Entrainment of the glossopharyngeal nerve in patients with Eagle syndrome: surgical technique and outcomes in a series of 5 patients

Clinical article

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Object. Eagle syndrome is characterized by unilateral pain in the oropharynx, face, and earlobe, and is caused by an elongated styloid process or ossification of the stylohyoid ligament with associated compression of the glossopharyngeal nerve. The pain syndrome may be successfully treated with surgical intervention that involves resection of the styloid process. Although nerve decompression is routinely considered a neurosurgical intervention, Eagle syndrome and its treatment are not sufficiently examined in the neurosurgical literature.

Methods. A review was performed of cases of Eagle syndrome treated in the Department of Neurosurgery at the University of Illinois at Chicago Medical Center over the last 7 years. The clinical characteristics, radiographic imaging, operative indications, procedural details, surgical morbidity, and clinical outcomes were collected and analyzed.

Results. Of the many patients with facial pain treated between 2001 and 2007, 7 were diagnosed with Eagle syndrome, and 5 of these patients underwent resection of the elongated styloid process. There were 4 women and 1 man, ranging in age from 20 to 68 years (mean 43 years). The average duration of disease was 11 years. In all patients, a preoperative workup revealed unilateral or bilateral elongation of the styloid process. All patients underwent resection of the styloid process on the symptomatic side using a lateral transcutaneous approach. There were no surgical complications. All patients experienced pain relief immediately after the operation. At the latest follow-up (average 46 months, range 7 months to 7.5 years) all but 1 patient maintained complete pain relief. In 1 patient, the pain recurred 12 months postoperatively and additional interventions were required.

Conclusions. Eagle syndrome may be considered an entrapment syndrome of the glossopharyngeal nerve. It is a distinct clinical entity that should be considered when evaluating patients referred for glossopharyngeal neuralgia. The authors’ experience indicates that patients with Eagle syndrome may be successfully treated using open resection of the elongated styloid process, which appears to be both safe and effective in terms of long-lasting pain relief. (DOI: 10.3171/2009.1.JNS08485)

KEY WORDS • Eagle syndrome • glossopharyngeal neuralgia • styloid process

EAGLE syndrome is a pain syndrome characterized by unilateral pain in the oropharynx, face, and earlobe, caused by an elongated styloid process or ossification of the stylohyoid ligament. This condition is often considered in the differential diagnosis of glossopharyngeal neuralgia; it is suspected whenever such symptoms are accompanied by documented elongation of the styloid process.1,3–5,7,11 In the past, we suggested that Eagle syndrome be considered an entrapment syndrome of the glossopharyngeal nerve caused by compression of the nerve by bone elements as it travels medial to the styloid process in the neck.11 Treatment of Eagle syndrome involves decompression of the glossopharyngeal nerve by resection of the elongated styloid process, which may be accomplished from either the transpharyngeal or extrapharyngeal direction. In this paper, we present our experience with the extrapharyngeal approach. To the best of our knowledge, this is the first report in the neurosurgical literature of a series of patients who underwent operations for Eagle syndrome.

Methods

With appropriate Institutional Review Board ap-
proval, we performed a retrospective review of patients evaluated for pain in the glossopharyngeal nerve distribution at the University of Illinois at Chicago between 1999 and 2007. Patients diagnosed with Eagle syndrome were identified and their clinical characteristics, imaging findings, operative indications, procedural details, surgical morbidities, and clinical outcomes were assessed.

Results

Of the hundreds of patients with facial pain who visited our clinic over this time frame, 7 were diagnosed with Eagle syndrome, and 5 of these 7 patients underwent resection of the elongated styloid process on the symptomatic side. These patients who underwent operations included 1 man and 4 women, between the ages of 20 and 68 years (Table 1). Two of the 5 patients had previously undergone a tonsillectomy. All patients underwent a preoperative glossopharyngeal nerve block, suggesting a potential benefit to operative intervention; 3 of these patients underwent these nerve blocks at our institution. Each patient had radiographic evidence of a styloid process > 2.5 cm long documented in all cases using 3D CT of the skull base (Figs. 1–3).

The procedure of styloid process resection was well tolerated in all patients with no morbidity. In all cases, elongation of the styloid process was confirmed at surgery. One patient (Case 3) was evaluated at the University of Illinois and had the surgery performed at Massachusetts General Hospital by one of the authors (E.H.E.). Clinical follow-up ranged from 7 months to more than 7 years, with only one recurrence of pain at 12 months (in Case 2). The other 4 patients have experienced complete resolution of their original pain symptoms.

Operative Technique

After the induction of general anesthesia, patients were positioned supine with the head slightly turned to the opposite side. The incision was marked over the projection of the styloid process halfway between the corner of the mandible and the tip of the mastoid process (Fig. 4). Dissection included opening of the fascia, identification and gentle retraction of the parotid gland superiorly, and proceeding anteriorly to the anterior border of the sternocleidomastoid muscle. The facial vein was identified, preserved, and retracted. Blunt dissection ended with palpation of the sharp underlying bone structure. The elongated styloid process was detached from the stylohyoid ligament distally. The muscles attached to the styloid process were separated from the bone surface by gentle subperiosteal dissection. The proximal end of the styloid process was then mobilized and broken off; it was removed as a specimen in 1 or 2 pieces (Fig. 5). Subsequent inspection of the area allowed us to confirm removal of the styloid process and complete elimination of all tethering elements. The integrity and an adequate decompression of the glossopharyngeal nerve could only be assumed because the nerve itself was not visualized directly.

<table>
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<th>Case No.</th>
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<td>5</td>
<td>20, M</td>
<td>5</td>
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Illustrative Cases

Case 1

This 35-year-old woman presented with intractable pain at the angle of her right jaw radiating toward her right ear for 18 months. Her pain was described as spontaneous, intermittent, and sharp. These symptoms were not related to any provocation or facial activity such as speech or swallowing. She had no previous surgical or dental interventions. She had been administered multiple medications previously, including anticonvulsants (carbamazepine, gabapentin, and phenytoin) and an antidepressant (amitriptyline) with minimal or no relief.

On physical examination there was no evidence of a palpable mass, but she experienced tenderness between the mastoid process and the angle of the jaw. A radiographic evaluation revealed normal MR imaging results of the brain and orthopantogram. A CT scan of the patient’s head using 3D reconstruction of the skull base showed an extremely elongated styloid process on the right side consistent with Eagle syndrome (Fig. 1, left). The styloid process measured 4 cm. The CT scan also showed a partially ossified left stylohyoid ligament (Fig. 1, right).

The patient subsequently underwent a glossopharyngeal nerve block to determine if the nerve was involved in the generation of symptoms and in an attempt to predict if her symptoms would improve with surgery. The injection eliminated her pain and soon thereafter she underwent extrapharyngeal decompression of the glossopharyngeal

![Fig. 1. Case 1. Three-dimensional CT scans of the right (left) and left (right) sides of the skull base obtained in a 35-year-old woman. The elongated styloid process on the right side most likely represents ossification of the most cephalad part of the stylohyoid ligament, whereas on the left side this ossification occurs a few millimeters away from the tip of the otherwise normal styloid process.](image-url)
nerve with resection of the elongated styloid process (Fig. 5A). The patient required no further pain medication and has remained pain free for more than 7 years.

**Case 4**

This 43-year-old woman presented with severe pain on the left side of the throat, which radiated to the ear and neck. The symptoms began more than 20 years ago following a tonsillectomy. Her pain started with swallowing, opening of the mouth, cold weather, and episodes of pharyngitis. Her medications included quetiapine, clonazepam, and valproic acid. Neurological examination results were unremarkable except for the pain upon palpation of the inside of her mouth next to the tonsillar fossa. She underwent a thorough workup and was found to have bilaterally elongated styloid processes on CT (Fig. 2). The patient's pain was worse on the left side, and even though she was diagnosed with bilateral Eagle syndrome, we chose to operate on the more symptomatic left side. Surgery was performed using the extrapharyngeal approach. The removed styloid process measured 5 cm in length (Fig. 5C). The patient remains pain-free after 29 months.

**Case 5**

This 20-year-old man had a 5-year history of intermittent pain in both ears and on both sides of his throat. He described a sensation of having something stuck inside his ear. The patient had tried using multiple analgesics and, most recently, oral gabapentin without improvement. He was referred to us for evaluation of possible glossopharyngeal neuralgia. His neurological examination results were unremarkable except for the pain. The maximum point of tenderness was located below the mastoid tip, although no mass was palpated. A CT scan of the skull base with 3D reconstruction showed an elongated styloid process on the right side (Fig. 3, left) and an ossified stylohyoid ligament on the left side (Fig. 3, right). The patient underwent decompression by resection of the styloid process. The specimen was removed in 2 pieces and measured 3.3 cm in total length (Fig. 5D).

**Discussion**

Eagle syndrome is a combination of multiple symptoms including headache, foreign body sensation, and pain...
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in the oropharynx, side of the face, and earlobe. These symptoms may arise due to multiple vascular, glandular, osteocartilaginous, soft tissue, and nervous structures in the neck area. In the differential diagnosis one should include laryngopharyngeal dysesthesia, dental malocclusion, neuralgia of the sphenopalatine ganglion, temporomandibular arthritis, glossopharyngeal and trigeminal neuralgia, chronic tonsillitis or pharyngitis, hyoid bursitis, histamine cephalalgia, esophageal diverticula, temporal arteritis, cervical spondylosis, neoplasms, and migraine headaches.

Eagle syndrome was originally described in 1937 by W.W. Eagle in patients who had undergone tonsillectomy and presented with an enlarged styloid process. The two types of Eagle syndrome are the classic and the carotid artery types. The first type is described as a dull, persistent, and spastic pain in the pharynx, and is observed in patients who have or have not had tonsillectomies, but do have elongated styloid processes. The other type is known as the carotid artery type and is not related to surgery. This type occurs when an elongated styloid process produces symptoms by compressing the internal or external carotid artery when the head is turned, which causes pain in the pharyngeal region due to compression of the perivascular sympathetic fibers.

Although Eagle syndrome is not believed to be a very common disease, the prevalence of styloid process elongation has been reported to be as high as 4%. Most reports of Eagle syndrome are found in the ear, nose, and throat and maxillofacial surgical literature. This syndrome appears to occur more frequently in women and most patients are in their 4th and 5th decades.

Embryologically the stylohyoid apparatus is derived from the Reichert cartilage of the second branchial arch. This cartilage has four portions that develop into the styloid process, stylohyoid ligament, and the lesser horn of the hyoid bone, and the final portion gives rise to most of the hyoid bone. The cause of the elongation is unknown, but multiple theories have been proposed, although none of them has been fully confirmed. The first two theories are related to the overgrowth of the styloid process versus ossification of the stylohyoid ligament via reactive hyperplasia and reactive metaplasia. These are believed to be secondary to injury or surgery. Others have postulated theories such as persistent mesenchymal remnants that produce osseous tissue, and ossification of liga
tments after menopause. Interestingly, although the radiographic features of Eagle syndrome have been repeatedly described in the past, no mention was given to different variants of the radiographic appearance of the styloid process. In our patient series, one can clearly notice the difference between Cases 1, 2, and 5 (Figs. 1, left, and 3, left), in which the styloid process seems to show a transition between what appears to be the normal styloid process and an ossified stylohyoid ligament, and the truly elongated styloid process as noted in Case 4 (Fig. 2, left). Also, in those cases in which the ossification of the ligament was the suspected cause of the Eagle syndrome, imaging of the opposite side did in fact show an incomplete union between the styloid process and the ossified stylohyoid ligament (Figs. 1, right, and 3, right), whereas in the case of genuine idiopathic elongation of the styloid process, such an abnormality was symmetric (Fig. 2, right). The shape of the surgical specimen was also markedly different between those with suspected ligation ossification (Fig. 5A, B, and D) and idiopathic styloid process elongation (Fig. 5C). It remains unclear, however, if this suspected pathogenetic difference has any significance to clinical presentation and/or surgical outcome.

**Conclusions**

Although Eagle syndrome has only been reported once in the neurosurgical literature, patients with its symptoms are often referred to neurosurgeons for operative consideration with a tentative diagnosis of glossopharyngeal neuralgia. Based on our experience, it is evident...
that a comprehensive evaluation of these patients should include a CT scan of the skull base, and some abnormally elongated styloid processes may account for these symptoms. The benefit of a local anesthetic block may support the bone decompression of the glossopharyngeal nerve for achievement of pain relief. We believe that Eagle syndrome is an entrapment of the glossopharyngeal nerve because the symptoms appear to be related to a single nerve involvement as it passes through an anatomical space, and because the surgical decompression results in complete resolution of the symptoms.

Disclaimer
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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References

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