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Eagle Syndrome: An Atypical Neurological Presentation

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1. Abstract

Eagle Syndrome occurs in approximately 4% of the population and is commonly asymptomatic. This disease occurs secondary to the elongation of one or both styloid processes, causing symptoms like dysphagia, a clicking sensation when turning the head, headache, otalgia, and other cervical symptoms. Here we present a unique case of a 43-year-old female who presented for ENT evaluation after undergoing bilateral carotid artery stent placement and treatment for a left frontal lobe stroke who subsequently developed otalgia and a clicking sensation when turning her head. The CT obtained upon her initial presentation for stroke symptoms was re-evaluated and noted to have bilateral elongated styloid processes. The patient elected to undergo styloidectomy which led to relief of symptoms. Although our treatment plan and surgical approach is not novel, we believe that this unique presentation is of benefit to the further understanding and work up of Eagle Syndrome.

2. Keywords: Eagle Syndrome; Styloidectomy; carotid artery stenosis

3. Background:

Eagle Syndrome is a rare condition believed to affect approximately 4% of the population1. Most of these affected individuals are asymptomatic, and many cases of an elongated styloid process are found incidentally on radiologic imaging. The diagnosis of Eagle Syndrome is not indicated based solely on the finding of an elongated styloid process. However, if this finding is also associated with patient symptomatology such as cervical pain, dysphagia, a sensation of foreign body in the throat, otalgia, or a sense of clicking when turning the neck, then a diagnosis is deemed appropriate. In this case report, we describe an atypical presentation of Eagle's Syndrome in which a 43-year-old female presented with bilateral stenotic carotid arteries.

4. Case Presentation

A 43-year-old female with a history of bilateral deep earaches and severe headaches for the past 6 months presented to the Emergency Center at Covenant Hospital in Lubbock, Texas, with unequally dilated pupils and was subsequently admitted for stroke workup. A CT evaluation was found significant for severely stenotic carotid arteries bilaterally; The right carotid artery was found to be 90% occluded and the left was 100% occluded. The patient then underwent bilateral carotid artery stent placement and was treated for a stroke in the left frontal lobe. At that time, there were no plaques noted within the arteries. After the stent placement, the patient reported mild improvement in her headaches but complained of left eye drooping throughout the day consistent with Horner syndrome on the left side. She also experienced residual right sided weakness secondary to the left frontal lobe stroke. The patient then began to complain of a "clicking" sound when she turned her head from side to side and described a sensation of something catching deep within her neck. Because of this new complaint, CT scans were re-evaluated and long styloid foramens bordering the carotid artery stents bilaterally were noticed at that time (Figures 1,2). She was then referred to Otolaryngology at University Medical Hospital in Lubbock, Texas, for further workup.

At the time of the Otolaryngology evaluation, the patient continued to complain of a catching sensation when turning her head bilaterally, but reported that the sensation was worse on the right side. She also complained of a sore throat and ear pain. Diplopia, hearing changes, or cough were denied by the patient. She had no past medical history of neck radiation or trauma to the neck. Physical exam findings were significant for left eyelid ptosis, facial numbness over V2-3 distributions of the Trigeminal nerve, right anterior nasal septum deviation, bilateral tonsillar hypertrophy (+1, tonsillar fossa palpated bilaterally, right sided tenderness, no tenderness on the left), Mallampati class [1], right neck incision from previous cyst removal, and mild hyoid level tenderness on the right side. The remainder of the physical exam was within normal limits. Due to the patient's complaints and findings on CT imaging, the option of exploratory surgery and potential styloidectomy

was discussed with the patient. Risks vs benefits of this procedure along with the differences between a transoral vs cervical approach were thoroughly discussed with the patient. The patient elected to undergo bilateral syloidectomy.

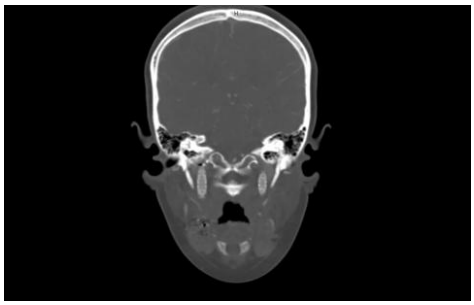

Figure 1: CT Angiography Head- Coronal view showing elongated styloid processes

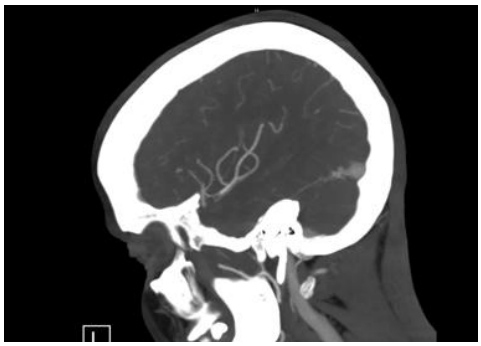

Figure 2: CT Angiography Head- Sagittal view showing elongated styloid processes

5. Operative Summaries

The patient was initially scheduled for bilateral styloidectomy. Consent was obtained and the patient was correctly identified and brought to operating room. Anesthesia was induced, and the patient was orotracheally intubated. The right neck skin fold marked and injected with 3cc 1:1000 units of lidocaine with epinephrine. An incision was created through skin, subcutaneous tissue, and platysma. Further blunt dissection was carried until exposure of posterior digastric and superior sternocleidomastoid muscle was obtained. The posterior digastric muscle was then retracted inferiorly. Palpation revealed a pointed portion of the styloid. The carotid sheath appeared 1cm medial to the styloid process while the patient's head was in a left turned position. The ligament attached to styloid process was peeled off, the styloid process was skeletonized, then 1.5cm distally was out fractured laterally. The dissected area was irrigated with saline and bacitracin. Evicel was applied to the cavity. The incision was closed by the platysmal layer then subcutaneous layer using vicryl. The skin was closed in a subcuticular fashion using 5.0 vicryl. Appropriate neck dressings were placed on right neck. Because the right styloid was not as calcified as originally anticipated, the surgeon did not proceed with a left styloidectomy, which was later discussed in length with the patient post-operatively.

At interval clinic follow up after her first surgery, the patient reported improvement in her symptoms. As such, she was interested to also pursue a similar surgery on the contra lateral side. The patient was then consented for left styloidectomy [2]. Operative steps and findings were similar to the initial surgery on the right side. The patient tolerated the procedure without any complications, and is doing well postoperatively with a similar improvement in symptomatology.

6. Discussion and Conclusions:

Although there is continued debate on the etiology of Eagle Syndrome, the most consistent symptomatology upon presentation consists of dysphagia, otalgia, and a sensation of a foreign body in the oropharynx. It is also thought that Eagle Syndrome can occur via irritation of the styloid process secondary to trauma or iatrogenic procedures, commonly after tonsillectomy [3]. The presentation of our patient is unique for several reasons. First, the neurological findings of this patient upon initial presentation are atypical for the classic case of Eagle Syndrome. The patient did have headache and otalgia, however due to her other neurological findings, these were originally attributed to be associated symptoms secondary to the stroke of her frontal lobe. When the original CT was obtained, elongation of the styloid processes was not noticed or if it was, it was not considered to be significant to the patient's pathology. Secondly, this patient had no history of surgery or trauma to the head or neck prior to her presentation. Although an atypical presentation, due to the elongated styloid processes and the patient's active symptoms, it was decided that the benefits of an exploratory surgery were warranted in order to evaluate the potential need of styloidectomy. After the procedure, the patient did report improvement of symptoms. At her follow up appointment a few months later, she was no longer experiencing a clicking sensation nor was she experiencing otalgia or the Horner symptoms on the left side. In conclusion, we deem it appropriate to consider Eagle Syndrome in the differential in a patient presenting with obstructive or stenotic carotid pathology and believe that this unique case is beneficial in further understanding uncommon manifestations of Eagle Syndrome.

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