallowing, turning the head , or opening the mouth, especially on yawning. When this entity is associated with discomfort and the patient has a recent history of neck trauma (e.g., tonsillectomy), the condition is called Eagle syndrome[10].others are vertigo, voice alteration, cough, dizziness, sinusitis, conjunctival injection, headaches. The symptoms often cause continuous discomfort over long periods of time and are often difficult to diagnose[8].

Table 1; Eagle's syndrome clinical presentation and patient demographics[4]

	Classic Type	Carotid Type
Sex	F > M	F > M
Age	5th and 6th decades (peak)	5th and 6th decades (peak)
History of tonsillectomy	Increased frequency	Increased frequency
Odynophagia	Frequent	Not observed
Dysphagia	Frequent	Not observed
Pain (present/absent)	Variable	Frequent
Pain (location)	Ipsilateral;parietal region to pectoral region	Ipsilateral;parietal region/superior periorbital region
Pain (quality)	Dull, constant	Variable (e.g. throbbing, stabbing)
Stridor	Rare (children)	Not observed
Dizziness	Not observed	Frequent
Syncope/TIA/Stroke	Not observed	Frequent
Flexion/extension	Aggravates symptoms	Aggravates symptoms
Contralateral head rotation	Minimal effect	Aggravates symptoms
Tonsillar pillar palpation (exam)	Aggravates symptoms	Minimal effect

The characteristic dull and nagging pain of an elongated styloid process that becomes worse during deglutition and can be reproduced by palpation of the tonsillar fossa is the hallmark. Duration of patients' symptoms ranged from 8 months to 37 months (mean, 14 months). There is no significant sex predilection in the occurrence of mineralization of the styloid process; however, symptoms are more common in females. Patients are usually older than 30 years and rarely younger.The pathophysiological mechanisms for the pain of ES include:

(1) Compression of the neural elements, the glossopharyngeal nerve, lower branch of the trigeminal nerve, and/or the chorda tympani by the elongated styloid process;
(2) Fracture of the ossified stylohyoid ligament, followed by proliferation of granulation tissue that causes pressure on surrounding structures and results in pain;
(3) Impingement on the carotid vessels by the styloid process, producing irritation of the sympathetic nerves in the arterial sheath;
(4) Degenerative and inflammatory changes in the tendonous portion of the stylohyoid insertion, a condition called insertion tendinosis;
(5) Irritation of the pharyngeal mucosa by direct compression by the styloid process;
(6) Stretching and fibrosis involving the fifth, seventh,ninth, and tenth cranial nerves in the post-tonsillectomy period[2].

Differential Diagnosis[5,6]

Trigeminal, sphenopalatine,glossopharyngeal, Superior laryngeal neuralgia, myofascial pain, Occipital neuralgia, Nervus intermedius neuralgia, Cephalalgia migraine,Cervicogenic headache,Oromandibular disorder temporoman-dibular joint disorders, impacted, Unerupted or distortedthird molar, Faulty dental prostheses, Sialolithiasis,Tonsillitis, Otitis Mastoiditis, temporal arteritis; dental pain; chronic tonsillitis or pharyngitis; submandibular sialadenitis or sialolithiasis; esophageal diverticulosis; benign or malignant neoplastic disease; and pharyngeal foreign bodies, Inflammatory and neoplastic processes in the oropharyngeal and esophageal areas and other psychosomatic diseases[5,6].

Table 2; Shows the differential diagnosis of Eagle's syndrome which may help the clinicians in practice[11].

	Eagle's syndrome	Pulpitis (Tooth pain)	Idiopathic trigeminal neuralgia	Temporo-mandibular disorder
Type of pain	Dull	throbbing	Current like pain	Dull stabbing
Intensity of pain	Medium	Slight to severe	Severe	Moderate
Duration of pain	Short(especially upon moving head)	Can be upto hours	Seconds	minutes to hours
Characteristic features	Dysphagia, pain upon head movement,pain in neck, throat, floor of mouth, ear	Sharp/ instantaneous, cold or hot, nocturnal/ diumal sensitivity	Trigger zone	TMJ/Muscle pain, inability to open mouth,pain during mastication
Local treatment	Corticoid injection surgery.I. Exremal approach, II. Internal approach	Dental treatment usually root carral therapy. pain may subside upon injection local anaesthesia		physical therapy, splints, anaesthetic injection
General Treatment	NSAIDs	NSAIDs	Anti convulsants	NSAIDs

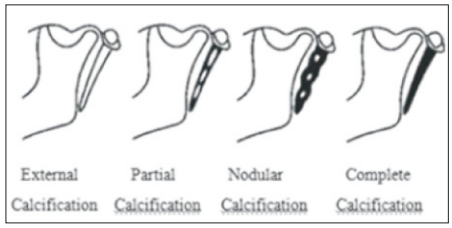
Diagnosis and Radiographic Evaluation[11]

Diagnosis is the pillar of any medical treatment. Without a proper diagnosis, the treatment success is impeded. A sound clinical knowledge of the condition makes diagnosis easier, and avoids unnecessary intervention of multiple medical specialities. As a rule, the workup of patients with suspected eagle's syndrome should include a detailed medical history, as well as a thorough head and neck examination to rule out other diagnoses. Attempts should be made to reinitiate the symptoms by manual manipulation over the stylohyoid complex cautiously. This can localize the pain trigger point while the patient performs cervical and oral movements. Palpation of the tonsillar fossa may reveal the tip of the styloid process as a hard bony spicule that aggravates symptoms with local tenderness. Transpharyngeal palpation ascertains the presence of a bony projection and gives rise to the characteristic Eagle's pain[11].

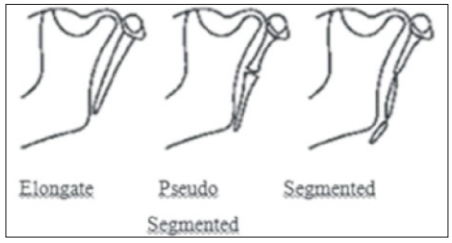
ES can be asymptomatic, while some patients present with cervicofacial pain in the distribution of the carotid artery, neuralgia of the pharynx, dysphagia, and alteration in taste. Other patients present with chronic headaches with pain in the ophthalmic and the occipital regions when they turn their heads. This pain is classically seen due to bilateral internal carotid artery compression.The key diagnostic presentations of ES according to otolaryngologists include a patient with throat pain radiating to the ear post tonsillectomy and a patient with a throbbing pain through either the external or the internal carotid artery distribution[12].

Classification

Figure 1[11]



The classification according to calcification proposed by Langlais (1986)



The morphological classification as proposed by Langlais (1986)

Pattern of calcification[13]

Calcified outline : This pattern describes a thin radiopaque border with a central radiolucency

Partially calcified : This pattern shows a process that has a thicker radiopaque outline, with almost complete opacification but a small, sometimes discontinuous,radiolucent core

Nodular complex : This pattern has a knobby or scalloped outline. it may be partially or completely calcified with varying degree of central radiolucency.

Completely calcified : This pattern is totally radiopaque,with no evidence of a radiolucent interior.

Morphologic classification[13]

Type1: Mineralized complex is characterized by uninterrupted integrity of styloid image

Type2: The styloid process joined to the mineralized stylomandicular or stylohyoid ligament by a single pseudoarticulation.

Type 3: consists of short or long noncontinuous portions of the styloid processor interrupted segments of mineralized ligament.

Radiologic features[11]

With classic presentations of eagle's syndrome, radiograph is not imperative for diagnosis. However, sometimes in case of misleading clinical presentation and missing physical findings, imaging can be very helpful in diagnosing a clinical suspicion of Eagle's syndrome. Radiographic evaluation is the ultimate step to confirm diagnosis and subsequently precedes the treatment required. Different types of imaging have been used to diagnosis Eagle's syndrome which include panoramic radiograph, lateral head and neck radiographs, Towne's radiographs, lateral oblique radiograph of mandible, antero-posterior head radiograph and computed tomography.

In some cases, lateral neck view is enough to confirm the diagnosis. Nonetheless, the advantage that modified Towne's view and orthopantomogram (OPG) have on it is that the entire length of the process can be seen very acutely and hence its deviation can be made out clearly . However, sometimes superimpositions of anatomical structures often hamper the visual clarity of the process. A CT scan usually allows for the precise measurement of the styloid process length, direction, and anatomic variance and a three-dimensional (3D) volume-rendering CT scan add value to the modern diagnostic methods. Three-dimensional CT is considered by most to be the radiologic test of choice for diagnosis of Eagle's syndrome because it is the most advanced technique available to accurately measure the length of the styloid process and hyoid bone.

Treatment

Once diagnosis is established, the mode of treatment can easily be chosen. Eagle's syndrome can be treated either medically or surgically. In medical therapy, oral medication like non-steroidal anti- inflammatory drugs (NSAIDS) with antidepressants and anticonvulsants like (gabapentin,amitriptyline,valproate, carbama-zepine).can be given. Several alternative conservative methods such as transpharyngeal infiltration of steroid or local anesthetics have been used on the tonsil or tender areas[4,11].

Scheme of a Complex Treatment[14]

1. One of the main components of this treatment is the use of glucocorticosteroid hormones. Most frequently we prescribed hydrocortisone. 25 mg (1 ml) of this preparation were diluted with 1 ml 0.25% of lidocaine or other local anesthetic solution and injected close to the styloid process.
2. Anti-epileptic preparations compose the second component of a complex conservative treatment. On the first day 400 mg of carbamazepine (Finlepsin, Stazepin, Tegretol) are prescribed; later the dose is increased up to 600-800 mg.Such treatment lasts from 2 to 3 weeks.
3. Anti-histamine preparations. For example, 0.025 g of diprazine 3 times a day.
4. The supporting component consists of the medicaments widening the blood vessels, neuroleptics, antidepressants and tranquilizers.The results of a complex conservative treatment were evaluated in the following way: good effect when fits of pain do not repeat; improvement when fits repeat rarely and the pain is not strong or nagging.

Surgical treatment was applied to those patients for whom a conservative treatment appeared to be inefficient.[14]

The most satisfactory and effective treatment is surgical shortening of the styloid process through either an intraoral or external approach . The most significant advantage of

an external approach is enhanced exposure of the styloid process and the adjacent structures, and this outweighs all other considerations.[2]

Intraoral approach	Extraoral approach
• No external scar • Shorter operative time • Restricted operative field • Increased risk of deep cervical infection (cleancontaminated wound)	• Cutaneous scar • Risk of facial nerve injury • Better visualization of the surgical field • Decreased risk of deep space neck infection (clean wound)

Table 3 Comparison of the intraoral and extraoral surgical approaches to the styloid process for Eagle syndrome

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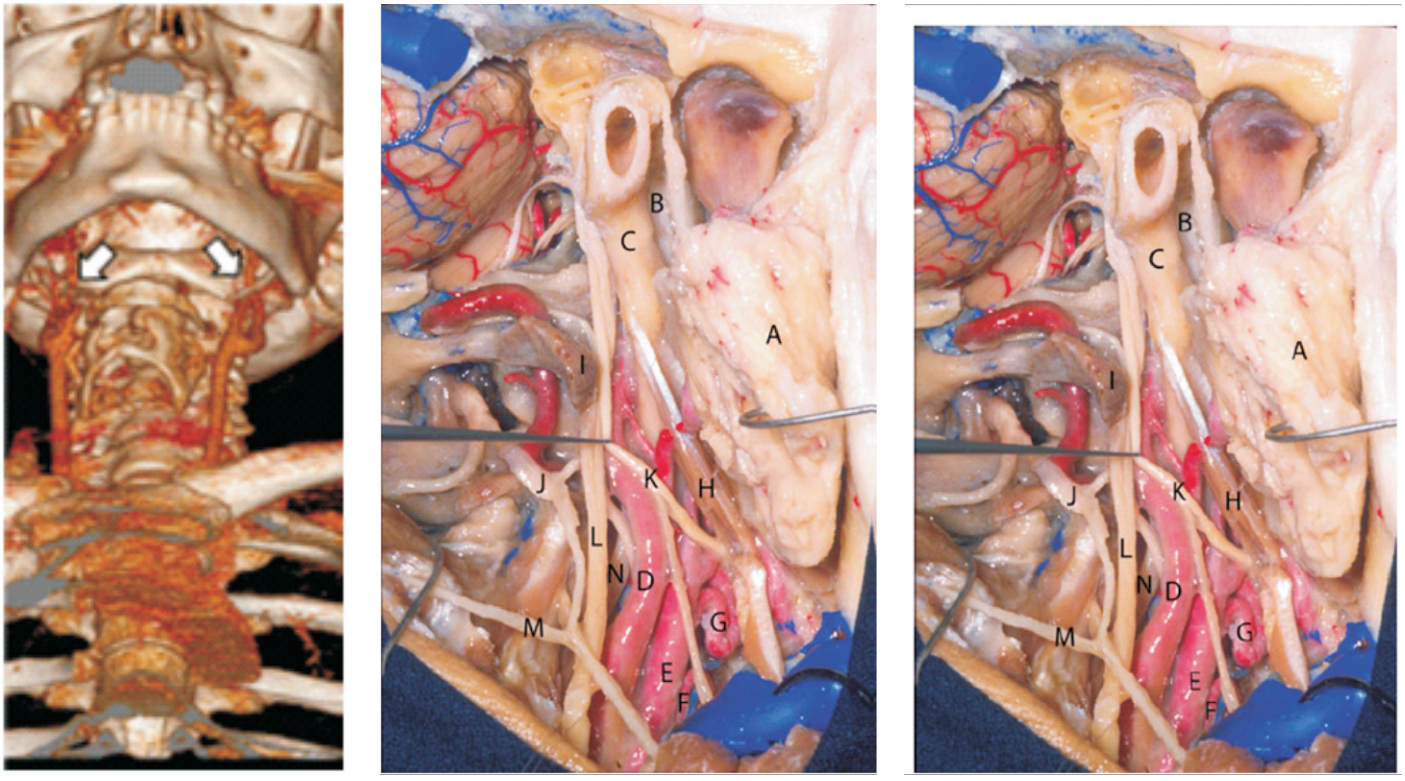

Relationship of the styloid process to critical infratemporal fossa neurovascular structures. The mandibular ramus and condyle, mastoid part, ipsilateral occipital bone, and posterior belly of the digastric muscle have been removed to expose the styloid process, which is lateral to the jugular foramen. The jugular vein has been removed The internal carotid artery ascends to enter the carotid canal in front of the jugular foramen. The vagus, accessory, and hypoglossal nerves descend between the carotid artery and internal jugular vein in the area immediately below the jugular foramen. After the glossopharyngeal nerve (not shown here) exits the jugular foramen, it turns forward, crossing the lateral surface of the internal carotid artery immediately medial to the styloid process. A parotid gland (retracted), B temporal bone, tympanic part, C styloid process, D internal carotid artery, E external carotid artery F superior thyroid artery, G lingual artery, H styloglossus muscle, I C1 transverse process, J C2 ventral ramus, K hypoglossal nerve, L cervical sympathetic chain, M ansa cervicalis, N vagus nerve.