Surgery is a safe option in management of severe stylo-carotid syndrome

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Abstract

Eagle syndrome is a rare though well-known condition defined by elongated or misshapen styloid process with or without calcification of the stylohyoid ligament. Although it is most often an incidental radiographic finding, the syndrome can present with neck and/or throat pain. Rarer still is a vascular variant of the disorder known as stylo-carotid artery syndrome. It is categorized by compression of the internal or external carotid by an enlarged styloid process. Here we present the case of a 53-year-old male who presented with retro-orbital headaches, left upper extremity weakness, falls, and neck pain. He was found to have a severe case of stylo-carotid artery syndrome with complete obstruction of the right internal carotid artery and 50-69% stenosis of the left internal carotid artery. The patient underwent right styloidectomy and right carotid endarterectomy (CEA) with pericardial patch angioplasty. His post-operative course was complicated by post-operative cervical cellulitis that was successfully treated with operative debridement and wound vacuum dressing placement. The patient is currently two months status post-surgery without any major complications. We present a case of severe stylo-carotid artery syndrome with significant carotid stenosis bilaterally managed successfully with open CEA and styloidectomy with excellent clinical outcome

Introduction

Eagle syndrome first described by Watt W. Eagle MD in 1937. The syndrome is characterized by an elongated styloid process, greater than 30mm in length.[1] Prevalence ranges from 4% as originally described by Eagle to as high as 18.2% in a radiographic prevalence study.[1-2] Symptomatic Eagle syndrome is much less frequent and has been divided into two distinct groups.[1,3] Classical Eagle syndrome presents with Symptoms of throat and neck pain, globus sensation, and referred otalgia. It is theorized to be largely due to compression of the sympathetic nerve plexus and the glossopharyngeal nerve.[3-5] Stylo-Carotid Syndrome is a much less common variant that presents with pain along artery course as well as neurovascular symptoms including TIA, syncope and stroke. It is felt to be due to direct compression as well as dissection and thromboembolism of the carotid artery due to direct mechanical injury by the enlarged styloid process.[3-5] In fact, multiple studies have shown that increased styloid process length is related to increased risk for carotid dissection.[6,7] One study found increasing odds ratio (OR) of dissection with both increased styloid process length and decreasing styloid process and carotid artery contact distance. Both intraoral and extraoral styloidectomy are a well described and accepted treatment of symptomatic Eagle syndrome that has been shown to be safe and efficacious in symptom improvement.[8] However, there is no general consensus on the treatment of stylocarotid syndrome and both single treatment or combination styloidectomy, endovascular stenting or thrombectomy, and open CEA have been proposed.[4,5,9]

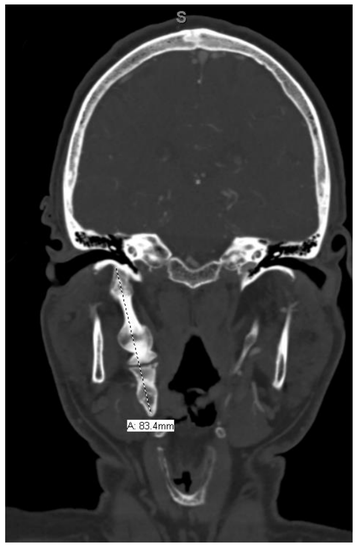

Figure 1. Right styloid process on CT imaging at initial presentation

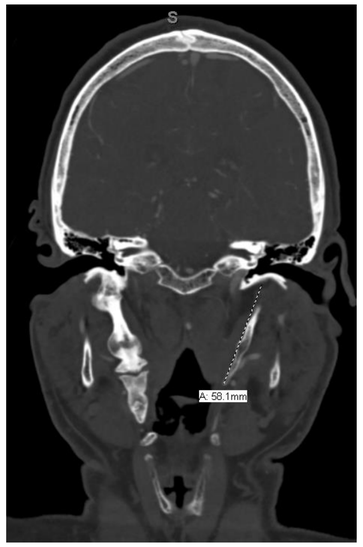

Figure 2. Left styloid process on CT imaging at initial presentation

Case Report

53 year old male with a past medical history of COPD, HTN, and alcohol abuse. He initially presented to the Emergency Department with chest pain but further questioning revealed one month history of right frontal / orbital / retro-orbital headaches, several falls and intermittent weakness of the left hand. Patient described a history of minor head trauma during physical altercations buts denied history of discrete neck trauma. CT angiography revealed right cervical / petrous internal carotid artery (ICA) stenosis with retrograde flow of the ophthalmic artery concerning for steal syndrome. CT also revealed significantly enlarged styloid processes bilaterally. Right styloid process measured up to 83 mm and left measured up to 54 mm. Though he did not have any previous angiographic imaging, the enlarged styloid processes are visible on plain film x-rays obtained 9 years prior to current presentation. Diagnostic angiogram confirmed near complete occlusion of the right ICA with cross flow through the anterior communicating artery (ACOM). MRI showed small late subacute right middle cerebral artery (MCA) ischemic infarcts consistent with thromboembolic infarcts. The patient was started on aspirin and clopidogrel prior to being discharged.

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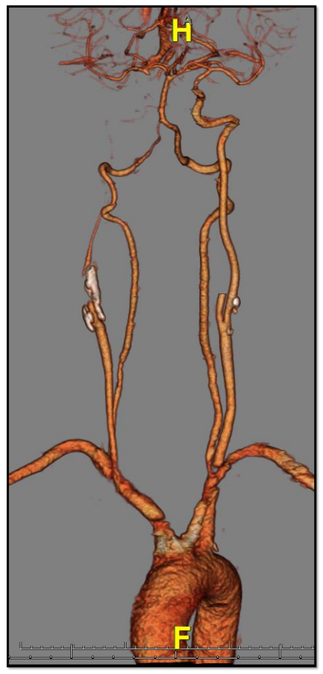

Figure 3. CT angiography at initial presentation

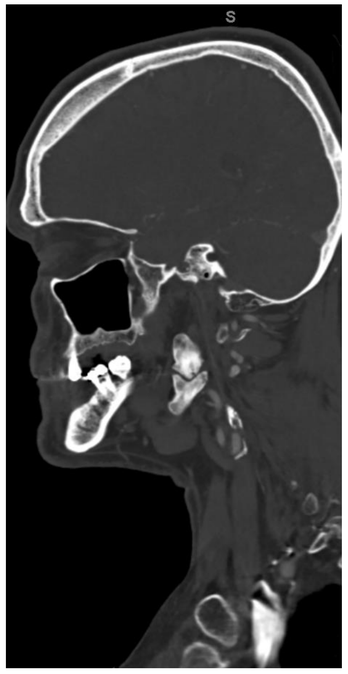

Figure 4. Sagittal view of left styloid process at initial presentation

Case Report - Cont

Patient returned for scheduled case. The patient was orotracheally intubated and extraoral styloidectomy performed. The facial artery was ligated due to its proximity to the styloid but CNXII was persevered throughout the case. The distal 2/3rds of the elongated styloid process was separated from the remainder of the styloid process as well as the hyoid bone using a powered oscillating saw. Vascular surgery then completed CEA with pericardial patch angioplasty and the patient was turned over to anesthesiology for emergence from anesthesia. Post-operative course unfortunately complicated by cellulitis without abscess. He returned to the ED for this on POD#7 and underwent incision and debridement in the OR with wound vacuum dressing placement. The wound was irrigated and closed on POD#9 in the OR. He recovered quickly with antibiotics.

He is currently 2 months status post-surgery and remains without any serious complications. As the patients left styloid process is 54mm the plan is for left styloidectomy in the future.

Discussion

There is no clear consensus on the treatment of stylocarotid artery syndrome. A variety of single and combined modality treatments have been published in the literature including anticoagulation, styloidectomy, endovascular stenting or thrombectomy, and open CEA.[4,5,7] We present a case of severe bilateral stylocarotid artery syndrome with massively enlarged styloid processes and unilateral complete ICA obstruction. In the presented case, the right styloid process measured 83 mm and the left measured 54 mm. Given the patient's right sided symptoms and complete obstruction invasive intervention was indicated. Bearing in mind the massive enlargement of his right styloid process, which to our knowledge is the largest recorded in the literature, open extraoral styloidectomy with concurrent CEA was performed. The patient recovered without any severe complications and is now asymptomatic 2 months status post-surgery. This technique may be applicable to other patients with severe bilateral stylocarotid artery syndrome with massively enlarged styloid processes

Conclusion

Stylocarotid syndrome is a rare variant of the more common Eagle syndrome and can have serious complications due to carotid artery injury. A variety of treatment methods have been proposed without any clear consensus on the optimal method. We present a case of severe bilateral stylocarotid artery syndrome with massively enlarged styloid processes and unilateral complete ICA obstruction. The patient was treated with open extraoral styloidectomy and concurrent CEA. He remains asymptomatic and without severe complication 2 months status post-surgery. Measured at 83 mm the styloid process removed in this case is among the largest published.

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