



Carotid artery type of Eagle syndrome: an uncommon cause of ischemic stroke

Katarina Lakner · Lina Savšek

Received: 4 January 2022 / Accepted: 20 July 2022

© The Author(s), under exclusive licence to Springer-Verlag GmbH Austria, part of Springer Nature 2022

Summary Acute ischemic stroke in patients younger than the age of 50 years is a rare occurrence that results in high mortality and substantial loss of functional years of life. Internal carotid artery dissection (CAD) presents a rare, but serious condition that needs to be fully evaluated and carefully treated, as it may lead to an acute ischemic stroke in all, but mostly in younger patients. A possible cause for CAD, the carotid artery type of Eagle syndrome (ESy), is atypical and underrecognized. In this case report we present a case of a young patient with carotid artery type of ESy, resulting in a severe acute ischemic stroke. Only recognition of such a syndrome in its early symptomatic phase could allow appropriate management to prevent this kind of a deleterious outcome.

Keywords Elongated styloid process · Internal carotid artery dissection · Acute ischemic stroke · Calcified stylohyoid ligament · Unilateral cervicofacial pain

Introduction

In patients under the age of 50 years, only 10–15% of all ischemic strokes occur [1]. Stroke incidence among this population is increasing, arguably due to many lifestyle changes and a concomitant increase in the burden of classical vascular risk factors [2], which remain similar regardless of age (atrial fibrillation, diabetes mellitus type 2, hypertension, hypercholesterolemia, coronary artery disease) [1].

Despite the low incidence in the general population, carotid artery dissection (CAD) is one of the most prevalent vascular diseases causing ischemic stroke in young adults [3]. It can be linked to trauma, infections, migraine, fibromuscular dysplasia or heritable connective tissue disorders, such as Ehlers-Danlos syndrome [4]. One of the rarest causes of CAD is Eagle syndrome (ESy), a condition in which the styloid process of the temporal bone is pathologically elongated, potentially causing physical impact on the carotid artery wall and thus dissection [4].

Case report

A 38-year-old right-handed motorbiker with no comorbidities experienced an episode of severe headache, with a subsequent onset of a weaker right side of the body and a lack of speech. He was brought to the ER, where global aphasia with a severe right-sided hemiparesis were present (National Institutes of Health Stroke Scale: 22 points; Glasgow Coma Scale: 12 points).

On CT imaging, an already demarcated left frontotemporal lobe ischemic infarction was present, which was later also confirmed on MR imaging (Fig. 1). CT brain angiography demonstrated a long left internal carotid artery (ICA) occlusion resulting in arterio-arterial embolism to the left anterior M1 and M2 middle cerebral artery segment. He was admitted to the stroke unit, where he received all the necessary and supportive treatment, while the search for the causative factor was continued.

Further information revealed that he had already reported left-sided facial and cervical pain within the previous week, with a missing history of trauma. He was a non-smoker and had a negative epidemiological history for COVID-19. On re-examination, a mild left-sided ptosis and miosis with absent signs of an-

K. Lakner, MD (✉) · Assist. L. Savšek, MD
Department of Neurology, General Hospital Celje, Oblakova
ulica 5, 3000 Celje, Slovenia

Faculty of Medicine, University of Ljubljana, Vrazov
trg 2, SI-1000 Ljubljana, Slovenia
katarina.lakner8@gmail.com

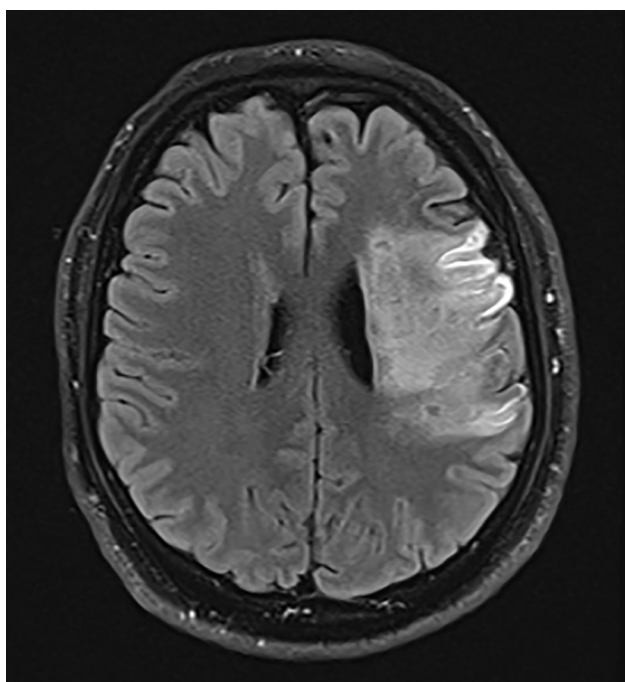


Fig. 1 Ischemic stroke in a major part of the left middle cerebral artery vascular bed in the frontotemporal lobe (T2-FLAIR weighted MRI, transverse section)

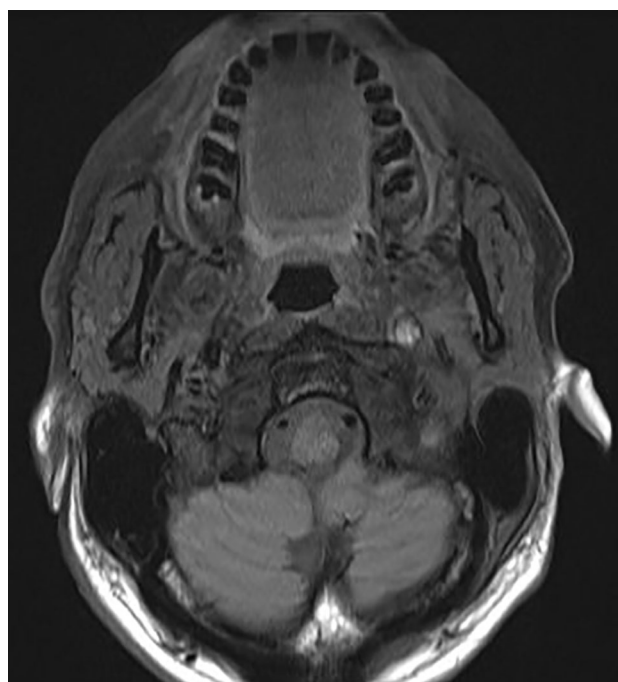


Fig. 2 Left ICA occlusion due to internal CAD (T1-VIBE weighted FS-MRI with contrast agent, transverse section)

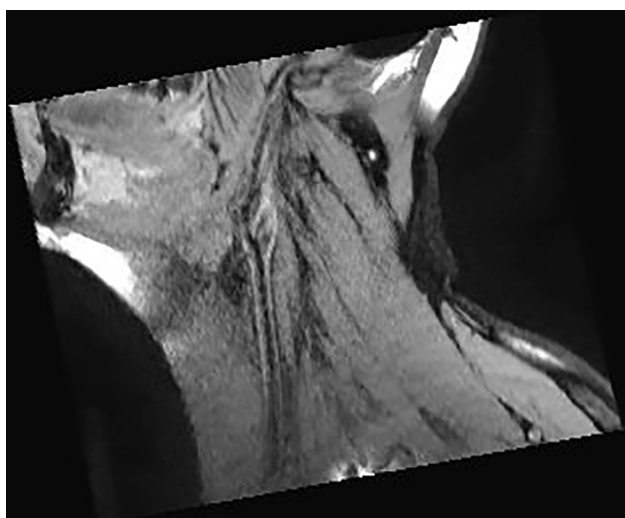


Fig. 3 Left ICA wall dissection with wall hematoma (T1-weighted FS-MRI with contrast agent, parasagittal section)

hidrosis were noted. A postganglionic Horner's syndrome suggested a possible left-sided internal CAD, which was subsequently confirmed with T1-weighted FS-MRI imaging (Fig. 2 and 3).

Additionally, a prominent styloid process was noted and later confirmed by a neck CT scan (Fig. 4) which showed elongated styloid processes on both sides (right: 45 mm, left: 50 mm), either in direct contact with the ICA wall (left side) or in its proximity (right side). Without any imaging signs of atherosclerotic changes or other pathology of the carotid endothelium, the dissection was speculated to be caused by

a mechanical pressure of the styloid process. Other causative factors for CAD as well as a comprehensive work-up for other possible causes of stroke were unremarkable. As such, carotid type of ESy was confirmed as the single cause for CAD.

The patient received anticoagulant medication with low molecular weight heparin (LMWH) agent in prophylactic doses (5.000 IU [five thousand International Units] subcutaneously per 24 h) as well as antiplatelet therapy with acetylsalicylic acid (100 mg/day) and clopidogrel (75 mg/day). Surgical correction of styloid processes on both sides was planned to prevent the recurrence of an ischemic event but as the process of early neurorehabilitation showed only slow signs of improvement (the movement deficits slowly improved, while global aphasia remained unchanged), a decision was made to postpone the procedure, to allow completion of the neurorehabilitation process, so that the patient would later better sustain the surgical procedure. After 1 month of treatment in the clinical department, he was transferred to the rehabilitation institute.

Discussion

Eagle syndrome (ESy), named after a renowned otolaryngologist Watt W. Eagle, MD, is classified by two types: the more common classical type, and the atypical carotid artery type [5]. The leading physical manifestation of the classical type is a sharp, unilateral cervicofacial pain, specifically close to the jaw. The main cause for this condition is the stretching of the cranial

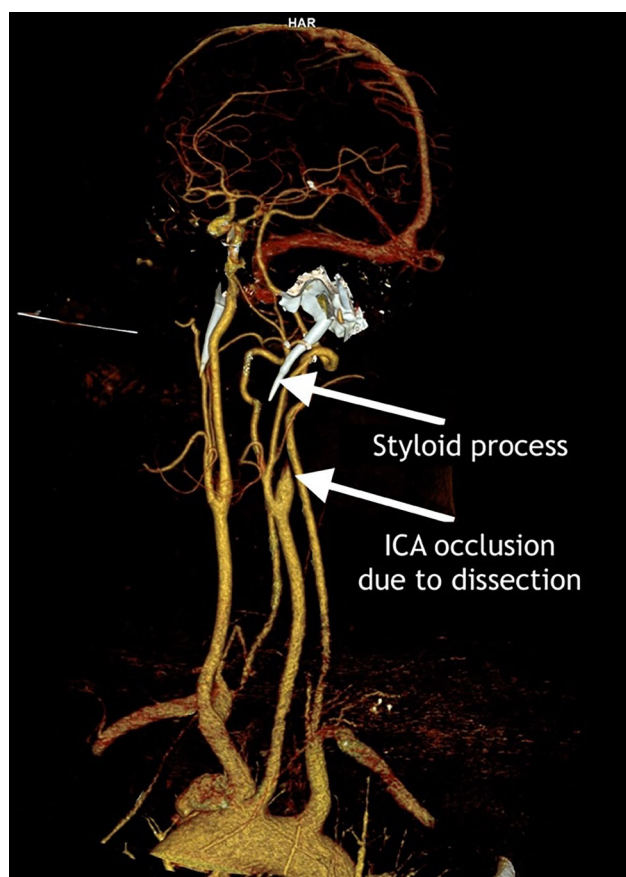


Fig. 4 Left ICA occlusion due to artery wall dissection caused by an elongated styloid process (3D-CTA model of neck structures)

nerves (V, VII, IX, X) by a calcified stylohyoid ligament due to a degenerative process or by a scarred ligament, usually seen as a complication after tonsillectomy. It is commonly accompanied by neck swelling, foreign body sensation and difficulty swallowing [5].

Even though an elongated styloid process can be found in about 4% of population, only 4% of these cases become symptomatic [4, 5]. The elongated process may put physical pressure on the sympathetic nerve plexus, as well as on the outer wall of the ICA [5]. Such irritation can cause general neurological symptoms (e.g., ipsilateral headaches), whilst in most severe cases the elongated process can cause a rupture or dissection of the carotid artery, thus promoting thromboembolisms in the subsequent cranial arteries, resulting in ischemic brain infarction [5]. On clinical examination, the elongated styloid process can be felt in the tonsillar fossa, and symptoms of irritation of the sympathetic plexus, such as Horner's syndrome (miosis, ptosis and enophthalmos) can be present [4].

The diagnostic procedure starts with a thorough patient history that includes family history, history of trauma and recent surgical procedures [4]. A detailed neurologic examination of facial and neck regions should be performed, as well as neuroimaging; panoramic radiography, head and neck CT scan, CT

angiography or MRI [5]. Out of all, 3D-CTA scan can most precisely measure the length of styloid processes (considered elongated, when longer than 3 cm) and determine the relation to the carotid arteries, as well as evaluate the ossification of stylohyoid ligament [5].

Depending on the type of the syndrome and the clinical picture, treatment can be either conservative or surgical. The decision about the most appropriate type of treatment depends on the symptom severity and the extent of adjacent tissue impairment. The surgical approach is mainly applied for severely elongated styloid processes and harmfully calcified stylohyoid ligament. When the patient presents only with orofacial pain, or cervical swelling, conservative management is the first line of treatment. Transpharyngeal application of analgesics, anaesthetic or steroid injection can relieve the pain and reduce the inflammation in the classical type [5]. In the carotid artery type the elongation of the processes is usually a bilateral manifestation, so once the relation to the adjacent neurovascular structures is determined, corrective as well as preventive surgical treatment is the best option [4].

Conclusion

Carotid artery type of Eagle syndrome (ESy) is largely an unknown condition affecting the cervical region [5]. While the classical type of ESy is more commonly seen in ENT or maxillofacial clinical setting, the carotid artery type falls predominantly in the neurological domain. Both can present with similar non-specific clinical picture, such as transient headaches or oropharyngeal pain, that may lead an inexperienced physician to dismiss the harmful symptoms, which can potentially result in severe and debilitating events, such as carotid artery dissection (CAD) and sequential transient ischemic attacks or acute ischemic stroke [4]. To prevent such deleterious consequences, physicians should remain vigilant as only timely recognition of such a syndrome allows appropriate (surgical) management to avoid possible premature death or loss of healthy life years due to ischemic stroke consequences.

Acknowledgements The authors would like to thank the neuroradiologist Igor Strahovnik, MD, for providing us with the relevant clinical images.

Conflict of interest K. Lakner and L. Savšek declare that they have no competing interests.

References

- Smajlović D. Strokes in young adults: epidemiology and prevention. *Vasc Health Risk Manag.* 2015;11:157–64. <https://doi.org/10.2147/VHRM.S53203>.
- Béjot Y, Delpont B, Giroud M. Rising stroke incidence in young adults: more epidemiological evidence, more questions to be answered. *J Am Heart Assoc.* 2016;5(5):e3661. <https://doi.org/10.1161/JAHA.116.003661>.

3. Sultan S, Elkind MS. The growing problem of stroke among young adults. *Curr Cardiol Rep.* 2013;15(12):421. <https://doi.org/10.1007/s11886-013-0421-z>.
4. Bokhari MR, Graham C, Mohseni M. Eagle Syndrome. In: StatPearls [Internet]. Treasure Island: StatPearls Publishing; 2022.
5. Moon CS, Lee BS, Kwon YD, Choi BJ, Lee JW, Lee HW, Yun SU, Ohe JY. Eagle's syndrome: a case report. *J Korean Assoc Oral*

Maxillofac Surg. 2014;40(1):43–7. <https://doi.org/10.5125/jkaoms.2014.40.1.43>.

Publisher's Note Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.