

Diagnostic and therapeutic dilemma in orofacial pain: A rare case of bilateral Eagle syndrome

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Abstract

Eagle's syndrome is a collection of symptoms due to abnormal and/or elongated styloid process. This can irritate the various neurovascular structures that lie in its close proximity, mainly the glossopharyngeal nerve, leading to odynophagia, dysphagia, foreign body sensation, cervicofacial pain, and headache. It is a diagnosis of exclusion and needs high degree of clinical suspicion. It is a rare condition with no reported incidence in the Bhutanese population so far. In this article, we present a case of elongated styloid process that was causing persistent and troublesome orofacial pain in a patient, which was not relieved by medication. The patient underwent trans-oral styloidectomy, which helped cure his symptoms.

Keywords

Styloid process, Eagle's syndrome, recurrent throat pain, neuralgias, Bhutan

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Introduction

The styloid process is a slender, cylindrical bone arising from the inferior aspect of the petrous temporal bone and its length in adults is around 20–30 mm.¹ It is a site of attachment for stylohyoid ligament, stylohyoid, styloglossus, and stylopharyngeus muscles. Medially it is related to internal jugular vein, internal carotid artery with sympathetic plexus, glossopharyngeal nerve, vagus nerve and accessory nerve, and laterally to the occipital artery and hypoglossal nerve.² The styloid process varies in length, angulations, and other morphological features between individuals.³ While these physiological differences are found incidentally, some present with a collection of symptoms known as Eagle's syndrome. These symptoms occur as a result of irritation and/or compression of surrounding neurovascular and muscular structures from an abnormal styloid process.

Eagle's syndrome was first described by Marchetti in 1652.^{4,5} In 1937, Watt W. Eagle started working on the clinical and radiological presentations, diagnosis, and treatment of elongated styloid process. He called it "Stylalgia" and defined it as an "abnormal elongation of styloid process (more than 30 mm) which irritates the adjacent structures, especially glossopharyngeal nerve, causing recurrent throat and neck pain that radiates to the auricular and mastoid region without any previous history of trauma."⁶

There are two types of Eagle's syndrome.⁷ The first type, classic Eagle's syndrome or stylohyoid syndrome, presents as a sharp pain in the neck or the ear extending to the maxilla, face, and oral cavity. It might appear exaggerated with head rotation, chewing, swallowing, tongue protrusion, or yawning. In addition, a mass might be palpable in the tonsillar fossa. Symptoms of classic Eagle's syndrome are usually unilateral but can rarely present bilaterally.⁸ These symptoms occur due to the irritation or possible entrapment of the nearby cranial nerves (CN V, VII, IX, or X). The irritation or entrapment that occurs may be secondary to the formation of local granular cells.⁹ The second type of Eagle's syndrome is known as stylo-carotid artery syndrome, which occurs when the styloid process impinges upon the internal or external carotid artery and the nerve plexus accompanying them. It presents as pharyngeal pain, orbital pain, or headache. Compression of the internal carotid artery might present with

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symptoms of internal carotid vascular insufficiency such as weakness, vision abnormalities, or syncope exacerbated with head movement. There is also the risk of the elongated styloid process causing carotid artery dissection, which can lead to transient ischemic attack or stroke.^{9,10}

Approximately 4% of the general population has an elongated styloid process but only 4% of these patients have related symptoms.^{11,12} It is mostly found in individuals aged 30–50 years with a female predominance. Although it is more common to have bilateral elongated styloid processes, most patients often complain of pain at only one side.^{13,14}

We present the case report of a patient with orofacial pain caused due to elongated styloid process that was misdiagnosed as pharyngitis/tonsillitis and received several courses of antibiotics and analgesics. After palpation of tonsillar fossa and radiography, he was diagnosed as Eagle's syndrome and the pain got relieved after surgical intervention.

Case report

A 33-year-old Bhutanese male presented to the ear, nose, and throat (ENT) outpatient clinic with recurrent right-sided throat pain for the past year. The pain was of pricking or stabbing type that originated at the right side of the throat and radiated to ipsilateral cheeks, forehead, and ears. It was exacerbated during chewing, swallowing, and occasionally during change of head posture. It was not associated with fever, sore throat, or cough. He had no history of local trauma. He had no underlying comorbidities or allergies. He was managed at the local hospital as a case of pharyngitis and tonsillitis with analgesics (paracetamol), antibiotics (amoxicillin), and antihistamines (cetirizine) but they were not of significant benefit and thus visited our tertiary care center.

On examination, the patient was clinically stable with normal vital signs. Intraoral examination showed unremarkable oral cavity and tonsils. He had no dental caries. On digital palpation, the patient complained of pain over the tonsillar bed radiating to the ears, and a hard, pointy styloid process could be palpated. Other ENT, head, neck, and systemic examinations were unremarkable. A noncontrast CT scan was requested which revealed bilateral elongated styloid processes measuring 42.7 mm on the right and 41.5 mm on the left (as shown in Figure 1). A diagnosis of Eagle's syndrome was made for the origin of orofacial pain. He was managed conservatively for a year after which a decision to perform styloidectomy was offered.

In the operating theater, after induction of general anesthesia, the patient was placed in Rose position. Routine bilateral tonsillectomy was done initially. After achieving adequate hemostasis, the tonsillar bed was irrigated with saline. Then the superior constrictor muscle fibers were dissected with artery forceps to expose the tip of the styloid process. After securing the location of the styloid process, Freer's elevator was used to gently scrape off and detach its

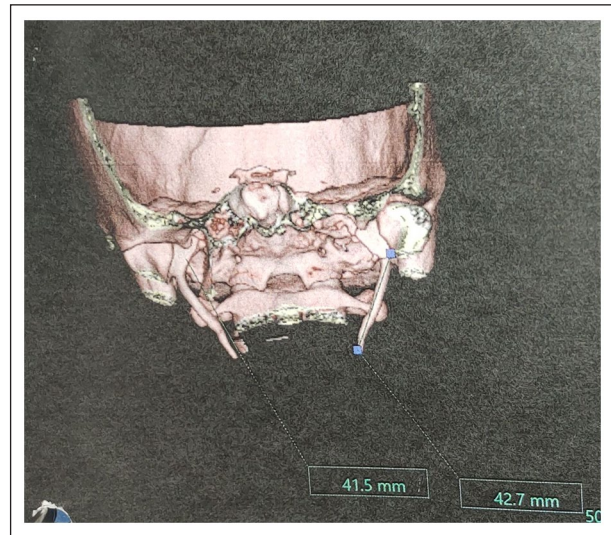


Figure 1. 3D reconstruction CT scan of a patient with chronic orofacial pain showing bilateral elongated styloid process.

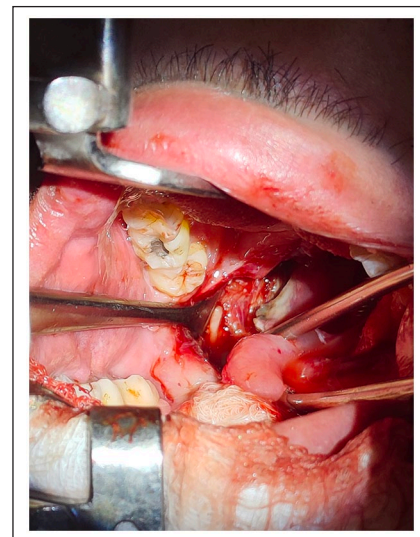


Figure 2. Intraoperative view of the left styloid process as seen in the tonsillar bed.

muscular attachments. The stylohyoid ligament was cauterized with bipolar first and then cut with tissue-cutting scissors (as shown in Figure 2). Finally, the styloid process was dissected from the soft tissue using ring curette and was firmly grasped with artery forceps and fractured manually. Similar steps were repeated on the contralateral side. The excised styloid processes each measured 15 mm in length (as shown in Figure 3). The surgical bed was irrigated with normal saline and hemostasis obtained. The patient was given intravenous antibiotics (ampicillin) and oral care (betadine mouth wash) and was discharged on the second postoperative day with oral antibiotics (amoxicillin). The patient was followed up after a week where he complained of dull pain



Figure 3. The resected specimen of styloid processes in a patient with Eagle's syndrome.

on swallowing, unlike the pricking type of pain he had before. There was healthy slough at the tonsillar bed with no clots or bleeding. The patient was given analgesics (ibuprofen) for symptomatic pain relief and asked to follow-up in 1 month. During the follow-up visit he reported complete disappearance of the pain that he used to feel before.

Discussion

Eagle's syndrome is a rare condition associated with abnormal and/or elongated styloid process. Diagnosis of Eagle's syndrome depends on the patient's clinical presentation, radiological investigation, and lidocaine infiltration test.⁹ The clinical symptoms of Eagle's syndrome are not specific and may be similar to several other diagnoses. A palpable mass in the tonsillar fossa might allow the clinician to narrow their differential; however, it is not always present in symptomatic Eagle's syndrome. Our patient presented with recurrent right-sided orofacial pain with no other symptoms suggestive of infectious or dental etiology. Previous efforts to provide conservative pain management had been unsuccessful. Our patient did have a palpable hard mass in the tonsillar fossa which helped narrow our differential diagnosis.

Radiographic imaging has an important role for establishing a definitive diagnosis. Orthopantomogram (OPG) X-ray of the oral cavity is a cost-effective investigation which can show the styloid processes bilaterally. However, nearly 4% of the population have elongated styloid process and an incidental finding on OPG done for other indications may not necessarily equate to Eagle's syndrome.^{11,12} The symptoms in Eagle's syndrome are highly influenced by head movements, thus various radiographic investigations should be performed in different head and neck positions to assess the relationship between styloid process and surrounding structures.¹⁵ Computed tomography (CT) allows for the evaluation of length and angulation of the styloid process. A three-dimensional CT is considered as the gold-standard

for radiological diagnosis and provides the best supplement to a plain X-ray. CT angiography is recommended in stylo-carotid syndrome to assess blood flow dynamics.^{16,17} The lidocaine infiltration test can be confirmatory for symptomatic patients. If a patient's symptoms are relieved after administering 1 mL of 2% lidocaine to the area surrounding the palpable styloid process, the test is considered positive and establishes the diagnosis of Eagle's syndrome.¹⁸ In our case, because of the high index of suspicion, we proceeded directly with CT scan of head and it did reveal the elongated styloid process. This further helped support our provisional diagnosis.

Conservative treatment options include oral antidepressants, anticonvulsant, opioids, and non-steroidal anti-inflammatory drugs. Transpharyngeal injection of steroid or local anesthetic agents may also be tried. Physical therapy and warm compression aid in relaxation of muscle spasm. These treatment options are less invasive and adequate for some patients. On the contrary, some patients may require surgery to alleviate the persistent pain.^{13,19,20} Intraoral or transoral approach to styloid process was first described by Eagle and is simple, easy, and less time-consuming.^{6,21} External scars can also be avoided but a downside of this approach is that it offers limited visualization and there are higher risks of injury to great vessels and deep neck infections.²² Extraoral procedure was explained by Chrcanovic et al.²¹ and Loeser and Cardwell,²² which provides better surgical access and visualization but external scars are difficult to avoid. Moreover, there is also the risk of facial nerve injury and a longer duration of surgery.²³

Our patient had been given symptomatic management with analgesics but the symptoms were not relieved. Hence, we opted for surgical management. We operated via intraoral approach because of its ease and precision. Postoperatively, the patient has had a long pain-free period without recurrence of the symptoms.

The decision to go with surgery is controversial and there is no consensus supporting a surgical approach. Holistic reasoning, taking into consideration the patient factors, the disease factors, and surgeon factors can aid in reasonable decision-making when managing such cases.

Conclusion

Eagle's syndrome presents with atypical pain at the orofacial and neck regions which can initially be diagnosed as pharyngitis, tonsillitis, eustachian tube infection, parotid gland infection, cluster headache, migraine, trigeminal neuralgia, temporomandibular joint disorder, or dental pain. Although Eagle's syndrome is a rare condition, a detailed history-taking augmented by physical examination by digital palpation of the styloid process in the tonsillar fossa can aid in localizing the cause of pain; presence of pain during palpation should prompt a thorough examination for signs of an elongated styloid process. Although rare, Eagle's syndrome can

be one of the possible etiologies. Due importance should be given to clinical history and examination, supported by radiographic investigations in order to make a timely diagnosis so that timely management can be initiated to treat the condition and its associated symptoms.

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Ethical approval

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Informed consent

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