Eagle Syndrome: Efficacies of Medical and Surgical Treatment

Kirklin Mitchell, MMS (c)

Faculty Advisor: Erin Wolf, BSN, MMS, PA-C

Department of Medical Science

Abstract

Eagle Syndrome is a rare syndrome caused by elongation of the styloid process of the temporal bone or calcification of the stylohyoid ligament occurring in a very small percentage of the population that can cause a wide range of common symptoms, leading to it being often misdiagnosed. While there are no set standards for treating Eagle Syndrome, both pharmaceutical and surgical options exist. Pharmaceutical options are often ineffective in long term treatment so surgery is usually required for full symptom resolution. Transoral and Transcervical surgeries have similar efficacies but each come with important drawbacks when considering which type of surgery is right for the patient. Novel methods of surgical approach are still actively being developed as well to give surgeons and patients more options for styloid process resection.

Introduction

Overview and History

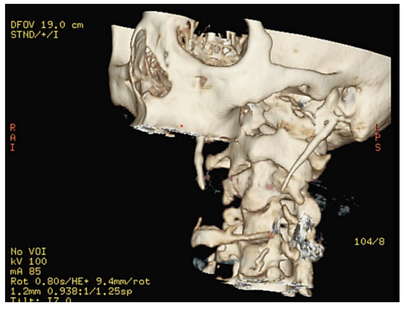
- Cascade of symptoms caused by elongation of the temporal styloid process/calcification of the stylohyoid ligament[1]
- First classified by Dr. Jon Eagle in 1937, but described as early as the 1870s[2]
- Minimal standardized diagnostic criteria, but the styloid is consider elongated if >3cm[2]
- Symptomatic in approximately 0.4% of the population[2]
- Various theories for why elongation happens, but no known cause[3,4,5,6]

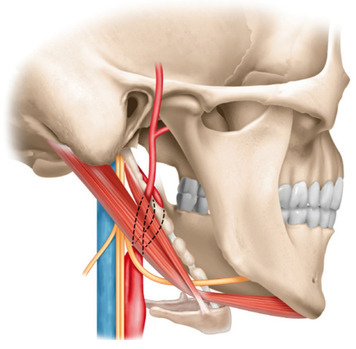
Classification and Symptoms

- Classic Type (Type I)
  - symptoms due to styloid interacting with CNV, CNVII,CNX, CNXII[2]
  - dysphagia, odynophagia, foreign body sensation, hoarseness, facial pain, facial paralysis, otalgia[2]
  - DDx: tonsillitis, pharyngeal cancer, otitis media, herpetic neuralgia, Bell's palsy, facial nerve palsy[7]
- Carotid Type (Type II)
  - Symptoms due to styloid interacting with carotid artery[1]
  - Recurrent TIAs/CVAs, syncope/dizziness/lightheadedness with head turning, visual disturbances, migraines[2]
  - DDx: carotid stenosis, hydrocephalus, brain cancer, TBI, encephalopathy, coagulation disorders[7]

Diagnostics

- Relief of pain with injection of lidocaine into the anterior pillar of the tonsillar fossa is diagnostic[4]
- Xrays, regular CT scans, MRIs to visualize structures and rule out other differential diagnoses[8]
- 3D reconstruction CT scan is gold standard for diagnosis as it allows visualization of the styloid and how it interacts with nearby anatomical structures[8]





Treatment Modalities

Pharmaceutical Management

- Classic Type (Type 1)
  - Initial treatment for pain symptoms with NSAIDs (Ibuprofen)[1]
  - Carbamazepine, an SSRI, or tricyclic antidepressant[1]
  - Combination of Pregabalin + Carbamazepine or Amitriptyline[1]
  - Weekly local fluoroscopy guided injection of Bupivacaine/Triamcinolone[9]
- Carotid Type (Type 2)
  - Warfarin or NOAC therapy for recurrent TIAs and CVAs[2]
    - Antiplatelet therapy with Aspirin + Clopidogrel[2]
  - Topiramate for migraine type symptoms[2]
  - Acetazolamide for increased intracranial pressure symptoms[2]

Transoral Styloid Resection

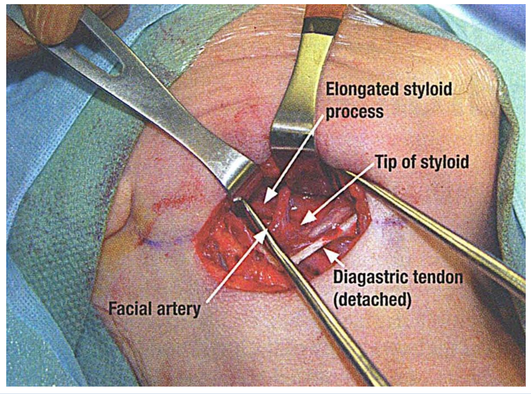
- Most often performed via a transoral robot system[12]
- A quicker surgery that has better cosmetic results[11]
- Little risk of damage to the carotid arteries[11]
- A "dirty" surgery which increases risk of deep tissue infection and risks damage to CNVII and airway compromise[11]

Transcervical Styloid Resection

- Allows for better visualization of the styloid and the surrounding anatomy leading to less risk of injury and infection[2]
- A longer surgery, increasing time under anesthesia, potential for blood loss, and longer post-op admission[2]
- Results in a large cosmetic scar on the anterior neck[13]

Retroauricular Styloid Resection

- Novel, still developing approach to resection by exposing the styloid via dissection starting from behind the ear[10]
- Quick surgery with that results in minimal cosmetic defects and no overnight admission or need for drains[10]
- Risk for damage to surrounding anatomy, particularly the carotids and CNVII, is still present with the added risk of damage to the auricular nerve and the parotid gland[10]



References:

1. Aydin E, Quliyev H, Cinar C, Bozkaya H, and Oran I. Eagle Syndrome Presented with Neurological Symptoms. *Turkish Neurosurgery*. 2016. Doi: 10.5137/1019-5149.JTN.17905-16.6
2. Martin TJ, Friedland DR, Merati AL. Transcervical Resection of the Styloid Process in Eagle Syndrome. *Ear, Nose & Throat Journal*. 2008;87(7):399-401. doi:10.1177/014556130808700714
3. Fitzpatrick TH, Lovin BD, Magister MJ, Waltonen JD, Browne JD, Sullivan CA. Surgical Management of eagle syndrome: A 17-year experience with open and transoral robotic styloidectomy. *American Journal of Otolaryngology*. 2020;41(2):102324. doi:10.1016/j.amjoto.2019.102324
4. Badhey A, Jategaonkar A, Anglin Kovacs AJ, et al. Eagle Syndrome: A comprehensive review. *Clinical Neurology and Neurosurgery*. 2017;159:34-38. doi:10.1016/j.clineuro.2017.04.021
5. Galletta K, Granata F, Longo M, et al. An unusual internal carotid artery compression as a possible cause of Eagle syndrome – a novel hypothesis and an innovative surgical technique. *Surgical Neurology International*. 2019;10:174. doi:10.25259/sni_317_2019
6. Papadiochos I, Papadiochou S, Sarivalasis E-S, Goutzanis L, Petsinis V. Treatment of Eagle syndrome with transcervical approach secondary to a failed intraoral attempt: Surgical Technique and Literature Review. *Journal of Stomatology, Oral and Maxillofacial Surgery*. 2017;118(6):353-358. doi:10.1016/j.jormas.2017.06.017
7. Taheri A, Firouzi-Marani S, Khoshbin M. Nonsurgical treatment of stylohyoid (eagle) syndrome: A case report. *Journal of the Korean Association of Oral and Maxillofacial Surgeons*. 2014;40(5):246. doi:10.5125/jkaoms.2014.40.5.246
8. Kamal A, Nazir R, Usman M, Salam B, Sana F. Eagle syndrome; radiological evaluation and management. *Journal of the Pakistan Medical Association*. 2014;64(11).
9. Maher T, Shankar H. Ultrasound-guided peristyloid steroid injection for Eagle syndrome. *Pain Practice*. 2016;17(4):554-557. doi:10.1111/papr.12497
10. Chen R, Liang F, Han P, Cai Q, Yu S, Huang X. Endoscope-assisted resection of elongated styloid process through a retroauricular incision: A novel surgical approach to Eagle Syndrome. *Journal of Oral and Maxillofacial Surgery*. 2017;75(7):1442-1448. doi:10.1016/j.joms.2017.01.016
11. de Souza Carvalho AC, Filho OM, Garcia IR, de Holanda ME, de Menezes JM. Intraoral approach for surgical treatment of Eagle syndrome. *British Journal of Oral and Maxillofacial Surgery*. 2009;47(2):153-154. doi:10.1016/j.bjoms.2008.07.195
12. Kadakia S, Jategaonkar A, Roche A, Chai RL. Tonsillectomy sparing Transoral Robot assisted styloidectomy. *American Journal of Otolaryngology*. 2018;39(2):238-241. doi:10.1016/j.amjoto.2018.01.007
13. Heim N, Faron A, Martini M, Reich RH. Eagle Syndrome: A follow-up examination of four patients after surgical treatment via cervical approach. *Journal of Craniofacial Surgery*. 2017;28(7):1683-1686. doi:10.1097/scs.0000000000003655
14. Lisan Q, Rubin F, Werner A, Guiquerro S, Bonfils P, Laccourreye O. Management of Stylohyoid Syndrome: A systematic review following Prisma Guidelines. *European Annals of Otorhinolaryngology, Head and Neck Diseases*. 2019;136(4):281-287. doi:10.1016/j.anorl.2019.05.002

Discussion

Pharmaceutical management

- Classic Type
  - 8.04% had satisfactory management with pharmaceutical management in one case study series[14]
- Carotid Type
  - 43.8% cure rate in one case study series[14]
  - 80% Cure rate in one case study series[1]
- The general consensus from multiple research publications is that medical management is more likely to be effective in carotid type

Transoral and Transcervical Styloid Resection

- Surgical management statistics per systematic research review;
  - Cure Rates
    - Transoral Surgery - 73.7%[14]
    - Transcervical surgery - 84.2%[14]
  - Complication Rates
    - Transoral surgery - no reported complications[14]
    - Transcervical surgery - 1.2% rate of facial paresis[14]
- Surgical management statistics per case study analysis
  - Cure Rates
    - Transoral surgery - 89.1%[14]
    - transcervical - 95.8%[14]
  - Complication Rates
    - Transoral surgery - 4.3%[14]
    - Transcervical surgery - 16.3%[14]
- Statistical analysis of outcomes in both reviews determined no statistical significance between the two surgical methods in terms of both cure rates and post-op complications (p = 0.005)[14]

Preauricular styloid resection

- 80.5% of patients had immediate complete resolution of symptoms[10]
- 15% of patients had immediate partial relief from symptoms[10]
- 4.5% had no symptom resolution[10]
- 91.7% of patients considered their post-surgical cosmetic appearance to be excellent[10]
- 3.7% complication rate with complications including facial paresis and ear paresthesia[10]

Conclusion

Eagle Syndrome is a rare and complicated disorder to manage and treat. It can present with a wide variety of symptoms across its two different classifications. It is important to consider Eagle Syndrome as the potential cause of countless neurological symptoms as well as facial and oral pains/sensations that fail to improve with therapeutic measurements for other symptom based differentials. There are limited diagnostic criteria for the syndrome and 3D CT imaging appears to be the gold standard for diagnosis because it allows for visualization of the elongated styloid and the surrounding anatomy. Medical management is often the first line treatment, but seems to mostly be efficacious in the case of specific carotid type presentations. Transoral surgery and transcervical surgery appear to be similarly efficacious with no statistically significant differences between their symptom relief or complication rate. Preauricular surgery is a newer approach to styloid resection that has promising results of being as efficacious as the other surgical methods while seemingly combining many of the benefits of transoral and transcervical approaches into one. More research is needed to confirm that this method does work as well as the small amount of current literature insists.