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The long styloid process syndrome or Eagle's syndrome

Giuseppina Fini,[1] Giulio Gasparini,[1] Fabrizio Filippini,[2] Roberto Becelli,[1] Dario Marcotullio[2]

[1]*Department of Maxillo-Facial Surgery, University of Rome, 'La Sapienza', (Director Prof. Giorgio Iannetti), V.le Regina Elena 287/a. 00196 Rome, Italy;* [2]*IV ORL Division, University of Rome, 'La Sapienza' (Director Prof. Marco De Vincentis), Rome, Italy*

SUMMARY. **Eleven cases of Eagle's syndrome or long styloid process syndrome are presented. It is a rare entity, which is not commonly suspected in clinical practice. Symptoms were dull and persistent pharyngeal pain, dysphagia, and facial pain.**

In addition to careful clinical examination, the use of plain radiographs, orthopantomograms or CT scan has allowed accurate preoperative diagnosis of this syndrome. Surgical treatment has positively resolved the symptoms in these cases. The International literature is discussed regarding anatomy, symptoms and treatment of the long styloid process syndrome. © 2000 European Association for Cranio-Maxillofacial Surgery

INTRODUCTION

In 1652, Pietro Marchetti, a surgeon from Padua, observed an elongation of the styloid process related to an ossifying process of the stylohyoid ligament. However, it was Eagle (1937 and 1949) who first defined stylalgia as an autonomous entity related to abnormal length of the styloid process or to mineralization of the stylohyoid ligament complex. Since then, several studies have been carried out some features of which are still not well explained. In 1988 the Headache Classification Committee of the International Headache Society, even defined the stylohyoid syndrome as 'not sufficiently demonstrated'.

The aim of this study is to report our experience with 11 cases observed during 4 years to review critically the International literature, and allow an analysis of the pathogensis, clinical and therapeutic aspects of Eagle's syndrome.

MATERIAL AND METHODS

Eleven patients with Eagle's syndrome have been treated with a mean age of 37 years (range 20–54 years) in the IV ORL Division and in the Department of Maxillo-Facial Surgery of Umberto I Hospital of the University in Rome, 'La Sapienza', from January 1995 to December 1998. Eight patients were female.

Symptoms were extremely variable with a mean onset of 4.5 years prior to diagnosis (Table 1). Accurate examination and intraoral palpation of the tonsillar area (Fig. 1) and radiological investigations including orthopantomogram and especially head and neck CT scan, facilitated the correct diagnosis. The treatment of choice in all patients was transoral styloidectomy (Figs 2 & 3), preceded in four cases by tonsillectomy.

All patients underwent antibiotic prophylaxis with Ceftriaxone perioperatively. We did not experience any postoperative infections.

RESULTS

Ten patients were followed-up for a period of 5 months to 4 years; one patient was lost to follow-up.

Our study showed a close correlation between Eagle's syndrome and previous tonsillectomy (seven cases). In two other cases the painful symptoms were related to previous trauma with fracture of the styloid process (Fig. 4). In one case, a 39-year-old woman, the only association noted was an early onset of menopause, with consequent endocrinological disorders.

In the last case, we were unable to determine a cause although surgical treatment led to a complete remission of symptoms. We mostly found pharyngeal pain with reflex otalgia (five cases); in three cases the patients had complained of a foreign body sensation and also painful swallowing (odynophagia). Dysphagia and cervical pain were present in two cases respectively. Pharyngeal paraesthesia, drowsiness, supraorbital pain and temporoparietal cephalgia were singularly present in just one case.

Complete remission of symptoms was achieved in five cases, while in five other patients partial remission was obtained (Table 1).

DISCUSSION

The stylohyoid apparatus (or stylohyoid complex) is made of: styloid process, stylohyoid ligament and the lesser horn of the hyoid bone. Embryologically, these structures are derived from Reichert's cartilage of the

Table 1 – Data of 11 cases of styloid syndrome

Patient	Sex	Age (years)	Symptoms	Possible cause	Results
1	M	24	Dysphagia, foreign body sensation for 2 years. Right otalgia	Tonsillectomy 3 years previously	Complete remission of symptoms
2	F	42	Dysphagia, foreign body sensation, right otalgia since 8 years	Tonsillectomy 20 years previously	Partial remission of symptoms
3	F	41	Pharyngeal pain and left otalgia for 1 year	Stylohyoid process fracture (trauma) 2 years before	Partial remission of symptoms
4	F	34	Cervical and right supraorbital pain provoked by cervical rotation for 11 years	Tonsillectomy 30 years previously	Partial remission of symptoms
5	M	52	Dysphagia for 7 years	Tonsillectomy 45 years previously	Partial remission of symptoms
6	F	38	Pharyngeal pain with left mandibular angle irritation and left otalgia for 2 years	Stylohyoid process ossification post-menopause? No tonsillectomy	Lost to follow up
7	M	43	Left cervical pain with foreign body sensation for 2 years	Tonsillectomy 8 years previously	Complete remission of symptoms
8	F	32	Pharyngeal and right otalgia for 3 years	Tonsillectomy 4 years previously	Complete remission of symptoms
9	F	54	Pharyngeal and right otalgia with pharyngeal paraesthesia for 6 years	Idiopathic ossification and medialization of the stylohyoid ligament complex. No tonsillectomy	Complete remission of symptoms
10	F	20	Pharyngeal and right otalgia for 3 years	Tonsillectomy 5 years previously	Complete remission of symptoms
11	F	27	Right temporo-parietal cephalgia with rotation of the head and painful swallowing drowsiness for 1 year	Stylohyoid process fracture (trauma) 2 years previously. No tonsillectomy	Partial remission of symptoms

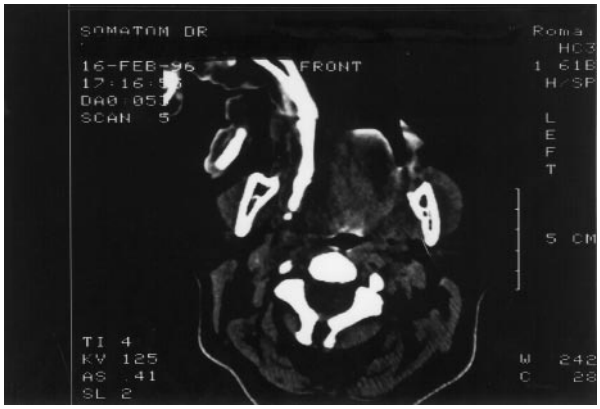

Fig. 1 – Intraoral palpation: CT imaging.

Fig. 3 – Stylohyoid process after styloidectomy.

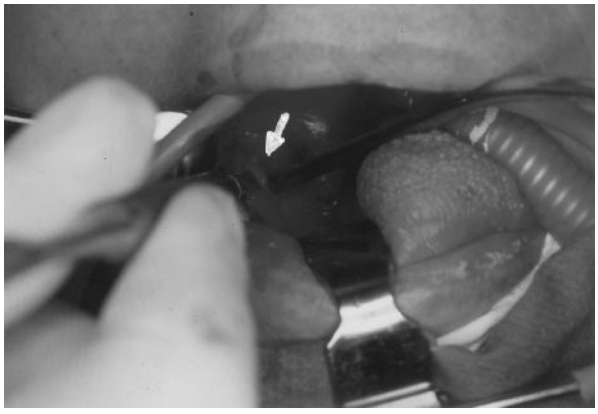

Fig. 2 – Oro-pharyngeal stylohyoidectomy.

Fig. 4 – Fracture of the styloid process lateral skull X-ray.

second branchial arch. Reichert's cartilage is made of four portions: an upper portion which develops into the styloid process, a central portion from which the stylohyoid ligament in derived; a lower portion from which the lesser horn of the hyoid bone originates; finally a basic portion from which part of the hyoid bone is formed (*Guerrier* et al., 1988). The styloid process appears as a narrow and enlongated

protrusion of the temporal bone, directed caudally, medially and anteriorly within the maxillo-vertebropharyngeal space, in which important anatomical structures are located such as: both carotid arteries the internal jugular vein, the facial, glossopharyngeal, vagus and hypoglossal nerves. Here the stylohyoid, the styloglossus and the stylopharyngeus muscles originate (which together form the so-called 'Riolano's bouchet') as well as the stylohyoid and stylomandibular ligaments (defined as 'the bouchet's white flowers'). The stylohyoid muscle originates from the base of the styloid process and is inserting on the hyoid bone near the greater horn. The styloglossus originates close to the apex of the styloid process and from the upper part of the stylomandibular ligament and is inserted in the septum of the tongue (*Motta* et al., 1987). The stylopharyngeus originates from the medial aspect of the styloid process close to its base and is inserted into the lateral wall of the pharynx. The stylohyoid ligament originates from the apex of the styloid process and is attached inferiorly to the lesser horn of the hyoid bone. The stylomandibular ligament originates near the apex of the styloid and is inserted medially into the mandibular angle.

The length of the styloid process is individually variable. *Moffat* et al. (1977) performed studies on cadavers and demonstrated that a normal styloid process is from 1.52 cm to 4.77 cm long; according to *Lindeman* (1985), it is 2–3 cm long (Fig. 3). Radiologic examinations (*Correl* et al., 1979; *Langlais* et al., 1986; *Montalbetti* et al., 1995) revealed that the length of the styloid process normally is less than 2.5 cm. However, according to *Monsour* and *Young* (1986) a 'long' styloid process is defined as more than 4 cm, since in this situation the highest incidence of Eagle's syndrome occurs. In a study carried out on 2000 cases only of cranial dissections, Gruber only found styloid processes longer than 4 cm in 11 cases (*Massey*, 1978).

The incidence of this syndrome varies between populations. *Eagle* (1948) found a long styloid process in 4% of the subjects he examined, whereas *Kaufman* et al. (1970) found incidence of 7%. However, just a very low percentage of those had complained of pain.

This condition is found more often in females and affects subjects more than 50-years-old most often. (*Glogoff* et al., 1981; *Montalbetti* et al., 1995). Although our sex ratio was the same, the mean age of our patients was lower.

Pathogenesis is still being debated. *Eagle* (1937; 1949) thought that surgical trauma or local chronic irritation could cause osteitis, periosteitis, or tendonitis of the stylohyoid complex with consequent reactive, ossifying hyperplasia. Later, different hypotheses have been formulated such as: persistence of mesenchymal elements able to produce osseous tissue in adults (*Lentini*, 1975); Reichert's cartilage residues undergoing osseous metaplasia as a consequence of trauma or mechanical stress during the development of the styloid process (*Laino* et al., 1987; *Roca* et al., 1992); abnormal development associated with malformations of the atlanto-occipital hinge (*Arnould*, 1969; *Carrella*, 1971); ossification of the stylohyoid ligament related to endocrine disorders in women at the menopause, accompanied by ossification of ligaments elsewhere (iliolumbar, thyrohyoid) (*Epifanio*, 1962).

However, the abnormal length of the styloid process by itself is not sufficient to explain the pathogenesis of Eagle's syndrome, as there is only a low incidence of symptoms. Eagle discovered a close correlation between the manifestation of the classical painful syndrome and tonsillectomy. He felt that this operation was responsible for the formation of scar tissue around the styloid apex in some cases with consequent compression and stretching of the nervous structures contained in the maxillo-vertebropharyngeal space, especially the glossopharyngeal nerve and perivascular carotid sympathetic fibres. However, *Fritz* (1940) reports that among his 43 cases only 11 had been subjected to tonsillectomy. Subsequently, other factors have been considered such as ossification of the stylohyoid ligament complex causing contraction of the stylopharyngeal muscle and consequently stretching of the XII cranial nerve (*Monsour* and *Young*, 1986); fracture and medialization of the ossified stylohyoid ligament (*Moffat* et al., 1977), with frustrated repair disturbed by the continuous hyoid bone movements, causing excessive proliferation of granulation tissue responsible for compression of the structures nearby; ossification of muscular tendons leading to irritation (*Kaufman* et al., 1970; *Lindeman*, 1985); and finally, the abnormal length associated with an abnormal angulation could be the reason (*Baddour* et al., 1978; *Balestra* and *Cherie-Lignerie*, 1903; *Frommer*, 1974; *Garel*, 1927).

In the original description, *Eagle* (1937) presented two possible clinical expressions of the syndrome:

(1) Classical stylohyoid syndrome
(2) Stylocarotid syndrome

The **classical stylohyoid syndrome**, almost always following tonsillectomy, is characterized by dull and persistent, pharyngeal pain (pharyngodynia), especially located in the tonsillar fossa, with radiation to the ipsilateral ear, accompanied occasionally by dysphagia and painful swallowing (odynophagia), foreign body sensation, as much as facial and/or cervical pain. Rarely the pain is very intense.

The **stylocarotid syndrome** is not correlated with tonsillectomy. It arises whenever the stylohyoid apparatus compresses the internal and/or external carotid arteries, and especially the perivascular sympathetic fibres. It is characterized by cervical pain arising when the internal carotid artery is compressed, provoked and aggravated by rotation and compression of the neck and radiates to the areas vascularized by the ophthalmic artery with involvement of the supraorbital and parietal regions. In contrast, if the external carotid artery is irritated, the

pain radiates to the infraorbital region. Drowsiness and visual disorders can be present. In order to diagnose Eagle's syndrome, it is necessary to obtain an accurate case history. The patient's description of symptoms is important. Even when local examination is negative, the styloid process may be palpated in the tonsillar fossa (Fig. 1) and this may initiate or aggravate the symptoms. Usually it is not palpable.

Radiological examination confirms the diagnosis. For visualization of the styloid process, orthopantomograms are more advisable than the standard radiological cranial examination with these latter views, identification of the stylohyoid process is not always possible as it is masked by other structures (*Lentini*, 1975). CT scans are better for defining length, angulation and anatomical relationships of the stylohyoid process (Fig. 5) (*Kaufman* et al., 1970).

Several disorders may simulate Eagle's syndrome: the glossopharyngeal and trigeminal neuralgias are characterized by a lancinating and sudden pain of short duration evoked by 'trigger zone' stimulation (*Mallardi*, 1995; *Montalbetti* et al., 1995) in contrast to Eagle's syndrome in which pain is dull and persistent. Temporomandibular joint disorders and pain from wisdom teeth are confirmed by radiological examination. Carotidynia, which seems to be caused by the pericarotid sympathetic plexus somehow irritated or involved with arteriitis manifests with symptoms similar to the stylocarotid syndrome (*Prendes*, 1983). The latter presents with hyperpulsation and dilatation of the external and common carotid arteries. Nimesulide and other anti-migraine drugs resolve the symptoms (*Raskin*, 1977). Neuralgia of the upper laryngeal nerve, oesophageal diverticula, neoplasia and migraine are also to be considered.

Eagle's syndrome can be treated both, surgically and non-surgically. *Evans* and *Clainnont* (1976) suggested transpharyngeal infiltration of steroidal drugs and local anaesthetics (lidocaine) in the tonsillar fossa whereas *Chase* et al. (1986) suggest manual fracture of the styloid process although long-term results have not been satisfactory (*Murthy* et al., 1990).

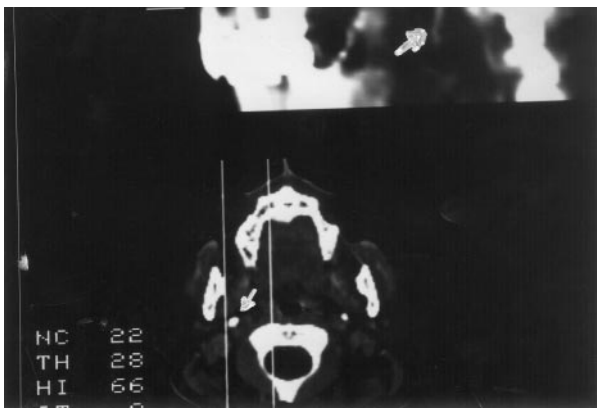

Fig. 5 – CT scan of head and neck, demonstrating length, angulation and anatomical relationships of the styloid process.

Styloidectomy is the treatment of choice. It can be performed transorally or by an extraoral approach.

The transoral approach was introduced by Eagle. The surgeon locates the styloid process by digital palpation of the tonsillar fossa, followed by surgical incision. Once the styloid process has been identified, it is stripped of the periosteum together with muscles and ligaments. Then styloidectomy is carried out. If the pharyngeal tonsil is still present, it is necessary to perform tonsillectomy first at the same operation.

The external approach was described by *Loeser* and *Caldwell* in 1942. A cervical incision is made from the proximal portion of the sternocleidomastoid muscle to the hyoid bone. The parotid fascia is reflected anteriorly, the carotid sheath and the sternocleidomastoid posteriorly. Aponeurotic and muscular insertions are separated from the styloid process which is then dissected and removed. This approach allows adequate visualization of the stylohyoid process and of the nearby structures, and enables greater intraoperative sterility. However, it requires a longer recovery and more intervention (drain, sutures) and of course results in a visible neck scar.

Transoral styloidectomy has been the preferred treatment in all our cases; the advantage of this method is brevity and the absence of a cervical scar. The disadvantage of the method is the poor visibility leading to major potential risks of iatrogenic injury to the main neurovascular structures. Furthermore, intraoperative contamination is possible.

We found a remarkable time link regarding the onset of symptoms and the timing of tonsillectomy: four of the seven patients who had undergone tonsillectomy and who had complete recovery had been subjected to tonsillectomy only 1–2 years before the beginning of symptoms. However, tonsillectomy had been carried out at 20 to 45 years earlier in the three patients with only partial remission. From the data it is clear that the organization of scar tissue following tonsillectomy probably was not complete in the four mentioned first, whereas in the three cases with partial remission, greater fibrotic organization had occurred resulting in less favourable results after styloidectomy.

A partial success has been also found in two cases with fracture of the styloid process. The reason may be the mechanism of fracture repair and the continuous movements of the hyoid bone, causing excessive granulation and fibrous tissue production.

CONCLUSIONS

We think that not only a long stylohyoid process, but also other factors are necessary to give rise to Eagle's syndrome.

CT scan is best for diagnosis, although also an accurate case history and the specialist's intuition are fundamentally important for the differential diagnosis regarding several other pharyngo-cranio-facial pain disorders.

In our opinion, surgical treatment is first choice and the transoral approach is preferable. This approach, avoiding injury to important structures contained in the maxillo-vertebro-pharyngeal space, is characterized by a short operation, and absence of visible scars and reduced hospitalization issue.

References

Arnould G: Appareil stylo-hyoidien et malformations de la chamiere occipito-vertebrale. Rev Otoneuroopathalmol 41: 190–195, 1969

Baddour HM, McAnear JC, Tilson HB: Eagle's syndrome. Oral Surg 46: 486–494, 1978

Balestra A, Cherie-Ligniere M: Di due apparecchi ioidei completamente ossificati. Comunicazione Assemblea Chirurgica di Parina, 3 Aprile 1903

Carrella A: Apparato stilo-ioideo e malformazione della cerniera atlo-occipitale. Acta Neurol Napol 26: 466–472, 1971

Chase DC, Zarman A, Bigelow WC, McCoy JM: Eagle's syndrome: A comparison of intraoral versus extraoral surgical approaches. Oral Surg 62: 625–629, 1986

Correl R, Jensen J, Taylor J, Rhyne R: Mineralization of the stylohyoid-stylomandibular ligament complex: A radiographic incidence study. Oral Surg Oral Med Oral Pathol 48: 286–291, 1979

Eagle W: Elongated styloid process. Report of two cases. Arch Otolaryngol 25: 584–587, 1937

Eagle W: Elongated styloid process: Further observation and a new syndrome. Arch Otolaryngol 47: 630–640, 1948

Eagle W: Symptomatic elongated styloid process: Report of two cases of styloid process-carotid artery syndrome with operation. Arch Otolaryngol 49: 490–493, 1949

Epifanio G: Processi stiloidei lunghi e ossificazione della catena stiloidea. Rad Prat 12: 127–132, 1962

Evans JT, Clainnont AA: The nonsurgical treatment of Eagle's syndrome. Ear Nose Throat J 55: 44–45, 1976

Fritz M: Elongated styloid process: cause of obscure throat symptoms. Arch Otolaryngol 31: 911–918, 1940

Frommer J: Anatomic variations in the stylohyoid chain and their possible clinical significance. Oral Surg 38: 659–667, 1974

Garel M: Angine styloidienne chronique. Lyon Med 9: 654–661, 1927

Glogoff MR, Gavan SM, Chiefetz J: Diagnosis and treatment of Eagle's syndrome. J Oral Surg 39: 941–944, 1981

Guerrier Y, Rabischong P, Guerrier B: Cours superieur d'anatomie chirurgicale de cou et des glandes salivaires. Ed. Lib. Arnett, Montpellier, 1998

Headache Classification Committee of the International Headache Society. Classification and diagnostic criteria for headache disorders, cranial neuralgias and facial pain. Cephalgia 8: 1998, Suppl. 7

Kaufman SM, Elzay RP, Irish EF: Styloid process variation: Radiologic and clinical study. Arch Otolaryngol 91: 460–463, 1970

Laino G, Ammirati G, Serpico S: Styloid-stylohyoid syndrome or Eagle's syndrome. Arch Stomatol 28: 183–190, 1987

Langlais R, Miles DA, Van Dis ML: Elongated and mineralizated stylohyoid ligament complex: a proposed classification and report of a case of Eagle's syndrome. Oral Surg 61: 527–532, 1986

Lentini A: Gli aspetti clinici e radiologici delle anomalie dell'apparato stilo-joideo. Radiol Med 61: 337–364, 1975

Lindeman P: The elongated styloid process as a cause of throat discomfort. J Laryngol Otol 99: 505–508, 1985

Loeser L, Caldwell E: Elongated styloid process: a cause of glossopharyngeal neuralgia. Arch Otolaryngol 36: 198–202, 1942

Mallardi V: Algie del distretto oro-faringeo. In: II dolore in otorinolaringoiatria. Editor A Di Girolamo, Roma 155–160, 1955

Marchetti P: Anatomia. Patavii 1652

Massey EW: Facial pain from an elongated styloid process (Eagle's syndrome). South Med J 71: 1156–1159, 1978

Moffat DS, Ramsden RT, Shaw HJ: The styloid process syndrome – aetiological factors and surgical management. J Laryngol Otol 91: 279–294, 1977

Monsour P, Young WJ: Variability of the styloid process and stylohyoid ligament in panoramic radiographs. Arch Otolaryngol 61: 522–526, 1986

Montalbetti L, Ferrandi D, Pergami P, Savoldi F: Elongated styloid process and Eagle's syndrome. Cephalalgia 15: 80–93, 1995

Motta P, Renda T, Marinozzi G: Anatomia Umana. Edi Ermes, Milano, 11: 209–216, 1987

Murthy PSN, Hazarika P, Mathai M, Kumar A, Kamatah MP: Elongated styloid process: an overview. Int J Maxillofac Surg 19: 23 0–23 I, 1990

Prendes J: Carotidynia or Eagle's syndrome? Headache 26: 26–46, 1983

Raskin H: Carotidynia. Neurology 27: 43–46, 1977

Roca A, Armengton M, Gimenez G, Barona R, Basterra J: Tratamiento quirurgico del sindrome de Eagle por via orofarigea. A propòsito de un caso. Acta Otorrinolaring Esp 43: 210–212, 1992

Dr. Giulio Gasparini
Via Pier Luigi Galletti 3
00135 Rome
Italy

Tel: 39 6 30816037
Fax: 39 338 2312492
E-mail: cichettol@yahoo.it

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