



ORIGINAL RESEARCH PAPER

Maxillofacial Surgery

BILATERAL EAGLE'S SYNDROME IN LUMBAR SPONDYLOSIS – A CASE REPORT AND REVIEW

KEY WORDS: Diffuse idiopathic skeletal hyperostosis, Eagle syndrome, hypopharyngeal compression, multidetector computed tomography

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ABSTRACT

There are Published case studies involving diffuse idiopathic skeletal hyperostosis (DISH) along with symptomatic ossification of the stylohyoid ligament, or "Eagle syndrome," are rare. Coincidence of the Eagle syndrome and diffuse idiopathic skeletal hyperostosis can very rarely result with compression of the hypopharynx. The three-dimensional volume-rendered air-specific Multidetector computed tomography MDCT image is accurately used to show the relationship between the Oro-hypopharyngeal air column and the elongated styloid processes. The authors here described a very rare case of hypopharyngeal airway compression and nagging pharyngeal pain secondary to cervical osteophytes in a patient with coincidence of Eagle syndrome and diffuse idiopathic skeletal hyperostosis. A known case of ankylosing spondylosis under treatment since last three years with degenerative changes in the cervical spine was taken up for removal of styloid process under General anesthesia. Follow up was done for one year and it was uneventful.

INTRODUCTION:

Eagle's syndrome is caused by an elongated styloid process or calcified stylohyoid ligament. It was defined by Eagle in 1937 (1,2) with characteristics related to an elongated styloid process, which is one of the numerous causes of pain in the craniofacial and cervical region. Eagle's syndrome has also been called: stylohyoid syndrome, styloid syndrome, elongated process syndrome, stylalgia, styloid-stylohyoid syndrome, styloid dysphagia, chronic styloid angina, temporal rheumatic styloiditis, stylocarotid syndrome and the Garel-Bernfeld syndrome. Eagle defined the length of a normal styloid process at 2.5 - 3.0 cm. The styloid process may vary in length and sometimes stylohyoid ligament may also ossify from its origin at the styloid process to its attachment at the hyoid bone mimicking elongated styloid process [3]. Eagle considered any styloid process longer than 25 mm in an adult to be abnormal which may be symptomatic or non-symptomatic. Non symptomatic elongated styloid process is encountered during head and neck radiological investigations, cadaveric dissections and autopsy leading to above estimations of prevalence. Nanjundaiah et al encountered elongated styloid process in two routine cadaveric dissection aged around 55 – 60 years [4]. The prevalence rate of elongated styloid is found to be 4% with symptomatic individuals (5,6,7). Eagle's syndrome is usually characterized by neck, throat, or ear pain; pharyngeal foreign body sensation; dysphagia; pain upon head movement; and headache. He divided the syndrome into two forms: classic type and carotid artery type. The classic type of Eagle's syndrome can develop after tonsillectomy, when scar tissue under the tonsillar fossa compresses and stretches cranial nerves V, VII, IX, and X (8,9). This type includes symptoms such as foreign body sensation, pain referred to the ear, and dysphagia. The carotid artery type of Eagle's syndrome presents with other symptoms, such as migraines, and neurological symptoms, caused by irritation of the sympathetic nerve plexus. Women and patients older than 50 years have a higher tendency to develop mineralization; moreover, the anomaly is often more symptomatic in females (10,11). There is always an association of Eagles Syndrome with other musculoskeletal disorder (12). A relation between diffuse idiopathic skeletal hyperostosis of the cervical spine and ossification of the styloid process has been previously reported (13). Coincidence of the Eagles syndrome and diffuse idiopathic skeletal hyperostosis can very rarely result with compression of the hypopharynx. Diffuse idiopathic skeletal hyperostosis (DISH), also known as Forestier's disease, described as senile ankylosing vertebral hyperostosis, a progressive noninflammatory entheses

disease and characterized by the ossification of ligaments and entheses in the prevertebral regions of the spine. Patients are often asymptomatic or are undiagnosed due to minor chronic and unrevealing symptoms, such as back pain or spinal stiffness. Some patients may present with more severe symptoms, such as dysphagia, spine fractures, and in rare cases, dyspnea. (14). This condition can very rarely present with dysphagia as a result of compression of the hypopharynx. Published case studies involving Diffuse idiopathic skeletal hyperostosis (DISH) along with symptomatic ossification of the stylohyoid ligament, or "Eagles syndrome," are rare.

Case Report

A 43 years old female was referred from the department of neurosurgery for evaluation pain in the floor of mouth and para tonsillar area. She is a known case of ankylosing spondylosis under treatment since last three years. The chief symptoms were vague nagging pharyngeal pain, cervicofacial pain, foreign body sensation in the throat, pain on swallowing, on changing head position, and radiating to the temporomandibular joint, and ear. There was no history of remarkable weight loss. She was examined thoroughly, and pain due to other factors, such as temporomandibular joint, dental, orthopedic, and pharyngoesophageal causes, were ruled out. She had pain on palpation of tonsillar fossa, on which a firm cord like structure was felt. On digital pressure the symptoms and local tenderness were aggravated. All these symptoms are accompanied dysphagia for the last 2–3 years. There is no history of diabetes mellitus, coronary artery disease, and chronic obstructive pulmonary disease. On radiological evaluation, orthopantomogram revealed the length of the styloid process was more than one third of the length of the ramus of the mandible, which was considered to be elongated.

Computed Tomography (CT) scan investigation confirmed the elongated styloid process bilaterally (Fig -1). The left side was 38 mm long and right side was 42 mm long. There was calcification of the stylohyoid muscle and also stylomandibular ligament. There were significant degenerative changes in the spine and its contents. She underwent MRI which also revealed lengthened stylohyoid muscle and stylomandibular ligament confirming the elongation of styloid process. There was also Lumber spondylosis with significant neural / thecal compression. There were degenerative changes of cervical spine. Bilateral uncovertebral osteophytes at C 5/6 level indenting the spinal contents were seen. Disc bulge was also present in relation to

C3/4, C4/5 levels. All cervical intervertebral discs appear degenerated. Craniovertebral junction was normal. Her blood investigations revealed that her Parietal cell antibody positive Titre 1:40.

After preanesthetic assessment she was taken up for resection of both the elongated styloid processes under general anesthesia. The styloid processes were exposed through transoral para tonsillar approach and resected. Hemostasis achieved and the wounds were sutured. The specimens were sent for histopathological examination (Fig-2).

The specimens were decalcified and stained with Hematoxylin and Eosin stain. The microscopic examinations revealed calcification and hyalinization of the muscle fibers thus confirming calcification of the stylohyoid ligament (Fig-3 and Fig-4).

Post operative recovery was uneventful. Post operative CT scan confirmed the excision of elongated styloid process (Fig-5). She was followed up for one year and was asymptomatic.

DISCUSSION

The styloid process is an elongated tapered projection that originates in the petrous portion of the temporal bone, lying medially and anteriorly to the stylomastoid foramen, between the internal and external carotid arteries, and laterally to the tonsillar fossa. The stylopharyngeus, stylohyoid, and styloglossus muscles are attached to the styloid process. This bony process supports the stylohyoid and stylomandibular ligaments. The stylohyoid ligament connects the apex of the styloid process and the lesser horn of the hyoid bone, and the stylomandibular ligament extends from the styloid process to the parotidomasseteric fascia between the mastoid process and the mandible (7). Recently, Murtagh et al. presented three etiologic theories to explain the development of Eagle's syndrome.

The first theory is congenital elongation of the styloid process due to the persistence of the cartilaginous precursor, the second is the calcification of the stylohyoid ligament by a mysterious process, and the third is the growth of osseous tissue at the insertion of the stylohyoid ligament. Steinmann proposed three mechanisms that might cause ossification: 1) Reactive hyperplasia, when trauma activates the remnants of the original connective and fibrocartilaginous cells 2) Reactive metaplasia, or an abnormal healing following a trauma that initiates ossification of the stylohyoid ligament and 3) Anatomic variance, which occurs without any distinctive trauma (7). In our case there was no history of trauma or any other systemic neurological disorder. The etiology of elongation cannot be ascertained. Literature reveals there was no recurrence after excision and our case was also followed up for one year without any recurrence.

DISH is a progressive noninflammatory disease characterized by ossification (bone tissue formation) of the anterior part of the vertebrae (8). It develops chronically and commonly presents with nonacute symptoms, such as back or neck pain, spine mobility restriction, foreign body sensation, and hoarseness. Dyspnea may occur in some cases but rarely has been reported to occur suddenly. In a review article of literature from 1980 to 2010, a total of 189 cases with dysphagia and 63 cases with airway obstruction (including 48 with both) were reported (15).

Coincidence of the Eagle syndrome and diffuse idiopathic skeletal hyperostosis can very rarely result with compression of the hypopharynx. A relation between diffuse idiopathic skeletal hyperostosis of the cervical spine and ossification of the styloid process has been previously reported (16). This condition can very rarely present with dysphagia as a result of compression of the hypopharynx. Hypopharyngeal airway

compression caused by massive cervical osteophytes a rare disorder (17). The Eagle syndrome often results with neck pain. It is rarely associated with vocal cord paralysis and pharyngeal airway compression. Multidetector computed tomography (MDTC) of the neck and head, a multiplanar reconstruction image at the coronal plane, demonstrates the bilateral elongated styloid processes at the level of the oro-hypopharynx.

The three-dimensional volume-rendered air-specific MDCT image is accurately used to show the relationship between the oro-hypopharyngeal air column and the elongated styloid processes. Multidetector computed tomography is the most important diagnostic modality because it can identify the presence of the ligamentous ossification and rule out associated complications such as oro-hypopharyngeal compression. Finally, the standard treatment for airway compression caused by diffuse idiopathic skeletal hyperostosis of the cervical spine or ossification of the styloid process is surgical repair, and the MDCT scan is the most useful diagnostic modality (18).

Diagnosis is guided by the clinical history and physical and radiographic examinations. The physical examination consists of palpation of the tonsillar fossa and local infiltration anesthesia. Trans pharyngeal palpation demonstrates a bony projection and reproduces the characteristic pain. Symptom relief should follow anesthetic injection in the tonsillar fossa. Radiologic evaluation, such as panoramic radiography, lateral cephalometry, Towne projection film or CT, may be used. In the panoramic view, the styloid process is visualized posteriorly to the external acoustic meatus with a descendant and anterior trajectory. When elongated, it attains over one third of the length of the mandibular ramus. CT allows for the precise measurement of the styloid process length, direction, and anatomic variance, in addition to evaluation of stylohyoid ligament ossification (6).

Eagle's syndrome can be treated non-surgically and surgically. A pharmacological approach includes trans pharyngeal infiltration of steroids or anesthetics into the tonsillar fossa. In addition, there are two surgical approaches, intraoral and extraoral. In the intraoral approach, the styloid process is found with palpation of the tonsillar fossa. The overlying mucosa and superior constrictor muscle are incised vertically, and the styloid process are dissected out and resected using a rongeur forceps. If necessary for exposure, a standard tonsillectomy should be performed first. The intraoral approach is good for aesthetic consideration and for shorter operative time. The intraoral approach also has the disadvantages of poor access, as in cases of trismus, and risk of intraoperative injury. An extraoral approach involves making a cervical incision from the proximal portion of the sternocleidomastoid muscle to the hyoid bone, and then dissecting and removing the styloid process. The external approach has the advantage of good anatomic exposure of the styloid process. However, it requires more intervention, results in a visible scar and risk of facial nerve palsy (8).

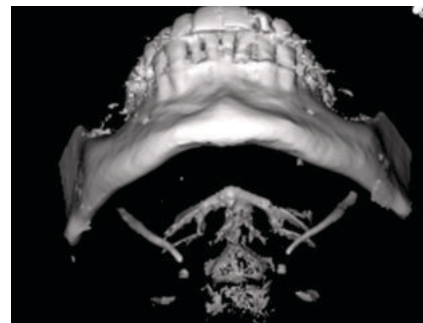


Fig -1 CT scan 3D reformatted image showing elongated styloid processes.



Fig-2 Resected specimen

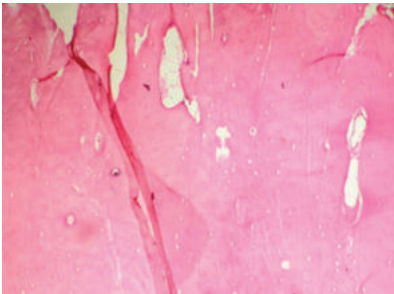


Fig-3 Microphotograph (10X) showing hyalinization

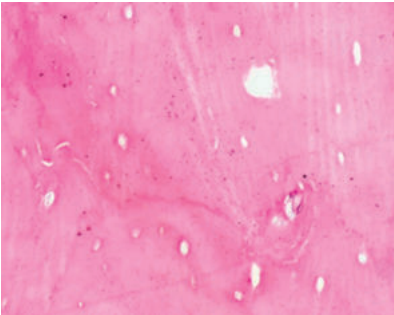


Fig-4 Microphotograph (40X) showing calcification

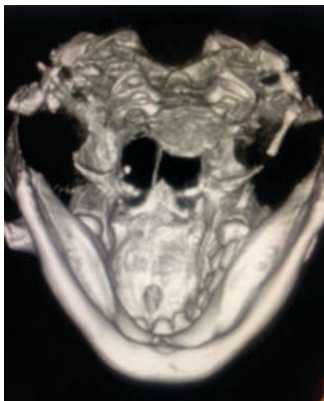


Fig-5 Post operative CT scan image

Conflict Of Interest – Nil.

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