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Case report

Battling the Eagle's sharp beak, Eagle syndrome; a case report

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ABSTRACT

Eagle Syndrome is a pain syndrome of rare and unwonted incidence. Forbearer has an elongated styloid process or a calcified stylohyoid ligament, suppressing glossopharyngeal nerve leading to a mélange of symptoms including sporadic cervicofacial pain, headache, and foreign body sensation. Here we present case of a 65 year old military man of south Asian origin, who presented with complaints of sudden episodes of blackouts for past five years and pain in neck while turning head to left for past two months.

Patient's ultrasound Doppler showed marked narrowing of proximal left internal carotid artery with approximate diametric stenosis of 70% according to The North American Symptomatic Carotid Endarterectomy Trial (NASCET).Further studies of MRI Brain was done,revealing small Foci of restricted diffusion along Territory of Left MCA along with age related Microangiopathic cerebral changes. CT Scan of neck was also done which showed Abnormal elongation of bilateral styloid process more on the left side.

The case was discussed in a Multidisciplinary Team Meeting comprising ENT surgeon, vascular surgeon and surgical excision was planned through trans cervical approach. Surgery was successful as seen by post op and follow up scans.

1. Introduction

An uncommon but well known otolaryngology entity eagle syndrome results, when an elongated styloid process compresses the cranial nerves resulting in a variety of symptoms including unilateral cervical and facial pain [1,2]. This sometimes also can lead to compression of the cervical carotid arteries leading to the “Carotid Artery Syndrome” with neurological symptoms due to the reduced blood flow in these arteries [3]. In our study we had a 65 year old male with no known co-morbids who presented with complaints of sudden episodes of blackouts for the past five years and pain in neck while turning head to left for the past two months. The sudden loss of consciousness would remain for 30–60 min and patient would attain consciousness after conservative treatments from a nearby local hospital. The principle for reporting this case not only lies with it being a rare presentation in our knowledge but also on the fact of it being reported in an elderly patient with no identified comorbids. The elongated styloid process syndrome can be managed both conservatively and surgically. Conservative treatments include analgesics, antidepressant, medications, anticonvulsant, trans pharyngeal injection of steroid and lidocaine. The most curative treatment is surgical resection of styloid process through trans cervical or trans oral approach. Trans cervical approach is more challenging and safe. [1,3]

2. Presentation of case

This case report covers a 65 year old male ex-military soldier belonging to a middle socioeconomic class of south Asian origin who is a known smoker with 20 pack years history with no known comorbids, presented in outpatient department with complaints of sudden episodes of blackouts for past five years and pain in neck while turning head to left for past two months.The patient was in usual state of health five years ago when he had sudden loss of consciousness which remained for

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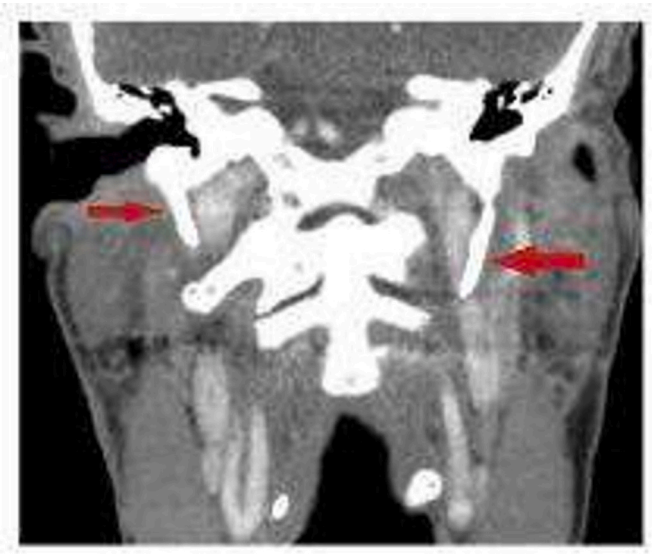

Fig. 1.1. CT Scan of neck was also done which showed Abnormal elongation of bilateral styloid process.

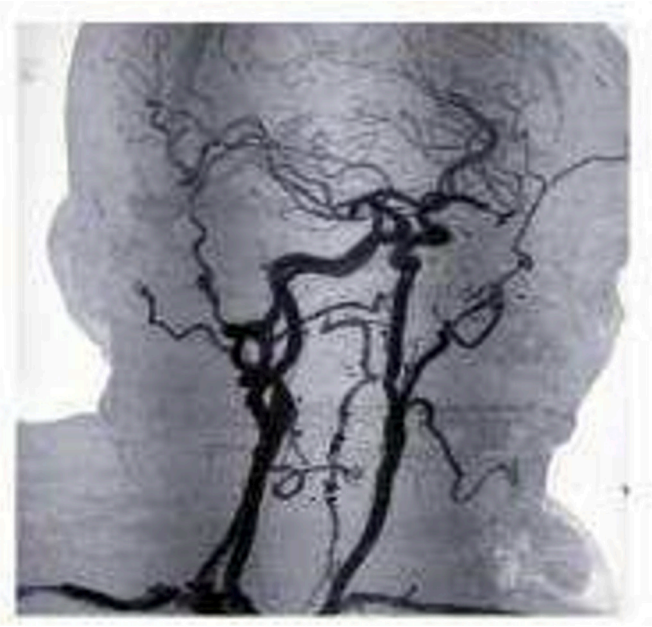

Fig. 1.2. CT angiogram was requested which showed marked stenosis with filling defect as we can see in ICA on Left side.

30–60 min then patient attained consciousness after conservative treatment from local hospital. Patient had 7–8 episodes in last five years and 4–5 months apart. According to patient he had spontaneous recovery from black outs without any specific treatment. He was never investigated. Patient had last attack last year. Patient was also having pain in neck while turning head to left for last two months which was diffuse and non-radiating and aggravated with movement of neck to the left side having score of 5–6/10according to numeric rating scale (NRS) without any relieving factors.Patient with these complains landed in Medical and ENT department of our hospital.

Patient's ECG, 2D ECHO and Holter test were done to rule out cardiac pathology which were normal. When patient's radiological evaluation was started X-ray neck was normal but Ultrasound Doppler neck showed marked luminal narrowing of proximal left internal carotid artery with approximate diametric stenosis of 70% & Peak systolic velocity up to 130 cm/s was seen. Furtherstudies of MRI Brain was done revealed Small Foci of restricted diffusion along Territory of Left MCA along with age related Microangiopathic cerebral changesCT Scan of neck was also done which showed Abnormal elongation of bilateral styloid process more on the left side (Fig. 1.1).

Later on patient's CT angiogram was requested which showed Marked Stenosis with filling defect as we can see in ICA on Left side (Fig. 1.2) and in coronal and sagittal section that styloid process is abnormally elongated greater on the left side and impinging ICA (Fig. 2.1). The report mentioned abnormal elongation of bilateral styloid process / calcification of stylohyoid ligament (left > right) (raising the possibility of eagle syndrome) causing compression and approximately 74% stenosis of left internal carotid artery according to NASCET criteria. After all the work-up the diagnosis of eagle syndrome was made.The case was discussed in a Multidisciplinary Team Meeting comprising ENT surgeon, vascular surgeon and surgical excision was planned through trans cervical approach as styloidprocess was lying deep and not palpable in tonsillar fossa. Vascular team was taken on board to cater for any vascular compromise. Patient's surgery was planned and Pre Anesthesia Assessment was done. During his admission period before date of surgery a counselling session was conducted in which the patient and the attendants were briefed about the nature of disease, the treatment options available with their merits and demerits and the possible risks involved in the surgery. Special emphasis was given on understanding potential damage to CN VII, IX, and XII during surgery. (See Figs. 3–4.4.)

Post-Operatively patient made an uneventful post-operative recovery and was provided with antibiotics, analgesics and wound care. Nerve function of Marginal mandibular along with Hypoglossal was completely functional. Drain was removed at 72 h and the patient was discharged by 4th post-operative.

Patient followed up after 4 weeks. He resumed normal lifestyle with 100% relief of symptoms. Post Op CTA done after 4 weeks which revealed reversal of stenotic effect. As we can appreciate that there is no filling defect or stenosis seen. Both external and internal carotid arteries are normal with smooth lumen and texture.

3. Discussion

The rationale for reporting this case lies in the fact that our case is a rare entity geographically and our patient had no prior surgical history, his plain radiograph of head and neck did not show an elongated styloid process as seen in previous case reports [4–6].Our case involves a 65 year old male with no known comorbids and otherwise healthy who presented to a tertiary care hospital with having multiple episodes of unconsciousness when turning the head to one side and ipsilateral sided neck pain, he was diagnosed as having Eagle's Syndrome by using different imaging modalities and correlating them to physical findings. Eagle's Syndrome also referred to as Stylohyoid syndrome or Styloid-carotid syndrome is an entity defined in 1937 by German doctor Watt Eagle as pain in laryngeal region, face or neck due to elongation of the styloid process with or without calcification of the stylohyoid ligament. The pathophysiology of this syndrome has been stipulated to be due to the close relation of the styloid process to the carotid canal and jugular foramen leading to compression of vessels and nerves.

Clinically Eagles syndrome is further subdivided into two main

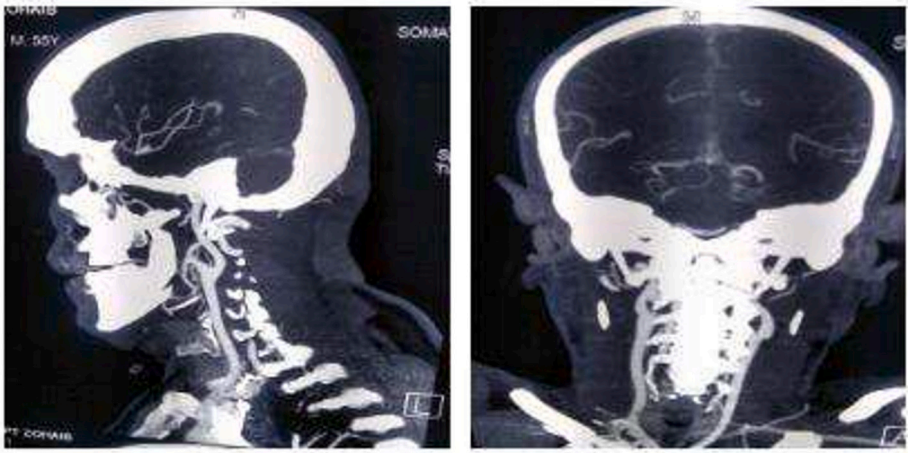

Fig. 2.1. CT angiogram in coronal and sagittal section that shows styloid process is abnormally elongated greater on the left side and impinging ICA.

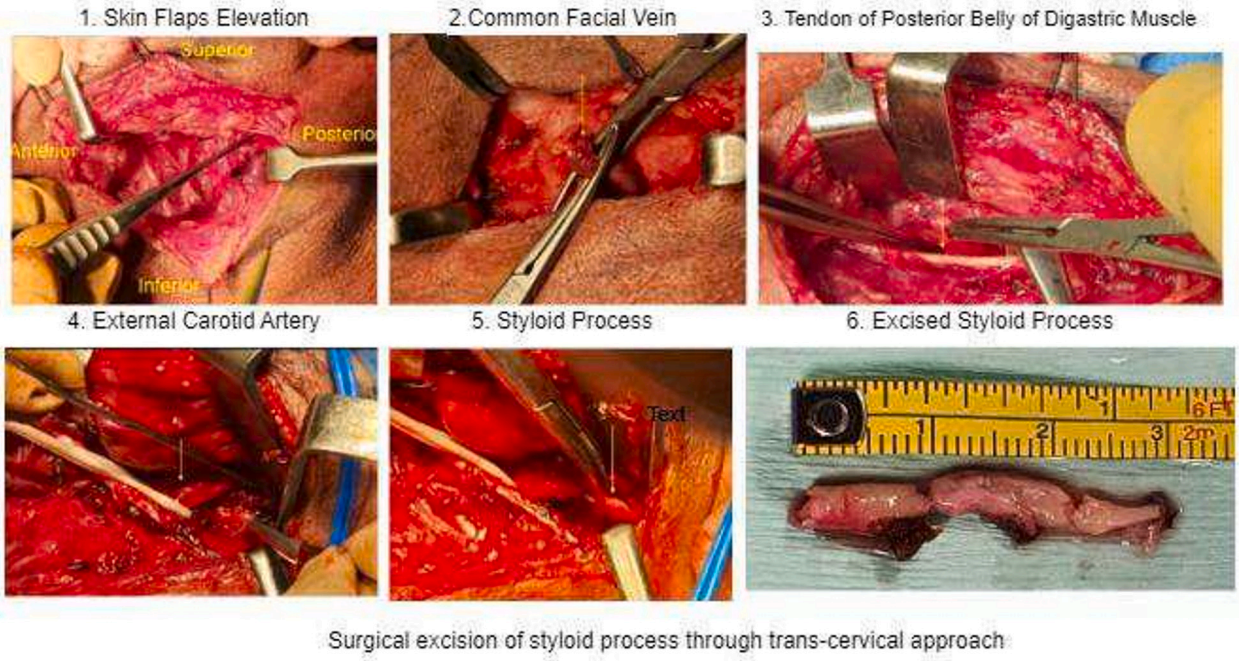

Fig. 3. 1. Skin Flaps Elevation. 2. Common Facial Vein. 3. Tendon of Posterior Belly of Digastric Muscle. 4. External Carotid Artery. 5. Styloid Process. 6. Excised Styloid Process.

classes, class 1 which is mainly neurological and due to compression of cranial nerves five, seven, nine or ten. Symptomatically patients present with odynophagia, dysphagia or foreign body sensation in the neck. Type 1 eagle's syndrome is also called the classical type, it is stipulated that type 1 Eagle's syndrome is more common in patients who have underwent tonsillectomy but in our patient there was no history of prior surgery or tonsillectomy.

Type 2 or Carotid type ofEagle's syndrome is due to compression of either of the two carotid arteries (internal carotid or external carotid) and presents as orofacial pain, neck pain, headaches or episodes of syncope on turning face to contralateral side of the pathology, it can also present as constant neck pain which is exacerbated by turning head to ipsilateral side. The compression can also lead to Transient ischemic attack or at times carotid artery dissection which is rare complication. In our case the patient had carotid type of Eagle syndrome which was proven by imaging. [4–6]

An enlarged styloid process can be found in approximately6 to 7% of the population but not all the people are symptomatic [4]. Eagle's Syndrome is diagnosed using physical examination along with imaging such as plain radiographs, CT scan and Ct angiography; some suggest 3D reconstruction with CT scan is the best imaging modality [4–6]. In our case plain radiograph of the neck showed no abnormality, CT head showed an elongated left styloid process, a styloid greater than 30 mm is considered abdnormal. CT angiogram was used which showed bilateral carotid artery stenosis which was greater on the left causing 74% stenosis which is diagnostic for eagle's Syndrome. An MRI brain showed

Fig. 4.1. Postoperative picture.

Fig. 4.2. Postoperative recovery picture.

microangiopathic changes inthe territory of the left MCA.

Management of Eagle's Syndrome is broadly categorized into two namely conservative management and surgical management. Conservative management is done using analgesics, steroids and antiepileptics but it is reserved for patients who do not wish to proceed with surgical management. Surgical management is done by either employing the intraoral approach which is done in patients in which the styloid process is palpable in the tonsillar fossa, the intraoral approach is beneficial as there is no external scar and hence a better cosmesis. A second approach is transcervical approach and is done in patients in whom the styloid process is not palpable through the tonsillar fossa and is lying deep [7,8]. In our case a transcervical approach was used.

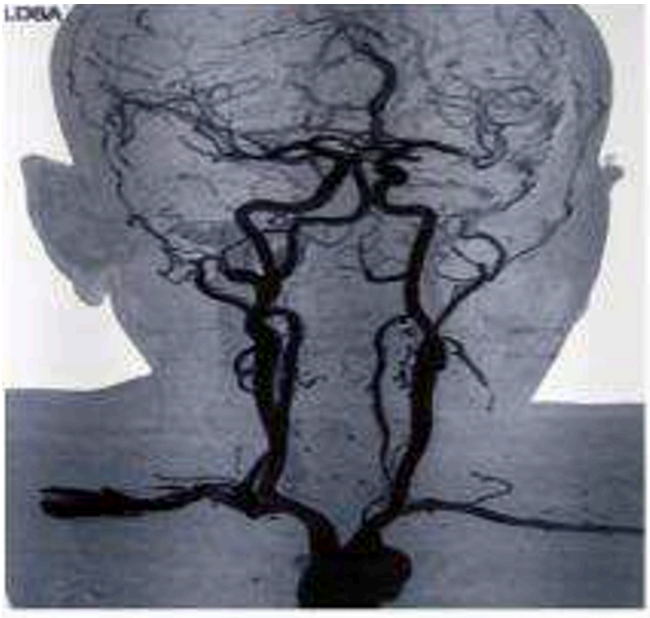

Fig. 4.3. Post operation CTA.

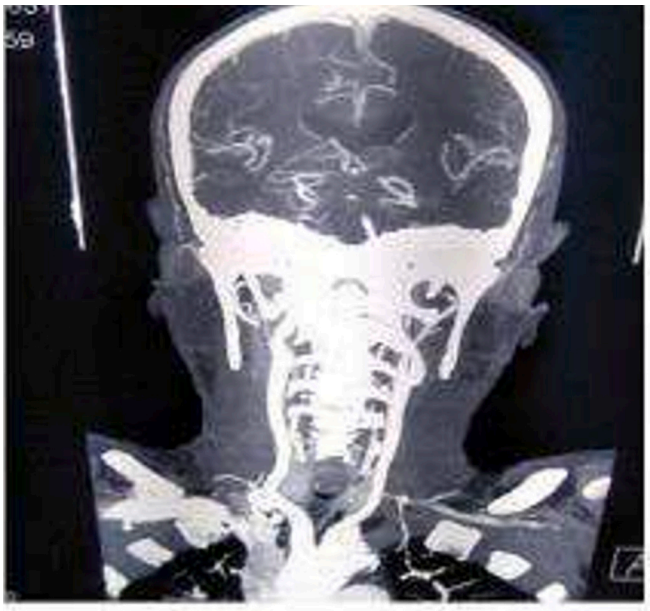

Fig. 4.4. Follow up CTA.

4. Conclusion

Eagle's syndrome is a rare entity which is due to an elongated styloid process, although an elongated styloid process is found a considerable percentage of the population but not everyone is symptomatic. Diagnosis of the syndrome is done using physical examination along with imaging. Physicians should be vigilant and have Eagle's Syndrome among the list of differentials in people presenting with syncopal episodes and neck pain.

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