



Clinical Study of Elongated Styloid Process/ Eagle's Syndrome in Five Cases with review of literature.

KEYWORDS

Eagle's syndrome, Styloid process, 3D CT, facial neuralgia.

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ABSTRACT *INTRODUCTION: Eagle's syndrome (ES) occurs when an Elongated Styloid Process or calcified stylohyoid ligament causes recurrent throat pain or foreign body sensation, dysphagia, or facial pain. The symptoms related to this condition can be often confused with a wide variety of facial neuralgias. Elongated styloid process occurs in 4 % of the general population, and only a small percentage of these patients are symptomatic. The normal adult Styloid process length is considered to be between 20mm to 30mm. The longest recorded elongated Styloid process so far that caused symptoms and hence underwent surgery was around 6.3 cms though in an adult human dry skull up to 8cms have been recorded. MATERIALS AND METHODS: Cases admitted in the Department of ENT & referred from neurology and medical ward, with neck pain, Ipsilateral earache and dysphagia from Jan 2015 To July 2015 at Government General Hospital ,Kurnool studied to relate our experience of 5 cases of elongated Styloid within a review of literature and discuss its diagnosis, treatment and follow up. CONCLUSION: An Elongated Styloid Process Syndrome can be diagnosed by a detailed history, physical examination, and radiological investigations x-ray skull, and 3DCT scan face and skull. The treatment of ES is primarily surgical, Resection/ Reduction of the elongated styloid process is the treatment of choice. The Styloid process can be shortened through an intraoral or external approach. In our series all the cases underwent Tran's pharyngeal intra oral tonsillar fossa approach with or without tonsillectomy and immediate postoperative period was uneventful with relief of symptoms with no complications.*

Introduction:

An abnormally long Styloid process or stylohyoid chain ossification producing cluster of symptoms gives rise to "Eagle's syndrome" (ES) or "Stylohyoid syndrome", (Winkler et al.,1981; Catelani and Cudia, 1989; Babad, 1995; Chouvel et al.,1996; Feldman, 2003) which is characterized by craniofacial or cervical pain. ES is a rare entity which is not commonly suspected in clinical practice (Fini et al., 2000), 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 (Murtagh et al., 2001). Patients with ES may present with a sore throat, ear pain, or even with foreign body symptoms in the pharynx secondary to pharyngeal and cervical nerve interactions. ES was first documented by Watt W. Eagle an otorhinolaryngologist in the year 1937 (Eagle, 1937). Over a twenty year period, Eagle reported over 200 cases and explained that the normal Styloid process is approximately 2.5–3.0 cm in length. He observed that slight medial deviation of the Styloid process, could result in severe symptoms of atypical facial pain (Eagle, 1949; Breault, 1986). We are herewith reporting our experience of treating 5 cases of elongated Styloid process at ENT Department, Government General Hospital, Kurnool.

Case Reports & Results:

CASE 1: A 43 years old female patient presented with chief complaints of pain in the right side of neck since 1 year, insidious in onset, initially was dull to moderate & gradually progressed to severe intensity for the past 1 month. Pain initially was intermittent & later on became continuous type. Pain increases on opening the mouth and during swallowing pain not radiating to ear and not

relieved by medication. Intraoral palpation over right Peritonsillar fossa revealed bony hard projection noted with circular outline and left tonsillar fossa was normal.

CASE 2 : A female patient aged 40 years old presented with chief complaints of pain in the right side of neck since 10 months, insidious in origin, initially was dull to moderate & gradually progressed to severe intensity for the past 3 months. Pain was continuous type and increases on opening the mouth and during swallowing. Intraoral palpation revealed tenderness over right Peritonsillar fossa.

CASE 3: A female patient aged 32 years old presented with chief complaints of pain in the right side of neck since 8 months insidious in origin was dull to moderate intensity, continuous type with pain radiating to right ear. Pain increases on opening the mouth and during swallowing. Intraoral palpation over right Peritonsillar fossa revealed tender bony hard projection.

CASE 4: A male patient aged 62 years old presented with chief complaints of pain in the left side of neck since 10 months, insidious in origin, initially was dull to moderate & gradually progressed to severe intensity for the past 3 month. Pain was continuous type and increases on opening the mouth and during swallowing. Intraoral palpation revealed tenderness over left Peritonsillar fossa with bony hard feel and right tonsillar fossa was normal.

CASE 5 : A male patient aged 66 years old presented with chief complaints of pain both sides of neck (right > left) since 6 months, insidious in origin, initially was dull to moderate & gradually progressed to severe type for

the past 2 months. Pain initially was intermittent & later on became continuous type. Pain increases on opening the mouth and during swallowing with radiation of pain to right ear and not relieved by medication. On extra oral physical examination, tender bony hard projection noted in right submandibular area at the anterior border of sternocleidomastoid. Intraoral palpation over right Peritonsillar fossa revealed tender bony hard projection and left tonsillar fossa showed only tenderness. In all the cases preoperatively (2% xylocaine) local anesthetic were injected into tonsillar fossa and had relief of symptom pain temporarily which was used as a diagnostic tool.

All the cases were planned for surgical removal of ES with thorough investigations like HB %, bleeding time, clotting time Blood Grouping & Rh typing, red blood cell count, white blood cell count, platelet count differential WBC count, random blood sugar, blood urea, serum creatinine,

serum electrolytes, screening tests for HIV, HbSAg, HCV, complete urine examination, ECG, diagnostic imaging procedures like x- ray skull and chest, 3D CT scan skull and face, Orthopantomogram. The details of the cases are shown in table 1 as per age, sex, side and duration of symptoms, size of styloid process and tonsillectomy procedure. In our series there were 3 female and 2 male cases and age ranging from 35 – 66 years. In four cases predominant symptom is on right side among this one case it is on left side and one case had bilateral symptoms. All the five cases were planned for reduction of ES and posted under general anesthesia through Trans pharyngeal intra oral tonsillar fossa approach ,only in the first case and fifth case tonsillectomy done to approach and to reduce size of elongated Styloid process and in other cases tonsillectomy was not done. The length of Styloid process ranged from 2.1 cm to 4.1 cm (Normal was 2.5 to 3.0 cm).The duration of symptoms ranged from 6 to 12 months.

TABLE 1 Showing details of cases of Elongated Styloid Process.

Name	Age	Sex	Symptom	Duration	Right	Left	Tonsillectomy
Case 1	43 Yrs	F	Right	12 m	3.62 cm	3.22 cm	Done
Case 2	40 Yrs	F	Right	10 m	3.2 cm	2.1cm	Not done
Case 3	35 Yrs	F	Right	08 m	3.8 cm	2.5cm	Not done
Case 4	62 Yrs	M	Left	10 m	2.5 cm	3.5 cm	Not done
Case 5	66 Yrs	M	Both	06 m	4.1 cm	3.4 cm	Done

Figure 1 Showing imaging of X- Ray Skull AP Lateral view and Orthopantomogram, figure 2 Showing an Elongate Styloid on 3D CT Scan skull and face, figure 3 Showing application of ring curette to base of Styloid during Surgery and figure 4 shows images of excised Styloid process variable sizes minimal 1 to 1.5cm is removed



FIGURE 1: Imaging showing x- ray skull (AP & lateral view) and Orthopantomogram

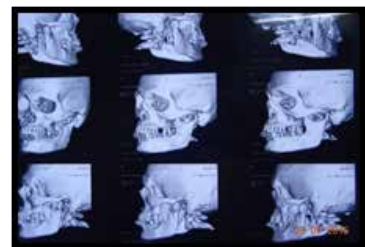


Figure 2 : 3D CT Scan of Face

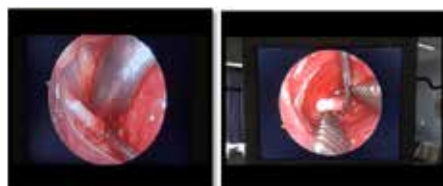


Figure 3: Surgical image application of ring curette to Styloid



Figure 4 : Images showing excised Styloid process

Discussion: The Styloid process, stylohyoid ligament, and the lesser horn of the hyoid bone form the stylohyoid apparatus, embryologically derives from the Reichert cartilage of the second brachial arch during embryogenesis. The Styloid process is the long and thin osseous part of the temporal bone, lies caudally, medially, and anteriorly towards the maxillo-vertebro-pharyngeal recess, which involves the carotid arteries, internal jugular vein, facial nerve, glossopharyngeal nerve, and the vagal and hypoglossal nerves Eagle's syndrome sometimes called as Styloid or stylohyoid syndrome, is defined as the symptomatic elongation of the Styloid process or mineralization (ossification or calcification) of the stylohyoid ligament complex (Monsour et al., 1985; Chauvel et al.,1996). From Eagle's early descriptions, patients were categorized into two groups: those who had classical symptoms of a "foreign body" lodged in the throat with a palpable mass in the tonsillar region following tonsillectomy; and those with pain in the neck following the carotid artery distribution (carotid artery syndrome).

The mere presence of an elongated styloid process or mineralization of the stylohyoid complex radiographically in the presence of cervico-pharyngeal pain does not automatically confirm a diagnosis of ES. There are reasons from three aspects. First, many patients with an ossified stylohyoid complex are asymptomatic. Second, there does not appear to be any correlation between the severity of pain and the extent of ossification of the stylohyoid complex. Finally, the majority of symptomatic patients have had no recent history of tonsillectomy or any other cervicopharyn-

geal trauma (Camarda et al., 1989).

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 and can be reproduced by palpation of the tonsillar fossa is the hallmark (Pierrakou, 1990). 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 (Keur et al., 1986; Bafaqeeh, 2000; Ceylan et al., 2008). *Embryological Basis of Styloid Process* 1) *Tympanohyal part - the base of the Styloid process* 2) *Stylohyal part - forms a large part of the Styloid process* 3) *Ceratohyal part - precursor of the stylohyoid ligament* 4) *Hypohyal part - development precedes the small horn of the hyoid bone. It is believed that ceratohyal part of the second branchial arch contains small parts of embryonal cartilage that may or may not, at a later stage, mature into bone*

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 tendinous 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 (Ceylan et al., 2008).

The diagnosis of ES must be based on a good medical history and physical examination. It should be possible to feel an elongated Styloid process by careful intraoral palpation, placing the index finger in the tonsillar fossa and applying gentle pressure (Montalbetti et al., 1995). If pain is reproduced by palpation and either referred to the ear, face, or head, the diagnosis of an elongated Styloid process is very likely. A Styloid process of normal length is usually not palpable. Injection of local anesthetic into tonsillar fossa relieves pain and can be used as a diagnostic tool (Prasad, 2002).

The diagnosis of ES can be ascertained with imaging which includes lateral head and neck radiograph, Towne radiograph, panoramic radiograph, lateral-oblique mandible plain film etc. In radiographs a threshold length of 3 cm is accepted as abnormal by current conventions. Plain radiographs are the commonest modality chosen. Lateral views are the best to show the length of the Styloid process, but antero-posterior views are also needed to determine whether there is bilateral involvement and the presence of lateral deviation. CT scans have been used in difficult cases to supplement diagnosis.

The 3-D reconstruction made it possible to visualize the exact spatial orientation of the Styloid process. Imaging only demonstrates elongation of the styloid process and

proximity to surrounding key structures. The diagnosis of ES is based on multiple clinical and radiographic factors.

One radiographic classification system includes three types of radiographic appearances. The Type I pattern represents an uninterrupted, elongated Styloid process. Type II is characterized by the Styloid process apparently being joined to the stylohyoid ligament by a single pseudo articulation. This gives the appearance of an articulated elongated Styloid process. Type III consists of interrupted segments of the mineralized ligament, creating the appearance of multiple pseudo articulations within the ligament (Langlais et al., 1986).

Another classification method is by means of calcification (4 types):- Surface calcified Styloid process -Partially calcified Styloid process- Nodular type calcification- Completely calcified Styloid process.

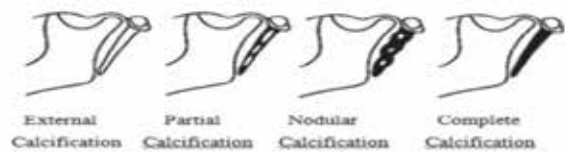


Figure 5: Calcification Classification proposed by Langlais 1986



Figure 6; Image pituitary ring curette

In differential diagnosis, Laryngopharyngeal dysaesthesia has to be considered as well as dental malocclusion, neuralgia of sphenopalatine ganglia, Temporomandibular arthritis, glossopharyngeal and trigeminal neuralgia, chronic tonsillopharyngitis, hyoid bursitis, Sluder's syndrome, histamine cephalgia, cluster type headache, esophageal diverticula, temporal arteritis, cervical vertebral arthritis, benign or malignant neoplasm's, and migraine type headache (Harma, 1966).

The elongated styloid process syndrome can be treated conservatively or surgically. Conservative treatment options have included transpharyngeal injection of steroids and lignocaine, nonsteroidal anti-inflammatory drugs, diazepam, the application of heat, and transpharyngeal manipulation with manual fracturing of the Styloid process. It should be noted that blind fracture of the Styloid process does not usually relieve symptoms and risks damage to nearby neurovascular structures. The most satisfactory and effective treatment is surgical shortening of the Styloid process through either an intraoral or external approach (Boedts, 1978; Zhang et al., 1987; Chase et al., 1986; Beder et al., 2005; Chrcanovic et al., 2009). The most significant advantage of an intraoral transpharyngeal tonsillar fossa approach over external is minimal trauma to surrounding structures with no scar and complete relief of symptoms and in all cases tonsillectomy is not mandatory to reach elongated Styloid. External approach is helpful and facilitates when the resection of a partially ossified stylohyoid ligament. Trans oral resection causes no outside scars, but in-

volves the risk of deep cervical infection and possible neurovascular injury (Chase et al., 1986;Ceylan et al., 2008)..

Conclusion : Elongated Styloid process/ Eagles syndrome causes recurrent throat pain and facial pain diagnosed by detailed history, physical examination, digital palpation in the tonsillar fossa, confirmed by radiological examination with 3D CT of skull and face. It can be confused with many other pain syndromes that must be excluded. Resection or reduction of the elongated Styloid process is the treatment of choice. Intraoral tonsillar fossa approach with are without tonsillectomy is a safe procedure with no scar and without any complications over external approach. The application and use of pituitary ring curette in all our cases, found helpful in stabilizing the base of Styloid process during surgical resection and can be used to avoid complications/injury to important structures.

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