

# The Eagle Syndrome : A Clinical Dilemma - Case Report and Review of literature

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**ABSTRACT** Elongated styloid process causing craniofacial symptoms is called as Eagle syndrome. We report a 45-year-old female with this syndrome. Clinically the elongated styloid process was mistaken for submandibular calculus. The diagnosis was made by computed tomography. Styloidectomy relieved her symptoms.

**Keywords:** Eagle syndrome

**Abbreviations:** USG - ultrasonography; CT - computerized tomography; ESP - Elongated styloid process

## Introduction

Eagle syndrome is a clinical condition characterized by craniofacial and cervical symptoms caused by elongated styloid process (ESP) or calcification of the stylo-hyoid ligament. It was first described by Eagle in 1937. According to him elongated styloid process is present in approximately 4% of the population.[1,2] However, other investigators have reported an incidence rate varying from 1.4 to 18.2 %.[3,4] Most of the patients with ESP are asymptomatic and only a small percentage of them (4 to 10%) actually become symptomatic.[5] Eagle syndrome is diagnosed only if the patient is clinically symptomatic and not merely by the presence of ESP. Patients may present with wide range of symptoms such as ear ache, sore throat, vague foreign body sensation in pharynx, hyper-salivation, painful movement of the neck and tongue,odynophagia and headache. These symptoms are due to pressure effect of ESP on adjacent soft tissues. In the absence of appropriate imaging technique clinical diagnosis is often missed due to the vague nature of symptoms.

## Case Report

A 45-year-old female presented with 3 months' history

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of pain on the right submandibular region. Neck movements were also painful. USG examination of the neck was unremarkable. Sialolithiasis of right submandibular gland was suspected clinically. Non-contrast spiral CT scan of the neck with three-dimensional reconstruction was done using 64-slice multidetector computed tomography scanner (Somatom Definition AS-Siemens). Coronal, sagittal images as well as thick multiplanar reconstruction image and volumetric rendering technique images were obtained. These images revealed dense ossification of the right styloid process and stylohyoid ligament. The grossly thickened styloid process was extending up to the hyoid bone (Fig. 1, 2). The right styloid process was measuring 7.4 cm whereas its left counterpart was normal in length (2.6 cm). The maximum thickness of the right styloid process was 1.2 cm. Bilateral submandibular salivary glands and other soft tissue structures of neck were normal. In addition there was presence of anterior bridging osteophytes and calcification of anterior longitudinal ligament of the cervical spine involving C3 to C7 cervical vertebrae. (Fig. 3) She was relieved of her symptoms after undergoing surgical removal of the right ESP.



Fig 1: Plain computed tomography showing elongated and thickened right styloid process (arrows) in coronal (A) and axial (B) sections.

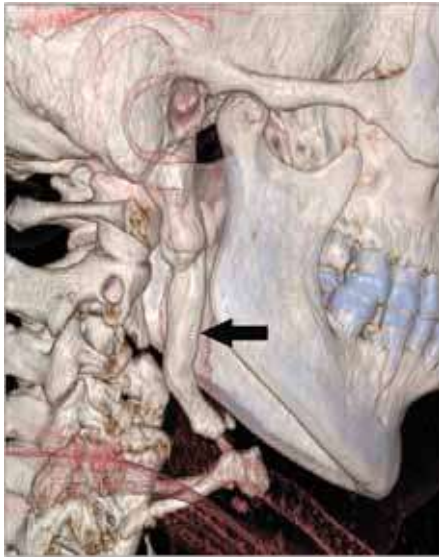


Fig 2: Volumetric rendered technique image showing elongated styloid process on the right side (arrow)



Fig 3: Mid-sagittal CT images of the same patient showing anterior bridging osteophytes and calcification of anterior longitudinal ligament of the cervical spine involving C3 to C7 vertebrae

## Discussion

Styloid process is a conical bone which arises from the mastoid portion of temporal bone just anterior to stylomastoid foramen and then courses downwards, forwards, and medially. The stylohyoid ligament connects the tip of styloid process and the lesser cornue of hyoid bone. The tip of styloid process is often cartilaginous. Ossification of the cartilaginous tip or calcification of the stylohyoid ligament causes an appearance as if the styloid process is thickened and elongated. Length of the styloid process tends to increase with advancing age and therefore, ESP is more prevalent in the age group of 31 to 50 years. [6] Symptomatic patients are usually females

above the age of 40 years. [7-9] However, few authors have reported a higher incidence of ESP in males. [6]

ESP exerts pressure effect on the adjacent soft tissue structures such as the internal jugular vein, carotid artery and cranial nerves V, VII, IX, X and XII. Pressure symptoms consequent to nerve compression constitutes the Eagle syndrome. In our patient the close proximity of the styloid tip to the submandibular gland was clinically mistaken for salivary calculus.

The styloid process normally measures 2.5 to 3 cm in length [2]; but it varies in length from person-to-person and even from side-to-side of the same person. Kaufman et al. have defined 30 mm is the upper limit for normal styloid processes. [3] Therefore, styloid elongation can be diagnosed if its length exceeds 3 cm. [2,3] Partial ossification of stylohyoid ligament is not uncommon, but complete ossification is quiet rare. [10] The actual cause of styloid process elongation is poorly understood and the various theories proposed are: 1) congenital elongation of the styloid process due to persistence of a cartilaginous analogue of the embryologic precursors of the styloid, 2) calcification of the stylohyoid ligament due to an unknown reason, and 3) growth of osseous tissue at the insertion of the stylohyoid ligament.[5]

Based on various radiological features, Langlais et al classified ESP into 3 types. Type-I is characterized by uninterrupted elongation of the bone; type-II is characterized by the styloid process apparently being joined to the stylohyoid ligament by a single pseudo articulation giving the appearance of an articulated elongated styloid process, and type III consisting of interrupted segments of the mineralized ligament, sometimes creating the appearance of multiple pseudoarticulations. [11] Our case belongs to type-I ESP with pseudoarticulation with hyoid bone.

Researchers have correlated the ligamentous ossification of the stylo-hyoid complex and osteophytes of the cervical spine, concluding that variable types of styloid process-stylohyoid ligament complex abnormalities have significant correlation with ligamentous ossification and osteophytes of the cervical spine including cervical diffuse idiopathic skeletal hyperostosis (DISH). Enlargement of this ligament were significantly correlated with transverse and antero-posterior dimensions of ligamentous ossification or osteophytes of the cervical spine at various levels.[12] This association was also present in our case.

There are two types of Eagles syndrome (not to be confused with the 3 types of ESP) as described originally by Eagle: (1) the classic styloid syndrome presenting as pharyngodynia localized to the tonsillar fossa due to a

post- tonsillectomy fibrous scar causing compression/ irritation of the cranial nerve endings against the ESPs; (2) stylo-carotid syndromes (Carotidynia and Ernst syndromes) are due to compression of the internal and/ or external carotid arteries and their perivascular sympathetic fibers, resulting in a persistent pain radiating to the carotid territory. [13,14] Such patient may complain of headache, chronic neck pain, pain upon turning the head, pain radiating to the eye, ear pain and vertigo. [13,14]

Eagle Syndrome can be suspected clinically on the basis of symptoms, diagnosed on physical examination and confirmed radiologically by conventional radiography or more sensitive CT scan. The elongated styloid process can be felt in the tonsillar fossa, and palpation may aggravate symptoms. Several radiographic projections have been used for visualisation of ESP and they include lateral head-and-neck view, Towne view, panoramic view, lateral-oblique mandible view, and postero-anterior head view. [15] Superimposed bony anatomical structures will make it difficult to visualize ESP in plain radiography. Reconstruction of three dimensional views using multidetector computed tomography is an extremely valuable imaging tool. Details of the length, angulation, and relation to adjacent structures can be obtained accurately from a 3D-CT scan. [15]

Differential diagnoses to be considered in patients with similar clinical picture include temporomandibular joint diseases, trigeminal, sphenopalatine or glossopharyngeal neuralgias, temporal arteritis, chronic pharyngotonsillitis, otitis media, external otitis, mastoiditis, dental pain, improperly fitting dental prostheses, submandibular sialadenitis or sialolithiasis, true pharyngeal foreign bodies, and tumors of the pharynx or tongue base. [14, 15, 16] Most of these causes can be diagnosed or ruled-out by CT.

Eagle syndrome can be managed by surgical and nonsurgical methods. Nonsurgical treatments involve reassurance to the patient, analgesics, and steroid injections. Surgical treatment option includes styloidectomy which can be performed either transpharyngeally or extra-orally. The latter is thought to be superior because of decreased risk of deep neck space infection and better visualization of the surgical field. [5,14,16]

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## References

1. Eagle WW. Elongated styloid processes: Report of two cases. Arch Otolaryngol 1937; 25: 584-587.
2. Eagle WW. Elongated styloid process; further observations and a new syndrome. Arch Otolaryngol 1948; 47: 630-640.
3. Kaufman SM, Elzay RP, Irish EF. Styloid process variation. Radiologic and clinical study. Arch Otolaryngol 1970; 91: 460-463.
4. Correll RW, Jensen JL, Taylor JB, Rhyne RR. Mineralization of the Stylohyoid – Stylomandibular ligament complex. Oral Surg Oral Med Oral Pathol 1979; 48: 286-291.
5. Murtagh RD, Caracciolo JT, Fernandez G. CT findings associated with Eagle syndrome. AJNR Am J Neuroradiol 2001; 22: 1401-1402.
6. More CB, Asrani MK. Evaluation of the styloid process on digital panoramic radiographs. Indian J Radiol Imaging 2010; 20: 261-265.
7. Desai VD, Maghu S, Sharma R, Koduri S. Unique asymptomatic long bilateral calcified styloid process: A case report. J Indian Acad Oral Med Radiol 2014; 26: 302-305
8. Ferrario VF, Sigurta D, Daddona A, et al. Calcification of the stylohyoid ligament: Incidence and morpho quantitative evaluation. Oral Surg Oral Med Oral Pathol 1990; 69: 524-529.
9. Okabe S, Morimoto Y, Ansai T, et al. Clinical significance and variation of the advanced calcified stylohyoid complex detected by panoramic radiographs among 80-year-old subjects. Dentomaxillofac Radiol 2006; 35: 191-199.
10. Bouzaidi K, Daghfous A, Fourati E, et al. Eagle syndrome. Acta Radiologica Short Reports 2013; 2: 14.
11. Langlais RP, Miles DA, Van Dis ML. Elongated and mineralized stylohyoid ligament complex: A proposed classification and report of a case of Eagle Syndrome. Oral Surg Oral Med Oral Pathol 1986; 61: 527-532.
12. Guo B, Jaovisidha S, Sartoris DJ, et al. Correlation between ossification of the stylohyoid ligament and osteophytes of the cervical spine. J Rheumatol 1997; 24: 1575-1581.
13. Eagle WW. Symptomatic elongated styloid process: Report of two cases of styloid process-carotid artery syndrome with operation. Arch Otolaryngol 1949; 49: 490-503.
14. Jain S, Bansal A, Paul S, et al. Styloid-stylohyoid syndrome. Ann Maxillofac Surg 2012; 2: 66-69.
15. Savranlar A, Uzun L, Ugur MB, Ozer T. Three-dimensional CT of Eagle syndrome. Diagn Interv Radiol 2005; 11: 206-209.
16. Beder E, Ozgursoy OB, Ozgursoy SK. Current diagnosis and transoral surgical treatment of Eagle syndrome. J Oral Maxillofac Surg 2005; 63: 1742-1745.

