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Case report

Stylohyoid complex syndrome: A report of two cases and review of the literature

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ABSTRACT

This article reports two patients with stylohyoid complex syndrome.

Cone beam computed tomography examination of the 37-year-old Caucasian woman with hemifacial pain, radiating to the left ear which intensified in response to opening the mouth revealed the ossified distal part of the left stylohyoid ligament, 23.85 mm long, which had a contact with the lesser horn of the hyoid.

A 43-year-old man with a 5-month history of pain in the anterolateral surface of the neck and temporomandibular joint on the right side when swallowing and yawning had the styloid process elongation with a "bayonet-like" deformity on the same side.

Ossification of the stylohyoid ligament, elongation and bending of the slyloid process, as a reason of the lateral neck and/or facial pain is not commonly suspected in clinical practice where diagnosis is often first made radiologically.

1. Introduction

The stylohyoid complex is composed of several structures such as the styloid process of the temporal bone, the lesser horn of the hyoid bone, and the stylohyoid ligament between these structures (Antić et al., 2016). Stylohyoid ligament is a fibrous remnant of the Reichert's cartilage of the second pharyngeal arch. Its ossification is more common in Caucasians. According to Morrison et al. (2012) more than 9000 case reports have been cited on PubMed since late 20th century. The styloid process and stylohyoid ligament ossification has been classified according to their shape and length as normal, elongated, bent, segmented, pseudoarticulated, and distally ossified and fixed to the lesser horn of the hyoid bone. The last type is the least frequently observed (Petrović et al., 2017).

Calcification of the stylohyoid ligament is often a benign accidental finding (Wu et al., 2018). In 1652, an Italian surgeon named Pietro Marchetti first described clinical symptoms of the intermittent respiratory distress associated with an elongated styloid process. The definitive syndrome, however, was defined by Watt W. Eagle in the late 1940s and early 1950s. Eagle described a number of common clinical features in about 200 patients, each with an elongated styloid process, stylohyoid ligament calcification, or both (Fusco et al., 2012).

By consensus, a styloid process measured more than 30 mm in length is considered to be elongated (Gracco et al., 2017). Based on radiographic data, the prevalence of an elongated styloid process in different populations ranges from 4 to 84.4% (Gokce et al., 2008; Ekici et al., 2013; Zokaris et al., 2019). Among those, it is thought that only a small percentage (from 4% to 10.3%) has symptoms suggestive of the anomaly (Antić et al., 2016).

There is a difference between Eagle syndrome (ES) and stylohyoid complex syndrome. ES describes patients in who elongated, ossified styloid processes develop after injury. Stylohyoid complex syndrome is applied when an elongated styloid process, or stylohyoid chain ossification, or both, develop early in life as an anatomical anomaly with no previous injury, surgery or trauma. Stylohyoid complex syndrome has symptoms similar to those of ES, which include dysphagia, headache, hemifacial, neck and pharyngeal pain, as well as pain when swallowing, speaking and opening the mouth, a sensation of a foreign body in the oropharynx, and pain radiating to the ear, which may intensify in response to rotational movements of the head (Ata-Ali et al., 2017). . Computed tomography (CT), either multidetector or cone beam CT, represents the gold standard for diagnosis of an elongated styloid process (Abdel Razek, 2019; Badhey et al., 2017).

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This case report presents two patients with stylohyoid complex syndrome due to the partial ossification of the stylohyoid ligament and styloid process elongation and bending. The study was approved by the Medical Ethics Committee of the Belarusian State Medical University, Minsk, Belarus.

1.1. Case report

A 37-year-old Caucasian woman presented to the private Minsk medical center for the temporomandibular joint CBCT investigation in March 2019 due to hemifacial pain, radiating to the left ear triggered by opening the mouth. Her medical history was uneventful for any significant trauma or ear-nose-and-throat pathology. Informed consent was obtained from patient for being included in the study.

CBCT scans were performed on Planmeca ProMax 3D Mid (Planmeca Oy, Helsinki, Finland) using face dental program (20 × 17 cm, 400 μm, 90 kV, 10 mA, 14 s). Axial, saggital and corrected sagittal CBCT scans were analyzed using Planmeca Romexis Viewer.

CBCT examination revealed the following findings:

- The ossified fragment of the right stylohyoid ligament was 17.86 mm long. It was located at a distance of 15.09 mm from the lesser horn of the hyoid. The length of the styloid process was 28.13 mm and its apex was located below the level of the mandibular foramen at a distance of 7.55 mm from the upper edge of the ossified ligament (Figure 1A).
- The ossified distal part of the left stylohyoid ligament, 23.85 mm long, which had a contact with the lesser horn of the hyoid. Calcified ligament had the radiographic appearance of a long bone with a thick radio opaque cortex and a central radiolucency that constituted the most of it. The rounded, sclerosed and thickened ends of the lesser horn and the lower end of the ossified ligament indicated the presence of "neoarthrosis", formed probably by the movement of these structures relative to each other. The length of the styloid process of the temporal bone was in the normal range (25.7 mm). Its apex lied at the level of the mandibular foramen at a distance of 22.36 mm from the upper edge of the ossified stylohyoid ligament (Figure 1B).
- From the central part of the back surface of the dorsum of the sellae, linear shadows, 1.26 mm thick, were traced dorsally to the upper borders of the temporal pyramids (Figure 1C, D). These bilateral shadows were defined as dural calcifications.

1.2. Case report

A 43-year-old Caucasian man was referred to the private Minsk medical center with a 5-month history of pain in the right anterolateral region of the neck aggravated by tilting the head to the left and back and radiating to the right half of the face. The patient was also complaining of pain in the throat on the right, sharply triggered by swallowing and yawning, and a foreign body sensation in the throat (VAS (visual analog scale) score = 7). Informed consent was obtained from patient for being included in the study.

There was no previous history of neck trauma. Physical examination revealed tension and pain of the sternocleidomastoid muscle and masticatory muscles on the right, as well as severe pain in the projection of the right stylohyoid ligament. Neck movement was within normal limits.

CBCT scans were performed on Planmeca ProMax 3D Mid (Planmeca Oy, Helsinki, Finland) using temporal bone program (7.5 × 7.5 cm, 200 μm, 90 kV, 12 mA, 12 s). TMJs and styloid processes were analyzed in opened and closed-mouth positions on corrected sagittal CBCT scans using Planmeca Romexis Viewer. The scans showed the bilateral recurrent temporomandibular joint dislocation, with the right and the left condyles being positioned anterior to the apices of articular tubercles by 8.22 mm and 8.03 mm respectively in the open mouth position (Figure 2A, B).

Morphological asymmetry of the styloid processes was also found. Within the field of vision, the length of the right styloid process was 33.01 mm, the left was 35.11 mm. No pathological changes of the left

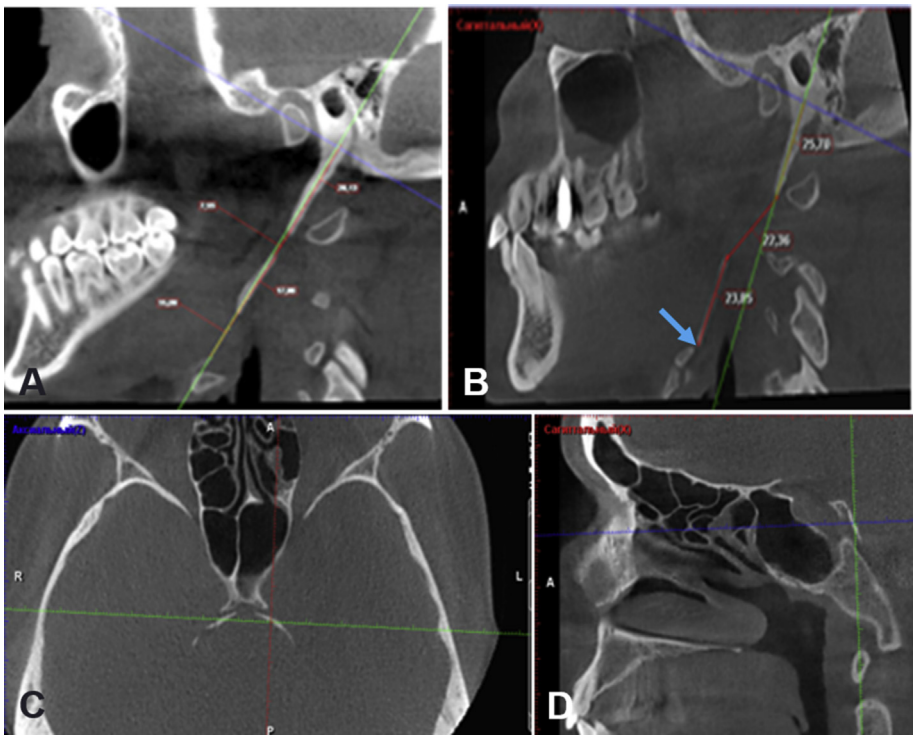

Figure 1. Bilateral stylohyoid ligament ossification. Corrected sagittal views show right (A) and left (B) styloid complex. The blue arrow indicates neoarthrosis between the lesser horn of hyoid and the lower end of the left ossified stylohyoid ligament. Axial (C) and sagittal (D) views illustrate the dural calcifications.

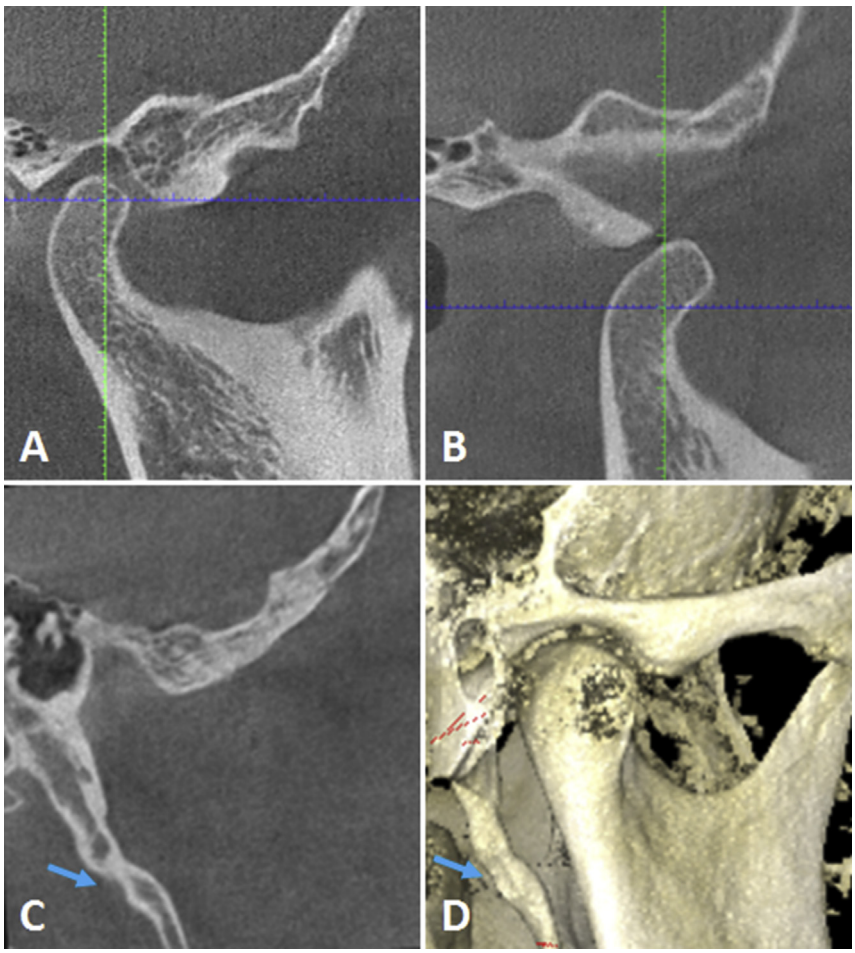

Figure 2. Unilateral elongated and bent styloid process. Corrected sagittal views show (A) condylar location in the closed-mouth position and (B) left condyle positioned anterior to the apex of articular tubercle upon mouth opening. Corrected sagittal limited field-of-view CBCT scan (C) and three-dimensional volume rendering image (D) illustrate the "bayonet-like" deformity (arrow) of the right styloid process.

styloid process were found. The right side had a "bayonet-like" deformity, located at a distance of 19.5 mm from its base (Figure 2C, D).

The patient was diagnosed with ES. The patient received centrally acting muscle relaxant, antiepileptic medication, shock wave therapy and high intensity laser treatment. Neck pain disappeared after 3 weeks of treatment.

2. Discussion

According to Langlais' classification there are three types of elongated and mineralized stylohyoid ligament complexes based on the radiographic appearance – Type I, elongated; Type II, pseudoarticulated; and Type III, segmented. The first case reported in this article can be classified as Type III elongated and mineralized stylohyoid complex, which is represented by two segments with interruption. Pain along the jaw and temple is an unusual but possible consequence of Eagle syndrome. An elongated styloid process should be considered as a possible reason for dull facial pain in the trigeminal distribution zone (Blackett et al., 2012). The stylohyoid complex is commonly known as the styloid process. This is a misnomer since this name ("styloid process") includes only one anatomical structure but no other components (Ledesma-Montes et al., 2018). Since long time ago, it was considered that the stylohyoid complex is an "elongated" structure, meaning that it "grows" during the postnatal life. Ledesma-Montes et al. (2018) believe that this is also a misinterpretation, since physical growth of the styloid complex was never proven. In fact, elongation occurs due to ossification of the stylohyoid ligament, attached to the apex of the styloid process. Thus the stylohyoid ligament seems to be mainly involved in ES or stylohyoid complex syndrome pathophysiology (Zokaris et al., 2019).

The etiology and pathogenesis of ES are still controversial. Several anomalies of development and bone homeostasis have been thought to contribute to the elongation of the styloid process, including the presence of two ossification centers in the styloid process, embryonic mesenchymal conversion to osteoid matrix, osteoarthritic changes, and diseases of calcium-phosphate maintenance (e.g., Paget's disease) (Fusco et al., 2012). For example, the theory of reactive hyperplasia states, that surgical trauma (tonsillectomy) or local chronic irritation cause ossification at the end of the styloid process down the length of the stylohyoid ligament (Bokhari et al., 2019; Hasan, 2018). Gokce et al. (2008) reported heterotopic calcification which led to elongation of the styloid process and thus the presentation of ES in patients with the end-stage renal disease having abnormal calcium, phosphorus, and vitamin D metabolism.

Several pathophysiological mechanisms have been proposed to explain the symptoms of ES: (1) traumatic fracture of the styloid process causing proliferation of granulation tissue, which determines pressure on the surrounding structures; (2) compression of adjacent nerves, the glossopharyngeal, lower branch of the trigeminal or chorda tympani; (3) degenerative and inflammatory changes in the tendinous portion of the stylohyoid insertion, called insertion tendonitis or "pseudostylohyoid syndrome"; (4) irritation of the pharyngeal mucosa by direct compression or post-tonsillectomy scarring (involves cranial nerves V, VII, IX, and X), and (5) impingement of the arterial vessels producing irritation of the sympathetic nerves in the arterial sheath (Scavone et al., 2019).

In the first case presented the styloid process was relatively short. We believe that hemifacial pain, radiating to the ear was associated with arthritis of the joint between the distal end of the left stylohyoid ligament and the hyoid bone, more than with the compression of adjacent nerves.

For many years, it was considered that the stylohyoid ligament "mineralizes" during the postnatal life. However, most likely, the formation of the bone tissue in the ligament is associated with the ossification of Reichert's cartilage remnants. Therefore, in the 1st case reported, only a partial ossification of the ligament has occurred. It resembles the radiographic appearance of a long bone with a thick radiopaque cortex and a central radiolucency. Mineralization (calcification) corresponds to calcium phosphate deposition constituting physiological or pathological structures, which do not have the same radiographic or biomechanical signification as bone (Hardy et al., 2014).

A retrospective study by Sekerci et al. (2015) indicated that the relationship exists between the presence of an arcuate foramen and an elongated styloid process. In the 1st case reported, in addition to the ossified stylohyoid ligament, an ossification of the cranial dura mater near to the sella turcica was found. Dural calcification and ossified stylohyoid ligament may be predictors of the bone mineral density and high serum calcium level.

Facial pain may be associated with the temporomandibular disc displacement (Razek et al., 2014). The second patient with ES showed bilateral habitual temporomandibular joint dislocation but pain in the anterolateral surface of the face and neck was revealed only on the right side were the styloid process elongation and bending were found. A potential cause of the "bayonet-like" deformity of the styloid process was its fracture associated with the sports injury.

In summary, ossification of the stylohyoid ligament, elongation and bending of the styloid process, as a reason for the lateral neck and/or facial pain is not commonly suspected in clinical practice where diagnosis is often based on radiological data.

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The authors declare no conflict of interest.

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