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Case Report

Horner Syndrome Secondary to Bilateral Spontaneous Internal Carotid Artery Dissection – First Manifestation of Fibromuscular Dysplasia

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Abstract

Horner's syndrome, also known as Bernard–Horner syndrome, is characterized by miosis, ptosis, with or without anhidrosis. This occurs due to affection of ipsi-lateral sympathetic trunk. Our patient, a 45-year-old healthy man, who presented with a Horner's syndrome, was found to have a bilateral spontaneous internal carotid artery dissection. Systemic disease investigation concluded of a fibromuscular dysplasia. Fibromuscular dysplasia is a recognized risk factor for multiple cervical artery dissection and the Horner's syndrome may be the presenting sign in cerebromuscular fibromuscular dysplasia. This case demonstrates that Horner's syndrome it's a relevant clinical sign and in its presence cervical artery dissections should be considered in the differential diagnosis and an underlying systemic abnormality should be investigated with appropriate diagnostic vascular imaging.

Keywords: *Horner Syndrome; Spontaneous Carotid Artery Dissection; Multiple Cervical Artery Dissection; Fibromuscular Dysplasia*

Abbreviations

FMD: Fibromuscular Dysplasia; ICAD: Internal Carotid Artery Dissection

Introduction

Fibromuscular dysplasia (FMD) is a non-inflammatory and non-atherosclerotic arterial disease that involves small and medium-sized arteries. Disease manifestation depends on the arterial bed involved. Arterial dissection may result in devastating outcomes for the typically young and otherwise healthy FMD patient, such as stroke or myocardial and renal infarctions [1,2]. There have been a number of publications in recent years showing it is not uncommon for FMD patients to have multiple artery dissection, including multiple spontaneous cervical artery dissections [2-5]. Clinical manifestations of internal carotid artery dissection (ICAD) include neck pain, headache, focal cerebral ischemic symptoms, cranial nerve palsies and also ocular complications such as Horner syndrome, ophthalmoplegia, ischemic optic neuropathy and arterial vessel occlusions. In patients with ICAD, Horner syndrome is thought to result from compression of the pericarotid sympathetic plexus [6].

We report a case of a man with a bilateral spontaneous carotid dissection due to FMD, which presented with a Horner's syndrome on the ophthalmology emergency department. This case shows the importance of an ophthalmological sign to raise the suspicion and further investigation of a systemic vascular disease.

Case Presentation

We examined a 45-year-old healthy man, ex-smoker, 3 days after appearance of palpebral ptosis and miosis of the right eye. He also reported prior episodes of right cervical pain, since 1 week ago. He denied physical efforts and jerky head movements. His visual acuity was of 10/10 in both eyes. There was no relative afferent pupillary defect and ocular motility was normal. He presented anisocoria with miosis and palpebral ptosis of the right eye accompanied by anhidrosis of the ipsilateral hemiface (Figure 1). Anterior segment and fundus examination was unremarkable in both eyes. There were no focal deficits on neurological examination.

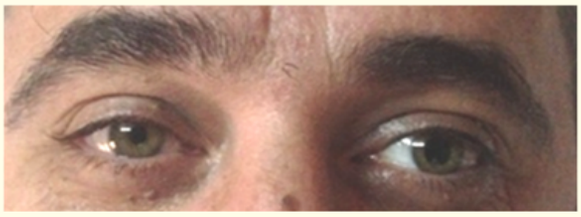

Figure 1: *Anisocoria with miosis and palpebral ptosis of the right eye.*

Supra-aortic trunks angiotomography revealed bilateral distal dissection of the internal carotid arteries (Figure 2). The patient was hospitalized, placed on strict bed rest and treated with dual antiplatelet therapy with aspirin 100 mg and clopidogrel 75 mg. Antihypertensive agents were also prescribed to lower high blood pressure that he presented during the hospitalization. No neurologic deficits developed. Transcranial doppler ultrasonography showed a collateral supply through the posterior communicating arteries and the external carotid arteries, with the inversion of the flow in the ophthalmic arteries.

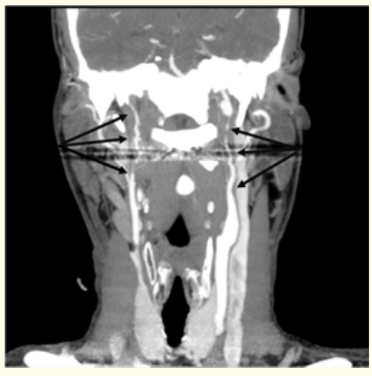

Figure 2: *Supra-aortic trunks angiotomography image showing distal dissection of the internal carotid arteries (arrows).*

During systemic investigation FMD was diagnosed based on renal magnetic resonance angiography that showed irregular aspect of the distal 2/3 of the right renal artery, with a moderate tapering of its distal third and proximal branches (Figure 3).

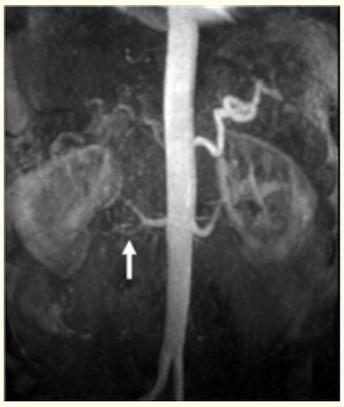

Figure 3: *Renal magnetic resonance angiography image showing irregular aspect of the distal 2/3 of the right renal artery (arrow), with a moderate tapering of its distal third and proximal branches.*

Due to the clinical stabilization, the absence of symptoms and the serial exams transcranial and cervical doppler ultrasonography showing a mild hemodynamic improvement, Horner syndrome was felt to be an isolated clinical finding, and so our recommendation was to keep treatment with dual antiplatelet therapy and with antihypertensive agents.

The patient opted for follow-up in a private hospital.

Discussion

The case reported of a patient who presented to the ophthalmologic emergency department with clinical findings of a Horner syndrome and which investigation concluded a bilateral spontaneous internal carotid artery dissection due to fibromuscular dysplasia is consistent with some previous reports. Although most patients with ICAD present with cerebral or retinal ischemic symptoms, suspicion of ICAD should also be aroused by local symptoms such as headache, neck pain and cranial nerve signs including hypoglossal nerve lesions or Horner syndrome [7]. Horner syndrome is thought to result from compression of the pericarotid sympathetic plexus [6]. Multiple studies showed that a significant proportion of patients with ICAD had Horner syndrome (8/32 = 25% [8], 29/78 = 37% [9], 234/533 = 44% [10], 191/496 = 38,5% [11]).

Early recognition and initiation of therapy may prevent or improve neurological deficits and avoid the potentially devastating complications of carotid artery dissection. Recent advances in noninvasive imaging have made the diagnosis easier, and magnetic resonance/computed tomography angiography are now emerging as the primary imaging modalities [12]. Antiplatelets and anticoagulation remain standard therapy for ICAD. However, in patients with either expanding pseudoaneurysms, severe flow compromise, worsening symptoms despite anticoagulation or contraindication to anticoagulation, endovascular stenting may be beneficial [13,14].

There is no explanation in the literature for carotid arteries to dissect bilaterally and spontaneously, with no obvious precipitant or risk factor [7]. FMD has been associated with multiple cervical artery dissections. A recent publication based on the U.S. Registry for FMD, with data from 921 patients, described that multiple cervical artery dissections were common in patients with FMD: of 177 patients with extracranial carotid or vertebral artery dissection, 66 experienced dissection of more than 1 cervical artery (carotid and/or vertebral) [2].

Our patient systemic investigation concluded of FMD as the main cause of the bilateral spontaneous ICAD, in which Horner's syndrome was the presenting sign and the first manifestation of the disease. Horner syndrome was reported as an uncommon manifestation of cerebrovascular FMD; it was the presenting sign in only 4,7% of patients in the first report of the U.S. Registry for FMD, involving 447 patients [15,16].

This case reports a bilateral spontaneous carotid dissection due to FMD, which presented initially with a Horner syndrome. We described this case to show that Horner syndrome may be a relevant clinical sign and the first manifestation of serious systemic diseases and ophthalmologists should be aware to make the correct diagnosis, warranting further investigation and diagnostic vascular imaging.

Conflict of Interest

None.

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