Don't look up, a rare but important cause of syncope: Eagle syndrome

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Introduction

Syncope is a common presentation to the emergency department (ED) that often requires a comprehensive workup to rule out life-threatening etiologies. Patients with an unremarkable initial history, electrocardiogram (ECG), and workup are often discharged home with no presumptive cause for their presentation. In this case report, we highlight a rare but important cause of syncope that can present similarly to a vasovagal episode, but could potentially cause serious complications if undiagnosed—Eagle syndrome.

Eagle syndrome was first described by Dr. Watt Eagle in 1937, and is characterized by an elongated styloid process or calcification of the stylohyoid ligament [1]. The most common clinical presentation generates symptoms of facial or throat pain, dysphagia, and a foreign body sensation [1, 2]. However, there are reports of cerebrovascular symptoms caused by compression of the internal carotid artery (ICA) in the setting of Eagle syndrome, leading to transient ischemic attack (TIA), syncope, carotid artery dissection, and stroke [3–5]. Here, we present a case of Eagle syndrome causing recurrent syncope due to ICA compression during yoga, and discuss important diagnostic and management approaches in this rare patient population.

Case report

A 29-year-old previously healthy male presented to the ED after a syncopal episode during a yoga class. He described a 2–3 s loss of consciousness after a hyperextension

movement of his neck, with no associated chest pain or shortness of breath, and he awoke without any signs of confusion, incontinence, tongue biting, or focal neurological deficits. He recalls multiple similar episodes over the past few years that were always triggered by the same hyperextension movement.

At triage, he looked well aside from a few superficial abrasions on the face, and his vitals were blood pressure 113/56 mmHg, heart rate 64 beats per minute, temperature 36.9 °C (98.4° F), respiratory rate 15 breaths per minute, and oxygen saturation 97% on room air. Cardiorespiratory and neurological examinations were normal. His ECG demonstrated no abnormalities. There were no concerns for a pulmonary embolism, or any signs of infection. He had no history of seizures, and he denied any alcohol or drug use.

Due to a history of syncope induced by sudden neck movements, a computed tomography angiography (CTA) of the head and neck was obtained to assess for a possible vertebral artery dissection or subclavian steal syndrome. The scan found no evidence of dissection, hemorrhage, or ischemia, but an “elongated calcified right stylohyoid ligament” was identified, consistent with a diagnosis of Eagle syndrome. A CTA image highlighting this finding is shown in Fig. 1. The patient was referred to an otolaryngologist and after reviewing his file it was felt that he would ultimately require surgical intervention for definitive treatment.

Discussion

Eagle syndrome is a poorly understood cause of syncope, as it is an uncommon presentation of an already rare condition [4]. Additionally, diagnostic criteria for Eagle syndrome remain unclear, as there is lack of consensus regarding the length of styloid process required for diagnosis [4]. Generally, it is accepted that a styloid process length > 30 mm is concerning for Eagle syndrome, however the extent of

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Fig. 1 Coronal view of a computed tomography (CT) scan of a patient demonstrating a calcified and elongated right stylohyoid ligament (arrow)

stylohyoid ligament calcification needed is uncertain [1, 4]. Nonetheless, it is clear from the above case, as well as others reported in the literature, that in addition to the original syndrome described by Eagle causing pain, dysphagia, and a foreign body sensation, Eagle syndrome can also present with cerebrovascular symptoms including syncope [3–5].

It is likely that Eagle syndrome is an under-recognized cause of syncope, especially given that symptoms can be transient and completely self-resolving. Furthermore, patients who present completely asymptomatic to the ED after a syncopal episode are often discharged without any neuroimaging, which is required for diagnosis of Eagle syndrome. This highlights the importance of obtaining a comprehensive history of events preceding a syncopal episode, including asking about any precipitating maneuvers. In the setting of syncope induced by any neck hyperextension, flexion, or cervical rotation, Eagle syndrome should be considered as part of the differential diagnosis. Further history of unilateral facial pain, dysphagia and/or a foreign body sensation in the throat can heighten suspicion for Eagle syndrome, but is not required for diagnosis. When Eagle syndrome is suspected, a CT scan should be obtained, as it is considered the gold standard diagnostic test, and has advantage over plain radiographs in appreciating the relationship between bony structures such as the styloid process, with surrounding neurovascular tissues [4]. CTA can also be useful in assessing for carotid flow, such as in this case report where concerns for dissection were present. There are reports of Eagle syndrome causing both carotid artery dissection and stroke, thus if there is suspicion for Eagle syndrome and the patient presents with any focal neurological deficits or neck pain, CTA would be indicated [3–5].

In the classical presentation of Eagle syndrome where pain is a prominent symptom, conservative management with anti-inflammatory or anticonvulsant medications are often trialed, but in the case where cerebrovascular symptoms are the primary concerns with no pain, definitive management with surgery is often the next step [4]. From the ED, a referral to an appropriate surgeon for consideration of a tonsillostyloidectomy should be initiated and education to avoid aggravating maneuvers, especially any neck adjustments, should be provided to the patient.

Conclusion

Eagle syndrome is characterized by an elongated styloid process or calcified stylohyoid ligament that most commonly presents with facial pain, dysphagia, and/or a foreign body sensation, but more rarely can cause cerebrovascular events such as syncope, TIA, stroke or carotid artery dissection [1–3]. In patients presenting with syncope precipitated by any neck hyperextension, flexion, or cervical rotation, Eagle syndrome should be considered and CT/CTA of the head and neck should be obtained to assess the styloid processes and stylohyoid ligaments, in addition to other neurological pathology. Definitive surgical treatment is required to prevent complications, thus prompt referral from the ED to an appropriate surgeon should be initiated once the diagnosis has been made [4].

Declarations

Conflict of interest Authors have no conflicts of interest to declare and received no funding support for this report.

Informed consent Written consent obtained from participant documented in this case report.

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