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Case Report

Eagle syndrome in a patient with Noma sequelae: A case report and a review of literature

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Abstract

Eagle syndrome (ES) is characterized by a group of radio-clinical features including elongation of the styloid process or the calcification of stylohyoid ligaments, headaches, facial pain, ear pain and, feeling of foreign body sensation in the throat. Eagle syndrome is rare and most of the time, it is diagnosed incidentally especially when no or few symptoms are present. We present a case of a 22-year-old patient with a complaint of inability to open the mouth of about 19 years duration, recurrent bilateral earache, and pain in the submandibular and cervical regions of about 6 months duration. Computed tomography (CT) scan revealed normal temporomandibular joint space bilaterally, elongated styloid process and calcified stylohyoid ligament. An assessment of Eagle syndrome on background was made and the patient had bilateral coronoidectomy, release of soft tissue contractures with favorable outcome. The stylohyoid ligament contributes to the mandibular movement and, its calcification or the elongation of the styloid process seen in patient with chronic Noma sequelae has been linked with long-standing immobility of the mandible following trismus release. More evidence is awaited to this to substantiate this finding.

Keywords: Noma, Stylohyoid ligament, Styloid process, Syndrome

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Introduction

The styloid process is an anatomically cylindrical, slender, needle-like projection of varying lengths averaging 2 to 3 cm. The styloid process projects from the inferior aspect of the petrous temporal bone and gives attachment to the stylohyoid ligament (1). The stylohyoid ligament also has its insertion into the body of hyoid bone at its junction with the greater cornu and is just superior to the omohyoid muscle (1). The styloid process facilitates the movement of the tongue, pharynx, larynx, hyoid bone, and mandible (2). Elongation of the styloid process or calcification of the stylohyoid ligament has been or both in association with certain clinical symptoms that have been termed Eagle syndrome (1, 2). The incidence of this syndrome has been reported worldwide as 4-8 cases per 10,000 population (3). It was found to be more common in females and, on average 40 years of symptoms presentation (4). The exact cause of Eagle syndrome is unknown however, several theories have been proposed, such as congenital elongation due to the persistence of an embryonic cartilaginous outgrowth, calcification of the stylohyoid ligament, and formation of bone tissue at the insertion of the ligament (4, 5). Eagle syndrome was also reported in the patient's post-tonsillectomy. In this report, a case of eagle syndrome in a 22-year-old patient with Noma sequelae has been presented.

Case Report

A case of a 22-year-old patient presented to Noma Children Hospital Sokoto with a complaint of inability to open their mouth for about 18 years duration. The patient had difficulty in feeding, speaking and, bilateral earache. The patient was treated for acute Noma when he was 2 years old and, has no history of tonsillectomy or any other surgery in the past. Examination revealed zero mouth opening, left oral commissure scarring Figure 1 (a), loss of left oral vestibule, adhesion of soft tissue of the inner cheek to the maxillary area, and, loss of upper anterior dentition (incisors). Computed tomography (CT) scan revealed normal temporomandibular joint space bilaterally, elongation of the styloid process, and calcification of stylohyoid ligaments bilaterally (Figure 2). There was also elongation of the coronoid process bilaterally. An assessment of Noma sequelae (trismus, bilateral coronoid hyperplasia, left commissure, and vestibular tissue scaring) and eagle syndrome was made. The patient had counsel and consented to trismus release under a

general anaesthesia. He had a bilateral coronoidectomy and release of soft tissue adhesion with a resultant mouth opening of about 4.5cm Figure 1 (b).

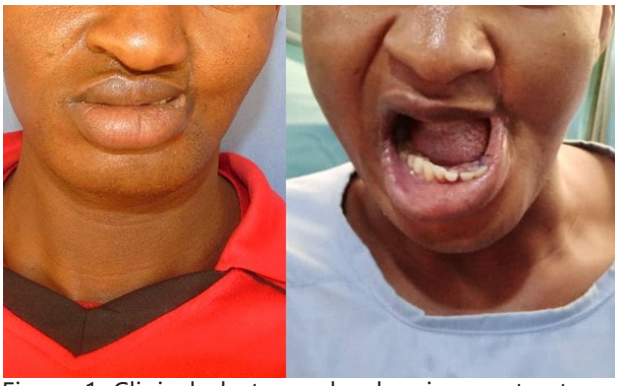

Figure 1: Clinical photographs showing contractures on the left side of the face (Blue arrow) in b and, the amount of mouth opening of about 4.5cm during the postoperative phase (in b).

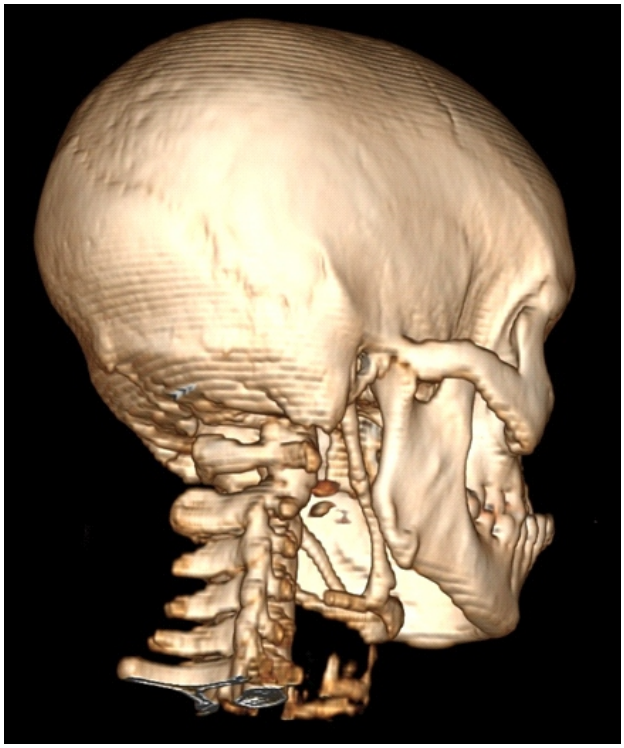

Figure 2. Reformatted 3D volume-rendered computed tomographic image of the skull and neck, right posterior oblique view showing bilateral elongation of the styloid process blended with calcified stylohyoid ligament (Blue arrows).

Discussion

The first mentioned cases of styloid process elongation with associated pain symptoms were reported by Watt Weems Eagle in 1937 hence, the term Eagle syndrome (5). The styloid process anatomically gives attachment to the stylohyoid ligament which serves as an accessory structure that participates in mandibular movement and, swallowing (1). In Eagle syndrome, there could be either elongation of the styloid process or the calcification of the stylohyoid ligament, or both (5). In this case report both the elongation of the styloid process and the calcification of the stylohyoid ligament were observed through the computed tomography scan.

The etiopathogenesis of Eagle syndrome is not fully understood, however, certain theories were put forward. The first theory was the "theory of reactive hyperplasia" which suggests that trauma to the pharyngeal area will activate reactive proliferation of the styloid process causing its elongation (1, 6). The second theory was the "theory of reactive metaplasia" secondary to trauma. The stylohyoid ligament may also inherently undergo metaplasia and, partial ossification (1, 7). The third theory is the "theory of anatomic variance," which proposes that the calcification of the stylohyoid ligament and the styloid process is a normal process that constitutes an anatomical variation (8). The fourth theory suggests that the elongated styloid process is a result of retained embryologic tissue from Reichert's cartilage (2, 8). Although few investigations suggest that dystrophic and degenerative changes in the hyoid complex of the styloid process might be the cause of ES, others suggest that cervicofacial inflammations, tumors, or tonsillectomies could play a major role in causing ES (7, 8, 9, 10).

Although these theories expressed some explanation for the possible etiopathogenesis of styloid process elongation or the calcification of the stylohyoid ligament seen in Eagle syndrome, a definitive accord has not been established. The calcification of the stylohyoid ligaments seen in the current case could have been triggered by the effect of the infectious and inflammatory process of acute Noma in the patient when he was 2 years as mentioned previously in the history.

A wide array of symptoms such as Earache, odynophagia, pain in the facial or cervical regions, the feeling of foreign body sensation in the throat, and recurrent headaches or vertigo could occur variably in patients with ES and these symptoms which are predominantly pain syndrome are attributed to the affectation of the anatomical structures (cranial nerves V, VII, IX and, X) surrounding or near the calcified stylohyoid ligaments or the elongated styloid process (2, 4, 9).

Two types of Eagle syndrome were recognized. These are the classic type and the carotid artery type. The classic type was said to arise following trauma to the pharyngeal region or post tonsillectomy where the scarring or calcification could compress or stretch the nerves structures in the space around the styloid process, whereas in the carotid artery type, the symptoms are due to compression of the carotid plexus and, this causes a feeling of foreign body sensation in the throat.

The current case presented with a bilateral earache, and pain in the submandibular and cervical regions. These are likely due to a referred pain from the affectation of sensory branches of the above cranial nerves that mainly occur in the classic type.

The diagnosis of ES has been based on clinical presentation, physical manipulation, and imaging modalities. The clinical presentation will only outline ES as a differential but confirmation depends on the radiographic evidence of styloid process elongation or the calcification of stylohyoid ligaments or both (11, 12). The elongation of the styloid process and calcification of the stylohyoid ligament has been seen bilaterally from the CT scan of this current case. In the absence of advanced imaging, plane radiographs such as a posterior-anterior x-ray of the skull could offer diagnostic benefits to some extent. Calcification of the stylohyoid ligament can be seen incidentally as a finding from imaging in the absence of any symptoms and that alone cannot qualify it to be ES. Other symptoms have to be explored by the patients. The non-surgical treatment of Eagle syndrome generally involves pharmacotherapy with anticonvulsants (e.g., gabapentin, pregabalin) or antidepressants, but results are short-lived (13). Long-lasting symptom relief requires the surgical removal of the elongated portion of the styloid process and, is related to symptom healing of patients with favorable outcomes (14).

In the current case, no treatment is offered yet for Eagle syndrome, since the patient's concern is to restore his mouth opening. The interincisal distance achieved was 4.5cm with bilateral coronoidectomy, and, scar excision. Coincidentally the patient reported symptoms resolution after the postoperative period but this may not rule out the need for surgical removal of the calcified ligament in the near future.

Conclusion

The stylohyoid ligament contributes to the mandibular movement and, its calcification or the elongation of the styloid process seen in patient with Noma sequelae has been linked with long-standing immobility of the mandible following trismus release. More evidence is awaited to substantiate this finding.

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