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International Journal of Current Research
Vol. 10, Issue, 01, pp.64186-64189, January, 2018

INTERNATIONAL JOURNAL OF CURRENT RESEARCH

ISSN: 0975-833X

CASE STUDY

ASYMPTOMATIC EAGLE'S SYNDROME – A RARE CASE REPORT

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ARTICLE INFO

Article History:
Received 18th October, 2017
Received in revised form
26th November, 2017
Accepted 21st December, 2017
Published online 19th January, 2018

Key words:
Styloid process,
Dysphagia,
Facial pain.

ABSTRACT

Eagle's syndrome is a condition where stylohyoid ligament gets calcified causing an elongation of the styloid process. In 1937, W. W. Eagle first summarised this disorder describing clinical findings related to an elongated styloid process. Review of literature suggests 4% of the adult population shows an elongated styloid process with 0.16% being symptomatic. It may develop inflammatory changes or impinge on the adjacent arteries or sensory nerve endings causing pain in the cervical region upon head movement. Other subjective symptoms include dysphagia, pharyngeal foreign body sensation and headache. It can be diagnosed with careful clinical evaluation and confirmed with radiographs showing an elongated styloid process. Severity of subjective symptoms dictates the need for surgical excision of the styloid process otherwise reassurance is the primary mode of management. Here, we report a rare case of elongated styloid processes bilaterally in a 52-year-old male.

Citation: Dr. Amit Kumar Roy, Dr. Santosh T – Reader, Dr. Sanjeet Kumar Das, Prof. (Dr.) Sk Abdul Mahmud and Dr. Neha Shah – Reader. 2018. "Asymptomatic eagle's syndrome – A rare case report", *International Journal of Current Research*, 10, (01), 64186-64189.

INTRODUCTION

An American otolaryngologist Watt Weems Eagle in 1937 defined a syndrome having an elongated styloid process related to ossification of the stylohyoid ligament. It was termed as Eagle syndrome (Eagle, 1937). It is also known by the name Ernest syndrome (Frommer, 1974). Pietro Marchetti first reported a similar disorder way back in 1652 (Eagle, 1937). It is a rare clinical entity that may be the cause of craniofacial and cervical pain (Ferreira, 2003). The styloid process is a small bony projection of temporal bone, which lies anterior to stylomastoid foramen. The normal styloid process is 25-30 mm long (Eagle, 1949). The dystrophic and degenerative changes in the hyoid complex of the styloid process is the cause of Eagle's syndrome (Baugh, 1993) Eagle described two types (Eagle, 1937) of clinical presentation:

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- Classical stylohyoid syndrome frequently follows tonsillectomy and is due to fibrous tissue causing distortion of nerve endings of cranial nerves (Vth, VIIth, IXth & Xth) in the tonsillar bed. There is sensation of foreign body in throat and pain on deglutition.
- Stylocarotid syndrome is not correlated to tonsillectomy and is due to compression of sympathetic chain in carotid sheath by the ossified ligament or tip of the styloid process, resulting in an incessant, radiating pain in the carotid zone.

It is usually seen in adult patients aged between 30 to 50 years (Holloway, 1991). No statistically significant association between age and elongation-calcification patterns has been reported (Balcioglu, 2009). There is no significant sexual dimorphism but according to some authors symptoms are more common in females (Balcioglu, 2009). The Symptoms range from as simple as perception of foreign body in the throat to as severe as stroke.

Other signs and symptoms include pain in the TMJ or preauricular region, cervical pain, headaches, tinnitus, restricted or asymmetric mandibular movements, vertigo/ dizziness, feeling of ear fullness and odynophagia (De Souza Carvalho, 2009 and Chuang, 2007). The diagnostic workup includes a detailed medical history and a thorough head-neck examination to rule out other diagnoses. Manual manipulation should be used as a guideline for producing symptoms similar to the subjective ones by palpating over the stylohyoid complex cautiously. Palpation of the tonsillar fossa will reveal the tip of the styloid process as a hard bony spicule that aggravates symptoms (Candice, 2011). If intraoral palpation fails to elicit any bony projection, transpharyngeal palpation can be used. The diagnosis of Eagle's syndrome can be established clinically if the classical signs are present. In case of misleading clinical presentations, radiograpghic imaging is of utmost importance. Radiographic evaluation is the final step to confirm diagnosis (Candice, 2011). The mode of treatment of Eagle's syndrome can be either medical or surgical management depending upon the severity of symptoms. Surgical correction includes excision of the styloid process and the ossified ligaments under general anesthesia. Medical management includes administration of steroid injection into the scar tissue and drugs to decrease muscle spasms (Palesy, 2000). Here, we report a rare case of the bilaterally elongated stylohyoid ligaments with no classical symptoms in a 52-year-old male.

Case Report

A 52 year old male patient reported to the Department of Oral & Maxillofacial Pathology with the chief complaint of burning sensation in bilateral buccal mucosa since last 3 months with previous medical history of tonsillectomy 15 years ago. Intra orally, palpating along the occlusal line posterior to the region of the tonsillar fossa a hard bony tip on the right side could be palpated (Fig.1.

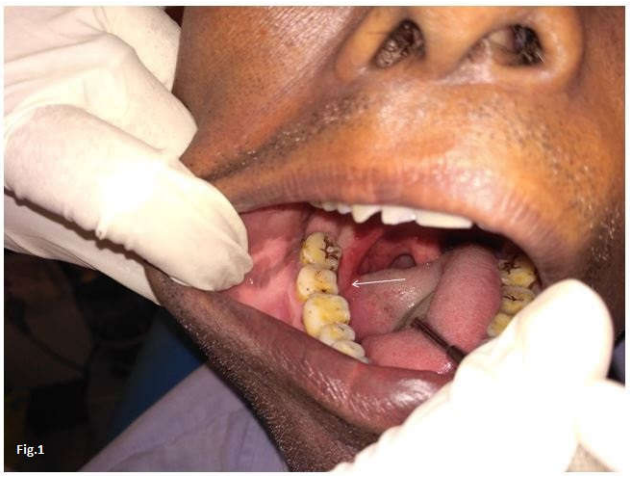

Fig.1. Intraoral clinical photograph showing the bony projection in the right tonsillar fossa

The contralateral side also revealed a hard bony tip in the left tonsillar fossa region but did not produce any pain on palpation. Orthopantomogram was advised for radiological evaluation (Fig. 2). Radiographic examination demonstrated elongated styloid processes measuring ~50mm on the right side (Fig. 3a) while ~47mm on the left side (Fig. 3b). On extra oral examination, no bony hard projection or tenderness on palpation in both right and left submandibular area at anterior border of sternocleidomastoid muscle could be appreciated.

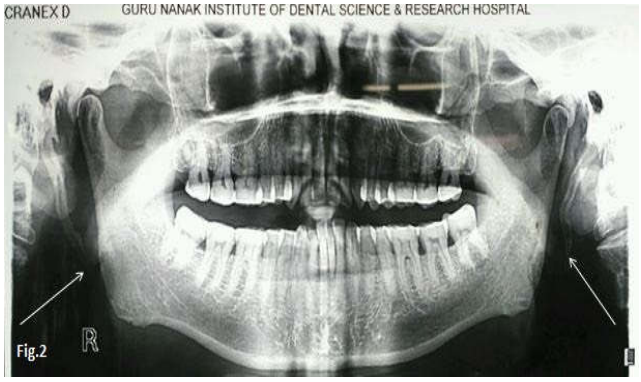

Fig. 2. OPG showing bilateral elongated styloid processes

The diagnosis of Eagle's syndrome was made based on the history of previous tonsillectomy with corroborative clinical findings, while confirmation was made with radiograph showing bilateral elongated styloid process. As the patient was asymptomatic, only palliative treatment for the burning sensation was prescribed and the patient was made aware of the consequences of the elongated styloid process. Reassurance was given to the patient and he was advised for close periodic follow ups. Subsequently, the patient was re-examined along with radiographic evaluation and no significant changes could be seen after a period of 6 months.

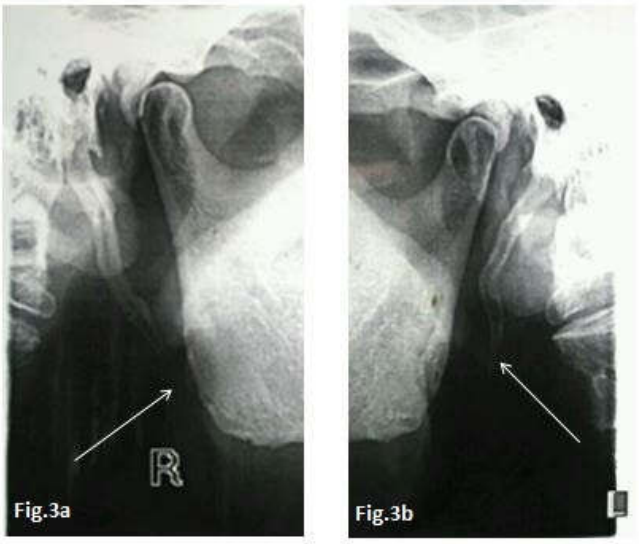

Fig.3a & 3b. OPG showing elongated styloid processes on both sides of Langlais type III morphological classification and type A radiological classification

DISCUSSION

In the 17th century, Pietro Marchetti (1652) first observed and recorded an elongation of the styloid process. In 1937, Watt W. Eagle first described the elongation of the styloid process which was later called the Eagle's syndrome (Eagle, 1937). Eagle's syndrome is defined as the symptomatic elongation of the styloid process or mineralization (ossification or calcification) of the stylohyoid ligament complex (Eagle, 1937). Several theories (Balbuena, 1997) about the cause of the elongation have been proposed:

- Congenital elongation of the styloid process due to persistence of a cartilaginous analog of the stylohyal (one of the embryologic precursors of the styloid)
- Calcification of the stylohyoid ligament by an unknown process

- Growth of osseous tissue at the insertion of the stylohyoid ligament.

In a study of over 200 cases, Eagle *et al* reported the normal styloid process to be in between 25 to 30 milimeters in length, beyond which can be termed as Eagle's syndrome (Eagle, 1937). The location of the tip is particularly important, which is situated between the internal and external carotid arteries, laterally from the pharyngeal wall and immediately behind the tonsillar fossa. The signs and symptoms of Eagle's syndrome are due to pressure of the elongated styloid process on the neurovascular structures situated around the styloid process like facial nerve, internal or external carotid arteries (Breault, 1989). Depending on the position of styloid process in relation to underlying anatomical structures, it could be symptomatic or asymptomatic. The symptoms vary such as pain in the TMJ or preauricular region, cervical pain, headache, tinnitus, restricted or asymmetric mandibular movements, vertigo/dizziness, feeling of ear fullness, odynophagia and foreign body sensation in the throat (De Souza Carvalho, 2009). A slight medial deviation of the styloid process, could result in severe symptoms of atypical facial pain (Breault, 1989), whereas the compression of the carotid artery by the tip of the styloid process can even lead to stroke (Bagga, 2012). However the above mentioned clinical features are not pathognomonic for Eagles syndrome because many patients with incidental findings of elongated styloid process are asymptomatic as was in our case. Patients can be categorized (Bagga, 2012) into two groups on the basis of their symptoms:

- Those who have classical symptoms of a "foreign body" lodged in the throat with a palpable mass in the tonsillar region following tonsillectomy
- Those with pain in the neck following the course of carotid artery distribution (carotid artery syndrome).

The age of presentation is between 30 to 50 years (Mortellaro, 2002). In our case the age of the patient was corroborative. There is no significant sex predilection however, symptoms are more common in females by the ratio of 3:1(F:M) (Balcioglu, 2009). In our case the patient is a male.

On the basis of radiographic findings, it is morphologically classified (Langlais, 1986) as:

Type I pattern represents an uninterrupted, elongated styloid process.

Type II is characterized by the styloid process apparently being joined to the stylohyoid ligament by a single pseudoarticulation. This gives the appearance of an articulated elongated styloid process and is the type present in our patient.

Type III consists of interrupted segments of the mineralized ligament, creating the appearance of multiple pseudoarticulations within the ligament.

Sometimes heterotrophic calcification such as abnormal calcium & phosphorus metabolism, chronic renal failure, rheumatoid diseases and some endocrinological disorders (Mortellaro, 2002), may lead into abnormal calcification of styloid process. It is classified radiologically[15] as:

Type A: Calcified outline of styloid process

Type B: Partially calcified styloid process with discontinuous radiolucent core

Type C: Nodular appearance of styloid process with varying degrees of central radiolucency

Type D: Completely calcified styloid process with no evidence of a radiolucent interior structure.

Our case falls under the type III Langlais morphological classification (Fig.3a & 3b) and type A Langlais radiological classification (Fig.3a & 3b) respectively, with little lateral deviation of the tip on either side. It can be attributed to the age of the patient which is 52 years and the changes could be a result of the normal process of aging. No significant changes could be observed during a periodic re-evaluation over a span of 6 months. This finding contributed to the fact that the elongated styloid process is not stretching the regional cranial nerves and showed no signs of fibrosis in the post-tonsillectomy period.

Conclusion

Accidental findings of elongated styloid process in routine examination has to be dealt with a detailed case history recording and thorough physical examination of the head and neck region.

Patients are to be made aware of the consequences of elongated styloid process and should be encouraged for the regular periodic evaluations. From the clinical and radiological evaluations, if the elongated styloid process is not disturbing any of the vital integrity, then it can be considered asymptomatic. Since it might be due to age related changes further studies are to be carried out for evaluating any significant changes in the styloid process and associated development of any symptoms.

Acknowledgements

The authors gratefully acknowledge the contributions made by Professor (Dr) RR Paul, Deputy Director cum-in-charge, Research and Development, GNIDSR, Kolkata, Professor (Dr) M Pal, HOD, Department of Oral and Maxillofacial Pathology, GNIDSR, Kolkata for their valuable guidance and help in every aspect.

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