AN INTERESTING CASE OF STROKE DUE TO EAGLE SYNDROME
Anil Kumar H¹, Swati Chouhan²

HOW TO CITE THIS ARTICLE:
DOI: 10.14260/jemds/2014/2877

ABSTRACT: Eagle Syndrome (ES) is a rare syndrome characterized by a specific orofacial pain due to a calcified stylohyoid ligament or an elongated styloid process. The ‘Classic’ Eagle Syndrome typically occurs after pharyngeal trauma or tonsillectomy. The second form, ‘Stylocarotid Syndrome’ is characterized by compression of internal or external carotid artery. Here, we present a case of acute dysphagia in a 48 years old male due to Eagle Syndrome. The patient presented with acute onset dysphagia of 1 week duration to both solid and liquid foods, associated with odynophagia and pain in the right oropharynx. Patient had a past history of left hemiparesis with facial palsy due to right sided cerebrovascular accident (CVA) 3 years ago, which had subsequently improved. Patient was investigated with Barium swallow, X-ray skull lateral view, MR Angiogram of neck and brain, Carotid Doppler study and finally Orthopantomogram (OPG). OPG showed an elongated styloid process measuring 3.8cms on the right side. Incidentally, MR Angiogram of neck and brain showed absent intracranial part of internal carotid artery on the right side (the main stem of the artery, starting from 2mm's from the common carotid bifurcation until the junction with anterior cerebral artery was missing). Rest all investigations were normal. Hence, the case was diagnosed as right sided Eagle syndrome, possibly of the stylocarotid variety.

KEYWORDS: Stroke, Dysphagia, Eagle Syndrome.

INTRODUCTION: Eagle Syndrome (ES) is a rare syndrome characterized by a specific orofacial pain due to a calcified stylohyoid ligament or an elongated styloid process.¹ Styloid process forms the stylohyoid apparatus, which arises embryonically from the Reichert cartilage of the second branchial arch. Length of a normal styloid process is 2.5-3.0 cms.² Elongated styloid process is seen in 4% of population. Majority of them are asymptomatic. Only a small percentage (between 4-10.3%) of these patients is symptomatic. So the true incidence is about 0.16%, with a female-to-male predominance of 3:1. Bilateral involvement is quite common but does not always involve bilateral symptoms.³

CASE REPORT: A 48 years old male presented with acute onset dysphagia with odynophagia to both solid and liquid foods and pain in the right oropharynx for one week duration. Swallowing was facilitated by tilting the head towards left side. He had a past history of left sided hemiparesis due to ischemic stroke involving right MCA territory 3 years ago, which had subsequently improved.

On examination mouth and oral cavity appeared normal. On deep palpation there was tenderness in the right tonsillar fossa with hard swelling palpable beneath it. Examination of cranial nerves was found to be normal. Indirect laryngoscopy revealed no abnormality.

Barium swallow, Upper GI endoscopy and X-Ray skull lateral view showed no abnormality. OPG showed an elongated styloid process of 38mm's length on the right side, while it was 25mm’s on the left side. Magnetic Resonance angiogram of neck and brain showed absent intracranial part of internal carotid artery on the right side (the main stem of the artery 2mm's after common carotid
artery bifurcation up to its junction with anterior cerebral artery was not visualized). Hence, the case was diagnosed as Right sided Eagle Syndrome possibly of the Stylocarotid variety.

**DISCUSSION:** Classic 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, exacerbating the patient's symptoms. Other symptoms include dysphagia, sensation of foreign body in the throat, tinnitus or cervicofacial pain.

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 process. It is related to a pain along the distribution of the artery, which is provoked and exacerbated by rotation and compression of the neck. It's not correlated with tonsillectomy. In case of impingement of the internal carotid artery, patients often have supra-orbital pain and parietal headache. In case of external carotid artery irritation, the pain radiates to the infraorbital region.

Symptoms of ipsilateral carotid artery compression secondary to an elongated styloid process or calcified stylohyoid ligament may be seen in Eagle syndrome; according to a case report by Chuang et al, where the patient can typically experience cervicofacial pain due to stimulation of the arterial nervous plexus. In addition, symptoms directly attributable to compression of the carotid artery may be seen, including visual symptoms and syncope.

The study reported a case of a patient who developed symptoms consistent with left hemispheric ischemia within 15 seconds of turning his head to the left. These symptoms were completely reversible on returning the head to the neutral position. No long-term sequelae were detected clinically or radiographically. In another study by Farhat et al, the authors discuss the case of a 70 years-old man who suffered from transient ischemic attacks on turning his head to the left, with immediate remission of symptoms when his head returned to the neutral position. Other atypical presentations of this disease include entrapment of glossopharyngeal nerve and even hypoglossal nerve palsy with Horner's syndrome.

In our study, the patient had almost near normal improvement of neurological deficits over few months after the stroke with physiotherapy and medical management. At the time of presentation to us, he had near normal power in all four limbs and mild facial palsy. This discordance between the clinical state and neurological deficit observed on imaging may be due to the fact that chronic vascular obstruction can over time lead to development of collaterals. This can reduce the neurological symptoms and can lead to significant improvement.

ES can be diagnosed radiologically and by physical examination. The elongated styloid process can be felt in the tonsillar fossa, and palpation can lead to an increase in symptoms. This elongation can be confirmed radiologically using conventional radiographs or CT scan. 3D CT helps in surgical planning and allows the physician to better explain the lesion and the surgical details to patient. It also helps to obtain measurements in three dimensions, along the plane of the styloid process being measured; in contrast there is underestimation of the length of the styloid process with two dimensional cross-sectional imaging, where even in the coronal plane, the images are usually not parallel to the styloid process.
ES can be treated by surgical and nonsurgical means. Nonsurgical treatments involve reassurance to the patient, analgesics, and steroid injections. Surgical treatment can be performed using one of two approaches: transpharyngeal or extraoral. The latter is thought to be superior because it is less likely to cause deep space infection of the neck.

CONCLUSION: Eagle syndrome is a rare condition seen in general population. It usually presents with only pain and odynophagia, although in this case it manifested as a serious condition in the form of ischemic stroke due to occlusion of the right internal carotid artery. Hence, aim of this case report was to make clinicians aware of this uncommon association of dysphagia and tendency for strokes seen in this case due to enlargement of styloid process.
BIBLIOGRAPHY:
**CASE REPORT**

<table>
<thead>
<tr>
<th>AUTHORS:</th>
<th>NAME ADDRESS EMAIL ID OF THE CORRESPONDING AUTHOR:</th>
</tr>
</thead>
</table>
| 1. Anil Kumar H.  
2. Swati Chouhan | Dr. Anil Kumar H,  
#306, 6th Main, 1st Cross,  
Remco Layout, Vijaynagar 2nd Stage,  
Bangalore – 560104.  
Email: dranilkh1@gmail.com | Date of Submission: 04/06/2014.  
Date of Peer Review: 05/06/2014.  
Date of Acceptance: 19/06/2014.  
Date of Publishing: 26/06/2014. |

<table>
<thead>
<tr>
<th>PARTICULARS OF CONTRIBUTORS:</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Associate Professor, Department of General Medicine, Rajarajeswari Medical College and Hospital.</td>
<td></td>
</tr>
<tr>
<td>2. Post Graduate cum Junior Resident, Department of General Medicine, Rajarajeswari Medical College and Hospital.</td>
<td></td>
</tr>
</tbody>
</table>