

The eagle is landing:

Eagle syndrome — an important differential diagnosis

CASE REPORT

A 35-year-old woman presented in the accident and emergency department with a 2-day history of right-sided Horner's syndrome. She had been seeing her GP over the last few months with right-sided neck pain, often brought on by chewing, and occasional tinnitus and dizziness, for which no cause had been found. Physical examination, other than right-sided Horner's syndrome, and chest imaging and head computed tomography (CT) were unremarkable. A 3D CT scan of the cervical region demonstrated elongation of the right styloid process, and bilateral ossification of the stylohyoid ligaments, more marked on the right, postulated to be impinging the sympathetic chain on the right causing ipsilateral Horner's syndrome. She was referred to an ear, nose, and throat (ENT) specialist for further management of suspected Eagle syndrome.

BACKGROUND

The rarely diagnosed Eagle syndrome, described in 1937 by the otolaryngologist Watt Eagle,¹ is due to elongation and/or ossification of the styloid bone and/or ossification of the stylohyoid ligament impinging on local structures. 'Stylalgia' describes the pain associated with this condition. The importance of this syndrome, lies in the fact that it may present with a range of commonly encountered, but occasionally potentially serious, symptoms.

ANATOMY

The styloid bone lies at the base of the temporal bone, posterior to the mastoid apex. The stylohyoid complex comprises the styloid process, the lesser horn of the hyoid bone, and the stylohyoid ligament. The normal length of the styloid process is about 2.5 cm, while an elongated styloid process is over 2.5 cm to 3 cm in length.

AETIOLOGY

The aetiology is unclear. Genetic

predisposition, trauma, and early onset of the menopause have been postulated. Symptoms may be due to rheumatic styloiditis and degenerative changes; compression of local structures by an elongated styloid process or fracture of the ossified stylohyoid ligament with scar tissue proliferation; tendinosis at the insertion of the ligament; local stretching and scarring post-tonsillectomy.

EPIDEMIOLOGY

There is a lack of robust statistical evidence regarding the true prevalence of Eagle syndrome. An elongated, ossified styloid process on imaging may occur in up to 28% of the population, but is a predominantly incidental finding.² Symptoms are generally unilateral, although styloid process elongation is often bilateral. There is a female to male ratio of 3:1, and it usually affects patients aged 30 to 50 years old. There is a paucity of clinical epidemiological evidence regarding the specificity, sensitivity, and predictive values of particular symptoms in suggesting a diagnosis Eagle syndrome.

CLINICAL FEATURES

Eagle syndrome is a symptom complex, and may present with a variety of symptoms, none of which is pathognomonic. The commonest symptoms are throat and neck pain, and the sensation of a foreign body in the throat. Less frequent symptoms are referred otalgia, headache, carotidynia, dizziness, and dysphagia. A range of cranial nerve palsies may occur. Pain is often unilateral and more than one symptom is usual. Precipitation or exacerbation of symptoms on turning the head and/or neck flexion has been noted in neck pain, transient ischaemic attacks³ and syncope, with surgical resection of the elongated styloid process proving curative in case reports.⁴ Sudden death attributed to compression of both carotid sinuses by bilaterally elongated styloid processes has been reported.⁵

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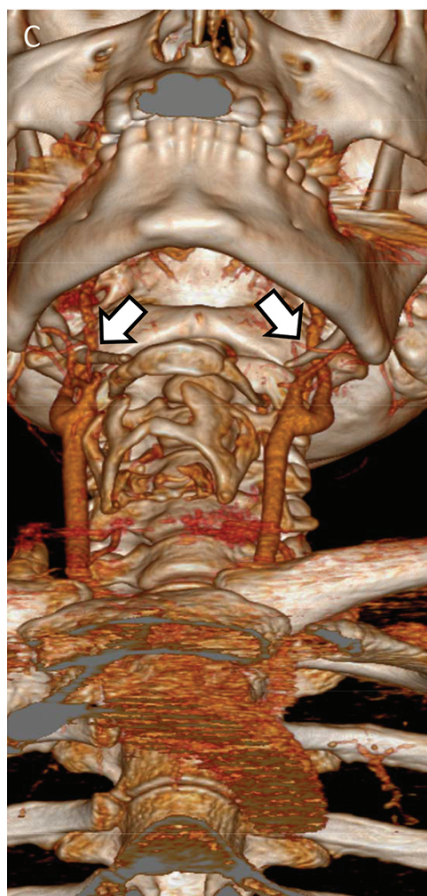


Figure 1. 3D CT scan demonstrating bilateral elongation of the styloid processes.

Eagle identified two types of Eagle syndrome: the 'classical' type, described as occurring after tonsillectomy, with symptoms due to proximity to local structures and the adjacent cranial nerves V, VII, IX, X, including sore throat, radiating to the ear, the sensation of a foreign body in the throat, and dysphagia. The second 'carotid artery' type, due to the elongated styloid process impingement on the carotid sheath and compromising blood supply and sympathetic innervation. Lateral deviation of the styloid process, compressing the external carotid artery, may cause pain below the eyes. Medial deviation, compressing the internal carotid artery, may lead to pain in the region of ophthalmic artery supply and parietal area.

DIAGNOSIS

A history of local surgery or trauma, and precipitation and/or exacerbation of symptoms on turning the head or neck flexion, compounding local pressure effects, may suggest Eagle syndrome. On examination, palpation of an elongated styloid process in the tonsillar fossa, not possible when the styloid process is of normal length, may reproduce, as well as exacerbate, the symptoms. Injection of local anaesthetic into the tonsillar fossa may alleviate and even eliminate symptoms, and may be administered in a secondary care setting to support the diagnosis, guiding the surgeon as to whether surgery may prove beneficial.

Investigation by imaging may be unreliable due to the high prevalence of incidental findings and the lack of evidence about the proportion of patients in whom typical imaging findings are responsible for the symptoms. While lateral plain cervical spine X-rays demonstrate an elongated styloid process, the gold standard is CT scanning, with 3D reconstruction, which displays the proximity of an elongated, ossified styloid process to adjacent structures.

MANAGEMENT

Management is medical or surgical but there is a lack of evidence, particularly on non-surgical options. These include analgesia, particularly nonsteroidal antiinflammatory drugs, carbamazepine, diazepam, heat application, transpharyngeal injection of local anaesthetic or steroids, and physiotherapy. Surgery is by the transpharyngeal or extra-oral approach and may be curative. The less invasive transpharyngeal route may predispose to injury of local structures, infection, and

post-procedure oedema, due to a limited surgical field. The extra-oral approach involves external scarring, but provides better visualisation of the surgical field and reduced risk of infection. While surgical failure rates of around 20% have been described, more recent studies demonstrate complete resolution of symptoms in most cases with few post-operative complications, chiefly a transient marginal mandibular nerve weakness.⁶

CONCLUSION

Until recently Eagle syndrome has remained predominantly in the clinical territory of ENT surgeons, but it constitutes a key cause of an unclear proportion of symptoms which are commonly encountered in primary care and are potentially serious, making it an important differential diagnosis for GPs to consider. It may be that many more patients have Eagle syndrome than have been recognised, although there is still a need for more research into this interesting condition.

Provenance

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