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Eagle's syndrome: Review of cases among Iraqi population

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ABSTRACT

Background: Styloid process is sparse bony protrusion situated between the external and internal carotid arteries just proximal to the internal jugular vein and facial nerve. Styloid process varies in length from absent to elongated process and styloid process longer than 25 mm is considered as elongated. Elongation of styloid process and calcification of stylohyoid ligament may cause Eagle's syndrome which can be diagnosed by high resolution computerized tomography. The most cases of Eagle's syndrome are idiopathic cause; however, the first case of Eagle's syndrome was diagnosed by Dr. Watt Weems Eagle as post tonsillectomy complication.

Aim of study: to detect the incidence of causes of Eagle's syndrome in the Iraqi society.

Material and Method: Thirty four patients with craniofacial pain, cervical pain, chronic dysphagia, and sensation of foreign body in the throat of both sexes aging between (10-58) years are diagnosed as Eagle's syndrome by high resolution computerized tomography.

Result: Five patients (15%) had undergone tonsillectomy, three patients (9%) had severe head injury and two patients (6%) had difficult prolonged dental extraction as causes of Eagle's syndrome, while the cause of Eagle's syndrome in the rest of patients is unknown (idiopathic).

Conclusion: Eagle's syndrome is a very rare and oftenly missed complication of tonsillectomy, severe head trauma and prolonged dental extraction that should be kept in mind.

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Introduction:

The styloid process is a sparse bony protrusion situated at the undersurface of the temporal bone, immediately posterior to the mastoid apex. [1] It is related to the vertebropharyngeal recess which contains the carotid arteries, and internal jugular vein.[2] Beside carotid arteries and jugular vein, the styloid process is in close relationship with five cranial nerves, namely the facial, glossopharyngeal, vagus, spinal accessory, and the hypoglossal nerves.[3] The normal length of the styloid process ranges from 20 – 25 mm and the styloid process is considered elongated if it measures more than 25 mm.[1,2]

The styloid process is cartilaginous in the first two years of life, but after the age of two years the cartilage of the styloid process starts to ossify and the cartilaginous band connecting the styloid process and the hyoid bone reabsorbs and is replaced by fibrous band (stylohyoid ligament). [1,2] In some individuals, ossification of the stylohyoid cartilage occur instead of resorption leading to ossified stylohyoid ligament and Eagle's syndrome. [4] The exact cause of elongation of styloid process is unclear, however some theories have been postulated to explain this phenomenon like:

1. Persistence deposition of cartilaginous analog in stylohyoid apparatus (congenital theory).
2. Idiopathic calcification of stylohyoid ligament.
3. Osseous growth in the vicinity of stylohyoid ligament insertion. [5]

Elongation of styloid process is due to either fracture of the process and its length is measured from the base to its tip with the fracture gap,[6] or due to increased length because of deposition of macromolecules made of fatty acid and protein with subsequent calcification.[7] Abnormal calcification of soft tissue occurs due to abnormality in metabolism of calcium, phosphrus or vitamin D and this can be seen in:

1. Parathyroid gland
2. Systemic lupus erythematosus
3. Scleroderma
4. Trauma
5. Dermatomyositis.[8]

Clinical features:

The clinical features of Eagle's syndrome are variable and confirming a clear relationship between the clinical symptoms and stylohyoid apparatus may be quite difficult. In classical cases of Eagle's syndrome, pain usually develops after tonsillectomy, because of distortion of local anatomy of the area. However, it can be encountered in patients who have not undergone head and neck surgery. The syndrome can be grouped into two main categories [9]

- ❖ Cranial nerves compression.
- ❖ Carotid artery compression.

Patients with cranial nerves impingement will suffer from symptoms due to irritation of trigeminal, facial, glossopharyngeal and vagus nerves. Palpation of the tip of styloid process will provoke the symptoms which include pain on extending tongue, facial pain while turning the head, foreign body sensation, dysphagia, sensation of

hypersalivation, change in voice, tinnitus or otalgia.[10] While arterial impingement will produce vascular/ischaemic symptoms with pain along the involved artery thought to be mediated by the sympathetic plexus. These symptoms are either due mechanical compression (syncope and visual symptoms) or due to sympathetic plexus irritation causing parietal pain or eye pain[11]

Patients and method:

A prospective study involving 34 patients aged 10 – 58 years is conducted during 5 years period from Jan, 1st 2013 till Dec, 31st 2018. Patients suffering from craniofacial pain, cervical pain, chronic dysphagia, and sensation of foreign body in the throat are carefully managed to confirm the diagnosis of Eagle's syndrome. Detailed medical history paying special attention to past surgical history including previous dental interventions and head trauma (even minor trauma) is sought. Meticulous otolaryngological examination is performed with referral to neurologist, rheumatologist, general surgeon and maxillo-facial surgeon to exclude other causes of headache and facial pain. All patients are sent for complete blood picture and biochemical analysis including liver function test, renal function test, calcium, vitamin D and phosphrus serum levels to exclude systemic disorder. Other investigations like paranasal sinuses radiology and cervical spine imaging are sent accordingly.

High resolution computerized tomography is done to detect styloid apparatus abnormalities (long styloid process more than 25 mm, fractured styloid process or calcification of stylohyoid ligament).

Result:

- Thirty four patients are diagnosed as Eagle's syndrome, 25 patients (74%) are females and 9 patients (26%) are males.
- Elongated styloid process is seen in 7 patients (21%), on the other hand 79% of patients (27 patients) have fractured styloid process.
- Eagle's syndrome is more common in young age group (65% of patients are between 21–40 year old group) (Table 1).
- Only 3 patients (9%) complained of bilateral symptoms of Eagle's syndrome while 31 patients (91%) have unilateral disease, however CT scan findings show bilateral styloid apparatus abnormalities in 44% of cases (15 patients).
- Five patients (15%) have fractured styloid process due to previous tonsillectomy, 4 of them are males (80%) and one female patient (20%) (Tables 2&3).
- Of the five Post tonsillectomy Eagle's syndrome, one patient is less than 10 years old, one patient is 10-20 years age group and 3 patients are 20-30 years age group (Table 3).
- Two patients (6%) have fractured styloid process due to previous dental extraction of lower wisdom tooth which was described by patients as difficult prolonged extraction.

❖ Three patients (9%) had history of severe head injury including fracture mandible or skull fractures.

Table 1: Age distribution of Eagle's syndrome

Age (Years)	Number of cases	Percentage
Less than 10 y	3	8.8%
11 – 20 y	4	11.8%
21 – 30 y	10	29.4%
31 – 40 y	12	35.3%
41 – 50 y	4	11.8%
50 – 60 y	1	2.9%
Total	34	100%

Table 2: Causes of Eagle's syndrome

Cause	Frequency	Percentage
Post tonsillectomy	5	14.7%
Head trauma	3	8.8%
Post dental intervention	2	5.9%
Idiopathic	24	70.6%
Total	34	100%

Table 3: Age and sex of Post tonsillectomy Eagle's syndrome patients

Patient	Age at Surgery	Sex
Patient 1	19 years	Male
Patient 2	27 years	Female
Patient 3	21 years	Male
Patient 4	8 years	Male
Patient 5	24 years	Male

Table 4: Symptoms of Eagle's syndrome patients in this study

Symptom	Number of cases	Percentage
Dysphagia	27	79.4%
Cervical pain	25	73.5%
Headache	11	32.4%
Facial pain	10	29.4%
Tinnitus or otalgia	4	11.8%
F.B. sensation	3	8.8%
Syncope	1	2.9%

Discussion:

Eagle's syndrome is combination of symptoms caused by elongated styloid process, calcified stylohyoid ligament or fractured styloid process which can be unilateral or bilateral. The most common symptoms are dysphagia, foreign body sensation in the throat, headache, craniofacial or cervical pain. [12] In our study, most patients complains of throat pain (80% - 27 patients) and cervical pain (74% - 25 patients) which are often misdiagnosed as chronic pharyngitis, inflammation of tonsillar remnant or cervical spine problems. On the other hand only 3 patients (9%) have foreign body sensation which is misdiagnosed as psychological problems.

Our study shows that male patients constitute 26% of patients and this is the same result of Karam and Koussa [13] from Lebanon who stated that Male: Female ratio is 3:1, and Cawich et al [14] showed that 80% of Eagle's syndrome are males in Jamaican population, thus imposing racial difference in incidence of Eagle's syndrome. However, different result was found by More [15] who compared gender distribution in different articles and concluded that there is female gender predominance.

Simone Moria [16] found that (41-50) year age group is the most common affected group while our study showed that Eagle's syndrome is more common among (31-40) year age group followed by (21-30) year age group; while Eagle [17] stated that his two cases were 26 and 28 years. This implies that Eagle's syndrome is a disease of young and middle age group, although it can occur at any age. [18] Our study showed that only 3 patients (9%) have bilateral symptoms of Eagle's syndrome while CT scan findings showed bilateral styloid apparatus abnormalities in 15 patients (44%), a similar finding was found by Karam and Koussa [13] and by Ryan [18] who stated that elongation of styloid process is bilateral and symptoms of Eagle's syndrome is unilateral and the cause behind this phenomenon is unknown.

Casale M, et al, [19] stated that 4% of population has elongated styloid process and only 4% of this group develops Eagle's syndrome, this is mainly because the pathophysiology of development of Eagle's syndrome depends on some sort of inflammatory and degenerative mechanisms (although the exact cause is unknown), and this is supported by most literatures [20, 21] which suggest that inflammatory and degenerative mechanisms are more important than trauma as a cause of Eagle's syndrome. Our study shows that fractured styloid process is seen in about 80% of Eagle's syndrome, supporting that is trauma is more important in pathogenesis of Eagle's syndrome which is the opposite of most results.

Five patients (15%) had undergone previous tonsillectomy and all of them has fractured styloid process. Two of them stated that their complaint started within the first postoperative day while the other three patient had their started in the first postoperative week, supporting that tonsillectomy may be the cause of Eagle's syndrome. Four of

post tonsillectomy patients are males suggesting that the stronger musculature of male may play a role in fracturing the styloid process. The same suggestion may be supported by the fact that four of these patients are aged 19 – 27 at time of surgery. The possible cause of fracturing styloid process is the strong muscle pull exerted on the process during surgery due to hyperextension of neck; therefore otolaryngologist should avoid vigorous neck extension during tonsillectomy. [22]

Three patients (9%) had previous severe head trauma and fractured styloid process raises the suspicion that styloid process injury is often overlooked in patients with serious head trauma. [22] Tow patients had undergone prolonged difficult extraction of wisdom tooth and started to complain of neck pain within the first postoperative week and CT scan showed fractured styloid process in both patients. The cause of fractured styloid process is the unsynchronized contraction of the muscles opening and closing the mandible due to repeated mouth closure during tooth extraction. [23]

The main objective of this study is to estimate the frequency of causes Eagle's syndrome in Iraqi population and elucidate the importance of styloid process, because medical practitioners do learn about it , but they do not apply this information into their clinical practice.[24] Finally in conclusion, otolaryngologists must pay attention to avoid vigorous neck extension during tonsillectomy to avoid fracturing styloid process, emergency room physician must keep Eagle's syndrome in mind and seek specifically for styloid process fractures in CT scan and dentist must avoid extraction of complicated wisdom tooth and switch to surgical extraction.

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