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Case Report

Recurrent stroke in young adults caused by atypical fibromuscular carotid dysplasia[☆]

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ABSTRACT

Background: Carotid Web and focal carotid diaphragm are atypical fibromuscular dysplasia. The bilateral stroke due to this dysplasia is extremely rare. We will report a series of three young patients, admitted for a bilateral ischemic stroke caused by carotid bulb web and internal carotid diaphragm. Also, we will discuss their manifestations and treatment modalities.

Case presentations: In our study, we will report a series of three North African patients, two females and one male, at the mean age of 37, admitted for an ischemic stroke caused by bilateral carotid bulb web and bilateral internal carotid diaphragm. All of our patients were young and didn't have a history of drug use.

Conclusions: In our series, only end-vascular treatment was performed which was necessary to prevent any recurrence. Antiplatelet therapy was used in all cases to prevent any stroke during the follow-up.

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Introduction

Bulbar carotid web (CW) - also known as carotid intimal variant fibromuscular dysplasia - and focal carotid diaphragm are

rare causes of ischemic stroke in young adults. The incidence of carotid ischemic stroke due to carotid dysplasia is approximately 3.8 per 100,000 person-years.

Bilateral stroke due to carotid focal dysplasia was never been reported before.

Abbreviations: CW, Carotid Web; NIHSS, National Institute of Health Stroke Scale; MRI, Magnetic Resonance Imaging.

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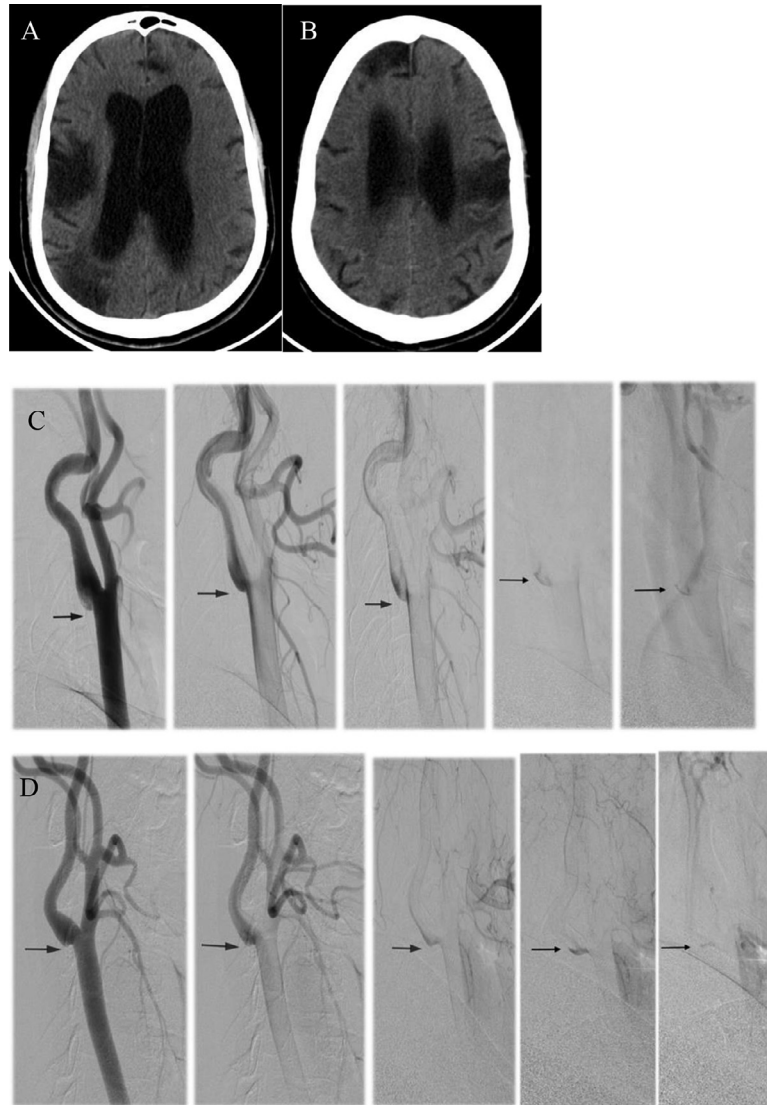


Fig. 1 – (A, B) Cerebral CT scan: bilateral frontal and right parietal low cerebral densities consistent with ischemic strokes. (C and D) Upper aortic vessels arteriography: “Shelf-like filling defect” in left (C) and right (D) proximal internal carotid artery consistent with Carotid Web. The contrast injection clears the internal carotid artery on sequential alternating frames, it shows the contrast stagnation (arrow) which is appreciated in the distality of the carotid Web.

We relate 3 rare cases of bilateral stroke in young adults caused by an atypical form of carotid fibromuscular dysplasia, we will also discuss their endovascular treatment and follow-up.

Case presentations

Case 1

Thirty-six years old male, with a history of unexplored facial paralysis for 2 months ago, was admitted to our neuroradiology service for investigation due to recurrent stroke.

Initial examination revealed left hemiplegia and dysarthria at T = 3 hours, Glasgow Coma scale was at 15. The National In-

stitute of Health Stroke Scale (NIHSS) score was 17. Emergency brain CT scan imaging showed cerebral low densities consistent with multifocal ischemic strokes (Fig. 1).

On the first investigations, the patient had benefited from an electrocardiogram and cardiac ultrasound which doesn't show abnormalities.

After, due to young age and the bilateral localization of ischemic lesions, cerebral vasculitis was suspected.

Inflammatory biological and spinal liquid analyses were done and were negative.

On the second line of investigation, cerebral and upper aortic vessels angiography showed a proximal internal carotid web revealed by a “shelf-like filling defect”. Contrast stagnation on the pocket of the carotid web determines its severity (Fig. 1 C and D). Furthermore, there was no circle of Willis abnormalities.

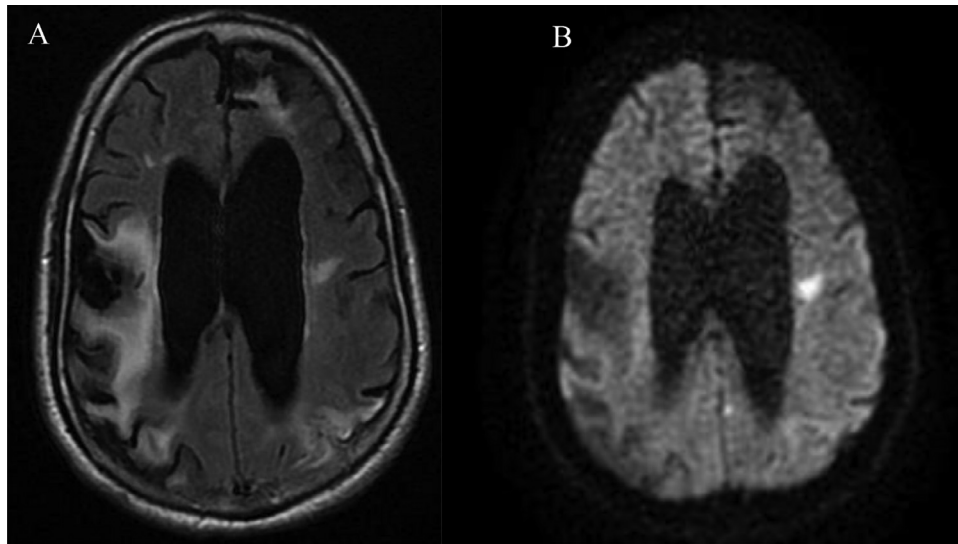


Fig. 2 – Cerebral MRI on FLAIR (A) and diffusion (B) sequences high signal intensity which was compatible with a significant ischemic lesion on the left semioval centrum.

In this case, after ruling out all the differential diagnoses, especially vasculitis, we concluded that multifocal ischemic strokes are caused by bilateral carotid dysplasia.

Initially, an intravenous recombinant tissue plasminogen activator (IV-rtPA) was administered without any improvement of neurological symptoms.

The patient has rejected endovascular treatment, so he has benefited from double antiplatelet therapy.

After 18 months, the patient was admitted for a second recurrence of right hemiplegia at T = 8 hours. Examination revealed a NIHSS of 13.

MRI showed significant ischemic lesions on the left centrum semiovale on FLAIR and diffusion sequences (Fig. 2). Arteriography showed a thrombus attached to the left bulb carotid web.

Thus, an emergency stenting on the carotid Bulb to exclude the dysplasia was necessary. A monorail delivery system Carotid WALLSTENT (Boston Scientific) covered device 7 × 40 mm was deployed. A satisfying result was obtained (Fig. 3).

After, the patient was admitted for second stenting after 10 days to treat his contralateral carotid Web (Fig. 4).

On his follow-up, the patient's neurological deficit gradually improved with a NIHSS of 5, he was transferred to the rehabilitation center. He also benefited from an antiplatelet therapy, which was 100 mg Aspirin and 75 mg Clopidogrel.

After 4 months of follow-up, no episodes of recurrence have been reported. However, he still has some residual neurological deficit.

Case 2

We report a case of a 36-year-old female, without a history of illness or drug use. She had a transient ischemic stroke 6 months ago.

She was admitted to the Neuroradiology department for right hemiplegia at T = 16 hours.

First, cerebral MRI showed a right and left parietal heterogeneous low intensity with negative mass effect, associated with some gliosis (Fig. 5A and B).

The patient was hospitalized and closely monitored. Oral double antiplatelet therapy was administered. She also had a cardiac ultrasound and an electrocardiogram to rule out an embolic cause due to cardiac vegetation or fibrillation.

Then, she had a cervical and cranial arterial CT scan, showing a thrombus attaching to the right bulb (Fig. 5C and D). On the left carotid bulb, the CT scan didn't show any abnormality which was a false negative.

After the patient was admitted to interventional radiology service, he benefited from a cerebral angiography which revealed a bilateral dysplasia of the carotid bulb (Fig. 6A and C).

Angiography also revealed digital endoluminal subtractions defect in the left proximal internal carotid bulb artery consistent with the carotid web. Thus, we confirmed that a CT scan with multiplanar reconstructions can be misinterpreted as a false negative (Fig. 6C)

On the first line of treatment, the patient had intravenous unfractionated heparin at 1000 U/H. After 20 days, medical treatment was followed by deferred endovascular stenting of the right carotid bulb with a Carotid WALLSTENT non-covered device 7 × 40 mm (Fig. 6B).

Scheduled stenting of the contralateral carotid bulb was performed after 1 month with satisfying clinical and angiographic control. After an improvement of his symptomatology, the patient was discharged with antiplatelet therapy of Aspirin and Clopidogrel with an NIHSS of 0.

Follow-up at 6 months didn't show any recurrence of ischemic stroke.



Fig. 3 – Angiography of the left carotid bulb. (A) Carotid web on the left bulb with irregularity in the carotid wall consistent with bulbar carotid Web (arrowhead). (B) Endoluminal reconstruction with a Carotid WALLSTENT: Angiogram revealing a post-stenting carotid wall. It shows the endoluminal reconstruction of the left bulbar carotid and the exclusion of the web and the thrombus attached to the carotid wall.

Case 3

We report the case of a 40 years old female, with a history of hypertension and right ischemic stroke dating from 1 year back, she was being treated with preventive antiplatelet and antihypertensive therapy.

She was admitted to neuroradiology service for a left hemiparesis at T = 2 hours.

The clinical exam revealed a National Institute of Health Stroke Scale (NIHSS) of 9.

An emergency cerebral MRI was performed and showed a left frontal high intensity on FLAIR sequences with a quiet mass effect (Fig. 7 A and B).

After this recurrence, a complete cardiovascular investigation has been done, it doesn't show any abnormality.

Angiography of the upper aortic vessels showed a bilateral defect of proximal internal carotids consistent with a focal diaphragm (Fig. 7 C and D).

An emergency endovascular stenting of the internal carotids, with a Carotid WALLSTENT non-covered device

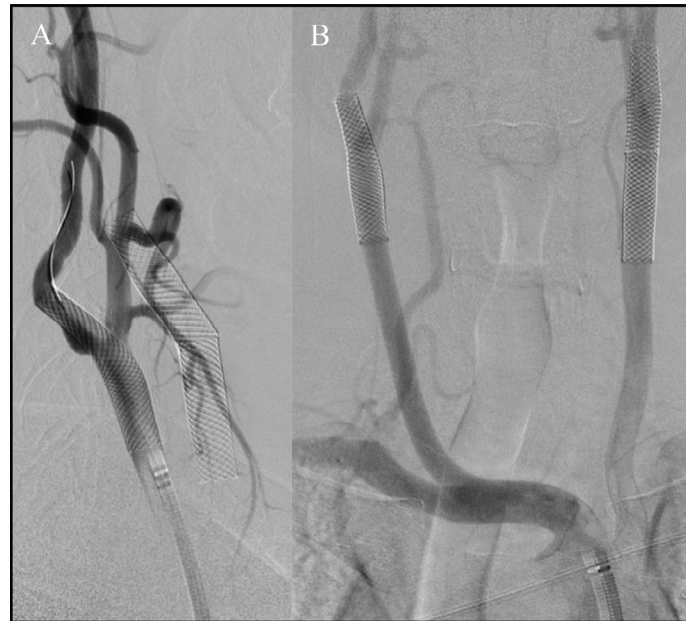


Fig. 4 – Angiography of the right carotid bulb. (A) Endoluminal reconstruction with a Carotid WALLSTENT deployed on the right carotid bulb. (B) Angiography reveals a bilateral post-stenting carotid wall in both carotid bulbs. It shows a satisfying endoluminal reconstruction wall.

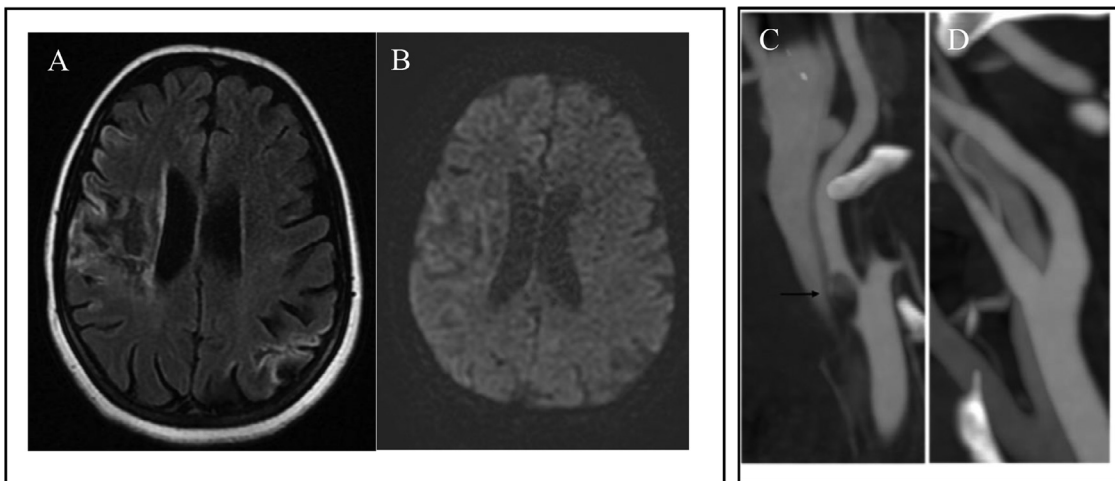


Fig. 5 – (A, B) cerebral MRI: right frontoparietal and left parietal heterogeneous low intensity with negative mass effect. (C, D) Upper aortic vessels CT scan: thrombus attaching to the right bulbar dysplasia. CT scan showed a normal left Bulb which was a false negative.

7 × 40 mm was done without any complications. Heparin with statin and antihypertensive treatment was maintained during her hospitalization, followed by a lifelong double antiplatelet therapy.

Follow-up after 3 months didn't show any recurrence, but the patient kept a left-sided weakness.

Discussion

Atypical fibromuscular carotid dysplasia is increasingly seen as a cause of ischemic stroke in young adults with a prevalence of 3.8 per 100,000 person-years [1,2]. Bilateral forms of

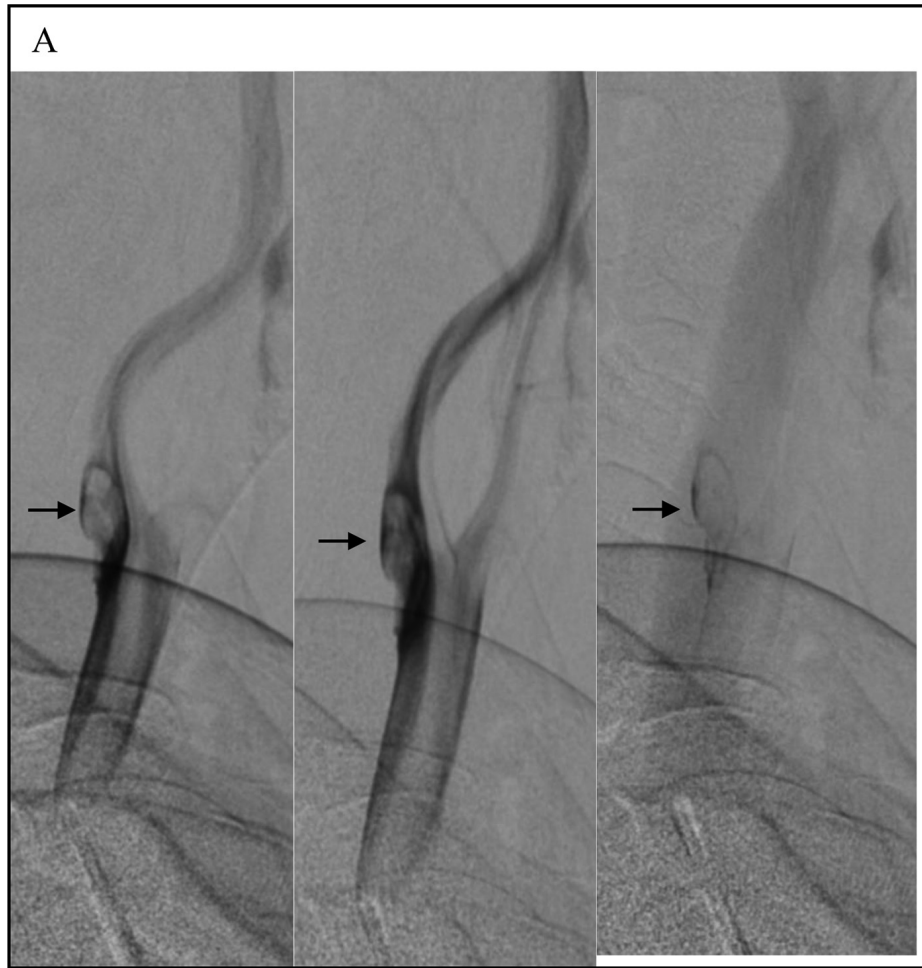


Fig. 6 – (A) Angiography of the right carotid bulb: showing a thrombus attached to the right bulbous dysplasia (arrow) with the stagnation of contrast. (B) Endoluminal reconstruction with a carotid wall stent: angiogram revealing a carotid poststenting which shows an endoluminal reconstruction of the right bulbous carotid (arrowhead) and the persistence of a minor contrast stagnation; (C) Digital endoluminal subtractions defect in the bulb of left proximal internal carotid artery consistent with CW (arrow), which were not detectable at CT scan.

focal carotid dysplasia leading to a recurrent stroke as we saw in the 3 of our cases have not been previously reported in the literature.

The most common form of FMD in typical cases is the “diaphragm”, the carotid web is a rare form of FMD. Demographic analysis shows that carotid Web occurs more in women with an incidence of 67%. This form was reported in recent series in Black or Asian patients with only 3 recent retrospective studies [1,3–5], also, in Canadian and Afro-Caribbean patients [6,7].

Carotid dysplasia is commonly associated with some risk factors such as hypertension, oral contraceptives, and smoking [5]. In our series, only one patient had hypertension and the mean age was 38.

Histopathology of the carotid web revealed intimal fibrous proliferation without atherosclerosis. (2.5.6)The ischemic stroke occurs due to flow stasis and thrombus forma-

tion in the pocket of the carotid Web, which may be the source of homolateral emboli [8,9]. The mechanism of stroke due to FMD appears to be an arterial embolism due to a thrombus that was expelled into the pocket of the FMD [6,7,10].

Imaging shows that carotid Web has a particular appearance, however, there is an important variability in their type and form. So, these lesions can be misinterpreted as atheroma. We note that these lesions may coexist in some patients. Studies reported some false negatives diagnosis, they showed that CT scan with MPR reconstructions may lead to misdiagnosis [11]

Endovascular treatment by stenting generates an immediate luminal reconstruction with a deviation and redirection of the carotid flow. It is followed by re-endothelialization because the excluded CaW pocket is resorbed gradually, probably due to laminar thrombosis and vascular remodeling [12,13].

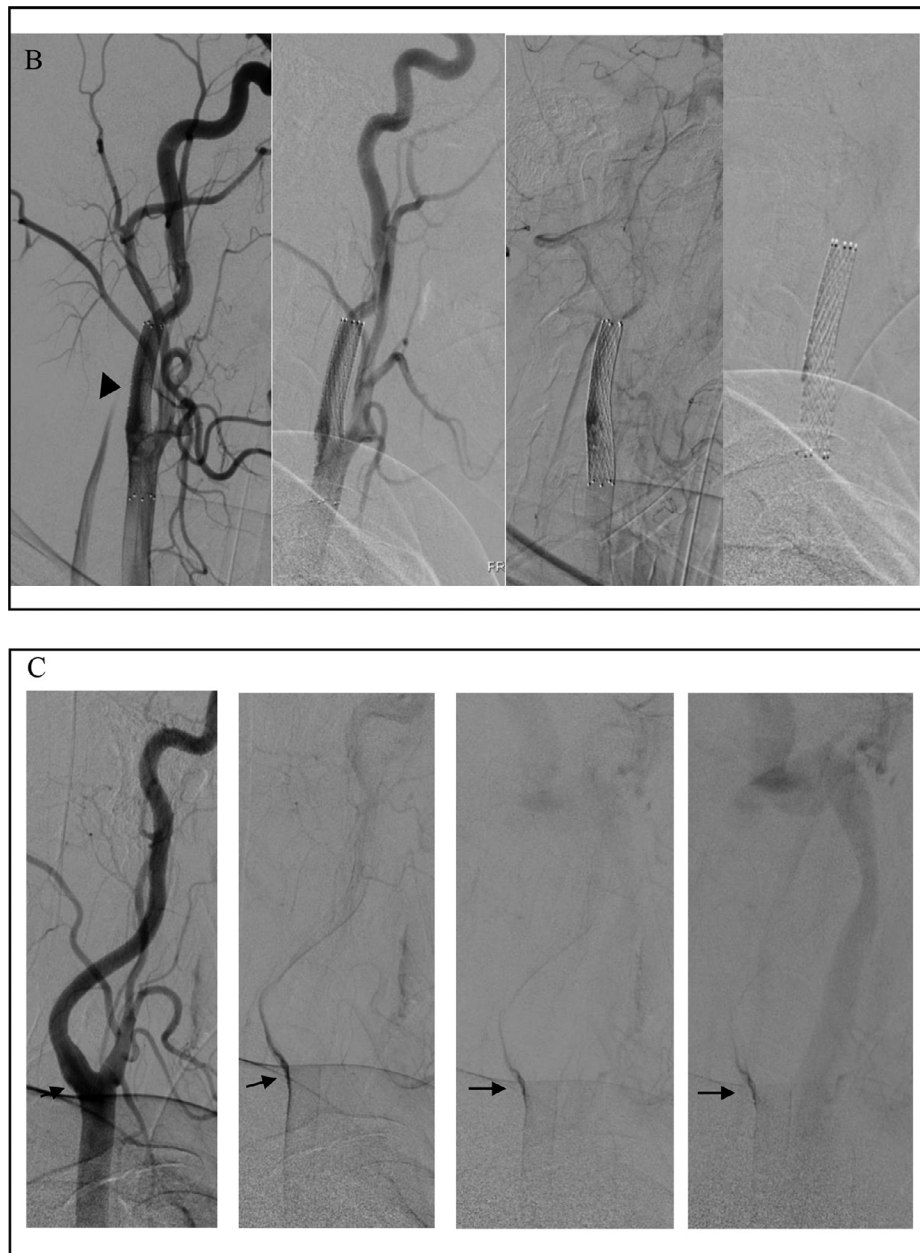


Fig. 6 - Continued

On the other hand, surgical resection of the CaW was firstly reported in 1968 and has been widely performed in cases of the carotid web [2-4,14,15]. It has been described as safe and potentially efficient. But stenting remains less invasive and does not require general anesthesia in most cases [16,17].

The most appropriate strategy of treatment for patients with symptomatic CaW still appears to be an unresolved issue. Medical treatment based on conservative antiplatelet

monotherapy is known to be linked to a high risk of recurrent stroke. Three studies in the literature have shown that recurrent ischemic stroke occurs in 30%-71% of patients on antiplatelet monotherapy [2,14,18].

Despite the high rates of stroke recurrence that have been consistently demonstrated in observational studies, many ambiguities exist. The difference between anticoagulation and revascularization is still unknown, despite the risk of recurrent events on antiplatelet therapy [18].

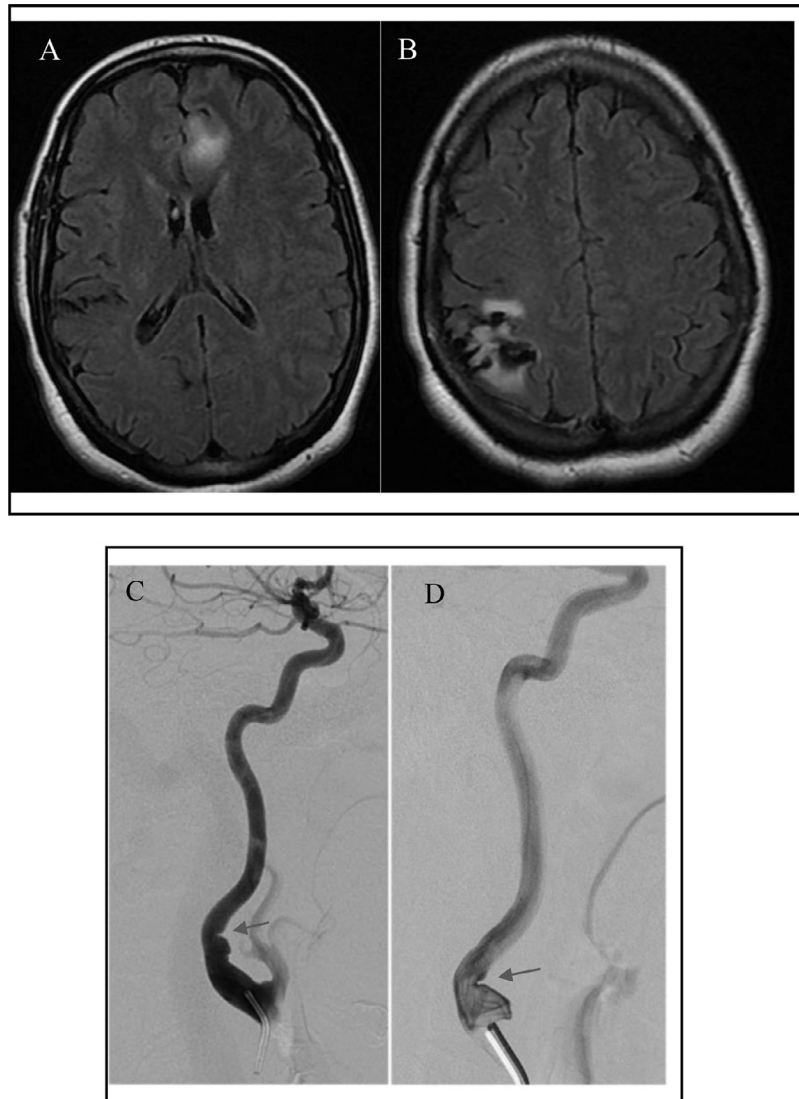


Fig. 7 – (A, B) Cerebral MRI in FLAIR sequences: another bilateral ischemic stroke that shows right frontoparietal heterogeneous low intensity with negative mass effect and left frontal median high intensity according to the new ischemic event. (C, D) Upper aortic vessels angiography: defect in the right and left proximal internal carotid consistent with the diaphragm (arrows).

Conclusion

The incidence of cryptogenic cerebrovascular stroke in young adults with FMD is high. Bilateral stroke due to FMD has never been reported before.

Our observations have shown the main characteristics of bilateral carotid Web and focal carotid diaphragm leading to ischemic stroke.

This dysplasia must be treated properly, either by antiplatelet therapy, stenting, or endarterectomy, or its recurrence is unavoidable.

Patient consent

Written informed consent was obtained from the patient's legal guardian for publication of this case report and any accompanying images.

Availability of data and materials

The data sets are generated on the CHU Hassan II data system of Fes, including the biological data and the interventional report.

Author's contribution

YC is the corresponding author, he participated in the organization and writing of the article and studying the cases with GS.

Professor MY.AL did the angiography of all the cases.

Professor SB, NC, M.F B, contributed to clinical examination hospitalization and follow-up of the cases, also medical treatment.

Professor NE, MH, BA supervised working and validated the figures.

Professor and chief of the department of radiology MB and MM read and allowed the article for publication.

All authors read and approved the final manuscript.

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Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:[10.1016/j.radcr.2022.07.081](https://doi.org/10.1016/j.radcr.2022.07.081).

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