EAGLE’S SYNDROME: A CASE REPORT.

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Abstract

Eagle’s syndrome (ES) occurs when an elongated styloid process or calcified stylohyoid ligament causes compression of adjacent anatomical structures leading to oro facial pain. Pain often improves with excision of styloid process.

Introduction:

Eagle’s syndrome was first described by an American Otorhino laryngologist Watt weems Eagle in 1937. 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. Eagle’s Syndrome is an 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 or a calcified stylohyoid ligament which manifests the symptoms (Murtagh et al., 2001). Patients with Eagle’s Syndrome may present with a sore throat, ear pain, or even with foreign body sensation in the throat due to irritation of pharyngeal and cervical nerves.

Case History:

A 45-year-old female patient presented to the ENT OPD with a complaints of pain over the left side of the throat for 4 months and ipsilateral ear pain for the last 1 month. 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 right side. The pain radiates to left ear and left side of neck. In addition, patient also had a sensation of foreign body in throat on swallowing. Examination of oropharynx showed enlarged tonsils on both sides (left > right) and tenderness over left tonsillarlingual sulcus. In addition, the patient presented with tenderness on palpation of the left paratonsillar fossa.

Diagnosis was confirmed with X-ray and CT scan. Radiographic examination demonstrated elongated styloid process, 4.5 cm on right side approximately along with tonsillar cyst on left side. Initially tonsillectomy was done on the both sides. The superior constrictor muscle was divided on the tonsillar bed. By intraoral approach styloid process identified and stripped of its periosteum up to its attachment to the base and excised. Muscle sutured with...
vicryl. Histopathology report of tonsil specimens showed features of tonsillar cyst on left side and chronic tonsillitis on right side. Post operatively the patient was relieved of pain.

**Figure 1:** CT scan slice showing bilateral elongated styloid process (left > right)

**Figure 2:** 3D reconstruction of CT scan showing spatial orientation of bilateral elongated styloid process

**Figure 3:** Photograph showing one of the excised styloid process
Discussion:

Eagle’s Syndrome, 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). Eagle’s Syndrome 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. Coorel et al. (1979) defined the normal length as < 2.5 cm. Lindemann considered normal as 2.5 to 3 cm is considered significant. 4% of the general population is affected by this and out of this only 4% are symptomatic. 1,4 Woolery stated that Eagle’s syndrome occurs more frequently in women (Woolery, 1990; Balcioglu et al., 2009) in a ratio of 1:3. Bilateral is quite common, but symptoms are mostly unilateral.

According to Eagle, 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. In the stylo carotid form compression of external / internal carotid artery by deviated elongated styloid process produces tinnitus, headache and orofacial pain. The pain aggravates typically on rotation of the head.

The pathophysiological mechanisms for the pain of ES
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 (Ceylan et al., 2008).

The diagnosis of Eagle’s Syndrome must be based on a good medical history and physical examination. Relief by injection of Xylocaine over the tonsillar fossa is also a simple bedside diagnostic procedure. 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 can also be used 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.

Radiographic classification system includes three types. The Type I pattern represents an uninterrupted, elongated styloid process and is the type present in this reported case. 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.

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 can be established only after the autopsy examination, which will reveal the elongation of the styloid processes compressing both carotid sinuses.

The elongated styloid process syndrome can be treated conservatively or surgically. Medical treatment includes analgesics, anticonvulsants, antidepressants. 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). Surgical excision can be done by intra oral or extra oral
approaches. The common complications are neck space infection and facial nerve involvement. Careful dissection and good antibiotic coverage pre and post operatively can avoid these complications.

Prognosis of Eagle’s Syndrome 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).

**Conclusion:**
The Eagle’s 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.

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