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CASE REPORT

Eagle's syndrome – A case report and review of the literature

Suneet Khandelwal ^{a,*}, Yajuvender Singh Hada ^b, Ashutosh Harsh ^c

^a Department of Oral Pathology & Microbiology, Jaipur Dental College, Jaipur, Rajasthan, India

^b Department of Conservative & Endodontics, Daswani Dental College, Kota, Rajasthan, India

^c SN Medical College & Hospital, Jodupur, Rajasthan, India

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Abstract 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. Additional symptoms may include neck or throat pain with radiation to the ipsilateral ear. The symptoms related to this condition can be confused with those attributed to a wide variety of facial neuralgias. ES can be diagnosed radiologically and by physical examination. The treatment of ES is primarily surgical. The styloid process can be shortened through an intraoral or external approach. In this paper a case of ES exhibiting unilateral symptoms with bilateral elongation of styloid process is reported and the literature is reviewed.

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1. 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. Since the symptoms are variable and non-specific, patients seek treatment in several different clinics such as otolaryngology, family practice, neurology, neurosurgery, psychiatry, and the last but not the least dentistry.

* Corresponding author. Tel.: +91 9785010007.

E-mail addresses: khandelwalsuneet@yahoo.co.in (S. Khandelwal), coolhadays@rediffmail.com (Y.S. Hada), harshcazri@gmail.com (A. Harsh).

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2. Case report

A 56-year-old male patient presented to the dental clinic with a complaint of pain in upper neck region of six months duration. The pain was insidious in origin, dull to moderate in intensity and intermittent in nature. The intensity of pain was exacerbated by movements such as looking up and turning face to left side. In addition, patient also had a sensation of foreign body in throat on swallowing. The patient gave a history of fracture in mandibular symphysis region 6 months back.

Extra-oral physical examination revealed a tender, small bony hard projection in right submandibular area at anterior border of sternocleidomastoid muscle, circular in shape and approximately 4 mm in diameter (Fig. 1). In addition, the patient presented with tenderness on palpation of the right tonsillar fossa.

Radiographic examination (Mandibular lateral oblique {ramus view} – Orthopantomogram – Fig. 2) demonstrated elongated styloid process, measuring 6 cm on left side and 5.5 cm on right side approximately. Subsequent to the radiographic findings re-examination of the patient revealed a hard bony mass which was palpated in the left tonsillar fossa but did not produce any symptoms.

The patient was given a primary diagnosis of ES. The patient underwent a chair side infiltration of 2% lignocaine in the right tonsillar fossa which led to immediate relief from the symptoms.

Finally, the elongated styloid process of the right side was resected surgically by the extra-oral approach through an incision that extends from the mastoid process along the sternocleidomastoid to the level of the hyoid then up across the neck to the midline of the chin under general anaesthesia (Figs. 3–5). Amoxicillin and ibuprofen were administered once preoperatively and postoperatively. The patient was discharged on the third postoperative day. The patient was



Figure 1 Clinical photograph showing enlarged styloid process on right side of patient.

asymptomatic at follow up at 1 year and did not have any complaint on the left side also.

3. Discussion

ES, sometimes called 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; Chouvel et al., 1996). 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).

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). Although these two types have a common etiology, their symptomatology differs (Breault, 1986). The presented case represents "classical syndrome" symptoms.

However, 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



Figure 2 Orthopantomogram – preoperative showing enlarged styloid process on both sides.



Figure 3 Excised styloid process (right side).

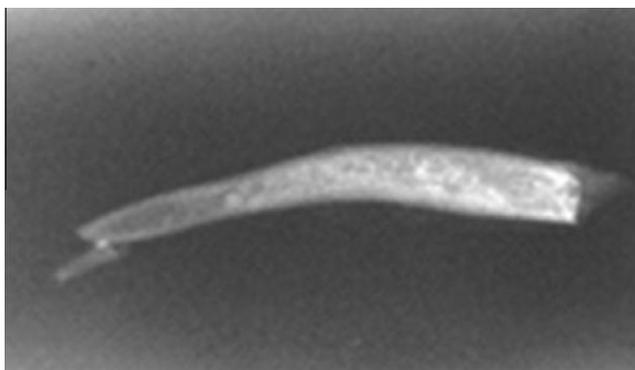


Figure 4 Radiograph of excised styloid process (right side).



Figure 5 Orthopantomogram – postoperative showing enlarged styloid process only on left side.

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 cervicopharyngeal trauma (Camarda et al., 1989). Despite this, the literature still categorizes patients into those with a pain pattern following the carotid artery distribution and those with a classical palpable mass in the tonsillar region.

In a review of 1771 panoramic radiographs, the incidence of mineralization of the stylohyoid complex was found to be 18.2% (Correll et al., 1979). The incidence of elongated styloid process was estimated at 3.3% out of which 55% bilateral cases in panoramic radiographs, and the male/female ratio was 1/9 in the radiographs. Average age was 43.35 ± 14.88 years and no significant difference was found in the ages of the participants, according to gender (Balcioglu et al., 2009). Despite these figures, only 1–5% of patients are symptomatic (Langlais et al., 1986; Fini et al., 2000). Literatures are not in agreement with sexual dimorphism of ES. Woolery stated that Eagle's syndrome occurs more frequently in women while others do not (Woolery, 1990; Balcioglu et al., 2009).

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 (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).

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 cause of onset of pain in patients previously free of symptoms is unknown, but several mechanisms have been proposed that include rheumatic styloiditis caused by pharyngeal infections, trauma, tonsillectomy, and involutional changes associated with aging (e.g., degenerative cervical discopathy, which may shorten the cervical spine and alter the direction of the styloid process) (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 anaesthetic 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 visualise the exact spatial orientation of the styloid processes. An ossification of the stylohyoid ligament could definitely be ruled out on the basis of the imaging procedures. Spiral-CT with subsequent 3-D reconstruction is the method of choice for exact determination of the localization. Also, barium swallow studies can show the indentation of the elongated styloid process as a filling defect (Dayal et al., 1971; Keur et al., 1986; Murtagh et al., 2001; Nickel et al., 2003). 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 and is the type present in this reported case (Figs. 2 and 3). Type II is characterized by the styloid process apparently being joined to the stylohyoid ligament by a single pseudoarticulation. 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 pseudoarticulations within the ligament (Langlais et al., 1986).

An enlarged styloid process may also compress upon the internal carotid artery, leading to transient ischemic attack and may pose a threat. Sudden death by vagus mediated cardiac inhibition due to ES has also been reported. The diagnosis was established only after the autopsy examination, which revealed the elongation of the styloid processes compressing both carotid sinuses. The anatomical changes were not visible in the radiological films. (Das et al., 2008; Ruwanpura et al., 2008). ES can be atypically present as exertional headache, beginning in the right ear and radiating to the neck and to the vertex (Maggioni et al., 2009). A rare case with diffuse bilateral stylohyoid chain ossification which can mimics symptoms of ES was diagnosed as temporomandibular joint disorder (Ramadan et al., 2007).

In differential diagnosis, laryngopharyngeal dysesthesia has to be considered as well as dental malocclusion, neuralgia of sphenopalatine ganglia, temporomandibular arthritis, glossopharyngeal and trigeminal neuralgia, chronic tonsillo-pharyngitis, hyoid bursitis, Sluder's syndrome, histamine cephalgia, cluster type headache, esophageal diverticula, temporal arteritis, cervical vertebral arthritis, benign or malign neoplasms, 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, traditional Chinese medicines, 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 external approach is enhanced exposure of the styloid process and the adjacent structures, and this outweighs all other considerations. It also facilitates the resection of a partially ossified stylohyoid ligament. Transoral resection causes no outside scars, but involves the risk of deep cervical infection and possible neurovascular injury (Chase et al., 1986; Ceylan et al., 2008). The reported case was successfully treated using an external approach.

Prognosis of ES is guarded by surgical failures (up to 20% of patients). This may be due to intraoperative injury, subsequent fibrous entrapment syndrome, or inadequate shortening of the process, assuming that the diagnosis was correct in the first place (Ghosh and Dubey, 1999).

4. Conclusion

The elongated styloid process syndrome can be diagnosed by a detailed history, physical examination, and radiological investigations. It can be confused or mistaken for many other conditions that must be excluded. Resection of the elongated styloid process is the treatment of choice. An awareness of pain syndromes related to the styloid process is important to all health practitioners involved in the diagnosis and treatment of neck and head pain to rationalize the line of management and the ultimate clinical outcome as a "cancerphobia" may cause great emotional distress.

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