
Julianna Rodin, MD; Ryan Rimmer, MD; David Cognetti, MD

Department of Otolaryngology- Head and Neck Surgery, Thomas Jefferson University Hospital, Philadelphia, PA

Introduction

- **Eagle's syndrome** is a rare condition associated with elongation of the styloid process or calcification of the stylohyoid ligament, first described by Dr. Eagle in 1940's.
  - Styloid process typically ranges from 2.5 to 3 cm
  - Associated with wide array of vague and atypical symptoms including: oropharyngeal/cevricofacial pain, dysphagia, dysphonia, globus sensation, headache
  - Often diagnosis of exclusion
  - Treatment usually involves conservative medical management or surgical resection of styloid through transoral or transcervical approach

- **Diffuse Idiopathic Skeletal Hyperostosis (DISH)** is a common but underdiagnosed disorder characterized radiographically by ossification along the anterolateral margins of at least four contiguous vertebrae, and the absence of changes of spondyloarthropathies or degenerative spondylitis.
  - Characterized by back pain, spinal stiffness, and sometimes dysphagia

Background

- 60 year old male was referred to clinic with persistent dysphagia and neck pain for several years. Originally dysphagia was thought to be due to anterior osteophytes which were removed surgically 3 years prior. Neck pain worsened with neck turning, especially to the left.
- Exam: Easily palpable styloid process in left tonsillar fossa. Prominence of posterior pharyngeal wall tissue on videostroboscopy
- Imaging: Massively enlarged unilateral left styloid process (measuring up to 2.2 cm in diameter and 8.8 cm in length), extending to the hyoid bone. Stable extensive calcification in the anterior longitudinal ligament from C2-C7 vertebrae (suggestive of extending to the hyoid bone. Stable extensive calcification in the ligament, first described by Dr. Eagle in 1940's.

Surgery

- Patient taken to operating room for transcervical excision of left styloid process
  - Horizontal incision in natural crease of left upper neck, carried down to subplatysmal plane.
  - Fascia elevated off of sternocleidomastoid muscle and digastric identified and transected to better expose large styloid process. Hypoglossal nerve identified and protected.
  - Drill used to transect styloid bone approximately 2 cm from hyoid. Exposed hyoid was massively thickened and divided near midline to included thickened portion with specimen. This portion was then freed and removed.
  - Proximal portion of styloid then drilled internally via an egg shell technique to protect surrounding structures and then cortex of bone was carefully drilled away. Resection of final portion of styloid bone was well above transverse process of C1.
  - Some branches of external carotid artery were exposed and clipped during procedure. CN IX was identified and preserved. Digastric muscle was reaproximated. Surgical drain was placed and the wound was closed primarily in multilayer fashion.

Postoperative Course

- Surgical drain was removed on postoperative day 1.
- Swallowing issues were noted postoperatively for which patient worked with Speech language pathology.
- At 6 month follow up, the patient no longer had dysphagia, but reported mild neck pain and stiffness.

Discussion and Significance

- The incidence of styloid process elongation or styloidhyoid ligament calcification is unclear. Eagle’s syndrome incidence ranges from 1.4-28%, most of whom are asymptomatic.
- Symptoms are theorized to be related to direct irritation or compression of nearby neurovascular structures including internal jugular vein, internal carotid artery, and cranial nerves V, VII, IX & X.
- Possible correlation between stylohyoid ligament calcification and/or osteophytes of cervical spine but few cases have been described.
  - Levy at al: DISH patients had significantly enlarged styloid processes compared to controls on CT imaging (34.4 mm vs 30.3 mm, p<0.05)
- Diagnosis of Eagle's syndrome or DISH might warrant evaluation for the other
- Few cases (4) have reported a unilaterally enlarged styloid process that is thick and contiguous with the hyoid bone
- Transcervical resection has been favored over transoral for clear visibility and allowing for larger resection. Yet, is associated with longer hospital stay, longer operating time, and scarring may occur.
- No cases have clearly discussed surgical removal techniques of a significantly enlarged styloid process or calcified styloidhyoid ligament in the setting of DISH or styloid removal alone using a drill.

Conclusion

This case report highlights a unique technique for removal of a significantly enlarged and elongated styloid process, and its postoperative course.

References


