

Eagle syndrome and carotid artery dissection: a rare skull base cause of stroke

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SUMMARY

Eagle's syndrome is a rare collection of symptoms that occur secondary to an elongated styloid process or calcified stylohyoid ligament irritating its surrounding structures. Classically, this presents as unilateral throat pain or rarely, as acute neurological symptoms secondary to compression of the internal carotid artery: so called 'stylocarotid syndrome'. Significant neurological events in teenagers, secondary to Eagle syndrome have not been reported. We discuss the rare case of a teenage boy, diagnosed with right internal carotid artery dissection and middle cerebral artery infarction, with no cause initially identified. Following further admission with a transient neurological episode, he was noted to have elongated styloid processes with the right abutting the site of carotid dissection. He underwent styloidectomy and has since remained symptom free. This case highlights the importance of considering anatomical variants when assessing young patients with neurological symptoms, and the potential morbidity and mortality benefit that early surgical intervention may have.

BACKGROUND

Eagle's syndrome is an unusual and rare group of symptoms that occur secondary to an elongated styloid process with a calcified stylohyoid ligament irritating the surrounding neurovascular and pharyngeal structures. The classical presentation was first fully described by Eagle and features atypical unilateral throat pain associated with a sensation of a foreign body in the throat.¹ Patients can also suffer from a persistent sore throat that is referred to the ear and becomes worse on swallowing. Other frequently associated symptoms include neck pain, dysphagia, syncope and tinnitus.²

Eagle later described the less common 'stylocarotid syndrome'³; wherein compression of the extracranial internal carotid artery (ICA) can result in neurological symptoms, stroke and transient ischaemic attacks. Patients with this syndrome can also experience irritation of the sympathetic chain when rotating their neck, causing pain in the distribution of the carotid artery.² Although more commonly seen in patients with connective tissue disorders and trauma, ICA dissection with acute ischaemic events secondary to stylocarotid Eagle's syndrome has been described in a growing number of case reports.⁴⁻³⁰ These cases share similar clinical presentations, however, given the rarity of the condition, there is no clear consensus on its definitive management. Resultantly, the phenomenon has been treated successfully with both conservative

medical and more invasive surgical management in the form of a styloidectomy.

In this report, we discuss the rare case of a teenage boy who underwent a styloidectomy by our ear, nose and throat (ENT) skull base team after experiencing repeat, acute neurological events, secondary to an ICA dissection and elongated styloid process. Significant neurological events in teenagers, secondary to Eagle syndrome have not been reported before. We review the current literature and discuss our management in relation to previously reported cases of this unusual phenomenon.

CASE PRESENTATION

A teenage boy, presented to hospital with acute onset left-sided hemiparesis, mild dysarthria and a severe headache after carrying a heavy rucksack. He had been admitted to hospital 18 months prior to this event after experiencing similar symptoms, where a right-sided ICA dissection, mural thrombus and middle cerebral artery (MCA) infarction had been found. He had been treated with acute thrombolysis, 6 months of anticoagulation (warfarin) and subsequent antiplatelet therapy. Despite extensive investigations, no cause for his initial presentation was found.

Repeat CT angiography (CTA) noted a reduction in the right MCA calibre, from the M2 segment onwards with a focal dissection of the cervical ICA (figure 1A). When compared with imaging from his previous admission, there had been minimal extension of his ICA dissection. MRI noted mature ischaemic changes in the frontal and temporal lobe, but no acute infarct was seen (figure 1B).

He underwent acute thrombolysis at his local hospital, and was transferred to our hospital with a view to perform thrombectomy. Before this was achieved, the patients' symptoms spontaneously resolved. He was reviewed by our ENT team and he was later discharged home with oral-anticoagulation.

The patient was subsequently discussed at our tertiary skull base multidisciplinary team (MDT) meeting. On further review of imaging, bilateral elongated styloid processes and calcification of the stylohyoid ligament was noticed with the right directly abutting the site of ICA dissection suggesting a diagnosis of Eagle syndrome (figure 2). It was felt that positional compression of the ICA due to this anatomical variant was the likely cause for his current transient neurological event. Furthermore, it is likely that chronic compression by this elongated styloid process



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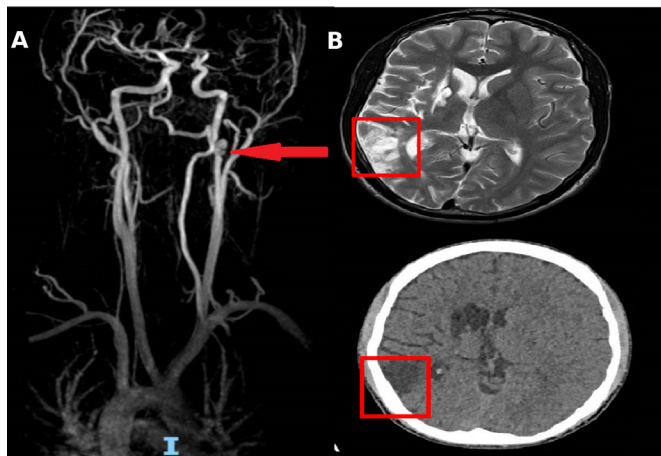


Figure 1 (A) CT angiography of right carotid. The internal carotid artery dissection is indicated by the red arrow. (B) MRI and CT images of mature middle cerebral artery infarct—indicated by red box.

prevented a natural resolution of his ICA dissection following his initial presentation.

TREATMENT

Following the diagnosis of Eagle's Syndrome in a young man with two cerebrovascular events in a short period, our skull base MDT consensus was to perform an elective right styloidectomy. Of note, the ICA dissection was managed medically, with the vascular MDT deciding not to stent in the acute setting due to the transient nature of the neurological episode.

An open approach was used through a right transcervical incision. Careful dissection was made through level II lymph nodes anterior to the sternocleidomastoid. A number of bulky lymph nodes were excised which subsequently revealed benign hyperplasia on histopathological examination. The styloid process was carefully delineated and surrounding neurovascular structures, in particular, the facial nerve, were preserved. The stylohyoid and stylopharyngeus were dissected free before final excision of the styloid process.

The patient had an uneventful inpatient stay and was discharged on day 2 postoperatively with no complications. He was reviewed in the outpatient ENT department 4 weeks after his procedure and had made an excellent recovery with

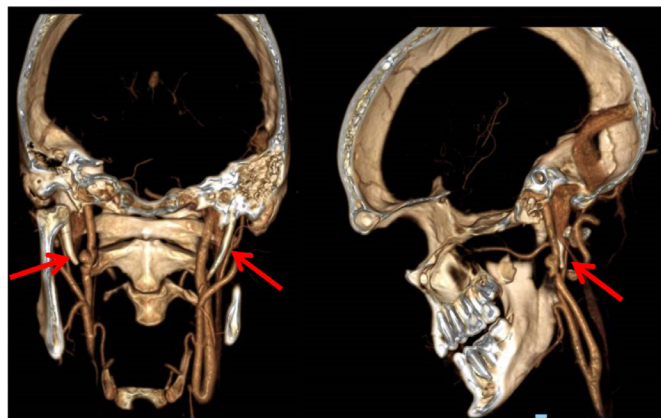


Figure 2 CT skull reconstruction. Left—coronal view with bilateral elongated styloid processes. Right—sagittal view showing 34 mm right styloid process in close proximity to Internal carotid artery dissection. Red arrows indicate styloid process.

no recurrent episodes of neurological symptoms. He remains on anticoagulation and is under close clinical surveillance by the vascular and stroke teams.

Though currently asymptomatic, due to the patient's young age and potential risk of further neurological sequelae, he has been considered for a prophylactic contralateral styloidectomy at a future date.

DISCUSSION

We highlight this unusual case of a young man with recurrent cerebrovascular events, secondary to an elongated styloid process causing ICA compromise.

The styloid process is a long bony prominence that usually measures 20–25 mm, and serves as an important point of attachment for three muscles (stylohyoid, stylopharyngeus, styloglossus muscle) and the stylohyoid ligament.² Traditionally, a styloid process of >30 mm is thought to be a risk factor for experiencing Eagle's syndrome, although Eagle found only 4% of patients with abnormal styloid processes noted symptoms.³¹ In two separate case series, patients with carotid artery dissection were noted to have a significantly increased length of styloid process resulting in a shorter contact distance between the artery and bone,^{32 33} strongly implicating Eagle's syndrome as an independent risk factor for carotid artery dissection. In addition to styloid length, Tardivo *et al* showed that the styloid angulation and its relation to C1 transverse process may also contribute to compression of the ICA and subsequent dissection, demonstrating a multifactorial pathogenesis to symptomatic disease.³⁴

Diagnosis of Eagle's syndrome is based on clinical findings and cross-sectional imaging. Classically, palpation of the tonsil fossa can demonstrate an elongated styloid process, with resolution of symptoms on injection of local anaesthetic being diagnostic for the condition.³⁵ Plain film radiography can occasionally demonstrate an elongated styloid, but is limited by calcification around the stylohyoid ligament and does not provide information on the adjacent soft tissues. CT scanning is the current imaging modality of choice for diagnosis of Eagle's syndrome, clearly defining the relationship of the styloid process to the surrounding neurovascular structures.³⁵ Where available, three-dimensional reconstruction of CT images can provide excellent information on the length and angulation of the styloid in relation to the carotid artery.³⁶ Additionally, several previous case studies have made use of CTA to diagnose aberrant flow and arterial dissection within the carotid vessels secondary to elongated styloid processes.

Surgical intervention for the classical description of Eagle's syndrome is indicated after failure of medical therapy. Several cases have used styloidectomy to prevent recurrence of stroke symptoms. External approach and intra-oral styloidectomy are the most frequently described surgical techniques.³⁷

The external approach provides the greatest exposure to the deep neck space and therefore facilitates the safest dissection of the styloid process and nearby neurological structures. Theoretically, it poses an increased risk of marginal mandibular nerve injury, however, Ceylan *et al*, found that in a large case series of 61 patients, only two patients suffered a temporary marginal mandibular nerve weakness with no other reported complications,³⁸ strongly supporting the safety and efficacy of this technique.

Intra-oral styloidectomy avoids the need for an external neck scar and has a shorter operating time, but affords only limited views of the styloid process and has an increased risk of retropharyngeal infection.^{37 39} Despite this, a number of alternative

intraoral resection techniques have been proposed including include a ‘tonsil sparing’ techniques as successfully described by Torres *et al* in a case series of 10 patients with no post-operative complications.⁴⁰ Endoscopic^{41,42} and robotic-assisted transoral⁴³ approaches have also been performed, but are isolated to small cases series and are not common practice.

On review of the literature over the last 20 years, to the best of our knowledge, there has been 34 cases of carotid artery dissection secondary to an elongated styloid process that have been published. The condition is more common in males with a ratio of 2.4:1. The patients were generally fit and well with 23 (68%) having no reported comorbidities. Twenty patients had medical or endovascular management only, and 14 underwent surgical intervention in the form of open approach styloidectomy (9 unilateral, 5 bilateral). Surgery was generally performed on a semi-elective basis, following initial medical or endovascular management. No recurrent clinical characteristics were noted in those selected for surgical intervention, however, there appears to be a trend for surgical management as a definitive treatment choice in recurrent and/or severe ischaemic disease^{4–30} (online supplemental table 1).

Excluding our patient, the mean age of cases is 49.3 years old (range 30–80 years old). Our case is the youngest by a considerable margin, demonstrating that ICA dissection due to Eagle’s syndrome can also occur in young patients. ICA dissection is implicated in 2.5% of all patients presenting with a stroke.⁴⁴ While little epidemiological data are available, case-series have shown ICA dissection to be present on imaging in 8%–20% of patients under 18 years of age presenting with acute neurological symptoms. Often, a history of minor trauma or recent infection are suggested as aetiological factors, however, most commonly ICA dissection occurs spontaneously. In these cases, connective tissue disorders such as Ehlers-Danlos syndrome or Marfans syndrome are commonly implicated.⁴⁵ Eagle’s syndrome as a contributing factor to ICA dissection in a paediatric population has not been seen.

Importantly, in our case, an elongated styloid process was missed on initial presentation, and was not considered as a possible causative factor for his repeated neurological events. This highlights the importance of considering anatomical variants such as those seen in Eagle’s syndrome in the differential diagnosis for young patients presenting with neurological symptoms. Similarly, young patients are more likely to develop future cerebrovascular events, and therefore definitive surgical management is likely to improve the patient’s morbidity and mortality. As such, we advise that all young patients with recurrent neurological episodes caused by elongated styloid process should undergo definitive surgical management in the form of a styloidectomy.

Learning points

- ▶ Although Eagle’s syndrome typically presents with sharp neurological pain in the throat and jaw, a long styloid process can cause carotid artery compression or dissection presenting with recurrent acute neurological symptoms.
- ▶ Unusual causes of stroke including unusual anatomy and trauma should be considered in young otherwise healthy patients.
- ▶ Definitive surgical management in the form of an elective styloidectomy is advised in young patients to prevent further cerebrovascular events.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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