



CASE REPORT

EAGLE SYNDROME: UNCOMMON CAUSE OF OROFACIAL PAIN; REVIEW OF 5 CASES

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SUMMARY

Eagle syndrome is an uncommon cause of orofacial pain. An elongated styloid process or calcified stylohyoid ligament triggers symptoms; most frequently as headache, facial pain, dysphagia and sensation of foreign body during swallowing. It is usually hard to diagnose if there is not enough degree of suspicion, cause the symptoms might be confused with facial neuralgias. The aim of this study is to show symptoms, steps of diagnosis and the treatment procedures of Eagle Syndrome as well as the literature review.

Keywords: Eagle Syndrome, Elongated stiloid process, orofacial pain, surgery of stiloid process

EAGLE SENDROMU: OROFASIAL AĞRININ NADİR NEDENİ; 5 OLGU

ÖZET

Eagle sendromu ağız ve yüz bölgesinde ağrı ile kendini gösterebilen nadir bir hastalıktır. Uzun stiloid kemik veya kalsifiye olmuş stilohyoid ligament semptomları tetikleyebilir. Baş ve yüz ağrısı başta olmak üzere disfaji ve yutma sırasında boğazda yabancı cisim hissi gibi semptomlarla karakterizedir. Şüphelenmediği takdirde tanısı zor olan bir durumdur ve semptomları trigeminal nevralji ile karıştırılabilir. Bu yazıda tanı konulabilmiş ve tedavisi yapılmış 5 olguda tanı ve tedavi prosedürlerini incelenmiştir. Bu çalışmanın amacı Eagle sendromunda semptomları, tanı basamaklarını ve tedavi seçeneklerini literatür ışığında ortaya koymaktır.

Anahtar Sözcükler: Eagle Sendromu, Uzun stiloid çıkıntı, yüz ağrısı, transoral stiloid çıkıntı eksizyonu

INTRODUCTION

Patients with pharyngodynia, neck and facial pain can lead to an extensive differential diagnosis, Eagle syndrome is one of the missed out causes. Misdiagnosis and multiple failed treatments are common in certain patient population¹. Eagle syndrome, is first described by Watt Weems Eagle in 1937, as a pair of case reports. Although etiology and pathophysiology of this syndrome was poorly understood, it was first defined as “stylalgia” and related to abnormal length of the styloid process or calcified stylohyoid membrane².

The styloid process is a cylindrical bony projection from temporal bone. It is derived from Reichter's cartilage of the second branchial arch. Typical styloid process is about 2.5- 3.0 cm in length³. Lengths greater than 3 cm are considered elongated. Many important neurovascular structures lie near the tip of the styloid process. The internal carotid artery, internal jugular vein and cranial nerves X, XI, XII lie on its medial side.

Compression of these structures is the main cause of the symptoms presenting. Bilateral elongation is commonly seen, but symptoms are typically unilateral.⁴

An elongated stylohyoid process occurs in about 4% of the general population as Eagle reported⁵, while a small percentage of (4%-10%) the cases are symptomatic. The true incidence is about 0.16% and there is a slight female predominance⁶. Variety of symptoms in eagle syndrome is wide; such as craniofacial pain, dysphagia,odynophagia, foreign body sensation, temporary voice changes, tinnitus, otalgia, syncope and focal cerebral symptoms⁷, it may also cause even stroke due to the compression of the carotid arteries and bilateral dissections of internal carotid arteries as aneurysms were described¹³.

Because of the wide range of symptoms, the differential diagnosis is broad and includes glossopharyngeal neuralgia, occipital neuralgia, sphenopalatine neuralgia, temporomandibular disorders, dental infection, tonsillitis, mastoiditis, and migraine^{11,12}. If appropriate clinical history and radiologic assessment is done, eagle syndrome can easily be recognized and treated.

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The aim of this study is to evaluate symptoms, steps of diagnosis and the treatment procedures of Eagle Syndrome as well as the literature review.

MATERIAL and METHODS

Five patients with elongated styloid process underwent transoral surgery in Hacettepe University, Department of Otolaryngology between 2007 and 2014. Medical records of the patients were evaluated for their age, sex, operative findings, preoperative and postoperative visual analog scale (VAS) for pain. The duration of follow up ranged from 6 months to 7 years. Patient were underwent general physical examination. Preoperative radiologic evaluation such as Towne graphy, lateral cervical graphy, CT, panoramic X-ray was made for each patient to identify elongated styloid process. Laboratory investigations was performed and total blood count and aPTT and INR levels were evaluated, patients were asked about other systemic symptoms of inflammatory diseases and there was none. Non of the laboratory assesment was performed for the systemic inflammatory diseases, that results in cartilage and bone distruction (such as Rheumatoid arthritis and Ankylosing spondylitis), because of the lack of systemic symptoms. All patients operated

transoral approach. Informed consent was taken from each patient. None of them had any complications such as airway edema or retropharyngeal infection After surgical exicion of styloid process cases were hospitalized for two days to observe the airway and there was not any sign of respiratory distress. All cases were given intravenous antibiotic therapy during hospitalization and after discharge oral antibiotic treatment was continued for 10 days. After begining this study procedure, the patients were asked to evaluate their preoperative and postoperative symptoms. They were instructed to rate their symptoms severity, by using a visual analog scale (VAS). The patients were talled that, according to VAS , (-) means no symptom, (+) referes mild symptom, (++) moderate severity, and (+++) referes severe degree of symptom. Preoperative and postoperative results of VAS, are shown on Table 1. As seen on the table 1, symptoms seen among our patients were dysphagia, orofacial pain, foreign body sensation. Among our patients; temporary voice changes, tinnitus, syncope and focal cerebral symptom did not occur. When estimated, all patients primary symptom was defined as being "severe". The dominant symptom seen between our patients was dysphagia, and it was seen that all the symptoms were releaved after surgical treatment.

Table 1: Comparison of preoperative and postoperative symptom severity scores

	Dysphagia		Orofacial pain		Foreign body sensation	
	PreOp	PostOp	PreOp	PostOp	PreOp	PostOp
Case 1	+++	-	+++	-	-	-
Case 2	-	-	++	-	+++	-
Case 3	-	-	+++	-	0	-
Case 4	+++	-	-	-	-	-
Case 5	+++	-	-	-	++	-

PreOp: preoperatively, PostOp: postoperatively, VAS (visual analog scale for the symptom severity), (-):none, (+):mild, (++)moderate, (+++):severe



CASE PRESENTATION

Case 1:

The patient was a 68-year-old man with a 3-year history of repetitive dysphagia and left neck pain, radiating to submandibular region. He had been diagnosed with dental disease and chronic pharyngitis and was frustrated with unsuccessful treatment trials. Before admitting to our clinic, he was evaluated by a dentist and suspected about elongated styloid process after a panoramic X-ray of the jaw. His medical and surgical history was non-contributory. He did not have any craniofacial trauma either. Physical examination was performed and showed an exacerbated pain by palpation of the left tonsillar fossa. Radiographic examination demonstrated styloid process extending down (Figure 1). Elongation of the styloid process was measured as 5.39 cm. His findings were compatible with Eagle syndrome and surgical removal by transoral approach was performed. He was followed up for 4 months postoperatively, without any complaint.

Case 2:

A thirty two-year-old female patient was referred to our department with a left cervical pain, foreign body sensation in the throat and dysphagia. She also complained about a pain radiating to the left ear. In her medical history, there was not any cervical trauma or surgery. She had been medically treated by several doctors with the diagnosis of chronic pharyngitis. Through the intraoral palpation, elongated styloid process was revealed in the left tonsillar fossa, and it was painful. Lateral cervical radiography confirmed the diagnosis of Eagle syndrome, with an elongated styloid process, measured as 4.06 cm (Figure 2). Transoral removal of the styloid process was performed and a fragment of 2.1 cm was excised. The patient was asymptomatic during the following up of 3 months.

Case 3:

A 33-year-old female patient admitted to our clinic with a continuous, daily, pressure-type pain on the right side of the neck radiating to frontal and temporal regions for 4 years. Her pain was ascending while opening her mouth. There was neither history of trauma or previous surgery, nor identifiable trigger points. Physical examination revealed pain on right tonsillar fossa with palpation, as well as on the right side of the neck. The neurological examination including the cranial nerves was normal. Towne and lateral neck radiography showed bilateral styloid process elongation (Figure 3-4). The length of the

styloid processes were 5.08 cm on right and 4.7 cm on left side. She was operated by transoral approach, 1cm long styloid process removed from the right side (Figure 5). The patient was asymptomatic during the following up of 2 months.

Case 4:

A 20-year-old female patient with repetitive dysphagia and sore throat applied our clinic. In her medical history, there was not any cervical trauma or surgery. She felt pain during the palpation of the tonsillar fossa and elongated styloid process was detected, bilaterally. Because of her pain, a neck MRI imaging was performed. Radiographic confirmed the diagnosis by showing the elongated styloid process, which was measured as 4,7cm. Transoral excision of the elongated styloid processes (bilaterally) was performed (Fig. 6-9). Through the five years of follow up, she did not have pain or dysphagia.

Case 5:

The patient was a 38-year-old male with repetitive dysphagia and foreign body sensation, especially ascending at night. These complaints, according to the patient, had been for 2 years. He had been administered non steroidal anti-inflammatory drugs and carbamazepine by some practitioners for facial neuralgia, without any response. Additionally, chronic pharyngitis and tonsillitis treatment had been given for many times. Elongated styloid process was revealed in the right tonsillar fossa that was tender via intraoral palpation. Styloid process elongation was detected through radiologic examination. It was measured as 5.1 cm long and excised transorally. He was followed up for four years and had never a complaint again, such as repetitive dysphagia.



Figure 1: Lateral cervical graphy of Case 1. It shows elongated styloid process



Figure 2: Lateral cervical graphy of Case 2. It shows elongated styloid process



Figure 5: It shows excised styloid process



Figure 3: Towne graphy of Case 3. It shows elongated styloid process



Figure 6: Elongated styloid process of case 4.



Figure 4: Lateral cervical graphy of Case 3. It shows elongated styloid process

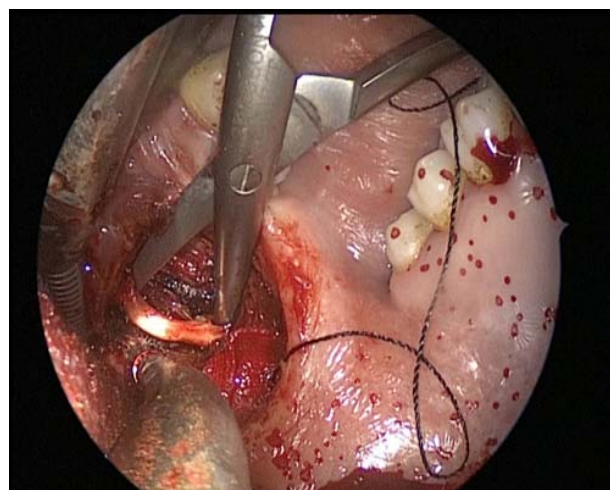


Figure 7: Dissection of styloid process in tonsillar fossa

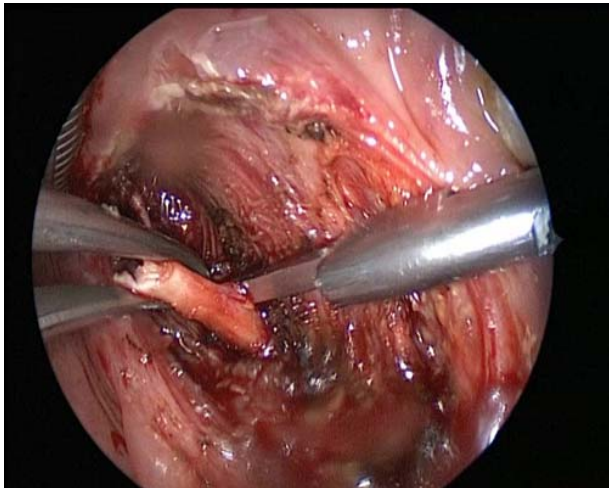


Figure 8: Styloid process excision via Kerrison forceps



Figure 9: Parapharyngeal space after excision

DISCUSSION

The “classical Eagle syndrome” is typically seen in patients after pharyngeal trauma or tonsillectomy, and it is characterized by ipsilateral dull and persistent pharyngeal pain, centered in the ipsilateral tonsillar fossa, that can be referred to the ear and exacerbated by rotation of the head. A mass or bulge may be palpated in the ipsilateral tonsillar fossa and the palpation can also exacerbate patient's symptoms^{7,8}. The “second form” of the syndrome (“stylocarotid syndrome”) is characterized by the compression of the internal or external carotid artery (with their peri-vascular sympathetic fibers) by a laterally or medially deviated styloid process⁹. It is related to a pain along the distribution of the artery, which is provoked by rotation and compression of the neck. It's not related with tonsillectomy. In case of impingement of the internal carotid artery, patients

often complain about supraorbital pain and parietal headache. In case of the external carotid artery irritation, the pain radiates to the infraorbital region¹⁰.

While evaluating the patient, history of symptoms should be taken carefully. Three main criteria have generally proposed for clinical diagnosis: a characteristic dull, nagging pain that is provoked during swallowing, a palpable elongated styloid process in the tonsillar fossa, and detection of a process 30 mm or longer on radiologic evaluation.

Diagnosis is classically based on the clinical signs and palpation of the styloid process in the tonsillar fossa. Radiological findings and pain loss after local anesthetic (lidocaine test) infiltration can also support the diagnosis. Lidocaine test was described as local injection of 1mL of 2% lidocaine into tonsillar fossa. Following the injection pain relieves and it can be used as a diagnostic test for elongated styloid process¹⁴. Panoramic radiographs, Towne graphs and lateral cervical graphs can be performed to see the elongated styloid process. Computerized tomography (CT) is to be considered the gold standard for diagnosis¹⁵ when it is performed three-dimensional. 3D-CT is a valuable diagnostic tool in the diagnosis of Eagle's syndrome because of its ability to facilitate accurate measurement of the length of the styloid process^{16,17}.

Treatment can be surgical and non-surgical. Non-surgical approaches consist of using non-steroidal anti-inflammatory medications, oral neuro-protective agents such as gabapentin, amitriptyline, valproate, carbamazepine and infiltration of steroids or anesthetics in the tonsillar fossa¹⁸. Surgical excision of the elongated styloid process with the external approach (transcervical) or transoral (transpharyngeal) approaches are often preferred¹⁹. As an otolaryngologist, we almost often perform the transoral approach. Through an oral corridor, afterwards tonsillectomy, an incision is made anterior to the tonsillar fossa and the tip of the styloid process is exposed via blunt dissection (Figure 6). This dissection is proceeded as proximally as possible along the styloid process, ultimately leading to removal of the process at the base (Figure 7). Complete exposure and thus complete excision of the styloid process are often not possible with this approach, although the excised component is almost always sufficient to relieve symptoms (Figure 8 and 9). The transoral approach is favored for its cosmetic by avoiding any external incision and for its potential for shorter operation time^{20,21}. Nevertheless, exposure of the retropharyngeal space to intraoral contents elevates the risk of infection. Further considerations include poor carotid artery access in case of



intraoperative injury²², airway edema, and trismus . In case of the elevated risk of airway edema, bilateral operations must be staged²⁰.

In the external approach, an oblique incision is made in the skin crease halfway between the angle of the mandible and the tip of the mastoid process. Dissection begins with opening of the superficial fascia and posterolateral retraction of the sternocleidomastoid muscle. With ongoing blunt dissection, superior retraction of the parotid gland, inferior retraction of the posterior belly of the digastric muscle, identification and preservation of the facial vein can be achieved. The elongated styloid process is identified and detached from the stylohyoid ligament distally. Muscular attachments are removed via subperiosteal dissection. The styloid process is then removed completely in a piecemeal fashion²³. The operating surgeon should take care to expose and then to avoid the mandibular branch of the facial nerve in the superficial fascia as well as external carotid artery branches within the deep fascia. The most common postoperative complication of the external approach is weakness of the mandibular branch of the facial nerve, which is usually transient. The primary advantage of this approach is the minimal generation of airway edema, and the consequent ability to perform bilateral styloidectomy in the same session. Minimally invasive external approaches involving styloid chain transection and resection of the lesser cornu of the hyoid bone have been used with the aid of stereotactic navigation. These approaches are rare, and there is not yet a consensus about their utility²⁴.

CONCLUSION

Although eagle syndrome's symptoms are similar to other pathologies and it creates a complex and difficult condition to diagnose. Physical examination and radiological evaluation are supportive to make the diagnosis. Eagle syndrome can be successfully treated with surgery and in most of the cases, pain relief can be provided. A surgical excision of this elongated styloid process is the main treatment option and transoral approach has lesser complication incidence.

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