MANAGEMENT OF EAGLE’S SYNDROME- CASE REPORT.

Dr. Chinmaya Sundar Ray¹, Dr. Krishna Arpita Sahoo², Dr. Kaushik Sarangi³ and Dr. Raisa Priyadarshini⁴.

1. Assistant professor, Department of ENT & Head & Neck Surgery, S.C.B Medical College, Cuttack-753007, Orissa, India.
2. Senior Resident, Department of ENT & Head & Neck Surgery, S.C.B Medical College, Cuttack-753007, Orissa, India
3. Postgraduate student, Department of ENT & Head & Neck Surgery, S.C.B Medical College, Cuttack-753007, Orissa, India
4. Postgraduate student, Department of ENT & Head & Neck Surgery, S.C.B Medical College, Cuttack-753007, Orissa, India.

Abstract

Eagle’s syndrome is the term given to the symptomatic elongation of the styloid process or mineralisation of the stylohyoid or stylomandibular ligament or posterior belly of the digastric muscle. Patients present with pain in throat localised in the tonsillar fossa or hyoid bone, hypersalivation, foreign body sensation, rarely voice change. The pain is triggered by head rotation, lingual movements, swallowing or chewing. Diagnosis is usually made on physical examination by digital palpation of the styloid process in the tonsillar fossa and radiographically by panoramic radiograph, computed tomography scan, magnetic resonance imaging. The clinical signs and symptoms are non-specific, so the correct diagnosis is most important.

Introduction:-

ES is a rare syndrome which is not commonly suspected in clinical practice (Fini et al., 2000). In 1652, Pietro Marchetti first described an elongated styloid process related to an ossifying process of the stylohyoid ligament. Eagle, an otolaryngologist, later defined “Eagle's syndrome” in 1937. [1] The styloid process is a protrusion of a long, thin, cylindrical bone from the petrous portion of temporal bone. It is located medially and anteriorly to the stylomastoid foramen, between the internal and external carotid arteries, and laterally to the tonsillar fossa. [2, 3] Typical styloid processes are between 2.5 and 3.0cm in length. Lengths greater than 3cm are considered elongated. Patient develops cluster of symptoms arising from an elongated styloid process and/or an ossified stylohyoid ligament, secondary to irritation or compression of the neurovascular bundle within the carotid sheath. Presentations can vary from craniofacial or cervical pain and a globus sensation to carotid dissection, transient ischemic attacks and stroke. [4] The prevalence of ES is 4% and 0.16% of these individuals are found to be symptomatic. [5] We present a case of Eagle’s syndrome describing its clinical presentation, diagnosis and management.

Case Report:-

A 43-year-old female patient presented to our ENT hospital with a complaint of pain in her ear & neck region on right side from past 8 months. The pain was abrupt in onset, mild dull aching to moderate in intensity and intermittent in nature. Pain was aggravated on tilting the head to right side and on mouth opening. Patient also
complained of foreign body sensation in throat. There was mild ill-defined tenderness on palpation over right retromandibular area. Examination of the TMJ was normal, with full range of jaw movement, no muscle tenderness, and no clicking sound from the joint. Face was apparently symmetrical bilaterally. There was no evidence of any mass in neck. Examination of oral cavity and oropharynx was normal. Tenderness was present on deep palpation of right tonsillar and para-tonsillar area.

In Radiographic examination, elongated right styloid process was seen in orthopantogram. CT scan of neck revealed elongated right styloid process measuring approx. 6 cm on right side and 3 cm on left side. (Fig.1 & Fig.2)

![Fig.1: Enlarged Right styloid process](image1)

![Fig.2: Normal Left Styloid process](image2)

We diagnosed this patient with Eagle's syndrome based on history, clinical features and radiographic examination. The patient underwent infiltration of 2% lignocaine in the right tonsillar fossa which led to immediate relief from the symptoms. Differential diagnosis for eagle’s syndrome includes temporomandibular disorders, carotid artery syndrome, tonsillitis, impaction, neoplasia, otolaryngeal infections, coronoid hyperplasia, neuralgias, oral submucous fibrosis etc. Patient had received conservative treatment with analgesics and antibiotics for 1 month. But as the pain was not relieved, we planned for complete removal of right elongated styloid process using an intra-oral approach under general anesthesia. A horizontal incision of 3 cm was given on the anterior tonsillar pillar. Right tonsil was dissected from the tonsillar bed and removed. Tonsillar bed was palpated to feel the styloid process. A longitudinal incision was given over the tonsillar bed and styloid process along with its attachments was exposed. (Fig.3) Stylohyoid ligaments along with muscular attachments and right styloid process were removed via subperiosteal dissection using a rongeur forcep and small osteotome. (Fig.4) Post-operative recovery was uneventful.
Fig. 3:- Exposure of right styloid process and dissection from its muscular attachment

Fig. 4:- Right styloid process after removal

The patient was discharged on the fifth postoperative day. The patient was asymptomatic at follow up at 3 months and did not have any complaint on the left side also. We determined that the patient's symptoms such as pain and odynophagia had resolved.

Discussion:-
An abnormally long styloid process or stylohyoid chain ossification produces a cluster of symptoms which 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.\(^1\,2\,3\) ES is a rare entity which is seen in 4% of the population (Murtagh et al., 2001).\(^6\)

The styloid process is derived from the second branchial arch of Reichert's cartilage.\(^7\) The stylopharyngeal, stylohyoid, and styloglossal 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 corn of the hyoid bone, and the stylomandibular ligament extends from the styloid process to the parotidomasseteric fascia between the mastoid process and the mandible.\(^8\) It is common in 3rd-4th decade age group because regional ligaments and the soft tissues of the styloid process become less elastic with age and offer more resistance to surrounding hard tissue structures. However, it has also been reported in children as a secondary pathology following traumatic fracture. It can occur as a consequence of a difficult endotracheal intubation.

It is usually asymptomatic. If various symptoms such as pain in the throat, dysphagia, otalgia, odynophagia or facial pain are present for which there is no other obvious cause, then a diagnosis of Eagle’s Syndrome can be made.\(^8\) An unusual presentation of a loud click following jaw movement has also been reported. In serious cases, continuous pressure to the carotid artery can lead to a stroke. Bilateral elongation is common, although symptoms are typically unilateral.
Symptoms present diversely according to the length and width of the styloid process, angle and direction of the curve, and degree of calcification of the stylohyoid ligament. The pathophysiological mechanisms for the pain of ES are: 1) Compression of the neural elements, the glossopharyngeal nerve, mandibular branch of the trigeminal nerve, and/or the chorda tympani by the elongated styloid process; 2) Fracture of the ossified stylohyoid ligament / styloid process, followed by proliferation of granulation tissue that causes pressure on surrounding structures; 3) Impingement on the carotid vessels by the styloid process, producing irritation of the sympathetic nerves in the arterial sheath; 4) The theory of insertion tendonitis due to inflammation of the tendinous part of the stylohyoid insertion; 5) Irritation of the pharyngeal mucosa involving V, VII, IX, X cranial nerves due to direct compression by the styloid process or post-tonsillectomy scarring. Diagnosis is confirmed through clinical symptoms, radiological findings, and physical examinations. Since the symptoms are variable and non-specific, patients seek treatment in several different clinics such as otolaryngology, family practice, neurology, neurosurgery, psychiatry and dentistry.

Mineralisation of the stylohyoid ligament is a well-recognised radiographic finding and an incidence of 18.2% has been reported on panoramic radiographs. However, in 1937, Eagle was the first to present two cases of pharyngeal discomfort associated with elongated styloid process. 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 V, VII, IX, and X. This type includes symptoms such as foreign body sensation, pain radiating 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 around the vessels. If the internal carotid artery is compressed, then ipsilateral headaches can occur. If the external carotid artery is compressed, then there can be pain in the temporal and maxillary branch areas.

Differential diagnosis of Eagle’s syndrome includes trigeminal neuralgia, migraine headache, TMJ disorders, temporal arteritis, carotid artery syndrome, unerupted or impacted molar teeth, faulty dental prostheses, tonsillitis, neoplasia, otolaryngeal infections, coronoid hyperplasia, hyoid bursitis, cervical vertebra arthritis, oral submucous fibrosis etc.[12] Diagnosis can usually be made by medical history and physical palpation of the styloid process in the tonsillar fossa. If pain is produced by palpation and if it radiates to ear, face, or head, the diagnosis of an elongated styloid process is very likely.[13] Injection of local anaesthetic into tonsillar fossa relieves pain and can be used as a diagnostic tool.[14]

Orthopantomography or a cranial radiograph using a lateral projection, and computed tomography or MRI confirms the diagnosis. CT scan gives measurement of the styloid process length, direction, and anatomic variation, in addition to evaluation of stylohyoid ligament ossification. Radiographically 3 types of appearance of elongated styloid process have been described. Type I: continuous uninterrupted styloid process, Type II: a single pseudoarticulation of styloid process to stylohyoid ligament. Type III: represents an interrupted mineralisation of ligament that gives the appearance of multiple pseudo-articulations within the ligament.[15]

Eagle’s syndrome can be treated both conservatively and surgically. In most cases we prescribe anti-inflammatory medications, Carbamazepine, Diazepam, Baclofen, short course of steroids, Chinese medications, transpharyngeal injections of steroids and analgesics to treat the symptoms. The most satisfactory and effective treatment is by surgical approach which is styloidecomy, performed through a trans-oral or extra-oral approach.[13,14,15,16] The trans-oral approach was introduced by W. W. Eagle. At first tonsillectomy is done and then the tonsillar bed is palpated for the enlarged styloid process. A longitudinal incision is given over tonsillar bed and styloid process with its muscular attachments is dissected and enlarged styloid process is removed via a small ostotome. Trans-oral resection produces no outside scars, but rarely causes cervical infection. The reported case was successfully treated using a trans-oral approach without any neurovascular injury.

In extra-oral approach, a horizontal incision was placed in a skin crease 5 cm inferior to the lower border of the mandible. A cervical incision was made from the proximal side of the sternocleidomastoid muscle to the hyoid bone, and the elongated stylohyoid ligaments were detached and the muscular attachments, styloid process removed via subperiosteal dissection. The most significant advantage of an external approach is enhanced exposure of the styloid process and the adjacent structures, and it facilitates easy resection of a partially ossified stylohyoid ligament.
Complications have been reported in 20% of cases which includes bleeding, partial or inadequate resection of styloid apparatus, temporo-mandibular joint arthralgia, post-operative neuralgia or subsequent fibrous entrapment syndrome.

**Conclusion:-**

Eagle’s syndrome can be diagnosed by a relevant medical history, physical examination, and radiological investigations. It can be confused or mistaken for many other conditions that must be excluded. It is mandatory to perform proper examination as because styloid apparatus is in a critically important & complex anatomical area. Surgical removal of the elongated styloid process is the definite treatment of choice.

**References:-**