A case of pseudarthrosis in Eagle Syndrome and review of the literature

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ABSTRACT

Objectives:
To describe a case of Eagle syndrome with extensive calcification of the stylohyoid ligament and elongated styloid process with the presence of a pseudarthrosis and to provide a review of the prior literature on Eagle syndrome

Study design:
Case report and review of the literature

Methods:
This is a retrospective description of a single case of Eagle syndrome at a single academic institution as well as review of the available literature in PubMed on Eagle syndrome.

Results:
Here, we describe a case of Eagle syndrome in a patient who had significant odynodysphagia due to compression of the lateral pharyngeal wall by a thick and densely calcified stylohyoid and elongated styloid. Due to the extensive calcification, a transcervical excision was performed. Intraoperatively, a pseudarthrosis was noted in the middle of the stylohyoid. A malleable retractor is usually placed underneath the styloid and stylohyoid to protect the pharyngeal wall and carotid artery. However, in this case, the thickness of the stylohyoid and styloid prevented placement of a malleable retractor. Therefore, we disarticulated the pseudarthrosis and were able to complete the excision without any complications. The patient subsequently did well postoperatively with resolution of his odynodysphagia.
Conclusions:
The presence of a pseudarthrosis in Eagle's syndrome should always be noted as disarticulation of the pseudarthrosis can be useful when excising a stylohyoid ligament that is densely calcified.

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INTRODUCTION

W. W. Eagle identified that an elongated styloid process has the potential to cause odynophagia in 1937 (1). A normal styloid process length is usually thought to be under 2.5 cm and once over 3 cm, it is considered elongated (1-4, 6). It is not clear what causes the styloid process to become elongated or the stylohyoid ligament to calcify. Different theories include: trauma, inflammation following tonsillectomy, chronic inflammation, congential, developmental malformations, disorders of calcium-phosphate homeostasis, and metaplasia (1, 2, 6, 8, 9). Up to 30% of the population has been found to have an elongated styloid process, but only a small percentage of people have symptoms, resulting in an incidence of 0.16% (1, 2).

There are two groups of symptoms. The first is the classic pharyngodynia, centered at the tonsillar fossae, which may be accompanied by foreign body sensation, or odynodysphagia (1-13). The second is the stylo-carotid variant, in which carotodynia, otalgia, headaches, syncope, strokes, aneurysms, and carotid dissections may be present (1-13).

Management of symptoms may be conservative (anti-inflammatories, anti-convulsants, psychotropics, anti-depressants), minimally invasive (anesthetic or steroid injections), or surgical (1-13). Surgical treatment is the gold standard for patients with moderate to severe symptoms and can be done via an intra-oral or external approach (1-13). In both cases, the styloid and stylohyoid complex are associated with important structures, as the external carotid artery is lateral and internal carotid artery, internal jugular vein, and the hypoglossal, glossopharyngeal and lingual nerves are medial (1). When the styloid and stylohyoid ligament are densely calcified, this can make both the intraoral and external approach quite challenging as it becomes difficult to see and protect those structures. Here, we describe a case in which the densely calcified styloid and stylohyoid ligament had a pseudarthrosis in the middle of the stylohyoid ligament which utilized for safe removal of the elongated styloid and stylohyoid complex.

METHODS AND MATERIALS

Institutional Review Board exemption was obtained by the State University of New York Upstate Medical University. A retrospective review of the patient's chart was conducted as was a literature search on PubMed.

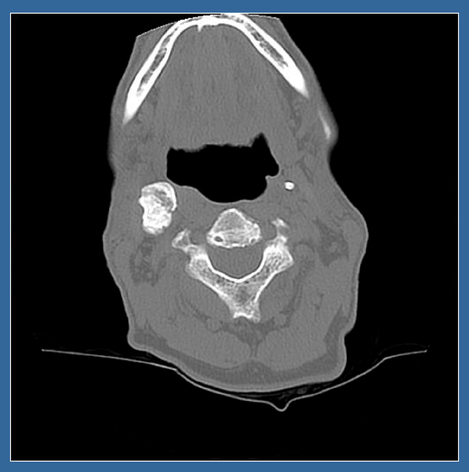

Figure 1. CT neck, axial view showing the calcified and thickened stylohyoid complex.

CASE REPORT

A 71 year old male presented with a 20-year history of cyclic episodes of voice loss followed by dysphagia for solid foods with difficulty initiating swallowing. Several years prior, he had been evaluated and treated by a Gastroenterologist with an upper GI evaluation and multiple dilations, with only periodic relief.

After failing an initial treatment for reflux, a modified barium swallow was then obtained which showed vallecular and piriform sinus pooling with laryngeal penetration of thin liquids, with a large osseous density extending from the right hyoid bone posteriorly and superiorly which appeared to articulate with the right side of C1. On examination he had noticeable a firm mass in right level 2. Subsequent CT imaging showed an ossified right styloid process articulating with the right aspect of a hypertrophied hyoid bone and ossification of the right stylohyoid ligament, consistent with the diagnosis of Eagle's syndrome (figures 1-3).

Given significant dysphagia, resection of the styloid and stylohyoid ligament was discussed with the patient and he elected to proceed. An external approach was performed due to the thickness of the styloid and calcified stylohyoid ligament. During the case, after sliding a malleable to protect the medial neurovascular structures, we made an inferior cut in the calcified stylohyoid ligament with the sagital saw and osteotome. We were unable to slide the malleable underneath the site of our planned superior cut in the styloid process due to the lack of space between the styloid and pharyngeal wall as the styloid was very thick. At this point we noted the pseudarthrosis and incised the pseudarthrosis and then disarticulated it with a freer and osteotome in order to avoid making our incision without any protection over the medial neurovascular structures and lateral pharyngeal wall. After removing the inferior segment of the pseudarthrosis, a size 5 cutting burr was used to drill down the styloid to the level of C2.

The patient did well in the immediate postoperative period, and was discharged on postoperative day 4. Prior to discharge he was advanced to a regular diet and tolerating it well. At his last follow up 6 weeks later, he had no residual odynodysphagia.

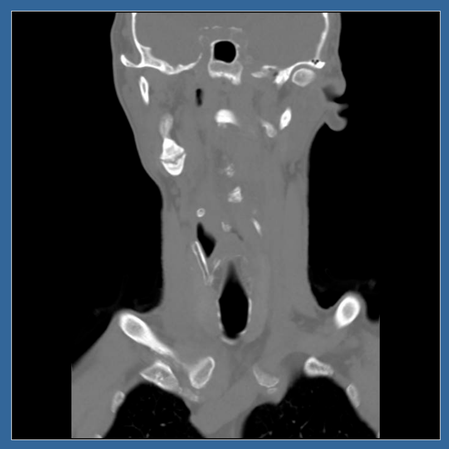

Figure 2. CT neck, coronal view, showing the calcified stylohyoid complex and pseudarthrosis.

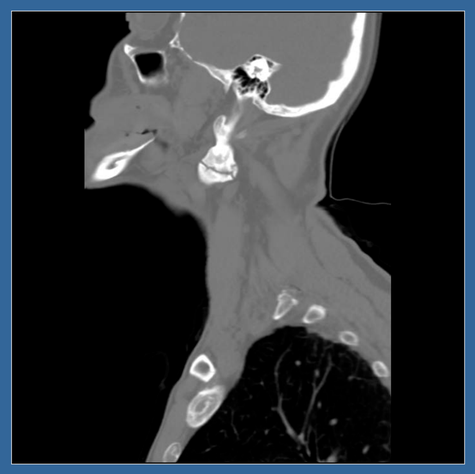

Figure 3. CT neck, sagital view, showing the calcified stylohyoid complex and pseudarthrosis.

DISCUSSION

While the overt presence of a pseudarthrosis in the stylohyoid complex or styloid process has not been previously described before, there have been suggestions in prior literature that this is possible. The stylohyoid ligament can have variable mineralization (9). This suggests a dynamic process of constant remodeling. It has also been suggested that the fracture of an ossified stylohyoid ligament or styloid process and movement causing a non-union is responsible for the pain experienced by these patients (3). While no authors describe a fracture site or fibrous union noted at the time of surgery, micromovement and nonunion leading to the formation of a pseudarthrosis is certainly possible as shown in the above case.

The development styloid and stylohyoid complex may also provide insight as to why a pseudarthrosis may form. The styloid and stylohyoid complex develop from Reichert's cartilage, a component of the second branchial pouch as four separate components (1-3). The four components are: the tympanohyal, stylohyal, ceratohyal, and hypohyal segments (1, 3). The tympanohyal and stylohyal segments form the styloid process (1, 3). The ceratohyal forms the stylohyoid ligament and the hypohyal forms the lesser horn and upper body of the hyoid (1, 3). The ceratohyoid segment is not usually ossified (1). Theories behind Eagle's syndrome rest upon the following three ideas. The first is the persistence of cartilaginous components, which later ossify (3, 6, 7). The second is that the ossification may be partial or complete (3). Third, the areas forming the connections between the each segment are prone to abnormal ossification (3, 7). Incomplete, absent or abnormal ossification may suggest why some patients, like in the above case, could be predisposed to formation of a pseudarthrosis.

While prior studies hint at the possible presence of pseudarthroses, neither has there been any mention of it in operative findings nor of utilizing its presence to remove the styloid or stylohyoid complex. Some patients with Eagle's syndrome have very thick and dense styloid processes and stylohyoid complexes, which prevent visualization of the medial neurovascular structures and can be hard to cut through. Often, a malleable retractor is placed just medial to the stylohyoid complex to protect the neurovascular structures, however this is not always possible in the case of a very thick stylohyoid complex. A pseudarthrosis represents a weak point in the stylohyoid complex, and can be easily disarticulated so that the styloid segment can be removed superiorly and the calcified stylohyoid ligament can be removed inferiorly as illustrated in the above case and minimize risk to the medial neurovascular structures.

CONCLUSIONS

While not previously described in the literature as a tool, the disarticulation of a pseudarthrosis can facilitate safe removal of the stylohyoid complex in cases where it is very densely calcified.

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