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*Case Report*

# Eagle's syndrome in tertiary health institution, southern region of Nigeria

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## ABSTRACT

Eagle's syndrome is a condition caused by elongation of the styloid process or ossification of the styloid ligament resulting in non-specific symptoms related to pressure on vital blood vessels of the neck, the internal carotid artery, internal jugular vein, and nerves; the glossopharyngeal and vagus nerves. Eagle's syndrome may have varied presentations with numerous symptoms: Pain in the throat/neck radiating to the ear and pharynx, foreign body sensation in the throat, dysphagia, otalgia, trismus, and intense facial pain. A high index of suspicion with a prompt resort to a three-dimensional CT skull could be helpful to give a quick diagnosis of Eagle's syndrome amid numerous differential diagnoses. We present a 59-year-old male with an 18-month history of left-sided neck pain, cheek pain, and the feeling of a foreign body in the throat. He had pain relievers and other drugs to no avail. A three-dimensional CT scan of his skull revealed features of Eagle's syndrome. The excision of the styloid process through the extraoral route relieves his symptoms. The second patient is a 42-year-old female with symptoms of peptic ulcer disease, dysphagia, pain in the throat, and sometimes fainting attacks when she moves her neck swiftly to the left side. A three-dimensional CT skull scan confirmed features of an elongated styloid process bilaterally suggestive of Eagle's syndrome. Treatment offered to her was conservative, with a diclofenac suppository only because she refused surgery. Restricted and limited neck movement relieved her of syncope attacks.

**Keywords:** Eagle's syndrome, Styloid process, Styloidectomy

## INTRODUCTION

Eagle's syndrome (ES) is a cluster of symptoms described by Watt W Eagle in 1937. These symptoms resulted from elongation of the styloid process or calcification of the stylohyoid ligament. It is also called styloid syndrome.[1] As the styloid process escapes from the stylomastoid foramen under the petrous bone, it bypasses several structures; internal carotid artery, maxillary artery, internal jugular vein, glossopharyngeal nerve, vagus nerve, and branches of the trigeminal and facial nerves.[2] Thus, you may have different shades of facial pain, cervicopharyngeal pain, dysphagia, foreign body sensation in the throat, and sometimes trismus. The abnormal styloid process may sometimes compress the cervical carotid arteries, leading to the dreaded carotid artery syndrome, or stylocarotid syndrome, which presents with transient ischemic attacks, especially when the patient turns the neck to the affected side. Rarely, death may occur due to vagus-mediated cardiac arrest. The styloid process elongation may be unilateral or bilateral, and most of the time, it is asymptomatic and often discovered during routine radiographic examination. The asymptomatic category requires no treatment. However, one should suspect Eagle's syndrome in the elderly, especially women of 60 years[2] and above who presented with unilateral symptoms that were not responsive to analgesics.

## CASE REPORTS

We report a 59-year-old male who presented to our facility with a 9-month history of left-sided neck and cheek pain and foreign body sensation in the throat. He had taken many painkillers, which include nonsteroidal anti-inflammatory drugs (NSAIDs) but to no avail. He reported no history of preceding trauma or throat surgery. He had a three-dimensional CT scan of the head and neck, which revealed the left styloid process elongation [Figure 1a and b]. The styloid process on the right was 28 mm in length, and on the left was 33 mm. The patient had left styloidectomy through the extraoral route with the resolution of his symptoms. Using the Radisson incision – a lazy-S incision from just before the tragus moves backward under the lobule and curves forward toward the neck in a skin crease in line with the hyoid bone; exploration continued by blunt dissection until the styloid process was found and cut at the root. The symptoms ceased 3 weeks after the operation, but the slight deviation of the angle of the mouth to the opposite side due to trauma to the ipsilateral facial nerve resolved after 8 weeks.

### Case 2

Our second case is a 42-year-old female with a 2-year history of left-sided pain in the face, neck, and throat. She can eat and swallow with occasional dysphagia. In addition, she complained of fainting attacks whenever the swift movement of her head from the right side to the left. The general practitioners placed her on NSAIDs and some self-medication without relief of her symptoms. Because she had been on NSAIDs repeatedly and was manifesting symptoms of peptic ulcer disease, she was placed on rectal diclofenac and requested to do a three-dimensional CT of the skull with the enhancement of the styloid processes on both sides, which confirmed Eagle's syndrome [Figure 2a-d]. She was treated conservatively with a diclofenac suppository because she could not tolerate oral diclofenac since she had peptic ulcer disease. She could not have surgery done on her because she refused consent for the operation. However, the restricted head movement reduced the incidence of syncope attacks.

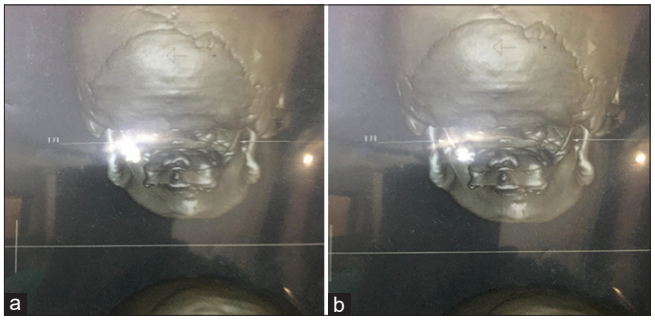


**Figure 1:** 3D CT SCAN of the Head and Neck of a 59-years old- man with 28mm and 33mm right and left styloid process respective.

## DISCUSSION

Eagle syndrome is usually a consequence of a long styloid process, ossification of the stylohyoid ligament, or both. The styloid process juts out from beneath the squamous part of the temporal bone, it crosses many structures; the internal carotid artery, the maxillary artery, the internal jugular vein, branches of the glossopharyngeal and vagus nerve, and those of the facial and trigeminal nerves.[2] A variant of Eagle's syndrome may have styloid elongation coursing through the transverse process of C1, causing significant compression of the internal jugular vein. The pressure or interference with these structures may give rise to a cocktail of symptoms such as cervicopharyngeal pain, foreign body sensation in the throat, vague facial pain, otalgia, dysphagia, trismus, odynophagia, sometimes Horner's syndrome, transient ischemic attack – the so-called stylocarotid syndrome, and rarely death due to vagus-related cardiac arrest. The symptoms may be unilateral or bilateral, and a long styloid process is usually an incidental finding in patients doing routine neck X-rays. Often time, patients with the long styloid process are asymptomatic.[2]

The incidence of Eagle's syndrome is said to be in the region of 4–8 in 10,000.[2] The etiology is not so clear, but Eagle himself thought that traumatic scarring and hyperplasia due to the previous surgery of tonsillectomy were the secondary causes of the elongation of the styloid process. ES is seen more commonly in females, especially from the fourth to the sixth decade, and *ossification of the styloid ligament is related to endocrine disorders in women at menopause*.[3] Our first case was a male in the fifth decade, while the second was female in her fourth decade. ES may be hereditary, likely

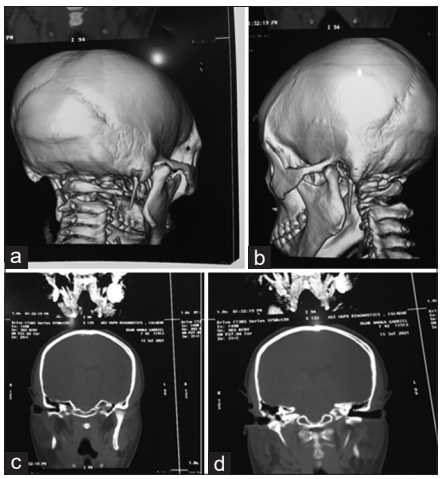


**Figure 2:** 3-dimensional CT of the skull (with enhancement) showing elongated styloid processes measuring 33mm and 31mm on the left and right respectively (a and b) with a normal brain study (c and d).

due to an autosomal dominant gene.[4] The length of the styloid process is from 25 mm to 30 mm, while 30 mm and above is elongated.[5] However, opinions are highly varied on the length of the styloid process and there seems to be no correlation between the styloid process length and the severity of symptoms in patients with Eagle syndrome.[5] In addition, many studies have shown that symptoms occur when the styloid process is about 40 mm.[6] Watt W Eagle stated that about 4% of individuals whose length of the styloid process is >25 mm in adults could be symptomatic.[7] Godden *et al*. reported that affected individuals with ES seldom develop symptoms.[8] The symptoms of Eagle syndrome are non-specific and the disease may run a protracted course. The diagnosis is basically by radiological features. The panoramic or plain neck lateral radiographs are sufficient for the diagnosis.[9] Furthermore, a 3D CT skull scan remains the gold standard for ES diagnosis. The two cases reported had a 3D CT skull scan and their styloid process on the affected side was 33 mm.

The modality of treatment includes medical and surgical therapy. The medical options involve pain control using oral analgesics, transpharyngeal infiltration with steroids, and local anesthesia in the tonsillar fossa.[9] However, the treatment of choice is styloidectomy, either transoral or extraoral route.

A 3D CT scan of the skull confirms the diagnosis of Eagle syndrome in both cases. In addition, a CT angiography is better for the jugular variant of Eagle's syndrome, which may be distinct. The Jugular variant more often presents clinically as a headache.[10] The first case was treated with both conservative and surgery, while the second opted only for conservative management.

## CONCLUSION

Eagle's syndrome is a rare cause of facial or cervical neuralgia. Therefore, it should be considered in differential diagnoses of facial or cervical neuralgias, especially when dealing with unilateral symptoms, with no apparent cause of neck pain that is unresponsive to painkillers.

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### Declaration of patient consent

Patients' consent not required as patients identity is not disclosed or compromised.

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### Conflicts of interest

There are no conflicts of interest.

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