Eagle syndrome and carotid dissection

Dear Editor:

Eagle’s syndrome (ES) is described as orofacial pain related to the elongation and calcification of the stylohyoid process. Eagle defined two clinical presentations: first, classic stylohyoid syndrome, in the form of cervicofacial pain accompanied by dysphagia and foreign body sensation, secondary to affection of lower cranial nerves, and second, stylocarotid syndrome, due to carotid artery compression, which may be associated with a neurological focus and even syncope. Different pathophysiological mechanisms have been proposed and the first type has even been related with amygdalectomy. We report a case of Eagle’s syndrome associated with carotid artery dissection.

The case in question is a 45-year-old woman, under immunosuppressive treatment due to ulcerative colitis, non-amygdalectomized, with a history of bilateral laterocervical pain episodes radiating to the orbital region, of several months duration. She attended consultation due to a sudden, intense pain episode with features similar to the usual, predominantly left-sided, in the context of neck rotation and forced extension. It was accompanied by right brachio-crural hemiparesis, language alteration and self-limited clonic movements of the right lower limb.

Neuroimaging studies (computed tomography [CT] and cranial magnetic resonance imaging [MRI]) were initially normal, with the exception of an elongated, intensely calcified left styloid process. The vascular study (Doppler and angio-CT) showed an image indicative of left extracranial carotid dissection (fig. 1), with intramural haematoma, which was confirmed in the axial T1-T2 sequences of the cervical MRI. The 3D reconstruction revealed an intimate contact between the styloid process and the left carotid artery (fig. 2).

With this diagnosis, anticoagulant treatment was initiated upon the appearance of repeated episodes of amaurosis fugax in the left eye. At discharge, the carotid Doppler showed partial resolution of the dissection, and the patient was asymptomatic.

This treatment was maintained for 6 months, at which time the control angio-MRI showed complete resolution of the condition. The patient was also referred for maxilofacial surgery to assess surgical treatment, which was rejected due to clinical stability.

Figure 1 Magnetic resonance angiography of supra-aortic trunks: the image shows irregularity and reduced calibre in the postbulbar segment of the left internal carotid artery, indicative of dissection.

Approximately 4% of the population has an elongated styloid process (> 3 cm), although only 4 to 10.3% of them present symptoms. Several cases of self-limiting neurological focus have been reported in the literature in patients with ES, particularly in relation to head movements. Two of these were also associated with carotid artery dissection after forced and sustained head rotation, as in the case described. It seems possible to attribute a traumatic origin to the dissection.

In general, ES is suspected by symptoms and physical examination. Elongation and calcification of the styloid process is diagnosed through craniocervical radiograph and CT with 3D reconstruction, which shows if there is contact with adjacent structures.
In cases where there is concomitant neurological focus, it is advisable to perform a Doppler or dynamic angiography, which shows the carotid deterioration induced by head movements. In our case, the ultrasound study made an early diagnosis of carotid dissection possible.

An elongated process can be treated by surgical excision, preferably through an extraoral approach\(^\text{10}\). As in spontaneous or non-traumatic carotid dissections, treatment should be anticoagulation unless it is contraindicated.

Our patient is an example that ES may occur as a complication of acute carotid dissection. When faced with cervicofacial pain related to a neurological focus, it is important to consider carotid deterioration due to an elongated styloid process.