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Surgical management of stylocarotid Eagle syndrome in a patient with bilateral internal carotid artery dissection: illustrative case

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BACKGROUND Eagle syndrome is characterized by an elongated styloid process, which can cause acute neurological symptoms when the projection impinges on local structures. One method by which Eagle syndrome can cause acute stroke is via internal carotid artery dissection.

OBSERVATIONS A patient presented with acute aphasia and right-arm weakness. Imaging revealed a left internal carotid artery dissection, which was treated with stenting. Three years later, the patient presented with left-sided weakness, and imaging revealed a new right internal carotid artery dissection. Closer review of the patient's imaging revealed bilateral elongated styloid processes. The patient subsequently underwent staged bilateral styloidectomy and returned to his prior baseline postoperatively.

LESSONS This case report describes a patient with Eagle syndrome who had two internal carotid artery dissections separated by several years. A literature review revealed that styloidectomy is well tolerated in patients with carotid dissection due to Eagle syndrome. Patients with carotid dissection due to Eagle syndrome remain at risk for contralateral dissection, and prophylactic contralateral styloidectomy should be considered.

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KEYWORDS Eagle syndrome; styloidectomy; internal carotid artery dissection

Eagle syndrome is characterized by an elongated styloid process, which causes cervical pain, odynophagia, or acute neurological symptoms when the projection impinges on local structures.[1,2] Approximately 4% of the population has an elongated styloid process, generally defined as longer than 25–30 mm, and it has been reported to cause symptoms in 4%–10% of those affected.[1–7] The most severe form of Eagle syndrome is the stylocarotid form, in which the styloid process contacts the internal or external carotid arteries, potentially causing a transient ischemic attack (TIA) or acute stroke.[8]

Stylocarotid Eagle syndrome can cause acute stroke via internal carotid artery (ICA) dissection.[9,10] Cervical artery dissection is a common cause of stroke in younger adults, with a mean age of 45 years at occurrence, and extracranial ICA dissection is the most common form of cervical artery dissection.[11] There have been several reported cases of TIA or acute stroke due to ICA dissection from stylocarotid Eagle syndrome.[1,3,7,12–17] These cases have reported a range of treatment approaches, from medical management to stent placement to resection of the styloid process, or styloidectomy.[14,17–20] Bilateral carotid occlusions as a result of elongated styloid processes are exceptionally rare, and there is a dearth of literature on this topic.

We report a case of a patient who developed acute stroke due to ICA occlusion with tandem middle cerebral artery occlusion, which was emergently treated with thrombectomy and carotid stenting. A dissection was noted immediately adjacent to the styloid process. The patient was noted to have bilateral elongated styloid processes, and he ultimately recovered completely. Years later, he presented again with sudden occlusion of the contralateral ICA with resultant stroke. He again underwent carotid stenting for symptomatic carotid occlusion and again made a favorable recovery. He then underwent staged bilateral styloidectomy. We also review the literature to describe the outcomes of styloidectomy for the treatment of ICA dissection due to Eagle syndrome.

ABBREVIATIONS CTA = computed tomography angiography; ICA = internal carotid artery; TIA = transient ischemic attack.

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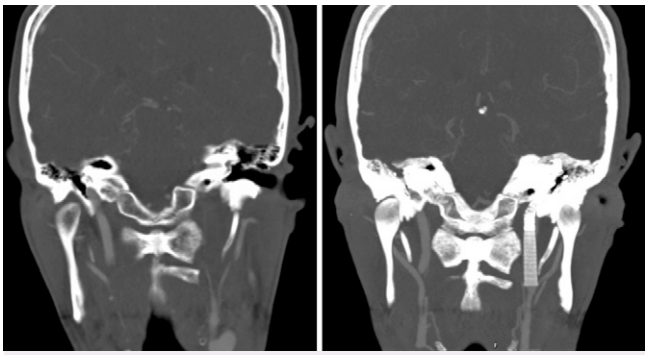

FIG. 1. Left: Preoperative CTA with left ICA occlusion. **Right:** CTA following left ICA stenting, showing proximity of the left styloid process to the ICA.

Illustrative Case

A 45-year-old male with no past medical history presented to the emergency department following the acute onset of aphasia and right-arm weakness. He had a National Institutes of Health Stroke Scale score of 7 upon arrival. Computed tomography of the head revealed no evidence of hemorrhage, and computed tomography angiography (CTA) of the head and neck demonstrated a left cervical ICA and intracranial middle cerebral artery occlusion. Tissue plasminogen activator was administered, followed by mechanical thrombectomy. During angiography, the cause of the cervical left ICA occlusion was identified as dissection, which required stenting to recanalize. The dissection site was noted to be immediately adjacent to an elongated styloid process, which had direct contact with the ICA on CTA (Fig. 1). He had no personal or family history of connective tissue disease and had no inciting head trauma. He was discharged 3 days later with deficits of mild aphasia and dysarthria and placed on 90 days of dual antiplatelet therapy with aspirin and clopidogrel. He returned to his prestroke baseline by his 1-year follow-up.

Three years later, the patient presented to the emergency department following two episodes of transient left-hand and left facial numbness that resolved after several minutes. He reported no recent trauma, neck manipulation, or other risk factors for blunt head and neck injury. He reported compliance with his antiplatelet therapy of aspirin. CTA revealed a new, complete right cervical ICA occlusion and widely patent left ICA stent. A small acute infarct was found in the right posterior frontal lobe on magnetic resonance imaging. Cerebral angiography revealed a right ICA dissection with complete occlusion of the vessel. Because the patient's symptoms completely recovered at the time of thrombectomy, thrombectomy and stent placement were not immediately performed. Review of the patient's imaging demonstrated bilateral elongated styloid processes (35 mm on the left, 25 mm on the right), both of which directly contacted their respective ipsilateral ICAs, leading to a diagnosis of ICA dissection and occlusion due to Eagle syndrome. The patient's symptoms ultimately worsened with intermittent episodes of hemiplegia and right gaze deviation, and imaging showed ipsilateral hypoperfusion; therefore, he underwent right carotid artery stenting for his dissection (Fig. 2). He was discharged after 3 days with plans for 6 months of aspirin and clopidogrel therapy, with a styloidectomy planned as the definitive treatment for Eagle syndrome following the cessation of clopidogrel therapy.

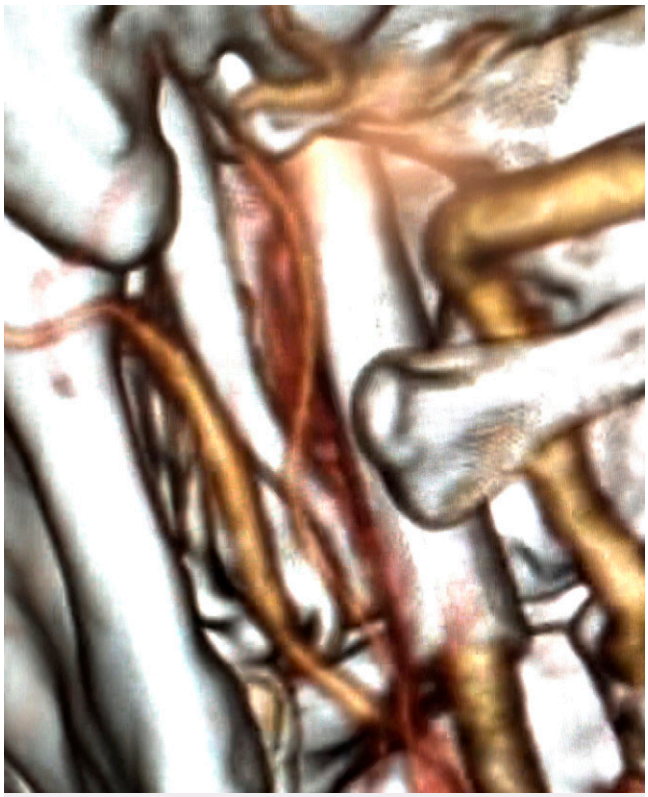

FIG. 2. Three-dimensional reconstruction of CTA, showing proximity of the right styloid process and stented right ICA.

The left styloid was removed transcervically 6 months following discharge, and the right styloid was removed in a similar fashion 2 months later (Fig. 3). Postoperative CTA revealed widely patent carotid stents with no residual styloid processes. At his 4-month follow-up visit, the patient reported resolution of his neurological symptoms and a complete return to his baseline.

Patient Informed Consent

The necessary patient informed consent was obtained in this study.

Discussion

Observations

We describe a case of bilateral ICA dissections due to elongated styloid processes, whose presentations were separated temporally by several years. The patient had two separate presentations 3 years apart with symptoms due to ICA dissection and subsequent occlusion, both of which required endovascular stent placement. Following the patient's second ICA dissection, he underwent an uncomplicated staged bilateral styloidectomy; he has since completely returned to his neurological baseline. This report highlights the need for early identification of patients with ICA dissection related to Eagle syndrome, particularly in patients without traditional stroke risk factors and in those who have dissections at the site of styloid process contact. This is particularly important because these patients may be at an elevated risk for contralateral injury, because both styloid processes are frequently elongated and early treatment may prevent a second neurological insult.

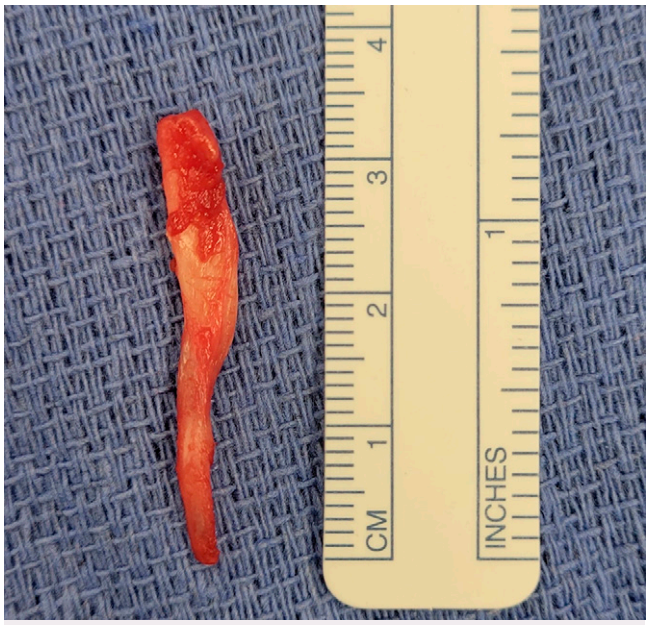

FIG. 3. Transected left styloid process following transcervical styloidectomy.

In 1937, Eagle first described his eponymous syndrome and described styloidectomy as the definitive surgical approach.[21] Two surgical approaches to styloidectomy, intraoral and external (or transcervical), have since been described. The intraoral approach begins with palpation of the styloid process within the tonsillar fossa, followed by an incision made in the oral mucosa.[9,22] The external approach begins with an incision in the neck below the angle of the mandible.[23] The external approach is generally considered more advantageous, because it allows better exposure of the styloid process and the adjacent anatomy.[3,23,24] There are no randomized trials comparing the two approaches.[9] One single-center study described 61 patients with Eagle syndrome who had undergone the transcervical approach and revealed that 57 (93%) of 61 patients were asymptomatic following styloidectomy.[23] However, none of the included patients had an ICA dissection.

Cervical artery dissection, although a common form of stroke in younger patients, affects patients across their lifespan, with the prevalence of hospitalization from dissection-related stroke increasing with age.[11,25] It is often associated with mechanical trauma, although less so in older adults, and can be associated with connective tissue diseases.[26,27] There are several prior reports of ICA dissection due to Eagle syndrome in the literature, with treatments ranging from medical management to ICA stenting to styloidectomy.[3,9,19,28] Although ICA stenting has generally been well tolerated, stenting on its own is not a definitive treatment, and the persistence of symptoms after stenting has been reported.[1,13,29] Additionally, stent fracture has been identified as a possible complication due to ongoing mechanical trauma to the carotid artery by the elongated styloid process.[30,31]

Dissection caused by Eagle syndrome is due to the physical proximity of the styloid process to the carotid vessels. Imaging in these

TABLE 1. Summary of reported cases of Eagle syndrome with carotid artery dissection treated with styloidectomy

Authors & Year	Age (yrs)	Sex	Dissection Laterality	Complete ICA Occlusion	Carotid Stenting Performed	Time From Presentation to Styloidectomy	Final Postop FU Duration	Patient Status at Final FU
Sveinsson et al., 2013[3]	38	M	Lt	No	Yes	NR	6 mos	mRS score 1
Razak et al., 2014[4]	41	M	Rt	No	No	2 mos	NR	NR
Naito & Yamazaki, 2014[7]	55	M	Bilat	Yes	Yes*	"Emergent"	1 mo	Improvement in neurological symptoms
Ogura et al., 2014[1]	55	M	Bilat	No	Yes*	6 days	3 mos	NR
	80	M	Lt	No	No	NR	NR	NR
Jo et al., 2017[32]	38	F	Rt	No	No	NR	6 mos	No symptoms
Torikoshi et al., 2019[14]	46	M	Bilat	Yes	Yes	8 mos	3 yrs	No symptoms
Baldino et al., 2020[24]	49	M	Bilat	No	No	13 days, 33 days	1 yr	No symptoms
	45	M	Lt	No	No	4 mos	5 yrs	No symptoms
	48	M	Lt	No	No	1 yr	2 yrs	No symptoms
Horio et al., 2020[20]	46	F	Bilat	Yes	Yes	NR	NR	NR
Xhaxho et al., 2021[35]	64	M	Lt	No	No	NR	3 mos	Fine motor difficulty in rt hand
Duarte-Celada et al., 2021[15]	43	F	Lt	No	Yes	8 mos	2 yrs	Headaches
Entezami et al., 2021[34]	46	F	Lt	No	Yes	6 mos	"Few weeks"	No symptoms
Selvadurai et al., 2022[33]	Teen	M	Rt	No	No	NR	4 wks	No symptoms
Present case	45	M	Bilat	No	No	3 yrs 7 mos (lt), 10 mos (rt)	6 mos (lt), 4 mos (rt)	No symptoms

FU = follow-up; mRS = modified Rankin Scale; NR = not reported.
* Carotid artery stenting performed after styloidectomy.

patients generally shows this proximity.[5,16] Dissection can occur in younger patients without other risk factors for dissection, as in our case.[3,32,33] Compression of the carotid vessels can also affect the sympathetic nerves that travel on those vessels, leading to symptoms of Horner's syndrome, which can improve following styloidectomy.[24,34]

A review of the literature revealed 15 cases of ICA dissection due to Eagle syndrome, which were treated with styloidectomy (Table 1).[1,3,4,7,14,15,20,24,32–35] The median age at presentation was 46 years (interquartile range 41–55 years), and 11 (73%) of 15 patients were male. Three cases noted possible triggers, two of which were related to neck motion, whereas the third case occurred in a patient with a recent tooth extraction with tonsillar swelling.[4,14,24] Symptoms at presentation varied and included headache, odynophagia, dysarthria, dizziness, and Horner's syndrome, as well as focal neurological deficits such as hemiparesis, hemineglect, and visual field deficits.[1,4,7,15,33] No cases reported recent head trauma. Of the 11 cases that reported a surgical approach, all used the external or transcervical approach. The last reported that postoperative follow-up in each case ranged from 4 weeks to 5 years. All cases that reported postoperative clinical outcomes (11 of 15) described an improvement in symptoms, with most (7 of 11) reporting a resolution of symptoms. These results reflect the present case, in which the procedure was well tolerated and resulted in the complete resolution of symptoms.

Lessons

Eagle syndrome can cause a range of clinical presentations, the most severe of which are attributable to impingement or dissection of the ICA. Eagle syndrome should be suspected in younger patients who present with dissection or carotid occlusion, particularly in cases in which there are no traditional stroke risk factors and in which the styloid process abuts or contacts the ICA at the site of dissection. Styloidectomy is the definitive treatment; in reported cases of styloidectomy in patients with ICA dissection due to Eagle syndrome, most patients returned to their prestroke baseline postoperatively. Furthermore, patients with ICA dissection due to Eagle syndrome remain at risk for contralateral dissection. In these patients, early contralateral styloidectomy can be considered to mitigate the risk of further neurological injury.

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Author Contributions

Conception and design: Tonetti, Vigilante, Khalife, Badger, Thomas, Swendseid, Jovin. Acquisition of data: Tonetti, Khalife. Analysis and interpretation of data: Tonetti, Vigilante. Drafting the article: Tonetti, Vigilante. Critically revising the article: Tonetti, Badger, Shaikh, Thomas, Swendseid, Jovin, Siegler. Reviewed submitted version of manuscript: Tonetti, Vigilante, Badger, Swendseid, Siegler. Approved the final version of the manuscript on behalf of all authors: Tonetti. Statistical analysis: Vigilante. Administrative/technical/material support: Tonetti. Study supervision: Tonetti, Jovin.

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