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Case Report

Atypical fibromuscular dysplasia or carotid web revealed by cerebral infarction: A review of 2 cases ☆

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ABSTRACT

Atypical fibromuscular dysplasia of the bulb or carotid web is a nonatheromatous pathology more common in African and African-American populations. It is implicated in the occurrence of cerebral infarcts of unknown causes. Its diagnosis is made by angio-CT of the supra-aortic trunks and is characterized by a defect in the posterior wall of the bulb. Treatment with antiplatelet agents prevents the occurrence of stroke, but radical treatment remains surgical and endovascular. We report 2 observations of carotid web diagnosed and medically managed at the regional hospital of Saint Louis.

Introduction

Carotid web, also known as atypical focal fibromuscular dysplasia of the carotid bulb is a proliferative, nonatheromatous, noninflammatory process involving the intima of the extracranial carotid system [1].

The condition is characterized by the presence of a linear membrane extending from the posterior wall of the carotid bulb into the lumen beyond the bifurcation [2]. Its prevalence ranges from 0.3% to 3.2% according to a study of supra-aortic angiograms [3]. It is increasingly described as a rare cause of cerebral infarction, particularly in African and African-American populations [2,3].

Indeed, atypical fibromuscular dysplasia is classified in the nosological context of undetermined causes of stroke, which represent 9.4%-37% of cryptogenic strokes [4,5]. The CT-angiogram of supra-aortic arteries is the best noninvasive imaging modality to perform and helps set out differential diagnosis [4].

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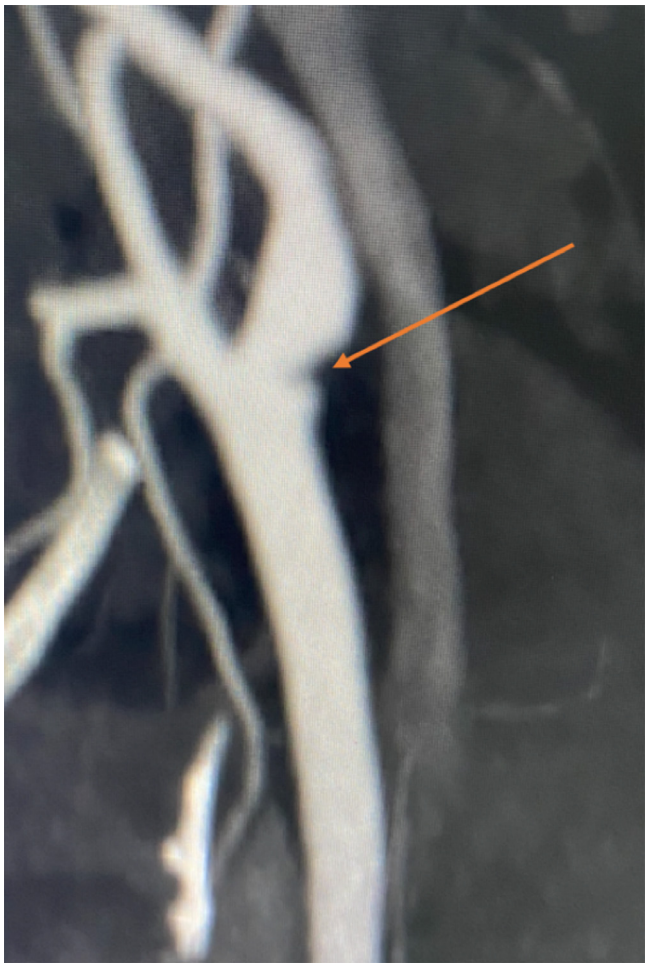

Fig. 1 – Maximum intensity projection reconstruction of the right internal carotid artery showing a bulbar diaphragm (orange arrow).

In this work, we report 2 cases of cerebral infarction occurring in women probably due to a carotid web.

Case 1

A 30-year-old woman with no particular medical history was admitted to the emergency room for aphasia and left upper limb weakness.

Neurological examination revealed a left monoparesis and motor aphasia, with an NIHSS score of 8. The rest of the physical examination was unremarkable.

The full blood count (Hb = 12.1 g/dL; platelets = 410,000/mm^3), prothrombin level (79%), and INR (1.16) were normal. The electrocardiogram and cardiac ultrasound were unremarkable. The complete cholesterol test was normal.

The CT scan showed a superficial right middle infarction. The multiphase CT angiography showed a bilateral bulbar defect involving the posterior walls of the internal carotid arteries with no evidence of plaque. This finding was suggestive of atypical fibromuscular dysplasia (Figs. 1 and 2).

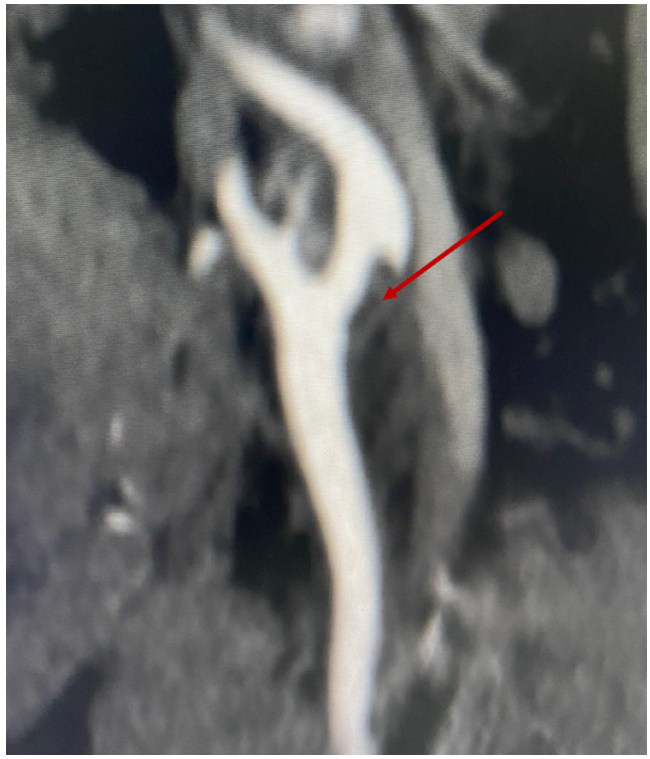

Fig. 2 – Sagittal reconstruction of the left common and internal carotid arteries showing a bulbar defect at the posterior wall of the carotid bulb (red arrow) with no visible plaque related to a carotid web.

Treatment with antiplatelet agents was initiated. The clinical outcome was unremarkable with an NIHSS score that had decreased from 8 to 5 at discharge.

Surgical treatment was proposed but the patient did not consent.

Case 2

A 54-year-old female under amlodipine 10 mg for 5 years had suddenly presented with a speech disorder and a right-sided weakness.

On admission, the neurologic examination revealed a right hemiplegia, expressive aphasia, and an NIHSS score of 11. The rest of the physical examination was unremarkable.

The full blood count (Hb = 13 g/dL; platelets = 236,000/mm^3), prothrombin level (87%), and INR (1) were normal. The electrocardiogram and cardiac echography were unremarkable, as well as the complete cholesterol test.

The CT scan showed a left middle cerebral artery infarction. The angio-CT of the supra-aortic arteries showed a left bulbar defect without visible plaque suggestive of a carotid web (Fig. 3).

Treatment with antiplatelet agents was initiated. The clinical evolution was unremarkable with an NIHSS score of 5 at discharge.

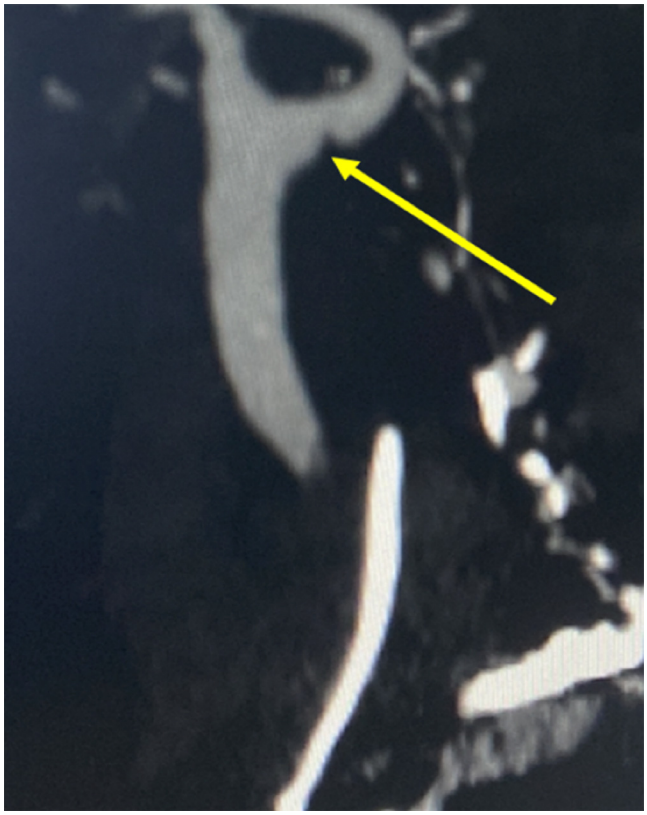

Fig. 3 – Parietal defect in the posterior wall of the left carotid bulb (yellow arrow) corresponding to a carotid diaphragm on a maximum intensity projection reconstruction.

Discussion

Atypical fibromuscular dysplasia was first described in 1968 by Rainer in a young woman with transient ischemic attacks [6]. Its prevalence ranges from 0.3% to 3.2% [3] and it has been implicated in undetermined causes of stroke accounting for 9.4%-37% of cryptogenic strokes [4,5]. However, data are scarce in sub-Saharan Africa, and the most consistent cohort published to date consists of 6 patients [7]. The related-risk factors to its occurrence are yet to be clearly understood, and the roles of endogenous (hormonal, genetic) or exogenous (tobacco, microtrauma) factors remain controversial [8,9].

It is a condition that is more common in young African or African-American subjects [2,10]. Dysplasia is increasingly recognized as a cause of cerebral infarction if it occurs in a relatively young person with few or no vascular risk factors, likewise in our 2 patients. Indeed, in our 2 observations, despite the first-line etiological assessment, only the carotid web was outlined.

Ischemic stroke manifestations may result from a thromboembolic or hemodynamic mechanism. CT angiography is a noninvasive imaging modality that can help to exclude alternative diagnoses such as carotid plaques and arterial dissection [4].

This abnormality appears as a defect in the posterior wall of the carotid artery on sagittal reconstructions.

Although interventional management includes carotid endarterectomy surgery and endovascular treatment (stenting) [2,4], our patients had been treated with antiplatelets meanwhile curative treatment.

However, stroke occurring in carotid web patients is often refractory to antiplatelet therapy, with a recurrence rate of 30% [10].

Conclusion

Atypical fibromuscular dysplasia of the bulb is a recently described carotid pathological entity that is specific to the young African-American subject and is a nonatheromatous pathology that can lead to recurrent cerebral infarction.

The CT-angiogram of supra-aortic arteries helps make the diagnosis and also sets out differential diagnoses. Antiplatelet therapy can prevent cerebral infarction but surgical and endovascular treatment are the best approach to avoid recurrence.

Patient consent

The patients' consent was obtained.

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