Case Report

A Case of Juvenile Stroke due to Carotid Artery Dissection from an Elongated Styloid Process—Revisiting Conservative Management

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Carotid artery dissection is a significant etiology of juvenile stroke. Blunt trauma from an elongated styloid process can rarely cause carotid artery dissection, which is one of well-known clinical presentations of Eagle's syndrome as known as stylocarotid syndrome. Growing number of publications contributed improved awareness and diagnostic modalities for this clinical entity, thus the carotid artery dissection from an elongated styloid process is often diagnosed appropriately. The management of carotid artery dissection in stylocarotid syndrome tends to be nonconservative (ie, removal of the process or carotid stenting) presumably due to a publication bias prone to surgical intervention. However, the compression of elongated styloid process to carotid artery is usually difficult or even dangerous to directly prove. Furthermore, stent fracture with subsequent stent and carotid artery occlusion has been reported as a complication of the treatment. Here, we report a male presenting with acute embolic stroke due to carotid artery dissection with the ipsilateral elongated styloid process who has been managed conservatively for more than 1.5 years without any sequelae. We will discuss the management strategy and emphasize the importance of patient education of daily life, since the surgical intervention seems not always necessary in this clinical setting.

Key Words: Carotid artery dissection—cerebral infarct—Eagle's syndrome—elongated styloid process—juvenile Stroke—patient education

Introduction

Carotid artery dissection in young adults is one of well-known clinical presentations of Eagle's syndrome as known as stylocarotid syndrome.[1,2] Growing number of publications contributed improved awareness and diagnostic modalities for this clinical entity.[2] Presumably due to publication bias, the management of carotid artery dissection in stylocarotid syndrome tends to be nonconservative (ie, removal of the elongated styloid process or carotid stenting) as discussed in this paper. However, it is usually difficult or even dangerous to directly prove the compression of internal carotid artery (ICA) by the elongated styloid process. Therefore, we will discuss management strategy of stylocarotid syndrome (ie, ICA

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Received June 24, 2019; revision received July 9, 2019; accepted July 21, 2019.

Financial Disclosure: None.

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1052-3057/$ - see front matter

https://doi.org/10.1016/j.jstrokecerebrovasdis.2019.104307

dissection from stylocarotid syndrome) to revisit conservative management by optimal patient education. It is important to emphasize postures to avoid, including looking-up too much (causing neck overextension) or looking-down too much (causing neck overflexion), both possibly resulting in the impingement on carotid artery by the elongated styloid process.

Case Presentation

A 49-years-old male with a past medical history of mild hypertension, developed sudden weakness in his left extremities and dysarthria immediately after looking overhead (ie, extended his neck) when he tried to swallow Japanese traditional sweets, "Yatsuhashi." Two days later, he visited our hospital. Extensive interview of anamnesis could not reveal any prior face/neck pain, dizziness in his daily life but he felt neck pain at the timing looking up and eating "Yatsuhashi" sweets 2 days ago. The neurological examination revealed dysarthria and weakness in his left hemiparesis (Manual Muscle Testing: 5-/5) with NIH Stroke Scale (NIHSS) of 4. Diffusion-weighted images demonstrated acute cerebral infarction of the right frontal lobe and MR angiography (MRA) showed right ICA occlusion at the distal cervical segment and ipsilateral middle cerebral artery occlusion at M3 branch (Fig 1). Further imaging work-up was made by 3-dimensional computed tomography angiogram, indicating cervical ICA dissection with proximity to an ipsilateral elongated styloid process as long as 3.1 cm (Fig 2). Thus, the proximity of the elongated styloid process to the ICA was confirmed; however, we could not directly show the evidence of impingement on the ICA because we tried not to overextend his neck to avoid cervical ICA compression.

He was conservatively treated with argatroban during the first week followed by oral antiplatelet therapy, as well as physical, occupational, and speech therapy. We also educated him and emphasized not to look up too much to avoid neck extension, which is a potential cause of ICA compression by the elongated styloid process. He discharged home at 30 days after admission without any neurological symptoms followed by antiplatelet therapy for 3 months. Over 1.5-year follow-up, he did not suffer from recurrent stroke. Repeated MRA check-up demonstrated chronological changes in the luminal diameter of the ICA, indicating complete luminal recovery at 6 months after the diagnosis and no recurrence of dissection afterwards (Fig 3).

Discussion

We have successfully treated a male with embolic stroke due to extracranial ICA dissection from the ipsilateral elongated styloid process without surgical intervention uneventful for more than 1.5 years. Although we confirmed the proximity of the elongated styloid process to the ipsilateral carotid artery as well as dissecting extracranial ICA occlusion by 3-dimensional computed tomography angiogram and MRA, we could not directly show the elongated styloid process to impinge on the ICA. We educated the patient to avoid torsion movement of his neck and head (such as look-up too much), which can

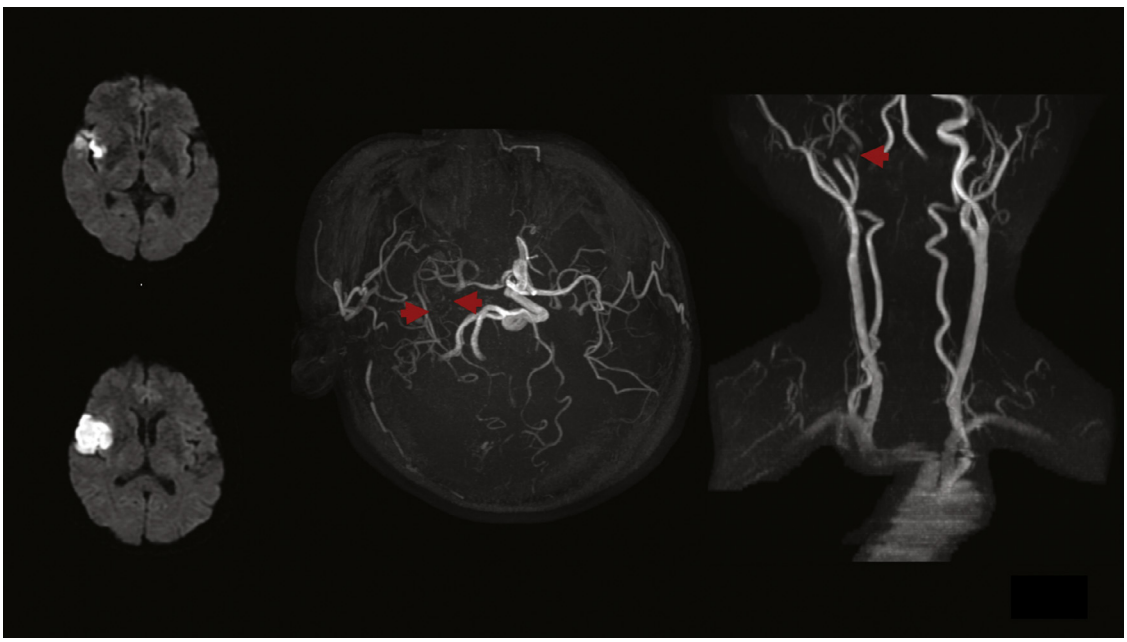

Figure 1. *Magnetic resonance images and angiographies (MRA) acquired on the admission day were demonstrated. Diffusion-weighted images show acute cerebral infarct in the right frontal lobe (left panels), which is the corresponding area of the occluded right middle cerebral artery, M3 portion as shown in MRA (middle panel). MRA of the head (middle panel) also demonstrate faint arterial flow signal in the right internal carotid artery (ICA). MRA of the neck (right panel) demonstrate disrupted arterial flow signal in the cervical segment of the right ICA.*

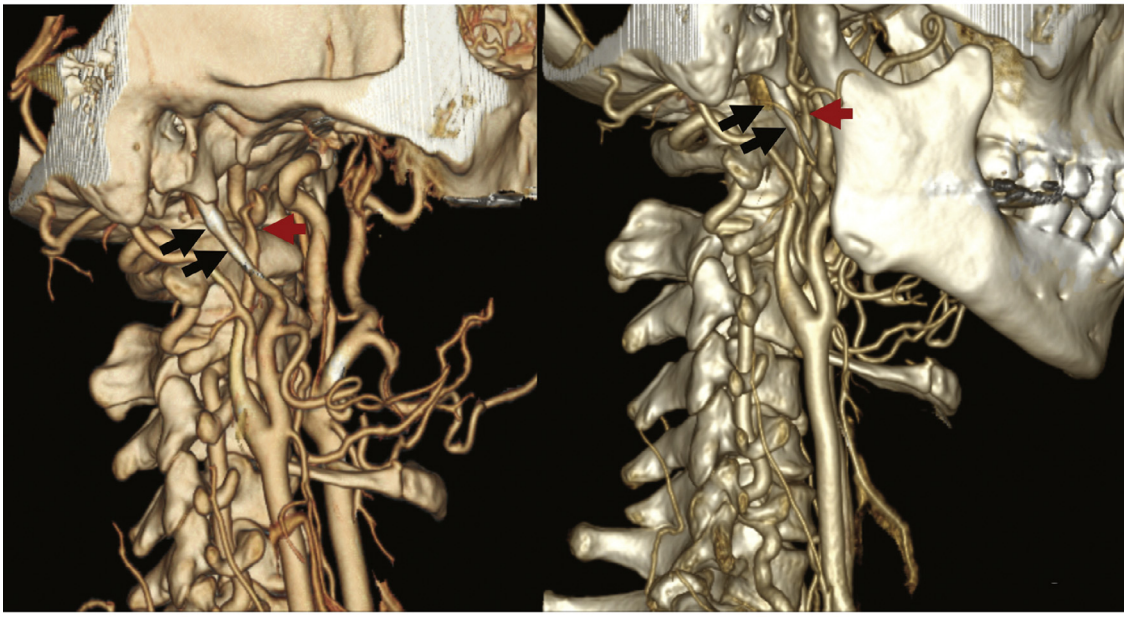

Figure 2. *Three-dimensional computed tomography angiography (3D-CTA) was acquired at 2 weeks after admission. The right styloid process is demonstrated as long as 3.1 cm (black arrows) with a proximity to the ipsilateral carotid artery. As well, the right cervical internal carotid artery appeared tortuous with severe luminal stenosis (red arrow), indicating arterial dissection. (Color version available online.)*

cause a deterioration of ICA dissection. Majority of recent literature reported and emphasized surgical intervention for this disease entity as shown in Table 1,[3-15] however, as we experienced in this case, patient education and conservative therapy can be a choice of management. Thus, we initially considered surgical intervention to prevent recurrence of ICA dissection by the elongated styloid process if we could directly prove the ICA compression by the elongated styloid process; however, we didn't try it. Hooker et al (2016) presented a case with ICA stenting in close proximity to a calcified stylohyoid ligament resulted in stent fracture within a year after the treatment.[16] Miyata et al (2016) also reported a case of stylocarotid syndrome presented with repeated unilateral cerebral infarct.[17] Authors underwent carotid artery stenting and demonstrated mechanical compression of the ICA stent by the elongated styloid process by rotating/tilting patient's neck and head. We thought this maneuver is dangerous and might deteriorate ICA dissection. In fact, the authors removed the styloid process afterwards with uneventful outcome. Moreover, we thought the evidence of surgical treatment effect, including carotid stenting and removal of

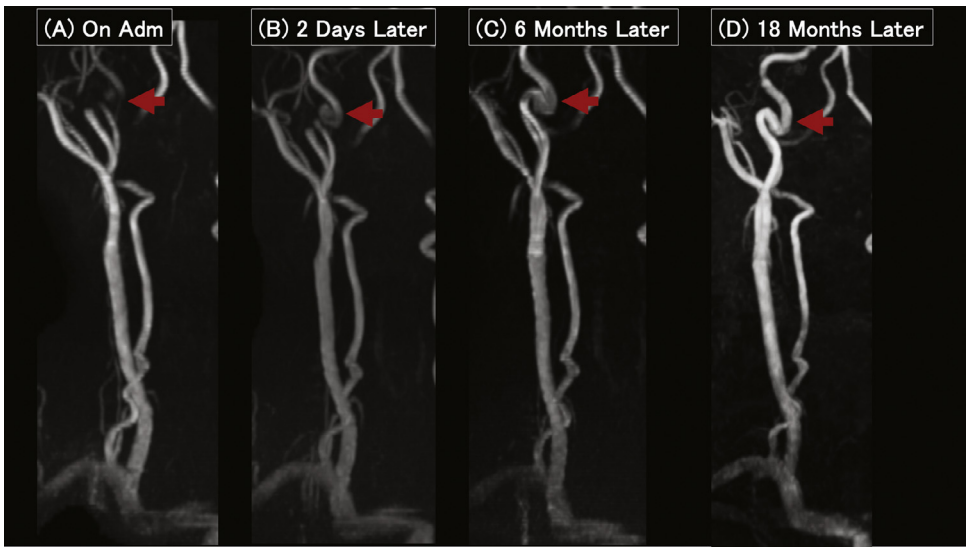

Figure 3. *MRA imaging of the neck was repeated on admission (A), 2 days (B), 6 months (C), and 18 months (D) after admission. These images demonstrate chronological changes in the dissected right cervical ICA. Of note, complete luminal recovery at 6 and 18 months was confirmed (C, D).*

Table 1. *A list of recent literature reported carotid artery dissection due to an elongated styloid process*

Year	Author	Age/Sex	Symptoms	Imaging	Side	Treatment
1999	Zuber M	43/M	Amaurosis fugax, tinnitus	DSA, CTA	Left	Conservative (anticoagulants, patient education)
2004	Soo OY	41/F	Blindness	CTA, MRA, US	Right	Conservative (anticoagulants)
2009	Faivre A	60/M	Confusion, hemiplegia, hemianopia	CTA, MRA, US	Bilateral	Surgical resection of SP
2012	Ohara N	43/M	Transient aphasia	DSA, CTA, MRA, US	Right	Conservative (antiplatelets)
2012	Todo T	80/M	Dizziness, syncope		Bilateral	Surgical resection of SP
		57/M	Transient aphasia	DSA	Right	Carotid stent
2013	Yamamoto S	51/M	Pain, Horner syndrome	CTA, MRA	Right	Conservative (antiplatelets)
2013	Sveinsson O	41/M	Severe headache	CTA, MRA	Right	Conservative therapy (anticoagulants followed by antiplatelets)
		38/M	Minor stroke	DSA, CTA, MRA	Left	Surgical resection of SP
2014	Razak A	41/M	Hemiparesis, hemianopsia	DSA, CTA	Right	Endovascular tPA injection
2014	Naito Y	55/M	Dizziness, visual loss	CTA, MRA	Bilateral	Carotid stent
2014	Ogura T	55/M	Amaurosis fugax	CTA, MRA	Bilateral	Surgical resection of SP
		55/M	Hemiplegia	CTA, MRA	Right	Carotid stent
		80/M	Vertigo	CTA, MRA	Left	Surgical removal of SP
2017	Smoot TW	60/M	Severe headache, hemiparesis	CTA	Bilateral	Carotid stent
2017	Benjamin H	57/M	Aphasia, hemiparesis, neck pain	CTA, MRA	Left	Conservative therapy (anticoagulants)
2018	Jeloder S	40/M	Hemiparesis, dysarthria	CTA, US	Bilateral	Surgical removal of SP

the styloid process, was weak in our case as we did not directly prove the mechanical ICA compression from the elongated styloid process. Zuber et al (1999) have previously claimed the importance of patient education not to flex the neck as a conservative treatment, as well.[3] Very recently, the cervical artery dissection in stroke study investigators revealed that there was no difference in recurrence rates or rates of angiographic recanalization with antiplatelets or anticoagulants in the randomized clinical trial.[18] Thus, current evidence suggests that antiplatelets or anticoagulants have similar effects at reducing the risk of recurrent stroke after extracranial carotid artery dissection. As shown in Figure 3, the dissecting extracranial carotid artery occlusion was recanalized with residual stenosis on day 2 after admission. Therefore, we conservatively treated this patient with anticoagulation therapy with argatroban (ie, a small molecule direct thrombin inhibitor) followed by oral antiplatelet therapy. Furthermore, we educated him not to look-up too much to avoid mechanical ICA compression by the elongated styloid process.

In 1937, Eagle W.W. reported the association between an elongated styloid process or calcified stylohyoid ligament and ipsilateral neck/facial pain, referred otalgia, or dizziness as known as "classical" Eagle's syndrome.[1] Eagle originally defined an elongated styloid process as one longer than 2.5 cm, whereas recent literature tends to define it as longer than 3.0 cm. The elongated styloid process can impinge on the surrounding structures and rarely shows aforementioned symptoms with a prevalence of 4%-10.3% of individuals with an elongated styloid process.[19,20]

It can also cause stylocarotid syndrome arising from interaction with the ICA and its sympathetic nerve plexus. As extensively reviewed by Badhey and Ducic et al, most individuals with an elongated styloid process are asymptomatic.[2] Even in cases showing with a symptom, stylocarotid syndrome is generally indicated by nonspecific cervical pain during torsion movements of the head and neck such as neck extension or look-up behavior, which could be a potential cause of ICA compression. Therefore, stylocarotid syndrome, a rare clinical manifestation of Eagle's syndrome, is often hard to diagnose until cervical carotid artery dissection presenting with stroke occurs. Cervical carotid artery dissection is one of the most common cause of juvenile stroke,[21] with an estimated incidence of 2·6 (95% CI 1·9-3·3) per 100,000 individuals per year at a mean age of 45 years.[22] Although the elongated styloid process is a known risk factor for cervical carotid artery dissection,[2] there is no reliable dataset for the prevalence of ICA dissection due to stylocarotid syndrome.[16] Moreover, the compression of ICA is usually difficult and even dangerous in some instances, to be directly confirmed. Although about 4%-7.3% of the general population can have an elongation in stylohyoid length, the elongated styloid process could rarely be symptomatic with a prevalence of 4%-10.3% of individuals with an elongated styloid process.[2] Thus, we experienced a very rare case of juvenile stroke with stylocarotid syndrome. Presumably due to the rarity of this disease, we found a lot of case reports and case series publications (please see Table 1), and most of recent literature reported successful surgical interventions, which might lead publication bias for the treatment strategy. We considered the weakness of evidence for surgical treatment of stylocarotid syndrome to prevent recurrence of stroke in our case, since it was

hard and dangerous to show the compression of the ICA by the elongated styloid process. As extensively reviewed by Badhey et al, surgical resection of the elongated styloid process has potential risks, including infection, poor exposure to control bleeding, postoperative airway edema in the traditional intraoral approach, as well as the injury of the marginal mandibular branch of the facial nerve in the external cervical approach.[2] Based on these facts, therefore, conservative management should be considered especially in case of initial stroke attack due to carotid artery dissection. More importantly, patient education is very important to avoid look-up too much (causing neck overextension) or look-down too much (causing neck over-flexion), both possibly resulting in the impingement on carotid artery by the elongated styloid process.

In summary, carotid artery dissection is one of rare manifestations of Eagle syndrome caused by blunt trauma by an elongated styloid process and should be considered in case of juvenile stroke. Conservative therapy can be a choice of initial management with an optimal patient education to avoid torsion posture of the neck and head.

Conflict of Interest

The authors have nothing to be declared.

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