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ORIGINAL RESEARCH

Heterogeneity in the Clinical Presentation of Eagle's Syndrome

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OBJECTIVE: Eagle's syndrome (ES) or symptomatic elongated styloid process is an uncommon but important cause of chronic head and neck pain. This study reports our experience in the diagnosis and treatment of a series of patients with ES.
STUDY DESIGN: Patient histories, radiographic tests, and operative reports of 3 patients over a 3-month period were prospectively collected.
SETTING: Tertiary referral otolaryngology service.
RESULTS: All patients had resolution of symptoms relating to their elongated styloid processes after surgical resection.
CONCLUSION: Although sometimes clouded by coexisting symptoms, ES can be easily diagnosed based on good history taking and physical examination. If diagnosed appropriately, surgical treatment can be administered promptly.
SIGNIFICANCE: Patients with ES commonly have a long history of chronic pain treated by multiple physicians. Appropriate diagnosis can lead to prompt treatment of this condition.
EBM rating: C-4

Patients with vague head and neck pain symptoms can lead to an extensive differential diagnosis. One easily overlooked but important cause of chronic pain is Eagle's syndrome (ES). Beginning in 1937, Dr. Watt Eagle published a series of articles describing a constellation of symptoms associated with an elongated styloid process. This syndrome that bears his name is associated with two classic presentations. The first, which the otolaryngologist is more familiar with, is throat pain radiating to the ear in a post-tonsillectomy patient. The second, and lesser-known presentation, is constant throbbing pain throughout either the internal or external carotid artery distributions.

We encountered a series of patients with ES whose pain complaints were misdiagnosed for many years. Each patient had classic ES symptoms within their multiple head and neck pain symptoms, yet it was only after a referral to a head and neck surgeon with a high index of suspicion and familiarity with ES did the patients receive appropriate treatment. Herein, we describe their presentations and management. We present these cases to illustrate the heterogeneity of clinical presentation of patients with ES, to point out that each patient with ES invariably will have classic symptoms within their multiple chief complaints, and that with proper diagnosis and prompt surgical management this condition is successfully treated.

University institutional review board approval was granted for the following case reviews.

CASE PRESENTATIONS

A 38-year-old female complained of left-sided throat pain for over 3 years. Intermittent pain radiated down her left shoulder and had intensified over time. She also experienced constant burning sensations over her left neck. She denied dysphagia or voice changes; however, some liquids such as soda would exacerbate her symptoms. Shortly after the throat pain started she noticed left ear pain that radiated upwards and caused severe headaches. She underwent a left tympanostomy and PET tube placement 2 years prior without relief. She denied trauma to the neck, but she was in a car accident 14 years prior to presentation. Past surgeries included a tonsillectomy and a cesarean section.

After her tympanostomy, the patient was referred back to her primary care provider. She also had a history of mold

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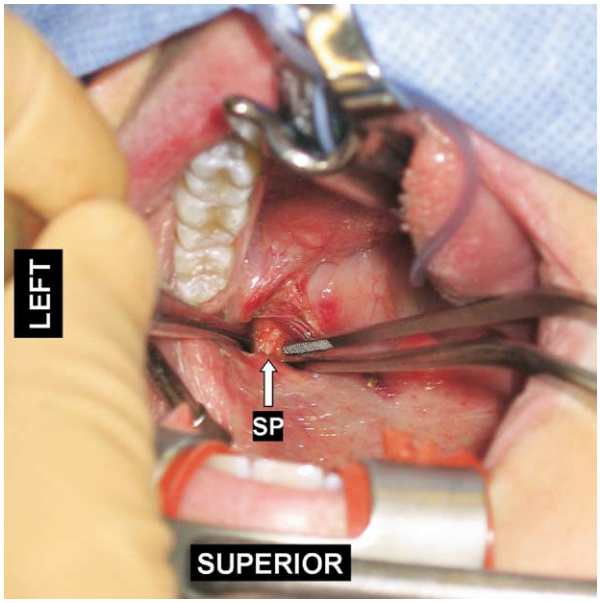

Figure 1 Case #1. Operative view of intraoral approach for resection of the left elongated styloid process (SP).

exposure and was diagnosed with pain related to mold toxemia. Subsequently, she was referred to multiple medical specialists and even initiated legal action against her residential management for the mold exposure. It was only later during a routine visit to her obstetrician that she was referred to our clinic. Oropharyngeal examination was remarkable for prior tonsillectomy scarring and an elongated styloid process that could be palpated intraorally at the left tonsillar fossa. Palpation of the tonsillar fossa elicited some of the painful burning sensation down her left neck. Skull plain x-rays were ordered and revealed a left elongated styloid process. She was taken to the operating room and the left styloid process was resected via an intra-oral approach (Fig 1). Approximately 1.5 cm of the styloid tip was removed. Four months postoperatively she is symptom-free and doing well.

A 45-year-old female had a 6-year history of chronic neck and facial pain. Her pain was bilateral, left greater than right, but either side could flare up independently. The pain was described as severe headaches initiating in the orbit radiating down the posterior neck. Exacerbations would occur when she would rotate her head at a specific angle. She denied any surgical history or history of trauma. Over the years she was under the care of various medical specialists. She received temporomandibular joint (TMJ) treatments, steroid injections, epidural injections, holistic medicine therapies, and radio frequency treatments to the neck. In addition, she was on oral prednisone for pain at the time of presentation.

On physical examination her oropharynx was unremarkable. Palatine tonsils were 1+ bilaterally. Styloid processes could not be palpated intraorally. Skull plain films and a neck MRI imaging were ordered (see Fig 2) and revealed bilateral elongated styloid processes both measuring approximately 3.5 cm. She was taken to the operating room where a bilateral external styloidectomy was performed (see Fig 3). Approximately 2.0 cm of bony fragments were removed from both sides. Four months postoperatively patient is symptom-free and doing well.

A 40-year-old male singer complained of a 12-year history of left-sided throat and neck pain with slight globus sensation and neck tightness. He also noticed a slow onset of hoarseness over that time period. Five years prior, the pain progressed to sharp-stabbing pain in the left throat radiating to the ear, with continued dysphonia. Drinking cold water and rotating his head to the left exacerbated the

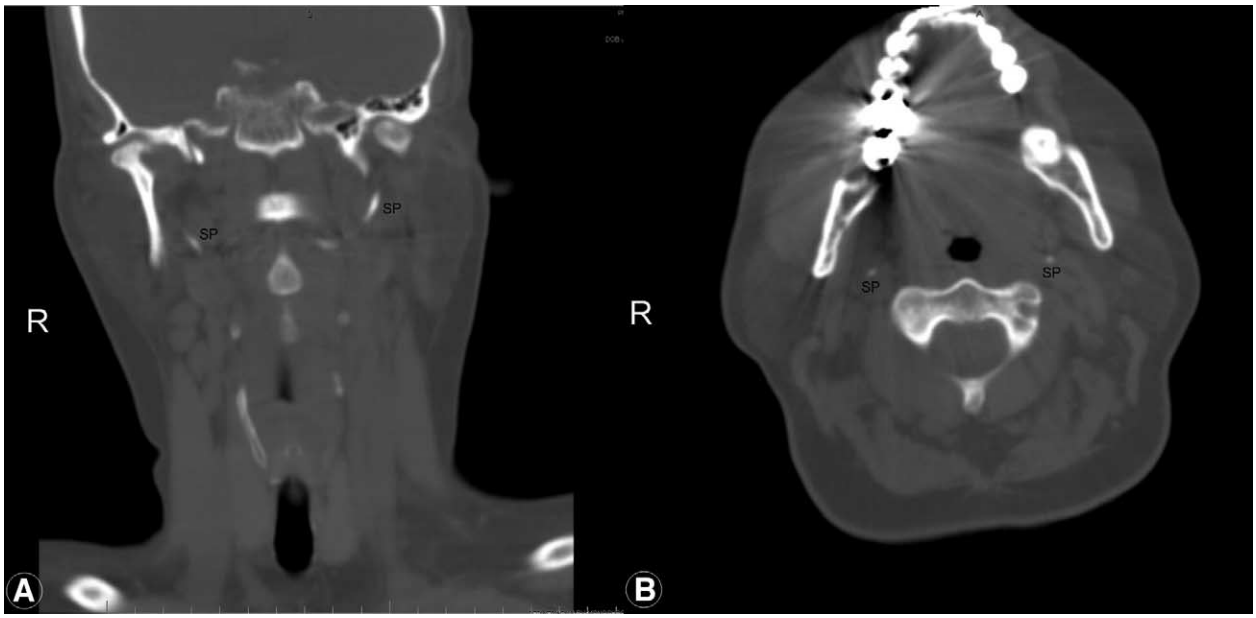

Figure 2 MRI imaging of bilateral elongated styloid processes, (**A**) coronal section and (**B**) Axial section. SP, styloid process.

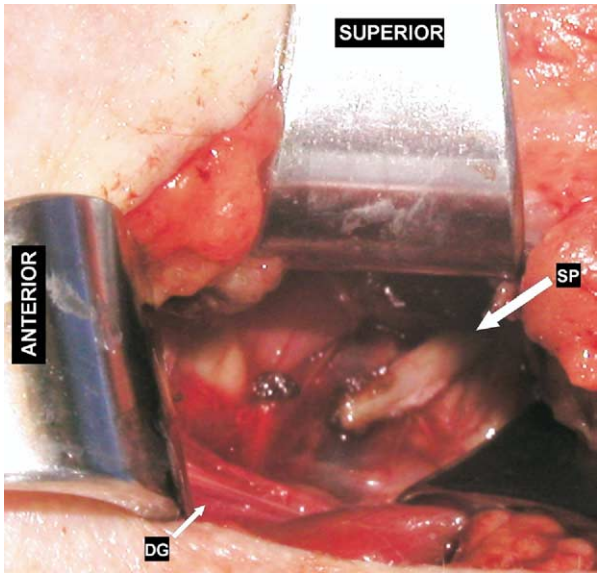

Figure 3 Case #2. Intraoperative view of external approach for resection of elongated styloid process (SP). DG, posterior belly of digastric muscle.

pain. After visiting a number of physicians, the pain was thought to be of tonsillar origin. Three years prior to presentation, many years after the start of symptoms, the patient underwent tonsillectomy without resolution of symptoms. Clouding the clinical picture, bilateral ear pain and mild hearing changes developed. The patient had bilateral tympanostomy tubes placed 1 year prior to presentation, with some relief of ear pain. He denied history of trauma or chronic headaches. Prior operations also included thyroid lobectomy 10 years prior to presentation.

After pain continued post-tonsillectomy, his otolaryngologist palpated the left tonsillar fossa and felt an elongated styloid process. The finding was confirmed with plain films of the skull. The patient was then referred to us. On physical examination palatine tonsils were absent bilaterally with some scar tissue noted in the left fossa. Styloid process was palpated deep to the left tonsillar fossa. Gastroesophageal reflux changes were noted on videolaryngoscopic examination. Plain films were obtained from the referring physician that revealed a left elongated styloid process. He underwent a left intra-oral styloidectomy (see Fig 4). A thick bony segment approximately 1.5 cm in length was removed. Three months postsurgery the patient's neck and throat pain have resolved.

DISCUSSION

The second branchial arch (Reichert's cartilage) gives rise to the styloid process and the lesser cornu of the hyoid bone. The greater cornu is a derivative of the third branchial arch. The styloid process originates from the temporal bone just medial and anterior to the stylomastoid foramen, where the seventh cranial nerve exits. The process runs anteromedially, with rare anatomic variation in course. It is bordered on either side by the internal and external branches of the carotid artery. Three muscles (stylopharyngeus, stylohyoid, and styloglossus) and 2 ligaments (stylohyoid and stylomandibular) originate at the styloid process.

Eagle originally quoted the normal process length at 2.5 cm.[1] Subsequent studies have stated normal lengths from 1.52 cm to 5.0 cm.[2-4] However, these studies look at the average styloid within the investigational population. Clinically, styloid processes greater than 2.5 cm in length should be considered abnormal. The etiology of an elongated styloid process has yet to be elucidated, and the reader is referred to Mortellaro[5] for a good review of current theories.

The typical patient with an elongated styloid process is a female between the ages of 30 and 50 years. There is a 3:1 female predominance in ES.[6] Eagle originally stated a 4% incidence of elongated styloid processes in the general population, and recently this percentage has been corroborated.[6] In addition it is not uncommon to find bilateral elongated processes. However, only 4% of patients with elongated processes will be symptomatic; thus the true incidence of ES is 0.16%.[7]

Diagnosis of Eagle's syndrome is based on a good medical history and physical examination. While there have been many reports of variations in presentation of ES,[5] the syndrome has been classically categorized into 2 groups. The first is found post-tonsillectomy. The pain in this case is usually referred to the ear, especially on swallowing. There can also be a foreign body sensation in the pharynx

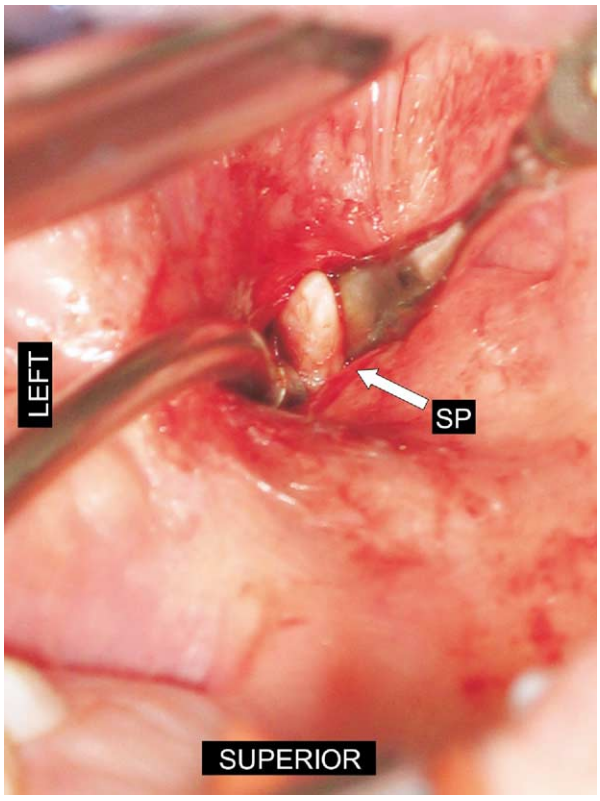

Figure 4 Case #3. Operative view of intraoral approach for resection of a robust left elongated styloid process (SP).

Table 1
Differential diagnosis of chronic pain (noninfectious, noncancerous)[11]

Condition	Diagnostic symptomatology	Other factors
HEAD PAIN		
Migraine headaches	Prodromal irritability, auditory or visual auras, unilateral throbbing pain	Can last 4-72 hours
Cluster headaches	Excruciating unilateral pain usually involving eye, temple, and/or upper jaw	Can last 15 min-2 hours
Chronic tension headaches	Continuous dull, band-like pain located in the bilateral forehead	Must occur more than 15 days/month
Cervicogenic headaches	Trigger points, moderate severity of pain, unilateral localization, causes pain at the occiput	
Paroxysmal hemicrania	Severe unilateral ocular and frontotemporal pain, provoked by specific neck movements	Can occur 10-30 times/day
Eagle's syndrome	Throbbing unilateral or bilateral eye pain radiating back to occiput	Styloid palpation can exacerbate symptoms
FACIAL PAIN		
Temporomandibular joint (TMJ) dysfunction	TMJ pain and tenderness, variable clicking sound	Umbrella term for many TMJ disorders
Myofascial pain dysfunction (MPD) syndrome	Pain and tenderness over the muscles of mastication, limited ability to open the mouth	Can have associated headaches or tinnitus
Trigeminal neuralgia	Sudden severe pain, feels like an electric shock or stab, unilateral cheek or jaw	Can last 20-30 sec, or occur in rapid succession
Glossopharyngeal neuralgia	Recurrent stabbing pains, frequently bilateral at the base of the tongue, tonsils, ear, or jaw	Similar to trigeminal neuralgia
Eagle's syndrome	Throbbing or stabbing pain radiating up the face to the level of the eye	Styloid palpation can exacerbate symptoms
NECK PAIN		
Degenerative disc disease	Dermatome distribution of pain, associated radiculopathy and/or weakness	Can have associated spinal stenosis
Chronic laryngopharyngeal reflux	Repetative throat clearing, hoarseness, heartburn	
Eagle's syndrome	Burning sensation down the neck, globus, variable dysphagia	Styloid palpation or cold liquids can exacerbate pain

with a persistent dull aching sore throat. There are many theories on the pathophysiology of the pain, but Eagle explained this as stimulation of the 5th, 7th, 9th, or 10th cranial nerves. The second group of symptom occurs irrespective of tonsillectomy and involves impingement or irritation of the carotid artery. The styloid process is found in between the internal and external branch of the carotid arteries. If there is a lateral deviation of a process, the resultant constriction and irritation will cause pain in the distribution of that vessel. With lateral deviation and external carotid impingement, pain can radiate up the face to the level of the eye. With medial deviation and internal carotid impingement, pain will begin in the ophthalmic vessel distribution and radiate back to the occiput. The pain is described as a persistent throbbing pain, which can be useful in differentiating other more common sources of headaches.

We found that the classic symptoms of ES are often found with variations in clinical presentation. Our patients had complicated medical histories and other associated symptoms that led physicians to other clinical diagnosis and made true diagnosis challenging. Such distractions in the history include concomitant head or neck pain from a second source, actual ear pathology (instead of simple referred pain), variable tonsillectomy history, and focusing on a single symptom alone. It is remarkable that 2 of our 3 patients complained of otalgia and received tympanostomy tube placement without relief of their symptoms. It is likely many other patients with ES carry the wrong diagnosis, and therefore ES should be kept in the differential diagnosis whenever examining a patient with head and neck pain syndrome. A differential diagnosis of head and neck pain syndromes is listed in Table 1. Two pain syndromes that can present similarly to ES are trigeminal neuralgia and glossopharyngeal neuralgia. Both neuralgias can have similar distribution of pain in the face as ES. However, these neuralgias typically have a history of sudden, severe, electric-type pain triggered by an event such as swallowing, brushing teeth, or touching the cheek. In contrast, a "trigger zone" does not typically elicit ES pain and an elongated styloid process accompanies it.

Physical exam is an essential component in the diagnosis of ES. The tonsillar fossa is digitally palpated. This procedure is usually tolerated well in the clinic, although some patients may gag. Having the patient extend the tongue during palpation may quiet the gag reflex. If a small bony prominence is palpated deep to the tonsillar fossa, by definition the patient has an elongated styloid process.[1] Next, if the pain is reproducible on palpation, either referred to ear, face, or head, then the diagnosis of ES is given. Care should be given not to cause an iatrogenic fracture of the process that will neither alleviate the symptoms nor help a future resection.

With classic presentations of ES, imaging is not necessary for diagnosis. However, presentations can be misleading and physical findings may be missing, and imaging can be very helpful in diagnosing a clinical suspicion of ES. In addition, imaging should be ordered before surgery. Plain skull x-rays are sufficient to find elongated processes. Panorex view will allow better visualization of the styloid process, but posterior-anterior skull films will show any medial or lateral deviation of the tips. CT films of the skull may also be ordered, and they have been described as an adjunct to plain films.[8] If a carotid impingement is suspected, angiography of the neck may be useful to fully visualize the anatomic relationships.

Once diagnosis is confirmed, treatment is surgical excision of the elongated styloid process. Two approaches have been described for removal of a styloid process: internal and external.[9] Eagle introduced the internal approach. A Crowe-Davis retractor is used for visualization into the oral cavity and the styloid tip is palpated. A mucosal incision is made immediately overlying the tip followed by blunt dissection until the styloid process is found (Figs 1 and 4). The muscular attachments are freed with bovie and freer-elevator and the elongated tip resected with ronguers. As much of the tip as can be safely removed should be resected. The pharyngeal incision is then closed in 2 layers using absorbable sutures. The advantages of this approach include no external scar, short surgical time, less surgical trauma, and a relatively straightforward surgical technique. Nevertheless, some studies have supported the external approach.[9,10]

The external approach to the styloid process is the transcervical approach to the parapharyngeal space. A horizontal incision is placed in a skin crease 2 to 3 cm inferior to the lower border of the mandible extending from just behind the submandibular gland to the anterior border of the sternocleidomastoid muscle. Subplastysmal skin flaps are then elevated, taking care to protect the mandibular branch of the facial nerve. The submandibular gland is reflected anteriorly and the anterior edge of the sternocleidomastoid muscle is retracted posteriorly, which allows identification of the posterior belly of the digastric muscle (see Fig 3). The parapharyngeal space is then entered between the mandible and the posterior diagastric muscle. An elongated styloid process can be invariably palpated and traced back towards its origin at the base of skull. As much of the styloid process as can be safely resected is removed. We routinely use intraoperative facial nerve monitoring during any approaches to the parapharyngeal space to guard against inadvertent excessive retraction of the facial nerve. The nerve monitor did not fire intraoperatively in case 2, and the facial nerve function was normal postoperatively. The cervical incision is closed in layers over a quarter-inch penrose drain. The external approach allows a much greater exposure of the styloid process than the internal approach. In addition, the parapharyngeal space is left unexposed to the oral cavity with the theoretic decrease in the risk of infection. The external approach is chosen in a non–post-tonsillectomy patient, or if the majority of the stylohyoid ligament is calcified, because it allows the surgeon to resect the majority of the process and its ligaments.

CONCLUSION

Eagle's syndrome is an uncommon diagnosis that can masquerade as common medical conditions such as headaches, earaches, temporomandibular joint disorders, and nonspecific pain syndromes. The classic presentation can therefore be obscured, but if a physician has Eagle's syndrome in the differential diagnosis it can be easily ruled in or ruled out by physical exam and, if needed, radiographic analysis. Once accurately diagnosed, resection of the elongated styloid process via an appropriate surgical approach appears to effectively treat the condition.

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