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CASE REPORT

Eagle's Syndrome: An Unusual cause of throat pain-Case Study

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INTRODUCTION

Eagle's syndrome or styalgia is characterised by the anatomical bone deformity of the styloid process, as identified by Wat W. Eagle in 1949[1]. An aggregate of symptoms caused by the elongation and severe calcification of the temporal bone's styloid process, as well as calcification of the ligaments attached to this process, including the styloid and stylomandibular ligaments, have been identified[2]. The styloid process is typically 20 to 30 mm in length, and patients with elongated styloid processes of 40 mm or longer have clinical distress [1,3] . This styloid process elongation could be unilateral or bilateral [4]. It affects around 4% of the general population, with 3:1 female predominance, however only 4% of these people experience associated symptoms [3]. Eagle mainly characterized two syndromes: "classic styloid syndrome" following tonsillectomy, and "stylocarotid syndrome" unrelated to tonsillectomy[6]. The "styloidogenic jugular compression syndrome", a third variation of Eagle syndrome, was recently identified, in which the jugular vein is squeezed, resulting in cerebral venous hypertension and, most commonly, headaches[1].

Cervicofacial pain, otalgia, foreign body sensation in the throat, pain when changing head position, cervical pain, and pain when swallowing have all been linked to an elongated styloid process. These symptoms emerge as a result of an abnormal styloid process irritating and/or compressing neighbouring neurovascular and muscle systems[5]. Eagle's syndrome is often diagnosed through a physical examination that includes palpation of the extended styloid process and radiography, including routine orthopantamograph or CT scan, which is now considered the "gold standard"[4].

We present the case of a 51-year-old woman who presented with left neuralgic pain in the throat, which radiated into the ear, and dysphagia for about a year.

CASE REPORT

A 51-year old married female presented to Shalamar Hospital ENT outpatient department with complaints of throat pain since one year and difficulty in swallowing for eight months. Patient was in usual state of health one year back when she developed episodes of left sided throat pain. This pain was sudden in onset and persistent in nature. It was radiating towards the left ear. It got aggravated by swallowing and left side neck movement. It was relieved by taking pain killers. There was no associated fever or malaise. She started having difficulty in swallowing eight months ago. This was mainly due to pain as she had no dysphagia. She had multiple outpatient consultations in different hospitals due to these complaints. She was prescribed repeated courses of antibiotics and pain killers.

She was a known case of hypertension since two years with poor compliance and newly diagnosed case of Diabetes at the time of consultation. She had a past surgical history of hysterectomy three years ago. Her family history was unremarkable and she was using over the counter painkillers.

On examination, the oral cavity and oropharyngeal mucosa were normal looking. Tonsils and tonsillar pillar area were also unremarkable. Patient pin pointed the location of pain, and upon deep palpation of left tonsillar fossa, it was found to be tender. Ear and Nose examination was unremarkable. Neck examination was normal.

On the basis of her presenting symptoms, several differential diagnosis were made including pharyngitis, tonsillitis, peritonsillar abscess, infectious Mononucleosis, retropharyngeal abscess, and diphtheria. CT scan head and neck was obtained to make the final diagnosis which revealed left more than right elongated styloid process along with focal non-contiguous bilateral stylohyoid ligaments ossification could be the cause of patient's symptomatology. Her elongated left styloid process measured 40mm and right styloid process measured 26mm. The complete blood picture, liver function tests, renal function tests, and coagulation profile were normal. The viral hepatitis markers as well as the COVID-19 PCR were negative, and her BSR (Blood sugar random) was in normal range.

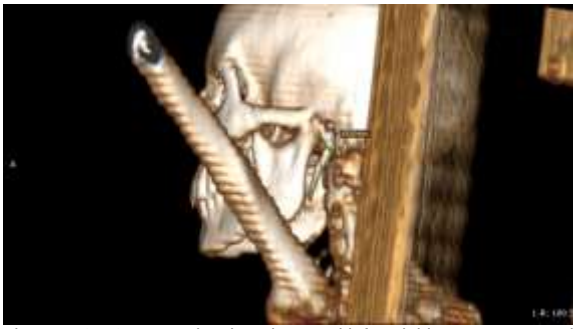

Figure.1: 3D CT scan showing elongated left styloid process measuring 42.0 mm.

On the basis of history, examination and radiological investigations, a final diagnosis of Eagle's syndrome and left tonsillitis was made. She was scheduled for left tonsillectomy and left styloidectomy. No medication was advised in TTR.

All baseline investigations were carried out as a part of anaesthesia evaluation, including, S/Electrolytes, Blood Grouping, ECG, and X Ray Chest. Patient had complete COVID-19 vaccination. Under aseptic measures, left sided tonsillectomy was done by harmonic method. Endoscopic transoral approach was used. After left stylohyoid ligament was dissected and styloid process was removed. Haemostasis was secured and stitches were applied. Patient was then handed over to anaesthesia team vitally stable.

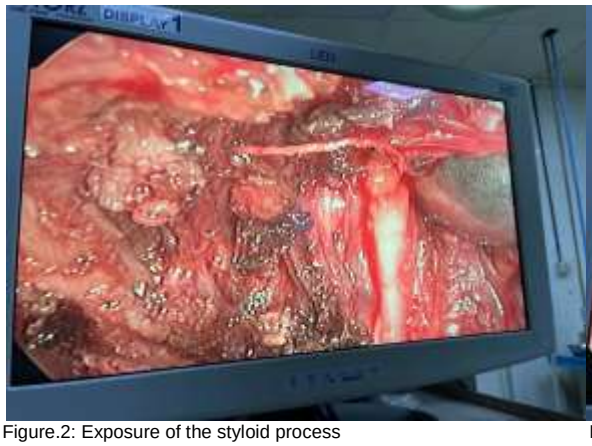

Figure.2: Exposure of the styloid process

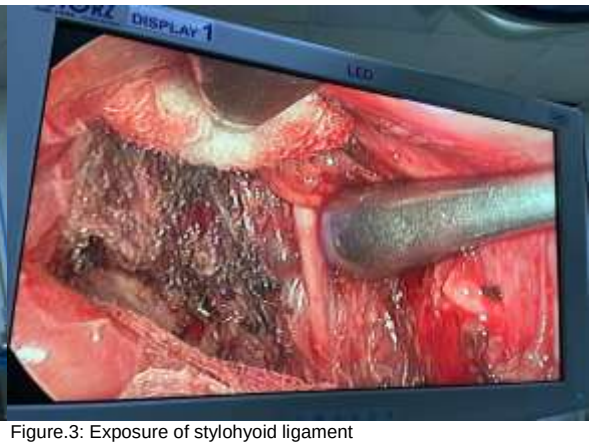

Figure.3: Exposure of stylohyoid ligament

Figure.4: Removed styloid process

Figure.5: Post Tonsillectomy

She was reported vitally stable with no active complaint on zero post-operative day, but complained of mild pain on operative site post-op Day 1, for which she was given analgesics as per TTR. The patient was then discharged as per protocol. A post-op follow-up of the patient was done after 2 weeks, which revealed a smooth recovery with no complications.

CASE DISCUSSION

When idiopathic unilateral pain occurs, especially in adult women, and the pain is not responsive to painkillers, Eagle's syndrome should always be suspected.[1]Eagle's syndrome is often misdiagnosed due to its rarity and non-specific symptoms, and delay to diagnosis has been reported to be between 10 and 27 years.[1,3,4] In our case report, it took almost an year for the patient to be diagnosed correctly. Similarly, it has been reported that despite that the patient having knowledge of Eagle's syndrome, it took 4 years to reach the correct diagnosis due to the unusual presentation of his symptoms.[3] The lack of a clear aetiological link and the lack of knowledge about this clinical entity contribute to misdiagnosis.[1] Patients with severe calcification of the SP have also been reported to be asymptomatic.[iii] Many times, the ambiguity of the presentation and the severity of the pain mislead people, resulting in the patient being evaluated by multiple consultants, including a neurosurgeon, an ear, nose, and throat surgeon, and, in certain cases, a psychiatrist.[4]

A detailed clinical history and a physical examination by digital palpation of the styloid process in the tonsillar fossa for localizing the source of pain, can be greatly helpful in diagnosis[1]

In such cases, a multidisciplinary approach is recommended, as is early referral to radiological investigation, particularly CT of the styloid process. CT has been recommended as the best imaging modality for assessing the styloid processes' location, length, thickness, and curvatures, along with the adjacent neuro-vascular structures.[2,4]

Eagle's syndrome treatment options include non-invasive treatment, as well as surgery, depending on the patient's symptoms.[2] Non-invasive treatment options include analgesic/opioid medications, steroid injections, non-steroidal anti-inflammatory agents, oral antidepressants, anticonvulsants, and local anesthetic agents.[3,5] Surgical styloidectomy has been reported to have an 80% cure rate.[1]Eagle first described an easy, quick, and less time-consuming intraoral or transoral approach to the styloid process[1,3] However, the final choice to proceed with surgery is contentious, and there is no prevailing opinion in the favour of a surgical approach, and the patient must be managed according to the presenting symptoms[5] The symptoms of our patient had not been alleviated despite administration of painkillers, as a result, we chose surgical treatment.

CONCLUSIONS

To date, there are limited studies on Eagle's syndrome, especially in Asian countries like Pakistan. This rare clinical entity, which is not frequently suspected in clinical settings but associated with a critical decline in the quality of life, should be made very clear to otolaryngologists, neurologists, and dental surgeons worldwide, particularly in Asian countries like Pakistan. Eagle's syndrome should always be included in the differential diagnosis of cervicofacial pain patients. To ensure successful identification and management of such patients, due importance should be given to the diagnostic tools including history and clinical examination, appropriate imaging techniques, and definitive treatment options. And awareness about these tools should be given to the concerned health specialists.

Recommendations: We recommend more studies to be conducted in this area in the light of the gaps identified, particularly in Asian countries. There is a need to understand the perception of both, the health consultants as well as the patients, about this rare entity.

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