A Prolonged Time to Diagnosis Due to Misdiagnoses: A Case Report of an Atypical Presentation of Eagle Syndrome

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Patient: Male, 23-year-old
Final Diagnosis: Eagle syndrome
Symptoms: Pain
Medication: —
Clinical Procedure: —
Specialty: Dentistry • Pathology

Objective: Unusual clinical course
Background: Eagle syndrome is an unusual condition in which the styloid process (SP) becomes elongated and causes different clinical symptoms due to pressure on adjacent anatomical structures. The symptoms are non-specific and include severe throat, facial, and ear pain, or headaches. They are usually exacerbated by head rotation, swallowing, yawning, or chewing, but atypical presentations exist. It is a difficult pathology to diagnose and it can take several years before a proper diagnosis is made.

Case Report: This report describes the case of a dental student presenting with an atypical presentation of Eagle syndrome. His styloid processes were 75 mm long and he was affected with severe pain to his throat, the anterior part of his ears, his submandibular area, and his molar teeth. The pain was exacerbated during maximal mouth opening, yawning, mandibular protrusion, and during downward head tilt, but not during the classically described movements of head rotation, swallowing, yawning, or chewing. Due to the absence of the classic symptoms, even with knowledge of the condition and unusual direct access to several oral and maxillofacial specialists, it took 4 years and multiple misdiagnoses to reach the final diagnosis. Following bilateral styloidectomy, all pain subsided.

Conclusions: The clinical presentation of Eagle syndrome varies, and the symptoms are non-specific. This greatly increases the complexity of diagnosing the condition and raises the time to diagnosis and the chances of misdiagnoses. It is, therefore, crucial to recognize the diagnostic tools, applicable imaging, and definitive treatment alternatives to successfully identify and treat patients affected.

Keywords: Diagnosis • Pathology • Diagnostic Errors • Neck Pain • Delayed Diagnosis • Eagle syndrome

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Eagle syndrome is a rare pathology characterized by elongation of the styloid process (SP) in association with a variety of symptoms [1-3]. The SP is a bilateral bony projection originating from the temporal bone, anterior to the stylomastoid foramen [1]. It is connected to the lesser horn of the hyoid bone by the stylohyoid ligament [1]. The SP normally measures between 20 and 30 mm in length [1]. The elongation of SP by calcification of the stylohyoid ligament can cause different clinical symptoms due to pressure on adjacent anatomical structures, including cranial nerves VII, IX, X, XII, the carotid arteries, and the jugular vein [1,4]. Eagle described 2 subtypes of the syndrome: the classic syndrome and the stylocarotid syndrome, depending on which structures are involved [3]. The most frequent symptoms reported are severe throat, facial and/or ear pain, and are usually exacerbated by head rotation, swallowing, yawning, or chewing [1,2]. Other symptoms reported also include foreign-body sensation in the pharynx, odynophagia, pharyngeal paresthesia, dysphagia, dysphonia, vertigo, syncope, and hearing problems [1,2,5]. Rare instances of cerebral ischemia and mandibular compression syndrome, has been identified [4,10]. In this variant, the jugular vein is compressed, resulting in intracranial venous hypertension and, most often, headaches [4,10-12].

The elongation of the SP classically occurs after a traumatic event such as the fracture of the SP, a minor trauma of the neck, or after tonsillectomy [1,2]. The incidence of elongated SP is thought to be between 4% and 15% of the population [3,13,14]. However, a minority of patients with elongated SP are symptomatic [3,14-16].

Diagnosing Eagle syndrome (ES) is complex due to its rarity and non-specific symptoms [5]. Patients usually seek diagnosis and treatment from different health professionals such as general physicians, otolaryngologists, oral surgeons, and dentists [17-19]. Misdiagnoses have been described [17,18], and time to diagnosis as long as 10 and 27 years have been reported [20,21]. It is important to better understand the condition and the symptoms associated to diminish the time to diagnosis of patients affected. In this report, we describe the case of a 19-year-old dental student affected by an atypical presentation of ES and exposing the complexity in reaching a proper diagnosis.

**Case Report**

This case report is unique in that it presents the lengthy diagnosis of a patient with an atypical presentation of ES. Even with unusual direct access to several specialists, it still took years to reach a final diagnosis. As a 19-year-old first-year dental student, the patient suddenly developed pain affecting his throat, the anterior part of his ears, and the bottom side of his mandible. Severe dental pain mimicking irreversible pulpitis was also associated with the right mandibular molars. Other than for his teeth, the pain was bilateral, but the magnitude varied on each side independently. The pain was constant with spontaneous exacerbations and worsened over the years. During the day, the level of pain reported was usually between 3 and 5 on a scale from 0 to 10. However, the pain was more severe in the morning and in the evening, when it usually reached a level of 8 out of 10. Pain was also exacerbated to levels of 7 to 9 during maximal mouth opening, yawning, mandibular protrusion, and during sustained downward head tilt such as when reading and studying. Submandibular palpation around the hyoid bone also worsened the pain. However, contrary to commonly reported symptoms of ES [1,2], the pain was not exacerbated by head rotation, swallowing, or chewing. The patient had no history of tonsillectomy or neck trauma, and he had no other medical condition except for von Willebrand disease type 1.

![Figure 1. Initial radiographs to rule out a dental origin of dental pain. (A) Periapical radiograph. (B) Bitewing radiograph.](image-url)
The patient initially consulted 2 different general dentists who diagnosed incipient and moderate carious lesions and proceeded to place several dental restorations. Because the dental treatments failed to mitigate his toothache, the patient consulted with an oral radiologist, but no treatments were prescribed, as pulp vitality tests and radiographs did not reveal signs of dental pathology (Figure 1). Following a lecture presentation on Eagle syndrome, attended as part of his dental curriculum, the patient consulted an oral pathologist, as some of the clinical description seemed to fit ES. It was concluded that even though his SPs were elongated (Figure 2), due to the absence of the classic symptoms of pain during head rotation, swallowing, or chewing, a diagnosis of ES could not be made. Further consultations were arranged with a physiatrist who ruled out structural neck problems, and an otorhinolaryngologist (ENT) who ruled out Eagle syndrome since there was no exacerbation of symptoms during transpharyngeal palpation. The patient was referred to a specialist in oral medicine to manage the pain pharmacologically using various drugs such as Ibuprofen, Tramadol, Celecoxib, and then Amitriptyline, but they failed to manage his pain.

During his fourth year as a dental student, the patient developed tics of mandibular protrusion and maximal opening and had suicidal thoughts. The patient was therefore referred to a second ENT. A CT scan was taken to evaluate his SPs. They both measured about 75 mm long with a width of about 8 mm (Figure 3). The calcification of the stylohyoid ligament also was found to immobilize the hyoid bone. The diagnosis of an atypical presentation of ES was made. It was therefore decided to proceed with bilateral styloidectomy using a transoral approach. Since the surgery was successful and pain decreased, no post-operative CT scan was taken. A panoramic radiograph (Figure 4) was taken instead as a baseline image for ulterior comparisons. Following surgery, the pain immediately subsided to a level of about 2/10, but moderate dental pain
persisted in the right mandibular molars. All associated pain completely subsided within 5 years, and 15 years later the patient is still completely asymptomatic. Figure 5 shows that no recalcification of the SPs occurred 13 years after the surgery.

**Discussion**

An elongated styloid process associated with cervicofacial pain and other symptoms was first described by W.W. Eagle in 1937 [3]. Eagle divided ES into the classic syndrome and the carotid artery syndrome. The 2 groups exhibit different clinical presentations such as chronic pain in the pharynx, ears, or throat for the classic group and migraines and headaches for the second group [3]. Recently, styloidogenic jugular compression syndrome, a third subtype, was described. In this variant, the jugular vein is impinged between the elongated styloid process and the lateral tubercle of vertebrae C1, resulting in obstruction of the venous outflow, which causes intracranial venous hypertension. Headaches are the most common
symptoms observed in this variant, but blurry vision and dizziness are also common [4,12]. Not all patients with elongated SPs have Eagle syndrome. Only 4-10% of patients with elongated SP are symptomatic [3,14-16]. It was reported that patients could remain asymptomatic even in cases of severe calcification of SP. An asymptomatic patients with over-elongated SP of 80 mm in length and 8 mm in width was documented previously [15]. Therefore, elongation of the SPs is not sufficient to diagnose the condition.

The diagnosis of ES is usually based on history, clinical symptoms, palpation in the tonsillar fossa, and pain relief following local anesthesia in the tonsillar region [1,16,22]. Radiographic assessment including panoramic radiographs and computed tomography (CT) scan presents useful information to diagnose ES [1,16,22]. CT has been recommended as the best imagery to evaluate the position, length, width, and angulation of the SPs, as well as their proximity to adjacent neurovascular structures [13,22,23].

Even though the patient described in this case report had knowledge of ES and privileged access to a wide array of oral and maxillofacial specialists, due to the atypical presentation of his symptoms, it still took 4 years to reach the correct diagnosis. Even with classic presentations of Eagle syndrome, it often takes many years for patients to be diagnosed. A time to diagnosis of 27 years in a symptomatic patient has been reported [20,21]. The patient presented in this report had pain that was extending to his posterior teeth and was therefore misdiagnosed by his dentist. As a result, teeth with doubtful or insipient caries were treated. Similarly, it was previously reported that with incorrect diagnoses of oral, dental or temporomandibular diseases, inappropriate dental treatments such as exodontia were performed in patients with Eagle syndrome [17,18]. We suggest that to avoid these situations, the list of clinical symptoms that may be associated with ES should include pain to mandibular molars in the absence of obvious dental pathology.

Severe chronic pain has been strongly associated with a 2- to 3-fold increased risk of death by suicide. As such, it is imperative that strong efforts made to correctly diagnose patients living with Eagle syndrome. In any situation of moderate to severe throat, facial, mandibular, or ear pain in association with elongation of the SP, we recommend that an anesthetic test in the tonsillar fossa should be conducted; it is a simple and cost-effective test that can help identify atypical presentation of Eagle syndrome.

Eagle syndrome can be treated surgically or non-surgically. Non-surgical approaches include various analgesic medications, steroid injection, antidepressants, and anticonvulsants [22]. Surgical treatment is a more definitive management with long-term symptomatic relief [24]. It can be done via an extraoral or an intra-oral technique [1,2,16]. An external, cervical method offers the best exposure to the entire SPs, with the caveat of a scar [24]. In the present case, since a proper diagnosis was not made for several years, an attempt was made to relieve the pain using various medications, but without success in managing his pain. Once the diagnosis was made, surgical resection using an intra-oral approach ultimately relieved him of most pain, but it took 5 years for all related pain to subside.

Conclusions

The clinical presentation of Eagle syndrome can vary greatly and it includes many non-specific symptoms, which makes it difficult to diagnose and properly manage the condition. Misdiagnoses are common and can lead to an extended time to diagnosis. Therefore, it is crucial to recognize the diagnostic tools, applicable imaging, and definitive treatment alternatives to successfully identify and treat patients affected.

Conflict of Interest

None.
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