Acta Otorrinolaringol Esp. 2010;61(3):233–235

Acta Otorrinolaringológica Española

www.elsevier.es/otorrino

CASE STUDY

Eagle's syndrome: transient syncopes as a clinical manifestation; complete stylohyoid ligament calcification

Juan José Sanz Gonzalo,* Javier Maiz Cal, Xavier Lao Luque, Guillem Viscasillas Pallàs, and Christiane Zschaeck Luzardo

Servicio de Otorrinolaringología, Hospital Mutua de Terrassa, Terrassa, Barcelona, Spain

Received January 26, 2009; accepted May 11, 2009

KEYWORDS
Eagle's syndrome;
Calcified stylohyoid ligament

Abstract
We report a case of a young man with transient syncopes as a first clinical manifestation. Neck radiographic examination showed a bilateral calcification of the stylohyoid ligament, doubly articulated in its extension and compressing the internal carotid artery.

PALABRAS CLAVE
Síndrome de Tagle;
Osificación del ligamento estilohioideo

Síndrome de Eagle: ictus de repetición como presentación clínica; calcificación estilohioidea completa

Resumen
Presentamos el caso de un varón joven cuya clínica inicial fue ictus de repetición y en el que el estudio radiológico mostró una osificación completa del ligamento estilohioideo bilateral, articulado doblemente en su extensión y que comprimía la carótida interna.

Introduction

Styloid syndrome was first described by Eagle in 1937 and he already reported two forms of clinical presentation: the classic and the carotid artery syndrome.[1,2]

The styloid process is defined as normal when it is less than 2.5 cm and as elongated when it is longer than 4 cm.[3] Women comprise 85%of cases and it is manifested between the second and third decades of life.[4,5]

The origin of the elongation or calcification of the styloid apophysis is unclear; it could be due to a retention of embryonic cartilage that maintains its potential for growth and bone maturation.[3,6] On the other hand, attempts have been made to link this syndrome, with no demonstrated evidence, with prior tonsillectomy, cervical pharyngeal trauma or tendonitis at the junction of the stylohyoid ligament and the lesser horn of the hyoid bone.[3,6-8]

Clinical presentation usually takes place with non-specific cervical pharyngeal pains (classic clinical manifestation) that are exacerbated by palpation of the amygdaloid fossa.[1,2] Clinical manifestation with ictus depends on the

*Corresponding author.
E-mail address: 32619jsg@comb.es (J.J. Sanz Gonzalo).

0001-6519/$ - see front matter

compression of the internal carotid, which in turn depends on the direction of styloid apophysis growth.

Differential diagnosis must take into account craniomandibular dysfunction, temporal arthritis, trigeminal and glossopharyngeal neuralgia, migraines, histamine headaches, pain secondary to impacted third molars, oropharyngeal or cavum tumours, carotid artery syndrome and Ernest syndrome.[3,4,6,7,9]

Case study

35-year-old male with no relevant medical history, who presented repeated ictus related to cervical rotations. The patient's cervical x-ray showed styloid apophyses that were elongated up to almost the hyoid bone (Figure 1). The patient underwent surgery through a U-shaped cervicotomy of both styloid apophyses, which were completely resected. It was found that the styloid apophysis were bilaterally elongated and articulated in their length. It was difficult to distinguish between elongation of the styloid and calcification of the stylohyoid ligament (Figure 2). The patient was asymptomatic and had no postoperative complications.

Discussion

The clinical presentation of Eagle's syndrome often goes unnoticed due to its non-specific symptoms, although it must be considered in the diagnostic process.

Computed tomography is the method of choice for radiological diagnosis (as it allows multiplanar and three-dimensional reconstructions).

In cases where carotid vascular symptoms predominate, the treatment of choice is surgery. In our view, the cervical approach is preferable in these cases, given that it provides a familiar area for the otolaryngologist, facilitates control of the vascular axis, allows a more radical "height" in the excision of the styloid and, in addition, facilitates the control of neighbouring neural structures. It is essential to have thorough knowledge of the stylomastoid region and its close relationship with the internal carotid.

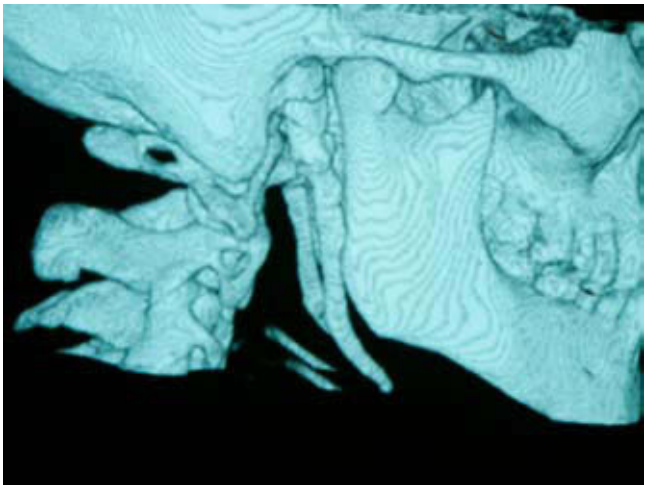

Figure 1 Elongated styloid apophysis with calcification of the stylohyoid ligament.

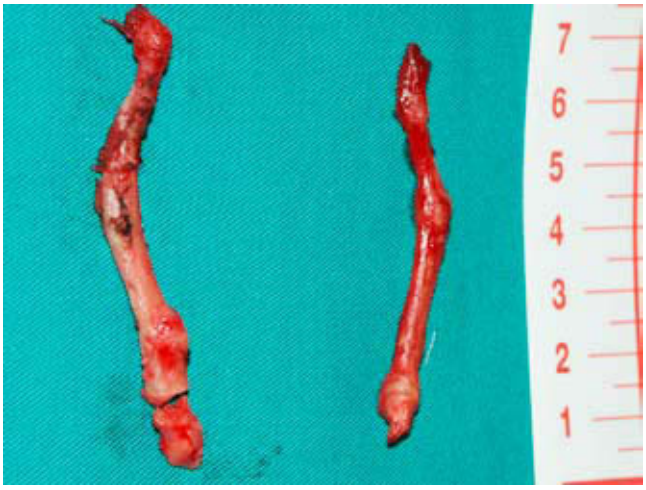

Figure 2 Styloid apophysis resected whole bilaterally and with joints along their length.

After a lateral cervicotomy, it is essential to identify the hyoid bone as a starting point for the dissection; the insertion into the lesser horn of hyoid is isolated and progress is carried out up to the styloid. Dissection following linear calcification and adhered to it avoids problems with the major vascular and neural structures in the vicinity. A posterior relationship is established with the hypoglossal nerve and sectioning the ossified ligament into fragments is advised, to facilitate total removal with minimal risk to the nerve.

The transoral approach offers greater postoperative pain and poorer intraoperative control. Severe arterial complications have been reported with this approach[10,11] and it would be limited to cases in which it was possible to detect the styloid apophysis by submucosal palpation.

It is important to take this syndrome into consideration in cases where the clinical signs feature non-specific pharyngeal pains, localised odynophagia or foreign body sensation.[11]

Prominent styloid apophysis, with or without apparent clinical signs, presents in the population in very variable manner, according to the studies reviewed; presentation ranged between 18% and 40% of the population in some series in which radiographic length alone was analysed.[4] Eagle, in his original publication, confirmed an elongated styloid process in 4% of his cases; these patients presented only 4% of accompanying symptoms.[12] The 1970 series of Kaufman revealed the elongation of the styloid process in 7.3% of patients.[13] This leads us to conclude that most patients with elongated styloid process are asymptomatic.[13,14]

Many authors believe that this syndrome is characteristic of adulthood, but we believe it is a continuous process of progressive ossification that begins in adolescence. At younger ages, greater ligament fibrosis is observed; this period is asymptomatic. As the process evolves, calcification takes place, with or without manifestation of classic symptoms.[5]

References

1. Chouvel P, Rombaux P, Philips C, Hamoir M. Styloid chain ossification: choice of the surgical approach. Acta Otorhinolaryngol Belg. 1996;50:57-61.
2. Aral IL, Karaca I, Güngör N. Eagle's syndrome masquerading as pain of dental origin. Case report. Australian Dental Journal. 1997;42:18-9.
3. Sá ACD, Zardo M, Paes AJO, Souza RP, Neto FB, Dreweck MO, et al. Alongamento do processo estilóide (síndrome de Eagle): relato de dois casos. Radiol Bras. 2004;37:385-7.
4. Neville BW, Damm DD, Allen CM, Bouquot JE. Patologia oral & maxilofacial. 2nd ed. Rio de Janeiro: Guanabara Koogan; 2004. p. 21-2.
5. Omnell KH, Gandhi C, Omnell ML. Ossification of the human stylohyoid ligament: A longitudinal study. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 1998;85:226-32.
6. Reis SSPM, Carvalho PL, Reis HSM. Processo estilóide alongado-relato de dois casos. J Bras de Oclusão, ATM e Dor Orofacial. 2001;1:296-300.
7. Milner E, Donato AC, Fregonesi M, Mistro FZ, Moreira CA, Kignel S, et al. Síndrome de Eagle-apresentação de 3 casos clínicos e revisão da literatura. Rev Paul Odontol. 1996;18:26-8.
8. Albuquerque RF, Müller K, Hotta TH, Gonçalves M. Temporomandibular disorder or Eagle's syndrome? A clinical report. J Prosthet Dent. 2003;90:316-9.
9. Shankland WE. Ernest syndrome as a consequence of stylomandibular ligament injury: a report of 68 patients. J Prosthet Dent. 1987;57:501-6.
10. Weidenbecher M, Schick B, Iro H. Styloid syndrome and its treatment. Laryngorhinootologie. 2006;85:184-90.
11. Nickel J, Sonnenburg M, Scheufler O, Andresen R. Eagle syndrome: diagnostic imaging and therapy. Rontgenpraxis. 2003;55:108-13.
12. Quereshy FA, Gold ES, Arnold J, Powers MP. Eagle's syndrome in an 11-year-old patient. J Oral Maxillofac Surg. 2001;59:94-7.
13. Moraes S, Nakonechny JP, Chaia A. Síndrome de Eagle. Relato de um caso. Rev Bras Odont. 1991;XLVIII:30-5.
14. Glogoff MR, Baum SM, Cheifetz I. Diagnosis and treatment of Eagle's syndrome. J Oral Surg. 1981;39:941-4.