

Eagle's Syndrome: Presentation, Diagnosis, and Management

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¹ Conception of study

¹ Experimentation/Study conduction

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¹ Manuscript Writing

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Abstract

Objective: To study the presentation, diagnosis, and treatment options in Eagle's Syndrome in a tertiary care hospital.

Material and Methods: The study was conducted at the ENT department of District Headquarter Hospital, Rawalpindi from 1st January 2016 to 31st March 2019. All the patients who presented in outpatient (OPD) of the ENT department with pain around the ear and throat and diagnosed as a case of Eagle's syndrome were included in the study. All the data of gender, age, CT scan findings, pain score, and management were noted on a predesigned proforma. Percentages and frequencies were calculated for all the data by using SPSS 21 version.

Results: A total of 12 patients were included in the study, with 5 (42%) females and 7 (58%) males. The patient's age range was from 24 to 68 years. Sharp, sudden onset, nerve-like pain was the major symptom in all cases. The length of the styloid process in 6 patients was between 3-4 cm and in 6 patients between 4-5 cm on a 3-dimensional computerized tomography scan. 100% of patients were managed surgically. In Group A (50%) styloidectomy was done after tonsillectomy while in other Group B (50%) patients styloidectomy was done by tonsil sparing technique. The relief of pain was a little better in patients of Group B.

Conclusion: Sudden onset, sharp, shooting pain in the jaw area, around-ear, and in the throat with no other co-morbid should be investigated for Eagle's syndrome. 3 dimensional CT scan is the best modality to see the length of the styloid process. Styloidectomy with tonsil sparing technique is a little better than tonsillectomy and gives early and permanent relief from this painful condition.

Keywords: Eagle syndrome, styloid process, styloid syndrome, styloidectomy, stylohyoid syndrome.

Introduction

Eagle's syndrome also called styloid syndrome or stylohyoid syndrome is a relatively uncommon condition.¹ Patient presents with unilateral or bilateral recurrent sharp pain in the throat and around the pinna due to mechanical compression of the carotid nerve plexus by elongated styloid process or calcification of stylohyoid ligament.^{2,3} Incidence of the disease is 4-8 per 10000 but most of the patients remain asymptomatic.⁴

The Styloid process is a bony slender structure attached at the base of the temporal bone posterior to the mastoid apex. Its anatomical location is in the maxilla-vertebra-pharyngeal recess which contains many important structures like carotid arteries, internal jugular vein, facial nerve, glossopharyngeal nerve, vagus nerve, and hypoglossal nerve.^{5,6}

The stylohyoid ligament and the small horn of hyoid bone along with the styloid process constitute the stylohyoid apparatus. It arises embryonically from Reichert cartilage of the second branchial arch. There are three muscles attached to the styloid process i.e. styloglossus, stylohyoid, and stylopharyngeus. Two ligaments stylomandibular and styloid are also attached to it.^{7,8}

The normal length of the styloid process is 2.5-3.0 cm (25-30 mm) but it can be of variable length according to different authors.^{9,10} Elongated styloid process is seen in about 4% of the population but very few patients are symptomatic.¹¹

There are different theories for the elongation of the styloid process. Eagle suggested that trauma due to any surgical procedure like a tonsillectomy or local irritation causes osteitis, tendonitis, or periostitis of the stylohyoid complex which results in reactive, ossifying hyperplasias.¹²

Lentini et al (1975) hypothesis was that during the development of mesenchymal remnants of Reichert cartilage persist undergo osseous metaplasia due to trauma or mechanical stress.¹³

Epifanio (1962) said that due to endocrine abnormalities at menopause there is the ossification of the styloid process along with other ligaments like iliolumbar and thyrohyoid.¹⁴

Chronic calcification may be a reason for elongation of the styloid process especially in patients with abnormal metabolism of calcium, phosphorus, and vitamin D.¹⁵

The patients with "typical Eagle syndrome" usually present with dull and persistent pain on the same side around the tonsillar fossa. The pain may radiate to the

ear and is aggravated on turning the head. Mass or bulge may be palpated in the ipsilateral tonsillar fossa. The patient may have dysphagia, foreign body sensation in the throat, tinnitus, or cervicofacial pain. The patient may have a history of tonsillectomy.

Another variant of this disease is "Stylocarotid syndrome" in which there is compression of the internal or external carotid artery by medially or laterally deviated styloid process. The patient feels pain in the carotid area in the neck which increases on head rotation. It is not associated with tonsillectomy.³

Some other conditions which can mimic Eagle syndrome are temporomandibular disorders, glossopharyngeal neuralgia, trigeminal neuralgia, impacted molars, cervical arthritis, tumors of the pharynx, and base of the tongue, and temporal arteritis.

One case of sudden death due to Eagle syndrome has been reported due to mechanical irritation of the carotid sinus by an elongated styloid process resulting in acute cardiovascular failure.¹⁶ The purpose is to study the presentation, diagnosis, and treatment options in Eagle's Syndrome in a tertiary care hospital.

Materials and Methods

After approval from the hospital ethical review board, this descriptive study was conducted from 1st January 2016 to 30 June 2019 at the ENT department of District Headquarter Hospital which is a teaching hospital of Rawalpindi Medical University. All the patients who presented with a history of pain in the throat and around-ear for a few months and no cause could be established were admitted and investigated for Eagle syndrome. A 3-dimensional computerized tomography (CT) scan was done to see the length of the styloid process. Patients having a styloid process longer than 30 mm of both gender and all ages were included in the study. Patients having malignancy, infections, or autoimmune disease were excluded from the study. Random sampling was done.

All patients were managed surgically. In Group A (50%) styloidectomy was done after tonsillectomy while in Group B (50%) patients styloidectomy was done by tonsil sparing technique. Patients having grade 3 and 4 tonsillar enlargements were done by tonsillectomy technique and those with grade 1 and 2 tonsillar enlargement were done by tonsil sparing technique. All surgeries were done under antibiotics and analgesic cover. The pain was assessed by the Visual Analogue Scale with 0 no pain and 10 worst pain. All the data of gender, age, CT scan findings,

complications, and management were noted on a predesigned proforma. Percentages and frequencies were calculated for all the data by the SPSS 21 version.

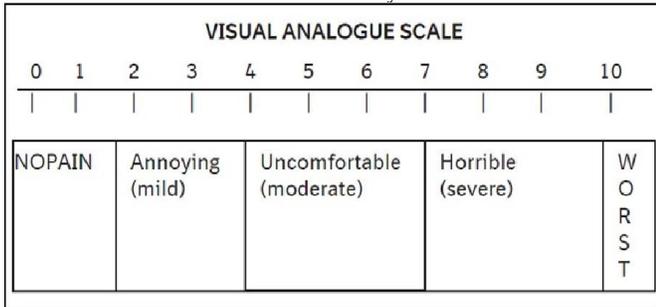


Figure 1: Visual Analogue Scale for pain

Results

A total of 12 patients were included in the study, with 5 (42%) females and 7 (58%) males. The patients' age range was from 24 to 68 years. Sharp, sudden onset, nerve-like pain was the major symptom in all cases. The length of the styloid process in 6 patients was between 3-4 cm and in 6 patients between 4-5 cm on a 3-dimensional computerized tomography scan.

Gender distribution

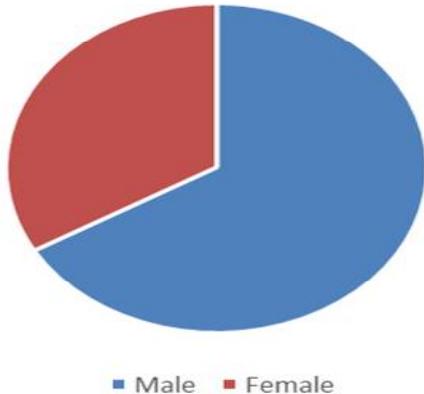


Figure 2: Gender distribution

Regarding the age of the patients, 4 (30%) patients were from the age group of 26 to 45 years of age, and 8 (70%) from 71 to 85 years of age.

Table 1: Comparison of pain in two groups before and after the procedure (n=12) after 2 weeks

| Groups | Pre-op Pain | Post-op Pain |
|---------|-------------|--------------|
| Group A | 06 (100%) | 01 (16%) |
| Group B | 06 (100%) | 00 (0%) |

There were no major complications in any patient. One patient belonging to group A had a reactionary hemorrhage within 24 hours of surgery and was managed by ligating the bleeding point in the tonsillar fossa under general anesthesia.

Discussion

Eagle syndrome is a clinical disease that manifests with pain in the throat and around pinna due to increased length of the styloid process. It was also described by Naik (2011) in the same manner.¹⁷

The elongated styloid process causes irritation of neurovascular structures in parapharyngeal space causing chronic throat pain and aggravated during swallowing which was seen in 3 of our patients. It is comparable to a study done by Maru and Yavuz (2003, 2008).^{18,19,20}

The exact cause of the syndrome is still unclear, although there are many theories by different authors.^{21,22,23} In our study a definite cause could not be established in any patient.

It was suggested by Frommer that it is not always the length of the styloid process that causes symptoms because many patients have an elongated styloid process and are symptom-free. The direction and curvature of the styloid process is a more important cause of symptoms. The angulated tip of the inferior end of the styloid process reaches the oropharyngeal wall and causes dysphagia and globus sensation.²⁴

The standard investigation for diagnosing this condition is 3 dimensional CT scan and is considered the gold standard. We also used 3 dimensional CT scan for diagnosis.²⁵

The management includes is both medical and surgical. Medical management includes analgesics, anti-inflammatory agents, steroids, and local anesthetic injections in the lower pole of the tonsillar fossa. Other general measures include soft diet, antibiotics, and/or maxillary mandibular fixation for 3-4 weeks.²⁶ In our study the patients who had already taken medical management and did not respond were included.

Definite management is surgical. There are two approaches transoral or transcervical. Different authors support different approaches like Golgoff et al²⁷ who support the transoral approach. This procedure has the benefit of no scar in the neck and short operation time. But there is limited exposure to the surgical area and little chance of infection in deep neck space. In our study, we operated all patients by the transoral route. In half patients, tonsillectomy was

done as part of the approach to the styloid process. In the other half, styloidectomy was done by tonsil sparing technique, an incision was given lateral to the palatoglossal fold. The rate of complications and post-op morbidity was almost equal in both procedures. The transcervical approach is also preferred by some surgeons. This approach has a good surgical exposure but operation times is increased a little and external scar is present.²⁸ This procedure should be reserved for longer styloid processes.

Conclusion

Patients of Eagle's syndrome present with pain around the ear and throat. It is diagnosed by 3 dimensional CT scan. When medical management fails then surgical management (styloidectomy) is the definitive treatment. It can be done by tonsil sparing technique and after tonsillectomy. Both procedures have merits and demerits but styloidectomy with tonsil sparing technique is a little better in terms of relief of pain. Styloidectomy is a safe and cost-effective procedure.

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