Radiologic Evaluation of the Stylohyoid Syndromes
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Citation

Abstract
The stylohyoid syndromes are a constellation of head and neck pain syndromes that are associated with an elongated styloid process or ossified stylohyoid chain. The reported prevalence of the syndrome varies widely between 1.4% and 30%. The clinical features are subtle and patients are frequently misdiagnosed. The clinician may be guided to the correct diagnosis by history and physical examination, but the condition is confirmed on radiographic investigations. We review the radiologic features associated with the stylohyoid syndromes.

INTRODUCTION
The stylo-hyoid syndromes are a constellation of head and neck pain syndromes associated with an elongated styloid process or ossified stylo-hyoid chain. Patients are often misdiagnosed because the clinical features are subtle. Physicians must be aware of the associated radiologic features in order to correctly make this diagnosis.

DISCUSSION
The styloid process (SP) is a slender projection of bone that arises from the inferior surface of the temporal bone just beneath the external auditory meatus (Fig. 1). The normal length in an adult is considered to be 20mm to 30mm.
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Figure 1
Figure 1: An adult human skull is viewed from lateral (1a) and posterior (1b) aspects. The normal SP (indicated by arrows) is seen originating from the inferior surface of the temporal bone. The SP measures 12mm on the left and 14.5mm on the right. The SP tapers toward its tip that lies in the pharyngeal wall lateral to the tonsillar fossa. Many important neurovascular structures lie near the tip of the SP (Fig. 2). The internal carotid artery, internal jugular vein and cranial nerves X, XI and XII lie on its medial side. Cranial nerve VII runs anterior and medial to the process. Cranial nerve IX exits the jugular foramen and curves in close proximity under the SP.

Figure 2
Figure 2: An adult human head transected in the median plane. Arrows indicate the stylohyoid ligament (SHL) extending from the SP to the hyoid bone. Several important neurovascular structures are concentrated near the tip, including internal carotid artery (B), and glossopharyngeal nerve (C). There are also several ligaments (stylohyoid and stylomandibular) and muscles (styloglossus, stylohyoid and stylopharyngeus) originating from the SP (Fig. 2). They maintain a delicate balance between supra- and infra-hyoid muscles in order to stabilize the hyoid bone during normal oro-pharyngeal functions. The stylohyoid ligament (SHL) arises from the tip of the SP and attaches to the lesser cornu of the hyoid bone. This anatomic arrangement, collectively referred to as the stylohyoid chain, is important in the etiology of the stylohyoid syndromes.

The components of the stylohyoid chain are derived embryologically from the first and second branchial arches in four distinct cartilaginous segments: tympanohyal, stylohyal, ceratohyal and hypohyal segments. These segments are derived from Reichert's cartilages that ossify in 2 parts. The SP develops from the tympanohyal (first) and stylohyal (second) segments that usually fuse at puberty. The lesser cornu of the hyoid bone arises from the hypohyal (fourth) segment. Connecting these two structures, the SHL originates from the ceratohyal (third) segment. It is normally composed of dense fibrous connective tissue in adults but may retain its embryonic cartilage and thus have the potential for ossification. Several theories have been proposed to explain the induction of ossification, including congenital factors, reactive metaplasia after local trauma or calcification and loss of pliability with the ageing process.
Excessive or abnormal ossification of stylohyoid chain components may result in several peculiarities including elongation of the SP (Fig. 3) and thickening of the hyoid bone\textsuperscript{4,5,6,7}. A solid stylohyoid chain results when the entire SHL ossifies, but many variations can occur including incomplete ossification, segmentation, pseudo-articulation and variations in thickness and/or angle\textsuperscript{4,5}. The resultant abnormal stylohyoid chain may then compress or irritate nearby structures and is believed to be responsible for the stylohyoid syndromes\textsuperscript{7,8,9}.

Figure 3
Figure 3: An edentulous adult human skull is viewed from the lateral (3a) and inferior (3b) aspects. Elongated SPs (arrows) are evident and measure 59mm on the left and 51mm on the right. Note the position of the SP in relation to the articulated mandible.

It has been estimated that between 1.4\%\textsuperscript{11} and 30\%\textsuperscript{12} of persons in the general population have radiographic evidence of stylohyoid chain ossification. In 75\% of cases the complex is affected bilaterally\textsuperscript{13}. Most patients are asymptomatic and the abnormal findings are only identified incidentally on plain radiographs or at post-mortem\textsuperscript{4,9,11}.

Only 1\%\textsuperscript{4,5} to 10\%\textsuperscript{14} of persons with an ossified stylohyoid chain will have related symptoms. Symptomatic patients tend to be older than 40 years, but there is no reliable correlation between the symptomatology and the SP length or the extent of stylohyoid chain ossification\textsuperscript{4,5,14}.

Many different names have been coined to describe the presence of head and neck symptoms associated with an elongated stylohyoid chain, including “Eagle's Syndrome”, “Elongated Styloid Process Syndrome”, “Carotid Artery Syndrome”, “Styloid Process Neuralgia”, “Stilalgia”, “Stylohyoid Syndrome” and “Pseudohyoid Syndrome”\textsuperscript{3,4,5,6,7,8,9,10,11}.

Dr Watt Eagle was the first to describe the clinical symptoms from an elongated SP, now commonly referred to as Eagle's syndrome\textsuperscript{10}. He described a group of patients who experienced a pharyngeal foreign body sensation, dysphagia and dull pharyngeal pain radiating to the ear, exacerbated by neck rotation or tongue protrusion\textsuperscript{10,15}. Most of these patients had tonsillectomy several years before, prompting his theory that the resultant scar tissue at the pharyngeal bed incorporated glossopharyngeal nerve\textsuperscript{10,15}. The symptoms may also arise when the scar tissue incorporates the nearby cranial nerves V, VII, X and/or XI\textsuperscript{1,2,14}. These patients tend to have worsened symptoms during physical examination when the physician palpates the tonsillar bed.

A related presentation, commonly called the carotid artery syndrome, occurs in patients who have not had tonsillectomy. It is believed to be due to subtle neck trauma that causes inflammatory changes, excess deposition of granulation tissue or even callus deposition from sub-clinical fractures around the stylohyoid chain\textsuperscript{15,16}. The result is direct mechanical irritation of the sympathetic nerves accompanying the internal and/or external carotid arteries within the carotid sheath\textsuperscript{16}. These patients may experience neck pain that radiates to the eye, ear, mandible, soft palate and nose when the external carotid artery is affected\textsuperscript{1,2,14}. In the case of impingement upon the internal carotid artery, the predominant symptoms are parietal headaches and pain along the distribution of the ophthalmic artery\textsuperscript{1,16}. The pain may be accompanied by hypersalivation, and in rare cases, a change of voice lasting for a few minutes\textsuperscript{16}.

These conditions are often misdiagnosed and many patients are treated for other problems before the correct diagnosis is made\textsuperscript{3,5,14}. Physicians should maintain a high index of suspicion to make this diagnosis in patients with suggestive symptoms. Directed physical examination requires trans-oral palpation of the tonsillar bed that will reproduce the symptoms\textsuperscript{1,2,14}. The symptoms may be relieved by a
diagnostic infiltration of local anaesthetic and/or steroids into the tonsillar bed. Alternatively, the normally impalpable SP may be appreciable at the tonsillar fossa.

Diagnostic confirmation requires visualization of the ossified stylohyoid chain on radiographs. Plain radiography is the commonest initial modality used to investigate these patients. The normal SHL is radiolucent on plain radiographs and a normal SP is not readily visible because the mandible and teeth will overlap it in most views (Fig. 4).

**Figure 4**

Figure 4: Plain anteroposterior (4a) and lateral (4b) radiographs of the adult human skull in Fig. 1. Here the mandible is articulated but the temporomandibular joints are hyper-extended to allow demonstration of the normal SP (arrows).

Although an abnormal stylohyoid chain may be more readily appreciated, it may also be overlooked due to superimposition of the teeth and mandible. When visualized, the SP length should be measured from its base at the temporal bone. A measurement of 3cm is considered to be the threshold of normal by current conventions. It is important to appreciate that position and contour may be as important as length when considering this diagnosis.

The ossified stylohyoid chain typically appears as an elongated slender bony projection that continues from the SP and extends towards, and occasionally appearing continuous with, the lesser cornu of the hyoid bone (Fig 5).

**Figure 5**

Figure 5: Plain lateral view radiographs of the cervical spine in an adult male patient. An ossified stylohyoid chain is present. The components are easily visible and include the styloid process (SP), stylohyoid ligament (SHL) and the hyoid bone (H).

Many variations in length, thickness, contour and segmentation are possible depending on the extent of stylohyoid chain ossification. The complex may be smooth and well corticated (Fig. 6) or it may be bulky with an irregular contour (Fig. 7).
Figure 6
Figure 6: Plain radiographs of an adult human skull in the lateral (6a) and anteroposterior (6b) views. The mandible has been disarticulated to facilitate examination of the elongated SP (indicated by arrows) that appears smooth and well corticated. There is bilateral elongation measuring 55mm on the left and 42mm on the right.

Figure 7
Figure 7: Plain radiographs of an adult human skull in the lateral (7a) and anteroposterior (7b) views, with mandible articulated. The ossified stylohyoid chain (indicated by arrows) appears irregular, poorly corticated and incompletely ossified. There is bilateral elongation measuring 54mm on the left and 68mm on the right.

Segmentation (incomplete ossification) and pseudo-articulations are variants of normal structure and should not be mistaken for a fracture of the process (Fig. 8).
Figure 8

Figure 8: Plain radiographs of an adult human skull in the lateral (8a) and anteroposterior (8b) views. The elongated SP (indicated by arrows) appears smooth and well corticated. There is bilateral elongation measuring 52mm on the left and 38mm on the right. Note that the SP is articulated at its base (A) and there is a pseudo-articulation (P) at the distal extent of the process.

Langlais et al. proposed a classification of these radiographic appearances based on the types of elongation and calcification patterns. Based on morphology, they described the stylohyoid chain appearing as a single continuous elongated SP (Type I: Elongated), a SP that is continuous with the stylohyoid ligament across a single gap (Type II: Pseudo-articulated) or an elongated chain that extends toward the lesser cornu of the hyoid bone in many discontinuous segments (Type III: Segmented). They also described four patterns of stylohyoid chain calcification: Calcified outline; Partially calcified; Nodular type; Completely calcified.

Patients suspected to be symptomatic of the stylohyoid syndromes are commonly investigated with plain radiographs using panoramic, antero-posterior and/or lateral views. Many other views have been described including lateral oblique, exaggerated Towne's projections and submento-vertex radiographs, but their benefits over the traditional views have not been demonstrated.

Panoramic plain radiographs are linearly distorted images that are designed to show the maxilla, mandible and dentition. They are commonly used in dental practice. An abnormal stylohyoid chain may be visible on these views, but there are several limitations to bear in mind. Firstly, the basal part of the stylohyoid complex cannot be properly visualized because there is still superimposition of the mandible and teeth. Additionally, the spatial relationship between the stylo-hyoid chain, mandible, maxilla and hyoid bones becomes inaccurate due to the linear distortion. This does not allow reliable assessment of the length or relations of the stylohyoid complex.

The commoner radiographic views used in medical practice are antero-posterior and lateral views. The length of the entire complex may be better demonstrated on lateral views because there is less superimposition of nearby structures. Supplemental antero-posterior views should always be requested to determine the presence of bilateral involvement and/or lateral deviation. Definitive measurements are not possible on these views because there is still potential distortion and magnification of the complex due to variations in patient position. There is also poor correlation between the radiographic appearances and the clinical scenarios.

Figure 9

Figure 9: Plain radiographs of the cervical spine in an adult male patient from the anteroposterior (9a) and lateral (9b) views. An ossified stylohyoid chain is indicated by arrows. The mandible and teeth overlap the stylohyoid chain in the antero-posterior views. Asymmetric elongation is noted on lateral views.

Computed tomography (CT) scans of the upper cervical region are being used more commonly to supplement diagnosis. Contrast enhanced images usually provide excellent definition of the complex and adjacent soft
Coronal images have been used to evaluate the length of the stylohyoid complex, but they may still be inaccurate because most times the images are not parallel to a laterally deviated complex, leading to under estimation of length. Axial images (Fig. 10) provide information about the location and immediate relations of the stylohyoid chain. They also allow an assessment of the extent of mineralization, thickness and pattern of the styloid process and ossified SHL.

**Figure 10**
Figure 10: Axial slices of CT scan of the upper cervical spine. The lateral mass of the atlas (A) and odontoid peg (OP) are readily visible landmarks. The ossified stylohyoid ligaments are indicated by arrows lateral to the pharynx (P).

Reformatting the raw data obtained from a spiral scanner allows the creation of reconstructed three-dimensional images (Fig. 11) that provide a wealth of information on the mandible, stylohyoid chain, cervical spine and base of skull. The reconstructed images allow more accurate length measurements that are impossible in two dimensional images in coronal or axial planes. The reconstructed images also allow definition of the extension, course, shape and segmentation and it can be used to identify any fractures along the bony elongation or ossified segments.

**Figure 11**
Figure 11: Reconstructed 3-D images of the base of skull and neck from a spiral CT scanner. Components of the ossified stylohyoid chain are visible and include the styloid process (SP), stylohyoid ligament (SHL) and lesser cornu of the hyoid bone (LC).

Currently, 3D spiral CT is the most advanced diagnostic imaging technique to evaluate the stylohyoid chain in spatial geometry, with accurate length measurements. The detailed information about the course and relations of the stylohyoid chain and the relation of important adjacent anatomic structures also allows pre-operative planning thereby reducing the potential for iatrogenic intra-operative injury. A potential limitation of CT is related to poor image quality that may be scanner related or due to image artifacts from motion or nearby metallic appliances. Nevertheless, 3D spiral CT is superior to conventional imaging modalities and many authorities now consider it the diagnostic modality of choice.

The diagnosis has occasionally been made with other imaging modalities such as barium meal or angiography. These modalities do not directly demonstrate the stylohyoid chain and the diagnosis is made by extrapolation. The barium swallow may show an upper oesophageal filling defect reflecting indentation from the stylohyoid chain. Similarly, there may be compression of the external carotid artery on angiography.

**SUMMARY**
Physicians should be aware of the clinical presentation of the stylohyoid syndromes in order to improve diagnostic accuracy. The condition is often confirmed on plain radiography but 3D CT with reconstruction is rapidly becoming the investigative modality of choice.

References
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