

Case Report

Unilateral complete articulated ossification and aberrant thickening of the stylohyoid chain

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Abstract: Eagle's syndrome is a rare clinical entity associated with craniofacial and cervical pain. It is caused by an elongated or deviated styloid process and/or calcification/ossification of the stylohyoid ligament. Complete ossification and aberrant thickening of the stylohyoid chain is the most unusual manifestation. We report a patient who presented with complaints of foreign body sensation, dysphagia and moderate submandibular pain caused by unilateral complete ossification, abnormal thickening and hypertrophy of the stylohyoid chain. This case was also unique in that histopathological investigation demonstrated mature bone with both compact and cancellous bone as well as bone marrow.

Keywords: Eagle's syndrome; complete ossification of the stylohyoid chain; orofacial pain.

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Introduction

Eagle's syndrome, also known as styloid-stylohyoid or styloid-carotid artery syndrome, was first described by Eagle, an otolaryngologist, in 1937 (1). The classic type involves a cluster of symptoms characterized by dysphagia, dysphonia, foreign body sensation in the neck and pharynx, oropharyngeal and cervical pain, and ear pain. The carotid artery type involves headache, orbital pain and transient ischemic attacks (1,2). It is caused by an elongated or deviated (anteriorly angulated) styloid process and/or calcification/ossification of the stylohyoid chain complex (1,3). Due to these anatomical changes, pain in the temporomandibular joint (TMJ) or pre-auricular area as well as limited or asymmetrical mandibular movements may also be reported by patients. As many dentists have limited experience of this diagnosis, symptoms may be attributed to other causes, leading to mistaken diagnoses and inappropriate management. While partial calcification (calcified tissue formation) of the stylohyoid ligament is often seen, complete ossification (bone formation) and hypertrophy of the stylohyoid chain is very unusual. Here, we report a patient who presented with a complaint of unilateral submandibular pain, and was eventually diagnosed as

having Eagle's syndrome. The present case of Eagle's syndrome was distinctive, as it was characterized by complete articulated ossification and aberrant thickening of the stylohyoid chain on the symptomatic side.

Case Report

A 49-year-old Chinese man was referred to the Temporomandibular Disorders and Orofacial Pain Clinic at Peking University School and Hospital of Stomatology by an oncologist. The patient complained of a six-month history of foreign body sensation, dysphagia and moderate pain in the right submandibular region. The symptoms were exacerbated when the patient turned his head to the right. Treatment with ibuprofen, a nonsteroidal anti-inflammatory drug (NSAID), did not alleviate the symptoms. The patient's general medical/dental history was unremarkable, and there had been no prior trauma or head/neck surgery. Digital palpation indicated a tender, firm, funicular mass with osteoid hardness in the tonsillar fossa and submandibular region on the right side. Pain in the right submandibular region was triggered by head movement to the right.

A panoramic radiograph revealed a large radiopaque mass running laterally and parallel to the ramus on the right side (Fig. 1). A computed tomographic (CT) scan was subsequently performed, and 3D reconstruction demonstrated an abnormally thickened and hypertrophic stylohyoid chain on the right side. The mass was about 10 mm in width, 90 mm in length, 3,650 mm² in surface area and 8,070 mm³ in volume (Fig. 2). The CT values varied from 1,500 to 2,400 Hu. No other abnormalities were observed.

Based on the information obtained, a clinical diagnosis of Eagle's syndrome was made. The patient was informed of his condition and the medical/surgical management options available. Surgical removal through an extra-oral approach from the neck was decided upon. Upon surgical exposure, a continuous hypertrophic bony mass was revealed. It was attached superiorly to the skull base, just in front of the mastoid process, and inferiorly to the hyoid bone. The bony mass articulated with the hyoid bone at the junction of the lesser cornu (Fig. 3). The superior resection border was planned to run underneath the facial nerve trunk to avoid possible nerve damage, and inferiorly above the body of the hyoid bone. The resected specimen was about 50 mm in length (including bony pieces removed with a rongeur) and 12 mm in maximum width (Fig. 4). The specimen was decalcified in 40% formic acid for 7 days, embedded, sectioned and stained using hematoxylin and eosin for optical microscopy. Histological examination showed mature bone without



Fig. 1 Panoramic radiograph showing a large radiopaque area (arrows) running laterally and parallel to the mandibular ramus from the skull base to the hyoid bone on the right side.

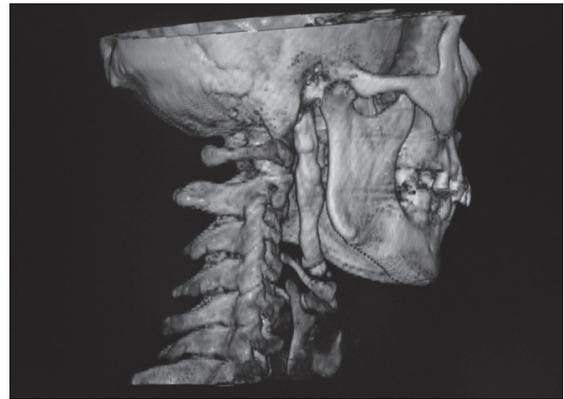


Fig. 2 3D reconstruction demonstrating an abnormally thickened and hypertrophic stylohyoid chain on the right side.

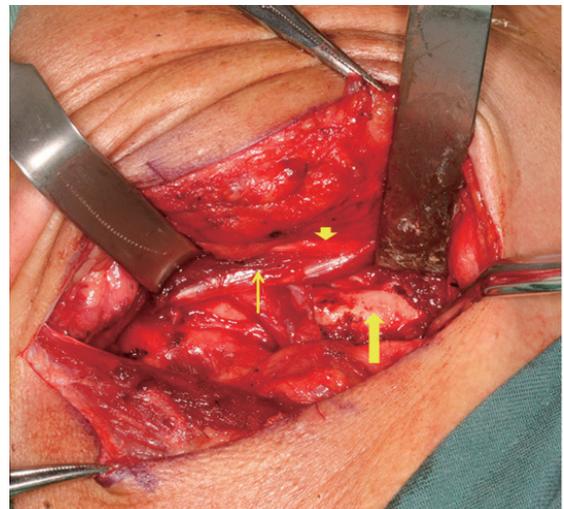


Fig. 3 A funicular bone attached superiorly to the skull base (immediately in front of the mastoid process) and inferiorly to the hyoid bone (widened arrow) was observed. Adjacent structures included the musculus stylohyoideus (arrow) and the posterior belly of the digastric muscle (arrow head).

cell atypia, consisting of compact and cancellous bone filled with red and yellow bone marrow (Fig. 5). This confirmed our clinical diagnosis of Eagle's syndrome caused by complete articulated ossification and aberrant thickening of the stylohyoid chain. On the second postop-



Fig. 4 The resected specimen, including segments removed with a ronguer, was submitted for histopathological investigation.

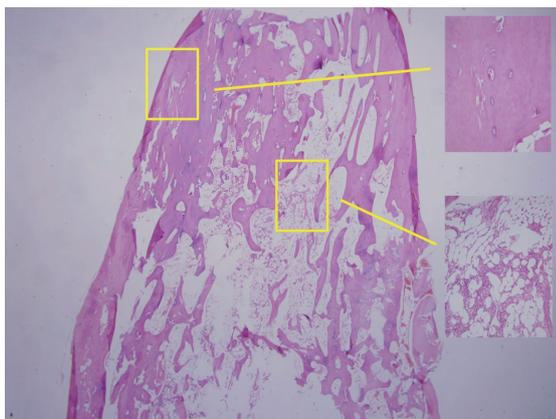


Fig. 5 Photomicrographs of the ossified stylohyoid chain (hematoxylin and eosin staining). The mature bone tissue comprised compact bone, cancellous bone and bone marrow (original magnification $\times 1.25$; Haversian system and cluster of osteocytes in the compact bone, original magnification $\times 20$; Bone marrow in the cancellous bone, original magnification $\times 20$).

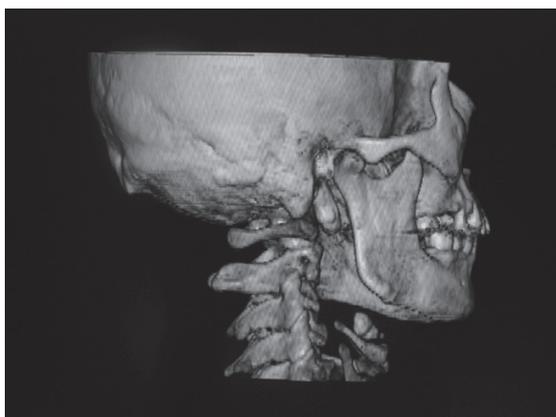


Fig. 6 3D reconstruction showing that most of the stylohyoid chain was removed.

erative day, the symptoms of foreign body sensation and dysphagia were significantly alleviated. Total symptom remission was achieved one month after surgery. The post-surgical 3D reconstruction image is shown in Fig. 6.

Discussion

The normal length of the styloid process varies from 25 to 32 mm in adults, and any structure longer than this is considered abnormally elongated (1). The reported incidence of elongated styloid process or a calcified stylohyoid chain varies from 1.4 to 84.4% in specific populations, of which only 4% result in Eagle's syndrome (4). Eagle's syndrome is more common in women and rarely occurs in individuals younger than 30 years of age (5). In addition to the classic type, a carotid type exists and this is characterized by headache, orbital pain and transient ischemic attacks (1,2).

Diagnostic criteria for Eagle's syndrome include: a) any of the aforementioned classic or carotid-type symptoms, b) a styloid process longer than 25-30 mm on radiographs, c) tenderness on palpation in the tonsillar area or a palpable styloid process, and d) transient symptom amelioration or disappearance under local anesthesia around the tonsillar fossa (5). A diagnosis of Eagle's syndrome can be made if three or more criteria are satisfied. In the present case, the first three criteria were met. The present case was unique in that the styloid process was not only long, but also completely ossified, attached superiorly to the skull base and inferiorly to the hyoid bone, restraining movement of the hyoid bone during swallowing.

MacDonald-Jankowski classified the stylohyoid chain based on four calcification centers: tympanohyal, stylohyal, ceratohyal and hypohyal (4,6). He reviewed 1,662 panoramic radiographs and categorized the patterns into 12 types according to the combinations of calcification centers involved. However, conjoined calcifications involving all four centers were not observed (4). In a series of 1,215 autopsies, Vougiouklakis reported that 0.9% (11/1215) of the cadavers had complete calcification of the stylohyoid ligament. Thickening of the stylohyoid was not detected in any of the completely calcified ligaments (7). These previous large series suggest that a fully ossified and abnormally thickened stylohyoid chain is atypical. To our knowledge, only two other cases of anomalous thickening of the stylohyoid chain have been reported (2,6). The calcification pattern in both cases lay outside the classification described by MacDonald-Jankowski. The present case differed histopathologically from these two isolated case reports in that only compact bone tissue was observed in the latter, whereas compact and cancellous bone as well as red/yellow bone marrow was present in the former.

The stylohyoid chain is derived from the second branchial arches and Reichert's cartilage. In a histological study of 15 embryonic and 35 fetal human specimens,

Rodriguez-Vazquez et al. found that Reichert's cartilage comprised two segments. The styloid segment (cranial part) was attached superiorly to the skull base whereas the hyoid segment (caudal part) was attached inferiorly to the body and greater horn of the hyoid cartilaginous structure. No cartilage had formed between these two segments except for one specimen in which a continuous cartilaginous structure was evident unilaterally (8). This unique variation and the theory that the styloid process continues to ossify slowly into adulthood may explain the complete ossification of the stylohyoid chain (2,6,8-10).

While some scholars have used the term "calcification" of the stylohyoid chain (4), others have deemed this label inappropriate and preferred the term "ossification" (2,3,6,10), as histological examination revealed hyperplasia of the stylohyoid process or metaplasia of the stylohyoid ligament into bone tissue (2,3,6). In the present case, the term "ossification" seemed more appropriate as histopathological examination established the presence of mature bone, rather than calcified tissue.

Eagle's syndrome can be managed medically or surgically. Medical treatment includes the use of NSAIDs, antidepressants and anticonvulsants for pain management. The outcome of transpharyngeal steroidal or local anesthetic injections in the tonsillar area as well as physical therapy has not been remarkable (5). As NSAID was not effective in our patient, a surgical approach was mutually decided upon considering the extent of the bony mass. Surgical interventions can be performed using an intra-oral (via the tonsillar fossa) or extra-oral (from the neck) approach. The extra-oral approach was favored in the present case because it allowed better visualization of the surgical field and the size of the bony mass. Possible disadvantages included a longer operation time, external scar formation and a risk of facial nerve injury. However, the surgery was uneventful and total symptom remission was achieved within a month after surgery.

Conflict of interest

None declared.

References

1. Eagle WW (1948) Elongated styloid process: further observations and a new syndrome. *Arch Otolaryngol* 47, 630-640.
2. Guimarães AS, Pozza DH, de Castro IC, Galdames IC, Palla S (2015) Complete ossification of the stylohyoid chain as cause of Eagle's syndrome: a very rare case report. *J Contemp Dent Pract* 15, 500-505.
3. Camarda AJ, Deschamps C, Forest D (1989) I. Stylohyoid chain ossification: a discussion of etiology. *Oral Surg Oral Med Oral Pathol* 67, 508-514.
4. MacDonald-Jankowski DS (2001) Calcification of the stylohyoid complex in Londoners and Hong Kong Chinese. *Dentomaxillofac Radiol* 30, 35-39.
5. Ceylan A, Köybaşıoğlu A, Celenk F, Yılmaz O, Uslu S (2008) Surgical treatment of elongated styloid process: experience of 61 cases. *Skull Base* 18, 289-295.
6. Kay DJ, Har-El G, Lucente FE (2001) A complete stylohyoid bone with a stylohyoid joint. *Am J Otolaryngol* 22, 358-361.
7. Vougiouklakis T (2006) Overview of the ossified stylohyoid ligament based in more than 1200 forensic autopsies. *J Clin Forensic Med* 13, 268-270.
8. Rodríguez-Vázquez JF, Mérida-Velasco JR, Verdugo-López S, Sánchez-Montesinos I, Mérida-Velasco JA (2006) Morphogenesis of the second pharyngeal arch cartilage (Reichert's cartilage) in human embryos. *J Anat* 208, 179-189.
9. Krmpotić Nemanic J, Vinter I, Ehrenfreund T, Marusić A (2009) Postnatal changes in the styloid process, vagina processus styloidei, and stylomastoid foramen in relation to the function of muscles originating from the styloid process. *Surg Radiol Anat* 31, 343-348.
10. Hardy H, Guichard B, Eliezer M, Choussy O, Péron JM, Trost O (2014) Unilateral complete articulated ossification of the stylohyoid apparatus: case report and review of the literature. *Surg Radiol Anat* 36, 941-945.