CASE REPORT

Carotid artery dissection: a rare complication of Eagle syndrome

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SUMMARY

Carotid artery dissection is a significant cause of ischaemic stroke in all age groups and accounts for a large percentage of strokes in young patients. Carotid dissection can be caused by trauma, underlying connective tissue disease, hypertension, mechanical injury or can be spontaneous. We present an exceedingly rare case of carotid dissection caused by an elongated styloid process, causing direct mechanical damage to the carotid artery.

BACKGROUND

Eagle syndrome is a group of symptoms characterised by recurring pain in the area of the pharynx and face, specifically the retromandibular area. It is due to an elongated styloid process or a calcified stylohyoid ligament, which interferes with the adjacent anatomical structures. Two forms of the syndrome have been reported. The 'classic' and more common elongated styloid syndrome usually presents with unilateral sore throat, neck pain, otalgia, dysphagia and tinnitus. Patients may also experience a foreign body sensation in the pharynx with persistent sore throat. The second form, stylocarotid syndrome, occurs when the elongated styloid process compresses or dissects the extracranial internal carotid artery (ICA) and can lead to transient ischaemic attack (TIA) or stroke. The latter can also present as pain along the course of the carotid artery as a result of stimulation of the sympathetic nerve plexus with rotation or compression of the neck.¹⁻⁴ The classic presentation is much more common than the vascular presentation. Even more uncommon is carotid artery dissection (CAD) caused by the elongated styloid, with only a handful of reported cases.²⁻⁵ We present an exceedingly rare cause of carotid dissection caused by elongated styloid process causing direct mechanical damage to the carotid artery.

CASE PRESENTATION

A woman aged 47 years with a medical history of diet-controlled diabetes mellitus presented to the emergency department after 1 hour of slurred speech, left arm tingling and weakness. The patient's symptoms started when suddenly looking up to the ceiling while talking over the phone. She also gave a history of pain on the left of her neck and eye on turning her head to the left side for the past 4 months. On examination, vital signs were within normal limits. Neurological examination revealed 4/5 left upper extremity weakness and sensory loss, along with a left facial droop.

INVESTIGATIONS

A CT of the head without contrast did not reveal any intracranial bleeding. MRI of the brain showed a small hyperacute right middle cerebral artery territory infarction involving the middle frontal gyrus (figure 1). Tissue plasminogen activator (t-PA) was administered; however, the left arm weakness worsened to 2/5. t-PA was stopped and a repeat CT scan was performed which did not reveal any bleed. CTA showed smooth tapering of the midright cervical ICA, ~3.5 cm above its origin, suggestive of carotid dissection and an associated elongated styloid process bilaterally, which was measured to be 4.2 cm in length (figures 2 and 3). It also showed that the dissection originated where the ICA was crossed by the styloid process. The distance measured between the right RCA and styloid process was 4.3 mm (figure 4). A cerebral and carotid angiogram confirmed acute dissection of the right ICA at the level of C2 with the narrowest segment measuring <1 mm resulting in 80–90% stenosis (figure 5). The proximal right ICA measured about 3 mm in diameter and the arterial diameter just distal to the dissection measured 4 mm in diameter. A lipid panel showed an LDL level of 103.

TREATMENT

Balloon angioplasty was performed with placement of a 4×20 rebel stent (figure 6). The patient was then started on aspirin 325 mg, clopidogrel 75 mg and atorvastatin 20 mg daily.

OUTCOME AND FOLLOW-UP

After placement of the stent, the left arm weakness improved to 5/5; however, a residual sensory deficit

Figure 1 Diffusion-weighted MRI of the brain showing acute infarction involving the right middle frontal gyrus.

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Figure 2 CTA of the neck showing internal carotid artery dissection starting at the site of crossing by an elongated styloid process. The length of the styloid process was 4.2 cm.

was observed. Of note, the patient was hospitalised again for recurrent left arm numbness and slurring of speech, which resolved spontaneously. Stroke workup at the time was negative. The patient is currently awaiting surgical intervention for the elongated styloid process because of her recurrent symptoms.

DISCUSSION

The elongation of the styloid process was first described in 1652 by Pietro Marchetti; however, it was not until 1937 when otolaryngologist, Watt Weems Eagle, first described the clinical findings of the syndrome, which now bears his name.⁶ Eagle defined the length of a normal styloid process to be between 2.5 and 3 cm, though some authors believe that the normal length may range up to 4 cm.⁷ About 4% of the general population

Figure 4 Axial CTA showing the distance between the internal carotid artery and styloid process to be 4.3 mm on the right and 7.4 mm on the left.

can have an elongated styloid process defined as >3 cm in length. Only 4% of patients with an elongated styloid process become symptomatic; hence, the actual population incidence of Eagle syndrome is about 0.16%.^{4 5 8} Women are affected three times more than men, most commonly between the ages of 30 and 50 years.

Figure 3 Axial CTA showing dissection of the right internal carotid artery at the level of C2.

Figure 5 Cerebral angiogram showing internal carotid artery dissection.

Figure 6 Cerebral angiogram of the internal carotid artery after the placement of the 4x20 rebel stent.

Anatomy

The styloid complex comprises the stylohyoid ligament, the styloid process and the lesser horn of the hyoid bone. The styloid process is embryologically derived from cartilage of the second pharyngeal arch.⁷ The styloid process originates from the base of the temporal bone, behind the mastoid and has an anterior and medial projection. The styloid process and stylohyoid ligament are in contact with the parapharyngeal space in which the common carotid artery is located. The most distal aspect of the styloid process is located between the external and internal carotid arteries, just lateral to the tonsillar fossa.^{1 9}

Three types of elongated styloid processes have been reported, the uninterrupted styloid process, pseudoarthrosis between the styloid process and the stylohyoid ligament, and segmental interrupted stylohyoid ligament.⁷

Pathogenesis

The classic variant of Eagle's syndrome is associated with pharyngeal injuries during tonsillectomy. Stylocarotid syndrome, the less common variant of the syndrome, is not associated with pharyngeal injuries, and symptoms are a result of mechanical irritation or compression of the perivascular nerve fibres in the external carotid artery and ICA. However, the exact aetiology of the elongated styloid process is still unclear, though there are many hypotheses. The styloid process may develop inflammatory changes or impinge on the adjacent structures, leading to the development of the classic or vascular symptoms of Eagle syndrome.⁹ Eagle believed that surgical insult (ie, tonsillectomy) led to osteitis, perititis or tendinitis of the styloid complex leading to ossification. Since the styloid complex is a derivative of Reichart's cartilage, it is hypothesised that the mesenchymal elements in the complex under stress or trauma may lead to reactive hyperplasia or metaplasia of the bone, leading to ossification.^{1 7 10} However, the syndrome can be observed in those without any surgery or trauma. An alternative theory is the ossification due to an endocrine disorder, where elevated serum calcium, phosphorous and vitamin D metabolism disorder may lead to stylohyoid ligament ossification.⁷

As above, CAD is not considered a part of Eagle syndrome. However, the elongated styloid process that has been described in Eagle syndrome is most likely a significant contributor of CAD. The carotid artery is subjected to the abutting elongated styloid process which may be driven into the artery with muscle contractions of the neck.⁵

Raser *et al* studied 38 patients with CAD compared with 38 control patients without dissection to evaluate the association of CAD with anatomical characteristics of the styloid process. The authors found that the styloid process was significantly longer on patients with dissection compared with control participants (mean length 30.3 vs 26.6 mm). They concluded that the elongated styloid process was a risk factor for cervical CAD.¹¹ Renard *et al* compared 88 patients with CAD with 88 control patients to determine whether the closer proximity of the styloid process to the ICA was associated with increased risk of dissection. They found that the styloid process was closer to the ICA in patients with CAD compared with those without dissection.¹²

Diagnosis

The diagnosis is based on a thorough history and clinical examination as well as imaging. A previous history of tonsillectomy along with exacerbation of pain on digital palpation of the styloid process in the tonsillar fossa can support the diagnosis. In addition, improvement or resolution of symptoms on injecting anaesthetic agents in the tonsillar fossa is highly suggestive of the diagnosis.⁹ Imaging with a lateral skull X-ray can show an abnormally elongated styloid process. A CT scan with 3D reformations is the gold standard investigation, which shows the anatomy of the styloid process in relation to the surrounding structures along with accurate measurement of its length and angulation, which is pivotal for surgical planning.^{2 8 13}

Management

Treatment of Eagle syndrome may be conservative or surgical. In our presented case, the patient suffered acute ischaemic stroke from a dissected carotid artery and was treated with stenting. Until now, there are no guidelines for the management of carotid dissection secondary to an elongated styloid process, though treatment often is surgical to prevent recurrences. On initial management, thrombolytic therapy with t-PA should not be withheld in patients with acute ischaemic stroke, even if carotid dissection is suspected. In theory, using thrombolytic therapy in patients with carotid dissection may lead to increased risk of bleeding. Paradoxically, there is strong evidence to support the safety and efficacy of using thrombolytic therapy for patients with ischaemic stroke secondary to CAD. Several studies have shown that there was no difference in the risk of bleeding and recurrent stroke in patients receiving thrombolytic therapy irrespective of the presence of underlying carotid dissection.¹⁴⁻¹⁶ Furthermore, the elongated styloid process may need to be surgically resected for prevention of recurrent carotid dissection, TIA or ischaemic stroke. Surgical resection can be achieved through an intraoral or extraoral approach, both of which have similar success rates.⁶ The intraoral approach, though simpler, is now used less frequently due to the increased risk of deep space neck infection and poor visualisation of the surgical field. The extraoral approach is favoured, as it allows better visualisation and thus is associated with fewer complications.^{1 2 17} It is also the favoured approach in those with extreme ossification, from the styloid process to the hyoid bone. In those with a 'classic' Eagle syndrome who wish to pursue conservative treatment, options include non-steroidal anti-inflammatory drugs, corticosteroid or anaesthetic injection, and anticonvulsants (similar to the treatment of trigeminal neuralgia and antidepressants).⁶

Learning points

- It is important to identify an elongated styloid process as a possible cause of carotid dissection since a transient ischaemic attack or stroke may not always present with the traditional risk factors for vascular disease such as advanced age, smoking, hypertension and hyperlipidaemia.
- A 3D CT scan is the gold standard diagnostic modality for Eagle syndrome.
- Treatment may necessitate surgical removal of the elongated styloid process to prevent recurrent disease.
- In a patient with suspected CVA, thrombolytic therapy should be administered, despite the presence of carotid artery dissection.

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