Case Report

A RARE CASE OF BILATERAL INTERNAL CAROTID ARTERY DISSECTION IN EAGLE SYNDROME: A MINIMALLY TRANSORAL SURGICAL APPROACH

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ABSTRACT

Eagle's syndrome is a condition associated with the elongation of the styloid process or calcification of the stylohyoid ligament, which is responsible for polymorphic head and neck symptoms, often resulting in delayed diagnosis. Two variants can be distinguished, as initially described by Eagle, the stylohyoid syndrome as a classic type and the stylocarotid syndrome as a vascular type; the latter can involve a dramatic evolution such as internal carotid dissection (ICD). The most commonly proposed curative treatment is styloidectomy, which allows complete resolution of symptoms in the great majority of cases and can be performed via a transoral or a transcervical approach. This paper aims to describe a rare case of bilateral internal carotid artery dissection (ICD) due to Eagle Syndrome and review available literature on the clinical features and treatment. We present the case of a 46-year-old male patient manifesting pharyngeal foreign body sensation, dysphagia, and neck and throat pain exacerbated by head movements, with evidence on CT scans of elongation of the styloid apophyses bilaterally in close proximity to the cervical portion of the internal carotid artery (cICA). Computer tomography angiography showed smooth bilaterally tapering of the mid portion cICA; therefore, ICD due to Eagle syndrome was diagnosed, and transoral styloidectomy was performed. In the literature, there are no cases of bilateral ICD derived from Eagle Syndrome treated with an intraoral approach.

KEYWORDS: *Eagle syndrome, styloid process, parapharyngeal space, internal carotid artery dissection, neck pain*

INTRODUCTION

The styloid process is a cylindrical, slender, needle-like projection of varying lengths, averaging 2 to 3 cm. It projects from the inferior portion of the petrous temporal bone, lying inferior and anterior to the external auditory meatus, anteromedial to the mastoid process, and anterior to the stylomastoid foramen; its tip is located between the internal and external carotid arteries, posterior to the tonsillar fossa. It offers attachment to the stylohyoid ligament, the

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stylomandibular ligament, and the stylohyoid, stylopharyngeus, and styloglossus muscles, grouped under the styloid diaphragm.

Significant vessels and nerves surround the styloid process: the internal jugular vein, internal carotid artery (well as its branches, the lingual artery, facial artery, superficial temporal artery, and the maxillary artery), and glossopharyngeal nerve (cranial nerve, CN, IX), vagus nerve (CN X), accessory nerve (CN XI) and a segment of the sympathetic chain lie medial to the styloid process; the external carotid artery and one of its branches, the occipital artery, hypoglossal nerve (CN XII) and facial nerve (CN VII) run along its lateral side.

Abnormalities in the stylohyoid complex were first identified in animals by Vesalius in 1543, while the first description in humans was published by Marchetti in 1656; the synthomatic elongation of the styloid process instead was first described in 1937 by Eagle, who first used the term ‘‘stylalgia’’ to assess a cervicofacial pain associated with abnormal length of the styloid process (1).

The cut-off value most commonly used to define a long styloid process is 30 mm. However, only 4–10% of patients with elongated processes report pain, and it is often an incidental finding on imaging (2). The clinical pattern could be explained by the peculiar anatomic relationship that SP acquires with many neurovascular structures and some muscles involved in swallowing. They are related to a long styloid process or ossification of the stylohyoid ligament, which leads to compression/impingement of adjacent neurovascular structures (3-4).

Eagle categorized the stylohyoid syndrome as classic and stylocarotid types (1). The former seems facilitated by fibrosis phenomena in the tonsillar fossa following tonsillectomy or minor cervical trauma, and it’s considered a whole intermittent compressive neuropathy involving different branches of cranial nerve (V, VII, IX, X); symptoms most commonly reported are cervicofacial pain with radiated otalgia or temporomandibular joint pain exacerbated by hyperextension, sudden neck movements or swallowing, migraine, hypopharyngeal globus sensation, dysphagia, odynophagia, tinnitus or even dysphonia or trismus (5). The latter is related to impingement on carotid vessels and associated sympathetic nerve plexus, causing referred pain (parietal/periorbital) in those areas supplied by the affected carotid branch,

Horner’s syndrome or even cerebrovascular manifestations such as visual disturbances, presyncope, syncope, dizziness or transient ischemic attacks, and cerebral ischemia due to direct arterial compression, arterial dissection, and thrombo-embolism of ICA (6). Furthermore, Zamboni et al. (7) describe a third type, the jugular variant, related to extrinsic compression of the internal jugular vein between an elongated styloid process and the transverse process of C1, in which symptoms such as headache, numbness, and dizziness may be related to impaired cerebral venous outflow and subsequent endocranic venous hypertension leading even to peri-mesencephalic sub-arachnoid hemorrhage.

A comprehensive anamnesis and accurate physical examination are essential elements in the diagnostic work-up. Pharyngeal palpation, especially of the tonsillar space, reveals an elongated styloid process and generally elicits existing pain. A reliable diagnostic test has been proposed, consisting of injecting 3 mL of 2% lidocaine into the tonsillar fossa, demonstrating Eagle’s syndrome when the pain is relieved. This test is an excellent predictive factor of a good response to styloidectomy (8).

Different imaging modalities have been advocated as diagnostic tools, such as orthopantomography and plain radiographs of the skull in L-L views. Still, the reference one is a CT scan with 1 mm sections and 3D reconstruction, which provides a detailed analysis of the styloid process, the degree of ossification of stylohyoid ligament, and neurovascular anatomical relations. CT-Angiographic can provide further information regarding carotid flow, especially if stroke or dissection is suspected.

Once the diagnosis is confirmed, the treatment of ES can be conservative or surgical. Medical management can be proposed when the patient refuses surgery and can give good but only temporary results and include analgesics such as non-steroidal anti-inflammatory agents, steroid injections, antidepressants, or anticonvulsants (9). Styloidectomy is the surgical treatment of choice for the alleviation of Eagle syndrome symptoms. It involves shortening the styloid process, leaving approximately 1 cm behind to minimize the facial nerve injury risk. If there is calcification of the stylohyoid ligament, then it is appropriate to remove a portion of that ligament as well. The surgical approach can be transoral or transcervical and aims to achieve partial or complete styloid resection.

The purpose of this article is to describe a clinical case that presented the first Eagle syndrome with bilateral internal carotid dissection (ICD) performed with a minimally invasive transoral technique.

CASE REPORT

We describe the case of a 46-year-old male patient addressed to the Maxillofacial Unit of IRCCS Istituto Ortopedico Galeazzi in Milan (Italy), complaining of polymorphic head and neck symptoms, such as pharyngeal foreign

body sensation, dysphagia, neck and throat pain exacerbated by head movements, even though neck pain was the major symptom. He experienced repeated vomiting episodes, which caused his neck to hyperflex. A detailed medical history and complete examination were performed, and the patient was classified as ASA 2. The head and neck clinical examination included careful evaluation of extension, flexion, and rotation neck movements and palpation of the tonsillar fossa, lateral pharyngeal wall, and the area between the mastoid apex and mandibular angle in an attempt to trigger the patient's discomfort. Palpation of the tonsillar fossa triggered the patient's pain.

A computed tomography (CT) and a 3-D reconstruction were performed (Fig. 1). This CT revealed an elongation of the styloid apophyses bilaterally, 4.5cm in length, bilaterally in close proximity to the cervical internal carotid artery (cICA). MRI did not show evidence of acute or prior strokes.

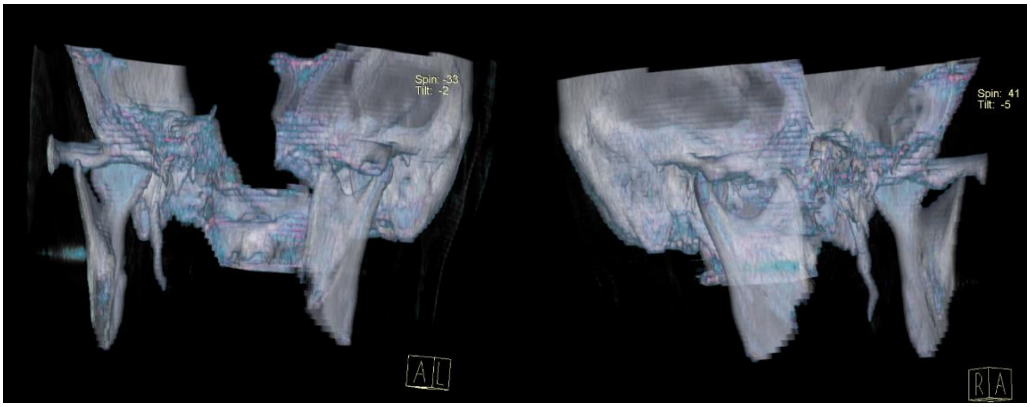

Fig. 1. *3D CT reconstruction revealing elongation of the styloid apophyses bilaterally, about 4.5cm in length.*

Finally, the computer tomography angiography (CTA) showed smooth bilateral tapering of the mid-cervical portion of cICA, 3 cm above its origin, which suggested carotid dissection (Fig 2).

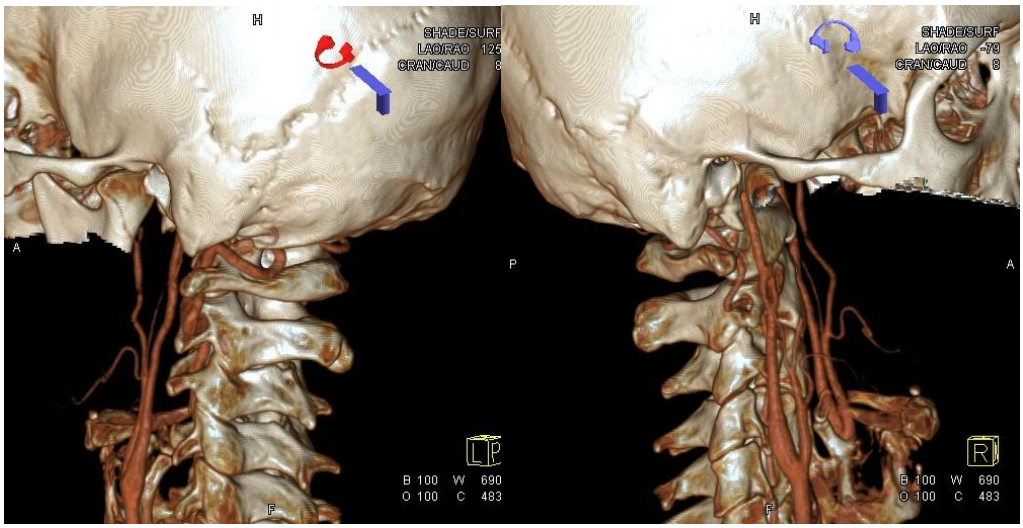

Fig. 2. *VR reconstruction of computer tomography angiography (CTA) showing smooth bilaterally tapering of the mid-cervical portion cICA, 3 cm above its origin, more evident on the right side, suggestive of carotid dissection.*

Dissections due to Eagle syndrome were diagnosed. The surgical approach contemplated a transoral approach using the technique described by Torres et al. (10). The patient underwent general anesthesia and was positioned with the head in extension. McIvor's retractor was used to perform a correct approach from the point of view of visualization of the surgical field, opening the mouth, and pushing the tongue down. Palpation of the region was performed to detect the

tip of the styloid process. After mucosa incision, dissection and exposure of the styloid process were carefully performed until the entire length of the styloid apophysis was clearly visible, preserving tonsils. Then, after its periosteum was incised and dissected, styloid apophysis was exposed, and the enveloping muscles were removed (Fig. 3).

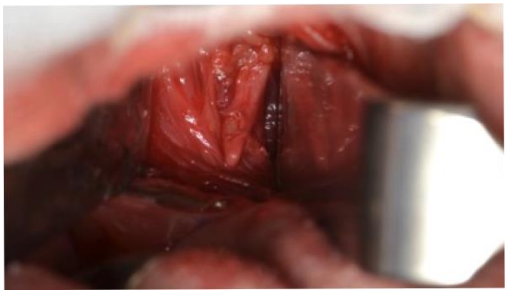

Fig. 3. *Surgical detail: transoral exposure of the left styloid process.*

Then, isolated styloid apophysis, detached stylohyoid ligament from the tip of the SP with Metzembaum scissors, clamped with Kelly hemostatic forceps, was subsequently resected close to its base (Fig. 4).

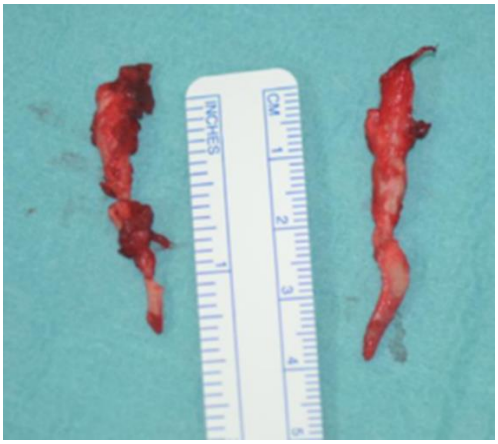

Fig. 4. *Surgical specimen: right and left styloid process resected.*

Bleeding was controlled with bipolar-diathermy forceps, and the mucosal incision closed with 3.0 Vicryl interrupted sutures. The patient was discharged after two days. The only complication during the post-operative period was a cervical subcutaneous emphysema on the right side, which resolved spontaneously.

Approximately four weeks and 3 months after discharge, the patient was evaluated in the clinic. The patient reported a complete resolution of symptoms.

DISCUSSION

Eagle syndrome is a rare pattern of symptoms (0.16% of the general population) due to the conflict with adjacent anatomical structures by an elongated styloid process or a calcified stylohyoid ligament. In 1986, Langlais described a radiographic classification: Type I - pattern represents an uninterrupted elongated SP, Type II, characterized by the SP apparently being jointed to a calcified SL by a single pseudo-articulation, and Type III – consists of interrupted segments (11).

There are no treatment guidelines for Eagle syndrome with carotid dissection (12-15). An indication for endovascular treatment can be extrapolated from the NASCET trial—Dissection causing greater than 70% stenosis. Three-dimensional reformatting of CT imaging is paramount in diagnosing stylocarotid syndrome (16). Some authors have proposed that the only definitive treatment for any symptomatic variant of Eagle syndrome is styloidectomy (17-18).

Two surgical approaches for treating Eagle's syndrome are described in the literature: cervical and intraoral. The cervical approach provides a vertical skin incision of 4 cm in length performed from the posterior border of the mandibular

angle along with the anterior edge of the sternocleidomastoid muscle, SCM. Once the platysma is dissected, the investing layer of deep cervical fascia is excised. The head of the submandibular gland is mobilized superiorly, while the SCM and posterior digastric muscle belly are retracted posteriorly to expose the styloid process (19). The advantages of the cervical approach are favorable surgical exposure and a low risk of infection. Possible complications are facial nerve damage and an unaesthetic cervical scar. The advantages of the intraoral approach include a small incision, limited dissection, shorter operative time, absence of drainage, and a short stay in the hospital. Disadvantages are limited surgical exposure, higher risk of infection, and subcutaneous emphysema.

In the literature, there are no cases of bilateral ICD derived from Eagle Syndrome that were treated with an intraoral approach. There is not yet a validated surgical protocol for these cases, and the intraoral approach has to be considered a convenient choice (20-22).

Disclosure

The authors have no financial interest to declare in relation to the content of this article.

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