Eagle syndrome: entrapment of the glossopharyngeal nerve?

Case report and review of the literature

KONSTANTIN V. SLAVIN, M.D.

Department of Neurosurgery, University of Illinois at Chicago, Illinois

Eagle syndrome is characterized by unilateral pain in the oropharynx, the side of the face, and the earlobe. It is caused by an elongated styloid process; resection of the elongated process eliminates the pain. Although quite rare, this syndrome is well represented in the oral, ear, nose, and throat surgery literature. In the neurosurgical literature, on the other hand, there is little if any mention of Eagle syndrome.

The author presents a case of a woman who suffered from severe pain in the throat, the side of the face, and the ear. After the diagnosis of Eagle syndrome was made based on radiographic findings and was confirmed using a local anesthetic block, resection of the elongated styloid process was performed, resulting in complete and lasting pain relief.

Eagle syndrome, which is caused by compression of the glossopharyngeal nerve as it passes the elongated styloid process, may be classified as an entrapment syndrome deserving of neurosurgical attention. The goal of this report is to familiarize neurosurgeons with Eagle syndrome and its diagnostic work up and treatment.

Key Words • glossopharyngeal neuralgia • entrapment neuropathy • Eagle syndrome • glossopharyngeal nerve

Case Report

An otherwise healthy 38-year-old woman presented to our neurosurgical clinic for evaluation of possible glossopharyngeal neuralgia. She had been suffering from severe pain in the right side of her face, throat, mandibular angle, and external ear area. The pain had started approximately 3 years before presentation and had gradually been getting worse. The intensity of the pain was graded at 8 or 9 on a scale of 10. There were no pain attacks, no trigger zones, and no association with speech, swallowing, or chewing.

The patient described a chronic intermittent sharp sensation that interfered with normal activities and resulted in partial disability.

Questioning revealed no history of trauma, and no previous surgical or dental interventions. The patient had been seen by a total of seven specialists in various fields, none of whom had come up with a definitive diagnosis or successful treatment. Multiple trials of antiinflammatory drugs, anticonvulsant medications, and antidepressant agents resulted in little or no reduction of the pain. Magnetic resonance imaging and magnetic resonance angiography studies of the brain yielded normal results.

On examination, there was no evidence of focal infection and no masses were palpable in the face or neck or seen in the oropharynx and tonsillar area. Results of neurological examination were normal. There was a mild focal tenderness on palpation between the right mastoid process and the corner of the mandible.

To narrow the large number of choices for the differential diagnosis, we performed a diagnostic radiographic work up that included a panoramic radiograph of the oral cavity and thin-slice CT scans of the skull base region. Although the panoramic film was read as normal, the 3D CT scan (Fig. 1) showed elongation of the styloid process on the right side, with partial ossification of the stylohyoid ligament on the left side.

A local anesthetic block was performed to evaluate whether selective blockade of the glossopharyngeal nerve eliminated the pain and whether the patient could tolerate complete interruption of the glossopharyngeal nerve. For this purpose, 3 ml of an equal mixture of 0.5% bupivacaine and 2% lidocaine solution was injected toward the tip of the styloid process from the lateral direction. Injury to the neighboring vascular structures was avoided by using an in-
Eagle syndrome

![Preoperative 3D CT scans of the skull base revealing the elongated styloid process on the right (symptomatic) side and the ossified styloid ligament on the left side.](image)

Eagle syndrome is a painful condition first described in 1937 in patients who had undergone tonsillectomies. The pain in Eagle syndrome resembles that of glossopharyngeal neuralgia, but in most cases it is more constant and dull in character. There are two distinct types of this syndrome: the classic and the CA type. The first type is characterized by a spastic and persistent pain in the pharynx; it is seen in tonsillectomized patients and sometimes in patients who have not undergone surgery, but who have a styloid process—usually more than 3 to 3.5 cm in length—or ossification of the stylohyoid ligament. The second type presents with pain in the pharyngeal distribution that becomes more prominent during head turning, headaches, and vertigo, and is not related to previous surgery. This type is thought to be caused by pressure exerted by the elongated styloid process on the CA, especially when the head is turned. Although it is part of the differential diagnosis of glossopharyngeal neuralgia, Eagle syndrome is a distinct pathological entity and may be ruled out with clinical examination and panoramic radiography of the mouth or, more recently, with 3D CT reconstruction of the skull base and neck.

There are four theories to explain the symptoms associated with elongation of the styloid process. The first two, the theories of reactive hyperplasia and reactive metaplasia, associate this elongation with either overgrowth of the styloid process itself or ossification of the fibrocartilagenous tissue of the stylohyoid ligament, respectively, usually in response to injury. This phenomenon may explain the occurrence of Eagle syndrome in patients after tonsillectomy, as it was originally described by Eagle. The third theory is that the pain is caused by compression of the glossopharyngeal nerve as it passes a styloid process elongated because of anatomical variability. Because the stylohyoid complex, which consists of the styloid process, stylohyoid ligament, and the lesser horn of the hyoid bone, develops from the second brachial (Reichert) cartilage, segmental ossification of this chain is frequently observed in asymptomatic patients. This theory could explain those cases of Eagle syndrome in which there is no history of tonsillectomy or other injury before the painful syndrome develops. According to some authors, this condition (pain from the elongated styloid process with no history of tonsillectomy) may be called stylohyoid or styloid syndrome to differentiate it from the true Eagle syndrome, but this differentiation does not alter the diagnostic or therapeutic decision-making process. According to the fourth theory, the symptoms may be a result of developmental anomalies of aging. Because normal aging is associated with a decrease in elasticity of soft tissues, tendinosis and some degree of reactive inflammation may cause pain in the glossopharyngeal nerve distribution, mimicking Eagle syndrome. To avoid confusion, it has been suggested that this condition be called pseudostylohyoid syndrome.

Diagnostic work up of Eagle syndrome is based on the clinical presentation. Although an elongated styloid process may cause episodic ticlike pain attacks that are typical of glossopharyngeal neuralgia, most patients present with a constant dull pain or a burning sensation. In some cases, the elongated styloid process can be palpated through the pharyngeal wall in the tonsillar area. Radiographic studies, especially thin-slice CT scans, provide an accurate assessment of the shape, size, and position of the styloid process, helping to determine the diagnosis and guide the subsequent choice of approach for surgical intervention.

Management of Eagle syndrome starts with standard medical treatment. In addition to regular analgesics, anticon-
vulsant, and antidepressant medications, local infiltration of the region of the styloid process with a mixture of steroids and local anesthetic agents may result in lasting pain relief. Patients who fail to respond to multiple medications may require surgery. It is possible to fracture the elongated styloid process manually by transpharyngeal manipulation, but surgical resection of the distal styloid process yields better long-term results. This resection may be performed via the transpharyngeal approach as originally described by Eagle or by using the external route as we did. The choice of approach may be based on the surgeon’s ability to palpate the styloid process in the tonsillar fossa; if the styloid process is not palpable, an external approach may be preferable. In most cases, however, this choice is based on the surgical specialty of the treating physician. Oral surgeons prefer a transpharyngeal styloidectomy, as do some ear, nose, and throat surgeons. Neurosurgeons, who are more familiar with neck dissection for CA exposure or skull base approaches, may feel more comfortable with an external approach to the styloid process.

Interestingly, we found no description of Eagle syndrome in the modern neurosurgical literature. The available reports and clinical series are almost equally distributed between oral surgery and ear, nose, and throat periodicals. In addition, none of the multivolume neurosurgery textbooks mention Eagle syndrome, even as a part of the differential diagnosis for neurosurgical conditions such as glossopharyngeal neuralgia.

In our opinion, Eagle syndrome may be considered to be an entrapment syndrome of the glossopharyngeal nerve. An entrapment neuropathy is defined as a region of localized injury and inflammation in a peripheral nerve that is caused by mechanical irritation from some impinging anatomic structure, which is what happens to the glossopharyngeal nerve due to the elongated styloid process in patients with Eagle syndrome. Because the management of peripheral nerve problems in general, and entrapment syndromes in particular, is within the scope of neurosurgical practice, we think that Eagle syndrome and its diagnosis and treatment should be made more familiar to neurosurgeons.

Acknowledgment

The author expresses his appreciation to Ms. Amy Akers for her help in the preparation of this manuscript.

References


Manuscript received November 26, 2001. Accepted in final form March 29, 2002. The case reported here was presented at the World Congress of Neurosurgery in Sydney, Australia, September 2001. Address reprint requests to: Konstantin V. Slavin, M.D., Department of Neurosurgery, University of Illinois at Chicago, 912 South Wood Street, M/C 799, Chicago, Illinois 60612. email: kslavin@uic.edu.