Eagle’s syndrome with facial palsy

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Abstract:
Eagle’s syndrome (ES) is a rare disease in which the styloid process is elongated and compressing adjacent structures. We describe a rare presentation of ES in which the patient presented with facial palsy. Facial palsy as a presentation of ES is very rare. A review of the English literature revealed only one previously reported case. Our case is a 39-year-old male who presented with left facial palsy. He also reported a 9-year history of the classical symptoms of ES. A computed tomography scan with three-dimensional reconstruction confirmed the diagnoses. He was started on conservative management but without significant improvement. Surgical intervention was offered, but the patient refused. It is important for otolaryngologists, dentists, and other specialists who deal with head and neck problems to be able to recognize ES despite its rarity. Although the patient responded to a treatment similar to that of Bell’s palsy because of the clinical features and imaging, ES was most likely the cause of his facial palsy.

Keywords:
Eagle’s syndrome, elongated styloid processes, facial nerve palsy

Introduction

The styloid process is a slender pointed piece of bone below the ear, just behind the neck of the mandible and in front of the mastoid process. The normal length of the styloid process varies, normally ranging from 2.1 to 2.9 cm. It is considered elongated when it exceeds 3 cm.[1]

There are two main types of Eagle’s syndrome (ES). In the first type known as the “Classic type” or otolaryngological syndrome, the styloid process causes mechanical irritation of the lower cranial nerves leading to dysphagia, odynophagia, foreign body sensation, and dull unilateral pharyngeal pain radiating to the ear and exacerbated by swallowing, yawning, and neck movement.[2] In the second type known as the “stylocarotid” syndrome, there is an irritation of the internal carotid artery with its sympathetic chain and usually presents with neck pain associated with parietal headache radiating to the periorbital region, and in rare cases may lead to a stroke.[3] Many physicians fail to recognize ES because of its varied symptoms which resemble symptoms and signs of many other diseases and consequently misdiagnose and mistreat. The goal of our study is to present a rare case of this syndrome where the patient presented with facial palsy along with other ES classical symptoms, to increase awareness of the clinical presentation of this rare syndrome, the best diagnostic modalities, the proper management, and avoid unnecessary medications or procedures.

Case Report

A 39-year-old male presented at the emergency department with a 1-day history of left-sided facial weakness and heaviness associated with the inability to close his left eye, sagging of the left corner of the mouth with drooling, and a flattening of the left forehead as well as nasolabial fold. His sense of taste and salivation were intact.

He also reported a 9-year history of difficulty in swallowing, painful swallowing, foreign
body sensation in the throat, change of voice, and constant left-sided neck pain radiating to the jaw and postauricular area and increasing with chewing or neck rotation. In the previous 9 years, he had sought medical advice in many hospitals and had been given antibiotics and analgesics, and he had his left wisdom tooth removed, but without any significant improvement.

He denied any history of trauma, recent upper respiratory tract infection, headache, otorrhea, tinnitus, vertigo, hearing loss, or any weakness in other parts of the body. There was no previous ear surgery.

On clinical examination, the patient was seen to have left facial nerve palsy Grade 4 on the House–Brackmann scale. Other cranial nerves examinations were normal. Tympanic membranes examination and nasal and throat examinations were normal. There were no palpable lymph nodes or masses.

The patient was started on a 3-week course of prednisolone 40 mg. The facial palsy improved from Grade 4 to Grade 2 on the House–Brackmann scale over several months. However, the patient still complained of left-sided neck pain, jaw pain, dysphagia, odynophagia, and foreign body sensation.

Brain imaging was normal. However, magnetic resonance imaging showed pathological enhancement of the descending segments of the facial nerve involving the mastoid segment. Computed tomography (CT) scan revealed bilateral elongation of the styloid processes (7.5 cm) which appeared to join the hyoid bone [Figures 1-3]. There was no evidence of vascular compression, and the visualized part of the internal carotid arteries was patent.

Diagnosis of ES was made, and the patient was started on medical treatment in the form of nonsteroidal anti-inflammatory drugs (NSAIDs) and acetaminophen, but there was no significant improvement. A decision was taken for surgical management, but the patient refused to have surgery.

Discussion

Facial palsy as a presentation of ES is extremely rare and was never heard of until Rosales et al. made the 1st case report of a 40-year-old male who presented at the emergency department with facial palsy and symptoms of ES. There are no previous reported cases in Saudi Arabia on facial palsy as a presentation of ES. Facial palsy in our patient can be explained by local compression and irritation to the facial nerve distal to the stylomastoid foramen which presented as facial muscle weakness but without a disturbance of taste, dryness of mouth, or hyperacusis which normally results when the damage of facial nerve is at a higher level.
The incidence of ES differs slightly from one study to another. In a study, Correll examined 1771 radiographs on panoramic views and concluded that the incidence of elongated styloid process is 18.2%. However, only 8 out of 1771 (0.4%) had symptoms of elongated styloid process, which is called ES.[5]

ES can be easily misdiagnosed because of the wide variety of symptoms involving the craniofacial and cervical region which can often be mistaken for glossopharyngeal and sphenopalatine neuralgia, migraine headaches, temporal arteritis, myofascial pain dysfunction syndrome, impacted molar teeth, faulty dental prostheses, and temporomandibular joint disorder causing the patient to undergo many needless procedures or be prescribed unnecessary medications.[6,7]

To diagnose ES, we depend mainly on 3 aspects:
1. The clinical presentation through careful history taking and physical examination including digital palpation of styloid process in the inferior aspect of tonsillar fossa
2. Radiological findings
3. Lidocaine infiltration test in the inferior aspect of tonsillar fossa. If the symptoms resolve with lidocaine injections, it is most likely to be ES.[5]

According to Chandramani B. More and Mukesh Khemchand Asrani,[8] the best imaging modality which shows the complete details of length, angulation, and relation to adjacent structures is CT scan with a three-dimensional reconstruction. However, several other imaging modalities have been used such as panoramic radiograph, lateral head and neck radiographs, Towne’s radiographs, lateral oblique radiograph of the mandible, anteroposterior head radiograph, and CT.

The management is determined on the severity of the case. The first line of management which is conservative includes analgesics, NSAIDs, and injection of steroid into the inferior aspect of the tonsillar fossa. However, long-term results have not been satisfactory. If the conservative management fails, the definitive treatment is surgical.[5] The procedure in which the styloid process is shortened may be done intraorally or with an external approach.[5,9] The advantages and disadvantages of each approach are mentioned in Table 1.

### Table 1: The Pros and Cons for two surgical approaches for Eagle’s Syndrome

|Pros| Transoral approach| External approach|
|---|---|---|
|No visible scar| Optimal visualization|
|Minimal risk of developing deep cervical infection| Visible postoperative scar|

### Conclusion

One cannot picture what the mind does not comprehend. It is important for otolaryngologists, dentists, and other specialists who deal with head and neck problems to be able to recognize ES even though the condition is rare. Although the patient responded to a treatment similar to that of Bell’s palsy, based on the clinical features and imaging, ES was most likely to have been the cause of his facial palsy.

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### Conflicts of interest
There are no conflicts of interest.

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