

Eagle's Syndrome: A Disease in Disguise

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Abstract

Background: Styloid process elongation or stylohyoid ligament calcification can cause multiple symptoms such as dysphagia, facial pain, globus sensation, referred otalgia and headache. Conglomeration of these symptoms is termed as Eagle's syndrome. It could be unilateral or bilateral. Since the symptoms mimic several other orofacial pains and neuralgia, the diagnosis must be made through a thorough history, adept clinical examination, and various imaging modalities.

Methods: This article reports 5 cases of Eagle Syndrome. All the patients presented with complaints of chronic throat pain radiating to neck which did not relieve on taking medications, for the same they have been consulting multiple specialities. The diagnosis of Eagle Syndrome was confirmed by palpation in tonsillar fossa and orthopantomogram revealed enlarged styloid process measuring more than 30mm. Upon confirmation, all the 5 patients underwent tonsillo-styloidectomy.

Result: All 5 cases diagnosed as Eagle's syndrome achieved a definitive benefit by tonsillo-styloidectomy surgery.

Conclusion: Eagle Syndrome is a diagnosis of exclusion and should raise high index of suspicion in patients with nonspecific throat and neck pain not responding to any conservative treatment. With an increasing incidence of side effects following injudicious treatment given in such cases, this condition requires attention, as it is a rare entity and often misdiagnosed.

Keywords: Eagle's syndrome, Elongated styloid process, Facial pain.

INTRODUCTION

Eagle's syndrome or stylalgia is one of the most common overlooked and missed condition in routine ENT practice. First defined by W. Eagle in the year 1937. Characteristic feature of Eagle's syndrome is morphological abnormality/

ossification of the styloid process. 20-30 mm is the normal average length of styloid process in the adult Caucasians and 15.4-18.8 mm in the Asian population. An elongated styloid process is usually defined as at least 30 mm long. The symptoms include recurrent throat and neck pain, referred otalgia and dysphagia. Symptoms can be bilateral or, more frequently, unilateral.

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It is usually underdiagnosed or misdiagnosed due to its vague symptoms. As a result, it becomes troublesome for the patient as he consults multiple specialities which leads to delayed diagnosis and treatment. Patient is treated with unnecessary analgesics and antibiotics which predisposes him to laryngopharyngeal reflux diseases.

However, palpable styloid process in tonsillar fossa with radiological evidence of the elongated styloid process accomplishes the diagnosis. Tonsillo-styloidectomy remains the main stay of treatment for Eagle Syndrome with a high success rate as there are no recurrences.

CASE REPORTS

Case 1

A 42-year-old female patient came to us with complaints of persisting nagging pain over the right side of the throat radiating to face and ipsilateral ear for the last 2 years and not relieved by any analgesics and antidepressants. She was examined by general surgeon and Neurologist elsewhere and found to be clinically normal. ENT examination here showed normal oropharynx on inspection. But tender bony mass was palpable in bilateral tonsillar fossa, which again confirmed the site of pain. The diagnosis of stylalgia was confirmed by taking an orthopantomogram which revealed bilateral enlarged styloid process measuring 40mm on right and 42mm left side respectively. She underwent tonsillo-styloidectomy (Fig. 1), (Fig. 2) after pre-operative evaluation. Patient was regularly followed up on day 7, day 14, and 1-month post-surgery was found to be symptom free.



Fig. 1

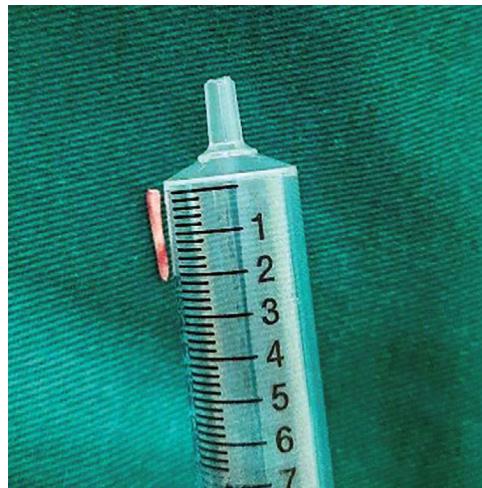


Fig. 2

Case 2

35-year-old female presented to us with complaints of right sided dull aching throat pain radiating to neck, right ear since 2 years. ENT examination revealed bilateral grade two tonsillar hypertrophy with right side palpable bony prominences. The diagnosis of Eagle Syndrome was thus accomplished. Initially patient was treated conservatively with analgesics but showed no improvement. Following which she underwent bilateral tonsillectomy with right styloidectomy (Fig. 3). Patient was discharged with oral antibiotics, mouth gargles and analgesics, followed up on 7th day, 14th day and 1 month post-surgery and was relieved of symptoms.

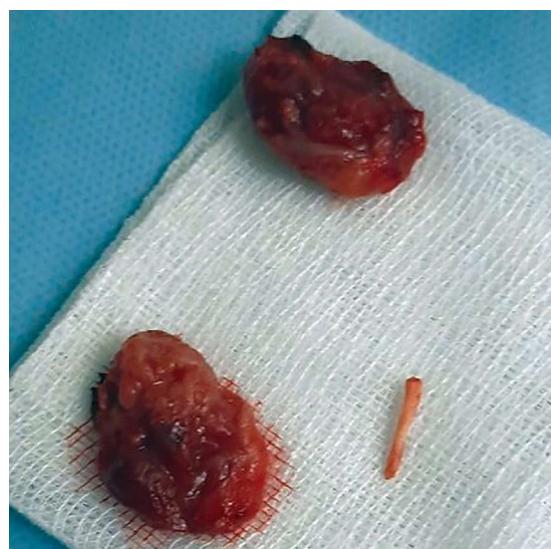


Fig. 3

Case 3

A 34-year-old male presented to us with complaints of intermittent dull aching throat pain associated with pain in the neck for 2 years.

Patient has been visiting various doctors for the same complaints before he came to us. On ENT examination, bilateral grade 2 tonsillar hypertrophy was noted, on bimanual examination right side bony prominences were palpable in tonsillar fossa. CT neck revealed bilateral enlargement of styloid process measuring 35.0 mm on right side and 25.0mm on left side. (Fig. 4).



Fig. 4

He was started on analgesics and preoperative evaluation was done, once diagnosis was confirmed. Following which the patient underwent bilateral tonsillectomy and right side styloidectomy by an intra oral approach and enlarged styloid process was removed. Following surgery patient had regular follow ups on day 7, day 14 and 1 month and had symptomatic relief of symptoms.

Case 4

A 36-year-old male presented to us with complaints of intermittent throat pain associated with pain in the neck for 3 years, with history of low-grade fever. Patient gave history of visiting multiple specialities for the same complaints before he came to us. ENT examination revealed bilateral grade 1 tonsillar hypertrophy, on bimanual examination bilateral bony prominences were palpable in tonsillar fossa. Orthopantomogram revealed bilateral enlargement of styloid process measuring 40.5mm on right side and 60.2mm on left side. Diagnosis of stylalgia was confirmed and was started on analgesics and preoperative evaluation was done. Following which the patient underwent bilateral tonsillo-styloidectomy by an transoral approach and bilateral enlarged styloid process were removed. (Fig. 5) On regular follow ups on day 7, day 14, and 1month post-surgery, the patient symptomatically improved.

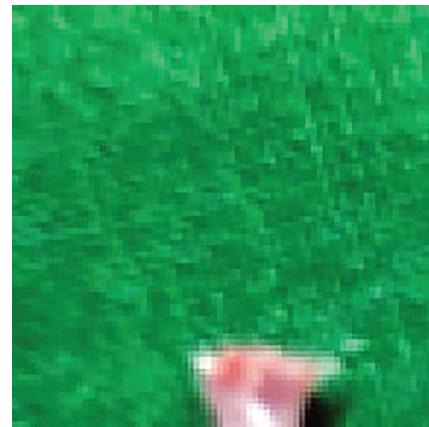


Fig. 5

Case 5

A 33 year-old female presented to us with complaints of dullaching throat pain radiating to neck, both ears and foreign body sensation in throat for 10 months which did not relieve on taking medications. ENT examination revealed bilateral tonsillar fossa to be tender with palpable bony prominences. Orthopantomogram was done and the findings were: bilateral elongated styloid process measuring right side 35.4 mm, left side 32 mm. Based on the imaging findings and clinical examination, the diagnosis of Eagle Syndrome was thus accomplished. Initially patient was treated conservatively with analgesics but showed no improvement. Following which she underwent bilateral tonsillo-styloidectomy which showed bilateral enlarged styloid process (Figure 3.6) Patient was discharged with oral antibiotics and analgesics, followed up on day 7, day 14 and 1-month post-surgery and she showed drastic relief of symptoms.

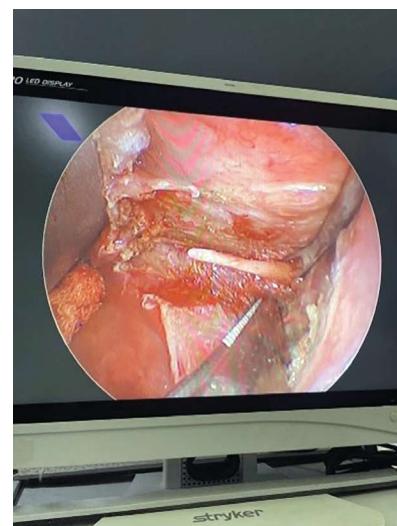


Fig. 6

DISCUSSION

Eagle syndrome presents as repeated pain in the oropharynx and along the face due to an elongated styloid process or calcified stylohyoid ligament. The styloid process is a sharp, pointed, bony projection on the temporal bone that extends from the bottom of the petrous part of the bone, just below the ear^[4]

Normal length of styloid process is 2-3 cm and is considered as elongated when it is more than 3 cm. It usually occurs as two presentation. First one being the classical type and the second is carotid artery type.^[1] Elongated styloid process can compress the adjacent neurovascular bundle consisting of internal carotid artery, V, VII, IX cranial nerves causing vague clinical presentation which includes throat and neck pain with referred otalgia to headache, neuralgic pain along the glossopharyngeal nerve, globus sensation, syncope etc.

The cause is not well known, various theories have been proposed, such as, stylohyoid ligament calcification and bone formation at the insertion of the ligament, the persistence of an embryonic cartilaginous outgrowth causing congenital elongation.^[2]

Clinical history and intra oral palpation for the elongated styloid process in the tonsillar fossa along with radiological investigations like Orthopantomogram or X-ray Townes view, Cone beam CT etc aides in diagnosis. Conservative management with analgesics, anticonvulsants, anti-depressants and local injection of steroids can be tried as first line of treatment and might help in alleviating symptoms in some patients.^[3] Surgery is the next alternative option. It can be done by either intra oral approach or extra oral approach. Intra oral approach has less morbidity and complications, without any visible scar. Hence intra oral approach is preferred by many otolaryngologists.

CONCLUSION

Eagle's syndrome is not uncommon but often clinically missed condition. Accordingly all cases of chronic throat and neck pain has to be evaluated in depth by taking thorough history and adept clinical examination, along with radiological investigations (orthopantomogram). Surgery is the best treatment

modality when conservative management fails, which helps in alleviating patient's morbidity due to chronic pain.

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