

CASE REPORT

Complete Ossification of the Stylohyoid Chain as Cause of Eagle's Syndrome: A Very Rare Case Report

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ABSTRACT

Aim: To report on a patient with Eagle's syndrome with a complete and very large ossification of the stylohyoid complex on the right side that to our best knowledge has never been published previously.

Background: Eagle's syndrome is characterized by a set of symptoms that are caused by the irritation of the neurovascular and soft-tissues caused by an elongated styloid process or ossification of stylohyoid ligament.

Case description: Because of the high discomfort and pain degree as well as limitations of mandibular and head mobility and also the thickness of the ossified stylohyoid chain, the patient was treated surgically by removing the hypertrophic segment.

Conclusion: These symptoms subsided completely after the surgical excision of the anomaly. The elongated styloid process on the left side was symptom free.

Clinical significance: Eagle's syndrome symptoms are not specific and can mimic those of other disorders, the syndrome must be included in the differential diagnosis of patients with pain in the orofacial, pharyngeal and cervical area.

Keywords: Eagle's syndrome, Styloid process, Ossification, Craniocervical pain, Stylohyoid chain, Case report.

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INTRODUCTION

Eagle's syndrome is classically characterized by dysphagia, foreign body sensation in the neck and oropharyngeal, cervical and craniofacial pain.¹⁻⁶ The pain is often exacerbated by rotation of the head to the contralateral side, swallowing, extending the tongue, and yawning and may radiate to distant areas. Indeed, a multitude of pain referral have been reported in the literature, for instance to the ear, neck, tongue, teeth, temporomandibular joint (TMJ) area and even the chest and upper limbs.⁷ Risk factors for the development of this syndrome are an elongated or more anteriorly angulated styloid process as well as ossified stylohyoid or stylomandibular ligaments.⁸⁻¹² Patients operated for an Eagle's syndrome had a significantly longer and more anteriorly angulated styloid process than controls.¹² A styloid process may be considered enlarged if it is more than 30 mm long. An elongated styloid process is found in 3 to 22% of the adult population^{10,13-16} and an ossified stylohyoid ligament in 2 to 20%.^{8,10,14,15} However, only a very small amount of subjects with an elongated styloid process (approximately 4%) are symptomatic, and in patients with bilaterally elongated styloid process the symptoms are in the vast majority of cases unilateral.^{17,18} The highest prevalence occurs in the age range 40 to 50 years¹⁴ and there is a female preponderance of about 3:1.^{14,17,19,20}

The Eagle's syndrome elicits symptoms that are not specific and that are most often caused by other disorders and pathologies of the orofacial and neck area. Consequently, it is often difficult to make a correct diagnosis. Classically, the diagnosis of an Eagle's syndrome is based on the chief symptoms presence, digital palpation of the process in the tonsillar fossa, elongated styloid process or partial/ complete ossification of the stylohyoid ligament as revealed by imaging. The diagnosis is greatly facilitated by the replication of the patient's symptoms during palpation of the tonsillar area and/or alleviation of the pain by injection of local anesthetic in the same region.^{1,3,5,10,21-23} Because the Eagle's syndrome symptoms are not specific and can mimic those of other disorders, the syndrome must be included in the

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differential diagnosis of patients with pain in the orofacial, pharyngeal and cervical area.^{6,21,24-27}

Eagle described two types: the classic and the styloid-carotid artery syndrome. The classic type is characterized by the symptoms just described while the styloid-carotid artery syndrome by ipsilateral headache, orbital pain and transient neurological symptoms in form of transitory ischemic attacks caused by a transient compression of the internal carotid artery and sympathetic chain.^{1,28-31} It was also suggested to distinguish three forms: the classical form, caused by trauma, the styloid syndrome and the pseudostyloid syndrome.²¹ The term 'stylohyoid syndrome', the most common form, defines the condition in which the patient's symptoms appear earlier in life owing to a non-traumatic developmental anomaly in the ossification of the stylohyoid ligament or to an elongated stylohyoid process. The 'pseudostyloid syndrome' on the other hand defines a tendinosis at the junction of the stylohyoid ligament and the lesser cornu of the hyoid in older individuals with no history of trauma and no evidence of styloid process elongation or stylohyoid ligament ossification on radiologic examination. Finally, owing to the fact that styloid process, the stylohyoid ligament and the lesser horn of the hyoid bone form the stylohyoid chain or stylohyoid complex, the term 'stylohyoid complex syndrome' has been proposed to classify all lateral neck and/or facial pain conditions resulting from an elongated styloid process, ossified stylohyoid ligament, or elongated hyoid bone.²⁹

In the following the two terms styloid chain and stylohyoid complex will be used interchangeably. In this article we report a patient with Eagle's syndrome. The particularity of the case resides in the complete ossification of the stylohyoid chain, which to our best knowledge has not been published previously. On the opposite side the patient had an elongated styloid process.

Case Description

A 55-year-old man was referred to the ambulatory services of the temporomandibular dysfunction and orofacial pain clinic at the hospital São Paulo – Escola Paulista de Medicina – Universidade Federal de São Paulo for orofacial pain. His main complain was severe pain in the jaw and submandibular right region on chewing and difficulty in opening the mouth, right otalgia, odynophagia and dysphagia since 1 year. The patient did not associate the symptoms beginning with a specific triggering event. The medical history was negative except for a prostate cancer treated surgically 3 years before.

The history revealed that symptoms started with a slight ear pain on the right side accompanied by a fullness or stuffiness sensation that lasted about 1 year. Thereafter, the ear

pain intensity increased. Despite the lack of a diagnosis he received an interocclusal appliance without any pain relief. On the contrary, this continued to increase. In addition, he began experiencing a mouth opening limitation. In the meantime a loud joint sound had developed that gradually decreased in intensity. This sound was accompanied by abrupt or jumping movements of the condyle when opening the mouth, but at that time he already had a mouth opening limitation.

These symptoms lead to several diagnostic hypotheses, for instance tumor in the pharyngeal/suprahyoid area, Eagle's syndrome, cervical disorders, temporomandibular disorders (TMD). A list of possible diagnoses in case of anterolateral neck and craniofacial pain disorders has just been published.²⁴ Difficulty in opening the mouth, severe pain in the jaw region and right side pain on chewing as well as otalgia are symptoms that could fit to the diagnosis of a TMD. However, the odynophagia, dysphagia and severe pain in the right submandibular region make this diagnosis unlikely. The pain in the right submandibular region could be due to gland pathology. The symptoms of odynophagia and dysphagia could be caused either by a tumor in the suprahyoid area or by an Eagle's syndrome. In addition, dysphagia could be due to neurological, muscular, anatomical, and/or psychological disorders.³² A cervicogenic problem could elicit pain when moving the head.

The clinical examination of the masticatory system revealed a maximum active and passive mouth opening of 26 mm with a hard end feel and, no tenderness on palpation of the TMJ areas but of the anterior, middle and posterior temporal muscle, of the masseter muscle of the sternocleidomastoid muscle and of the area behind the ascending mandibular ramus, all on the right side. On a 1 to 3 scale the patient graded the palpation pain as 3, 2, 1, 1, 2 and 3 respectively. There was a clicking sound in the left TMJ. The cervical screening showed that all head movements were limited and painful except for head extension and that some cervical muscles were tender to palpation. In addition, external palpation revealed a solid, hard mass in the right tonsillar region. This in addition to the reported symptoms reinforced the hypothesis of a tumor or of an Eagle's syndrome, thus the need of imaging of the neck area.

The orthopantomograph revealed an elongated styloid process and a large radiolucent area below the mandible on the left side and a large radiolucent area running laterally and parallel to the mandible on the right side (Fig. 1). In order to get a better definition of the radiolucent areas CT were requested. The 3D reconstruction showed on the left side an elongated styloid process and on the right side a complete and very thick, hypertrophic ossified stylohyoid chain (Figs 2 and 3).

Because of the high discomfort and pain degree as well as limitations of mandibular and head mobility and also the thickness of the ossified stylohyoid chain, the patient was treated surgically by removing the hypertrophic segment. The transoral technique was used.^{20,21,33} The resected piece had a length of 5 cm and a maximum width of 2.5 cm (Fig. 4). The elongated styloid process on the left side was not removed as it did not cause symptoms. The histopathologic analysis showed a mature compact bone tissue without

cellular atypia, confirming the diagnosis of ossification of the stylohyoid complex.

The surgical intervention leads to a total symptoms remission except for the dysphagia that lasted for about 3 months. There was also a recovery of neck movements and the mouth opening increased to 42 mm. The patient remained symptom free until he died on a cerebral aneurysm four years past surgery. The Escola Paulista de Medicina – Universidade Federal de São Paulo approved this case report and patient filled an informed consent.

DISCUSSION

The stylohyoid chain develops in the prenatal and perinatal period and originates from the second pharyngeal cartilage arch, which the proximal segment giving rise to the styloid process and the distal segment to the stylohyoid ligament. The lesser horn and upper part of the body of the hyoid bone originate from the ventral part.^{23,30} The ossification of the styloid process starts during childhood with a rather rapid and linear increase in the length of the ossified parts

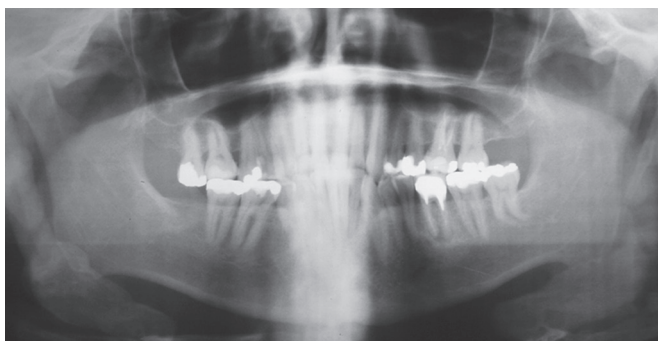
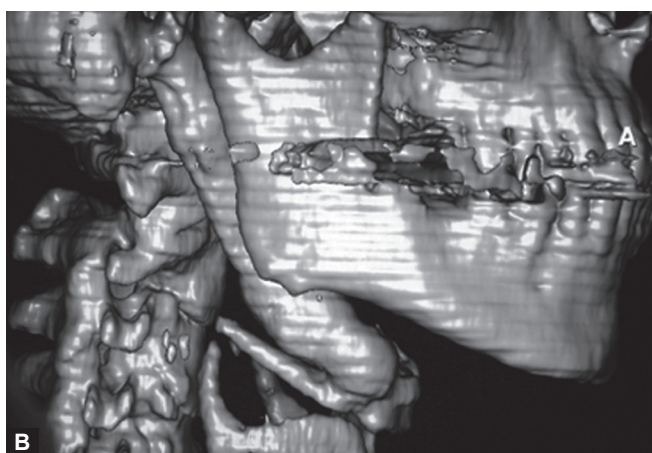
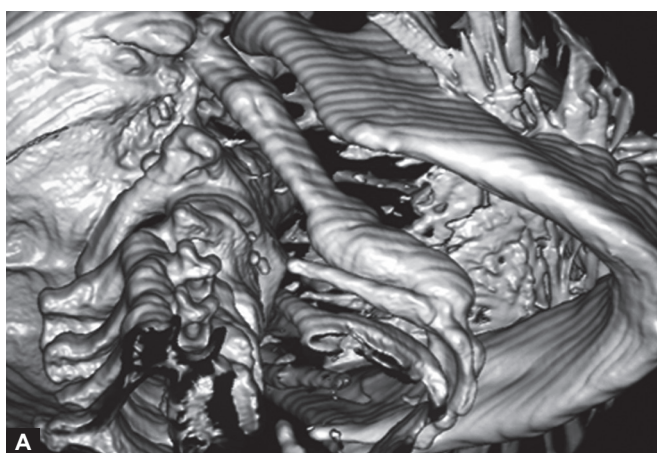


Fig. 1: Orthopantomograph demonstrating a large radiolucent area running below, laterally and parallel to the mandible on the right side



Figs 2A and B: Three-dimensional computed tomography confirming a complete and very thick hypertrophic ossified stylohyoid chain on the right side

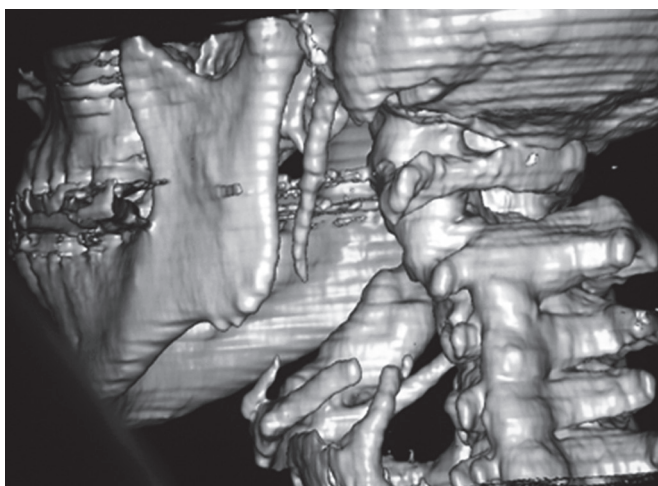


Fig. 3: Three-dimensional computed tomography reconstructions confirming an elongated styloid process on the left side

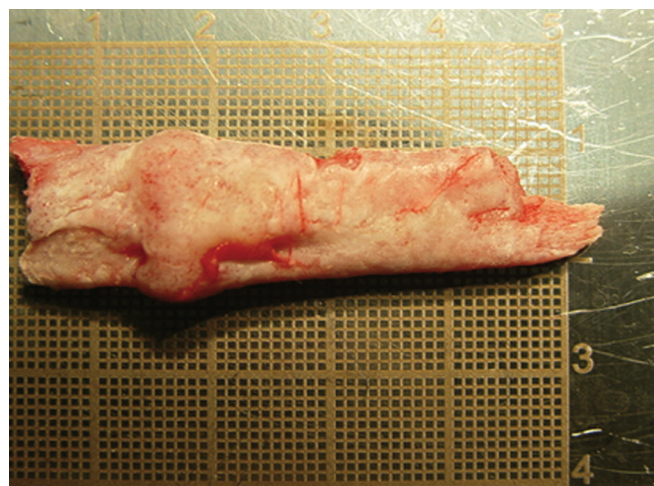


Fig. 4: The resected piece with a length of 5 cm and a maximum width of 2.5 cm that was sent to histopathologic analysis

up to the age of 20 years. Thereafter, the ossification speed decreases and becomes much slower continuing long into adult life. The reasons for this life-long ossification process are not understood yet, but may involve the variability in the length and shape of the second brachial arch cartilage,³⁴ which can explain the cases of styloid complex ossification in adults.^{25,28,34} However, the long lasting ossification process could be part of the normal aging process as it occurs also for other structure deriving from the brachial arches, such as the laryngeal cartilages.³⁵ As a matter of fact, the styloid process of 80 years subjects is longer than in younger adults.³⁶

Several theories have been proposed in order to explain the abnormal ossification found in patients with Eagle's syndrome: the theory of reactive hyperplasia, of reactive metaplasia, of anatomic variance,³⁷ and of aging developmental anomaly.²¹ Other authors hypothesized a genetic variance and an endocrinological dysfunction in postmenopausal women (details in Piagkou).⁷ For other possible causes the reader is referred to Montellaro.³⁸

There may pass several years before symptoms occur.^{21,25,30,39} It is thought that these are caused by the ossified structure irritating the tissues around it, including the carotid artery and the cranial nerves VII, IX and X^{10,17,19,28,29,33,40-42} or by the gradual loss of elasticity of the surrounding soft-tissues that develops over the years.^{8,29} The lack of elasticity could result in a more posterior position of the greater horn of the hyoid bone, displacing it closer to the neurovascular structures.²⁹

It is important to underline that, in spite of the fact that up to one fifth of the population has an elongated styloid process or an ossified stylohyoid chain,^{8,10,13-16} only a very small number of these subjects develop symptoms. Moreover, the symptoms are generally unilateral also in subjects having a bilaterally elongated process,^{17,18} as it was the case of our patient. These observations indicate that the presence of an elongated styloid process and/or an ossified stylohyoid ligament represents a very low risk factor for developing an Eagle's syndrome, and that the image of an elongated styloid process does not by itself justify the diagnosis of an Eagle's syndrome. Furthermore, care must be taken not to diagnose on orthopantomograms radiolucent areas behind the mandibular angle as an ectopic ossification of the stylohyoid chain while they in fact represent calcification of the carotid artery, phleboliths and lymph node calcification.³⁶ Therefore, some authors require a positive block of the tonsillar fossa in order to confirm the diagnosis of Eagle's syndrome.¹⁷

The particularity of this case consisted on one side in the large size of the ossification of the whole stylohyoid chain, and on the other side in the relatively low symptomatology in

relation to the huge size of the ossified structure. To the best of our knowledge this is the first description of a full ossification of the stylohyoid complex. Indeed, this ossification pattern is not included in previously published classification system of the stylohyoid complex.^{15,36}

It could be easy to explain, at least partially, the symptoms by the hypertrophic ossification of the stylohyoid chain that, by disturbing its functional mobility, may have caused the swallowing difficulty, the limited head rotation and the irritation of the surrounding structures. This pure mechanical explanation contrasts the fact that the patient was symptom free for a long period of time: the ossification process must have taken several years. This observation brings up the question which factor(s) may, suddenly, have triggered the pain and the other symptoms? It may be postulated that a nonwell recognized trigger elicited the symptoms that in turn lead to a vicious cycle in which the symptoms alters the soft-tissue equilibrium in the area, change the muscle contraction pattern of the surrounding muscles which in turn concurs sustaining the symptoms.

The Eagle's syndrome can be treated either conservatively or surgically by excising the elongated styloid process. Considering the fact that an elongated styloid process or ossified stylohyoid chain can remain symptom free for years and that we do not know what triggers the symptomatology, the initial treatment should be conservative and the surgical approach left to those cases in which the conservative therapy fails.⁸ A retrospective study in which 11 patients were placed on a stepwise therapy plan, which began with a pharmacological treatment, followed by a surgical treatment in case of symptoms persistence, showed, however, that only three out of the 11 patients did not require a further therapy after the pharmacological treatment.²⁰

The conservative treatment involves injections of long-lasting anesthetics and/or local corticosteroid into the tonsillar fossa as well as administration of nonsteroidal anti-inflammatory drugs.^{1,21} Manual fracture of the ossified styloid process under local anesthesia is another approach that has, however, unsatisfactory results.⁴³ Also, the fracture should be regarded critically because of its vicinity to the internal carotid artery.²⁰

Two different approaches have been described for the excision of the elongated styloid process: the extraoral or transcervical and the intraoral or transpharyngeal approach, each one with its advantages and disadvantages. The external approach has the advantage of a good visualization and reduced possibility of deep neck space infection, while the disadvantages are an external scar, longer duration of surgery, and risk of injury to the facial nerve. On the other hand, the intraoral approach is safe, simple and less time

consuming and does not cause external scar. However, is combined with a higher risk for infections of deep neck spaces and of injury of the internal carotid artery that may lead to cerebral infarct.^{17,20}

CONCLUSION

The present case report describes a patient with a complete ossification of the stylohyoid complex and that remained symptom free for several years before developing an Eagle's syndrome. The symptoms subsided after surgical removal of the ossified process.

CLINICAL SIGNIFICANCE

Eagle's syndrome symptoms are not specific and can mimic those of other disorders, the syndrome must be included in the differential diagnosis of patients with pain in the orofacial, pharyngeal and cervical area.

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