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Therapeutic and Diagnostic Implication of Inadequate Circle of Willis in a Patient with Acute Spontaneous Bilateral Internal Carotid Artery Dissection: Case Report and Review of the Literature

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Key Words

Spontaneous arterial dissection · Internal carotid artery · Cough · Stroke · Circle of Willis

Abstract

Background and Purpose: Emphasizing the therapeutic and diagnostic implications of concomitant inadequate collaterals from the circle of Willis in a rare case of spontaneous acute bilateral internal carotid artery dissection (BICAD) following 5 days of isolated rigorous cough (pertussis like).

Case Description: A 45-year-old male has been referred to our department with rapid neurological deterioration consisting of dysarthria and severe left hemiparesis following 5 days of isolated rigorous cough. CTA demonstrated BICAD, a tiny anterior communicating artery and no bilateral posterior communicating artery. The patient had no personal or familial risk factors. Infectious, traumatic, vascular and connective tissue diseases were ruled out.

Results: Neurological deterioration persisted despite immediate provision of continuous ‘full-heparinization’ with concomitant rigorous control of blood pressure. Endovascular treatment consisting of bilateral stenting was undertaken. Ten days later, the patient was discharged with mild hemiparesis and resuming normal activity after 3 months.

Conclusions: BICAD with concomitant inadequate collaterals from the circle of Willis may predispose to hypoperfusion which might not respond to the usual conservative treatment prompting for flow reestablishment. Moreover, isolated rigorous cough can cause acute spontaneous BICAD even among patients without any risk factors.

Introduction

Spontaneous cerebral arterial dissections (CAD) are more frequent among young patients (~40–45 years of age [1, 2]), of which the internal carotid artery (ICA) are one of the major sites for dissections [1]. CAD may be traumatic or spontaneous. Spontaneous CADs are generally related to: connective tissue diseases [fibromuscular dysplasia [3] in up to 15–50% of bilateral internal carotid artery dissection (BICAD) [4], Ehlers-Danlos syndrome, Marfan syndrome, polycystic kidney disease, osteogenesis imperfecta, and systemic lupus erythematosus] and/or preceding infectious diseases. The impacts of atherosclerosis and aneurysm are yet debatable [5]. Some spontaneous CADs follow mild efforts as neck maneuvering, forceful cough and vomiting [6].

Internal carotid artery dissections (ICADs) may account for up to 2.5% of all first strokes and are responsible for ~5–20% [4, 6] of all ischemic strokes (the commoner type among young patients [2]) in adults. Among all ICAD, BICAD is as frequent as 5–21% despite possible initial presentation with unilateral symptoms [7–9]. ICAD without subarachnoid hemorrhage (SAH) are more frequent in younger patients (<30 years of age), while vertebro-basilar dissections with SAH are more common in older patients (>30 years of age) [1]. Spontaneous acute BICAD is sparsely reported.

Common presenting features of spontaneous ICAD include: headache, ipsilateral neck pain, incomplete Horner's syndrome (ptosis and myosis), bruits, transient ischemic attack/cerebrovascular accident signs as amaurosis fugax, syncope, encephalopathy and focal signs. These might be explained by either embolic event and/or reduced flow distal to the dissected segment and/or concurrent SAH. Young patients might exhibit great variability [4, 7, 8]. Most cases can be diagnosed by MRI, MRA, and CTA; however, angiography is still needed for some cases.

Treatment consists of anticoagulation/antiplatelet therapy, while endovascular or surgical interventions generally are indicated for intradural cases with SAH or when persisting clinical deterioration is noted despite anticoagulation.

A case of simultaneous spontaneous acute BICAD concomitant with inadequate collateral blood supply from the circle of Willis, preceded by isolated rigorous cough in a patient without any risk factor or infectious disease was not reported previously and is hereby presented, emphasizing the therapeutic implication of these concomitant findings.

Case Report

A 45-year-old man who suffered from paroxysmal severe nonproductive cough lasting for 5 days before his admission was referred to our emergency room from another hospital. Family and personal history were noncontributory. By the fifth day, the patient developed acute dysarthria and left hemiparesis. A BICAD with severe stenosis was demonstrated on CTA. A tiny anterior communicating artery (ACOM) with no bilateral posterior communicating artery (PCOM) was demonstrated (fig. 1a, b, brain angiography). Following the CTA, immediate administration of heparin was initiated.

On his arrival to our emergency room, the patient's neurological findings consisted of dysarthria, left hemiparesis (hand pronation) and bilateral Babinski signs. The patient was alert, hemodynamically stable, and no fever was recorded. No regular medications were noted in his file. His blood tests were normal except for prolonged PTT (>120 s) due to previous heparin administration. The patient was transferred to the neurointensive care unit for the continuation of

heparin treatment, maintaining a PTT range of 60–70 s with concomitant rigorous regulation of blood pressure (systolic 110–140 mm Hg).

Persisting clinical deterioration for the next following hours despite anticoagulation was noted: the patient developed neglect and became hemiplegic. No signs of bleeding and/or infarcts were showed on CT. He was taken to the angiography room: under general anesthesia four vessels angiography was performed demonstrating near complete occlusion of the right ICA and severe stenosis of the left ICA due to BICAD in the cervical area ([fig. 2a, b](#)). Stenting of both ICA (two stents each side) was performed restoring flow bilaterally with normal parenchymal blush ([fig. 1a, b](#), [fig. 3](#)). A small ACOM with mild flow from left to right was demonstrated, while restoring the flow in the left hemisphere. The PCOM was not open in neither side. Following stenting, the patient received IIb/IIIa-antagonist further converted to clopidogrel and aspirin by the next day, and heparin treatment was discontinued. Two days following stenting, a subcortical small watershed infarct was demonstrated on a CT (in the para-midline ACA-MCA passage) further supporting that hypoperfusion was the underlying cause ([fig. 4](#)).

A thorough workup for venous-arterial and connective tissue diseases, both personal and familial, was noncontributory. Gradual improvement was noted and the patient was discharged after 10 days, showing further improvement: fully conscious, able to stand and walk with some support, less prominent neglect and dysarthria. The main neurological deficit was still his left hand weakness. He was referred to further rehabilitation and resumed normal activity as a physician by 3 months. Antiplatelet (aspirin and plavix for 6 months) and neuroimaging were prescribed on discharge. A CTA at 4 months post-discharge showed patent stents bilaterally. The patient did not show any new neurological event until now, a year from treatment.

Discussion

ICAD can account for 10–25% of strokes in young patients [[2, 8, 10–12](#)] and is considered as one of the main causes for stroke recurrence [[12](#)]. Among 1–5% of all spontaneous ICAD cases, a heritable connective-tissue disorder can be identifiable and ~5% may have one family member with spontaneous dissection of various arteries including the aorta or its main branches [[13–16](#)]. Women with spontaneous ICAD are on average younger [[15](#)].

Spontaneous BICAD was reported sparsely. Marshman et al. [[17](#)] reported a case of a previously healthy 29-year-old woman with bilateral vertebral artery dissection (VAD) and ICAD associated with multiple aneurysms (saccular and fusiform) that resolved spontaneously by 6 months. Abisaab et al. [[18](#)] reported a case of BICAD in a post-partum patient.

The natural course of ICAD seems to be composed of an acute phase during which conservative or more rarely endovascular/surgical treatments are undertaken followed by a chronic phase during which a recanalization occurs in the majority of patients during the following 6 months. Some have reported that delayed recanalization can occur after 16 months as well [[10](#)], a fact that might prompt individual consideration regarding the time for anticoagulation/antiplatelet therapy.

Recently, Schwartz et al. [[20](#)] reported that among 177 young patients, 8.5% had recurrent ischemic events unrelated to the type of antithrombotic agent (others have reported a recurrence of 15% [[19](#)]) and 1.1% had recurrent dissections (others have reported a recurrence of 3–8% after longer clinical follow-up of up to 7.4 years [[21–23](#)]).

The relation between infectious diseases and ICAD is debated. Skowronski et al. [16] reported a case of an adult male with CAD and massive right hemisphere infarct during the course of pertussis. This patient as ours had no predisposing risk factors (HTN, DM, dyslipidemias, connective tissue diseases or smoking). Their patient required 6 weeks of acute care hospitalization during which craniectomy and temporal lobe resection were needed followed by prolonged rehabilitation. Preceding respiratory tract infection has been proposed as a possible risk factor for ICAD supported by the fact that the occurrence of ICAD peaks during the fall [24–26].

Grau et al. [24, 25] reported that patients with CAD had a higher prevalence of infections compared with controls (62 vs. 22%), while excluding mechanical factors as cough, sneezing and vomiting. ‘Mechanical cough’ (naming, unrelated to infectious diseases) causing dissections is reported in just few case studies describing VAD rather than CAD. Nomura et al. [27] reported a case of a 45-year-old man who developed Wallenberg’s syndrome due to VAD following severe cough. Herr et al. [28] reported a case of a man with VAD presenting with signs of posterior-inferior cerebellar artery territory stroke, following severe cough.

We hereby describe a unique case of a patient with rigorous noninfectious cough followed by acute bilateral spontaneous dissection of the anterior rather than posterior circulation and with concomitant inadequate collaterals from the circle of Willis.

Therapeutic Considerations

It is generally accepted that most of the ICAD cases proceed to recanalization within 6 months after dissection. Progression of disease is believed to start a few days post-dissection and can take up to 6 months.

The treatment for ICAD consists of three possible approaches [29, 30]: conservative, endovascular or surgical. Conservative treatment consists of two steps: an acute anticoagulation with heparin followed by secondary prophylaxis by oral anticoagulation/antiplatelet that normally is sufficient for 3–12 months (however, delayed recanalization as described by Vicenzini et al. [10] can prolong this period).

Although anticoagulation is recommended, the question whether warfarin or aspirin is more effective for recanalization is yet debatable. Once recanalization is documented and arterial flow is reestablished, anticoagulation continuation should be reconsidered. The patient must therefore undergo neuroimaging follow-up (MRA and MRI) during the following 3–6 months and avoid forceful efforts.

Surgical and/or endovascular treatments are indicated for spontaneous ICAD with either persisting severe stenosis (mainly for intradural ICAD), progressive neurologic symptoms despite conservative treatment (as in our patient) or BICAD that result in severe cerebral hemodynamic impairment. Patients with ICAD and SAH require either surgery or endovascular treatment due to recurrence tendency [1].

Conclusions

To our knowledge, this represents the first reported case of spontaneous BICAD following isolated rigorous cough with inadequate circle of Willis that seemed to have important therapeutic implications. BICAD with concomitant inadequate circle of Willis (tiny ACOM with no PCOM as in our patient) can prone for hypoperfusion which might not respond to conservative treatment and can lead to rapid deterioration and therefore prompt to earlier endovascular intervention as was done in this case.

Rigorous cough can solely cause acute spontaneous BICAD that should be suspected even in a patient with no other risk factors as in our patient. Moreover, unilateral symptoms cannot exclude BICAD.

We would therefore recommend a thorough evaluation of the collateral flow in patients with uni-/bilateral ICAD, which might facilitate the decision in whom to perform an early arterial flow reestablishment.

Fig. 1. **a** Angiography: post-stenting right common carotid artery (RCCA; lateral view). **b** Angiography: post-stenting left common carotid artery (LCCA) with injection to the left ICA demonstrating opacification of the right anterior cerebral artery due to a tiny ACOM (anteroposterior view).

Fig. 2. **a** Distal LCCA dissection (lateral view). **b** Distal RCCA dissection (anteroposterior view).

Fig. 3. Post bilateral ICA stenting.

Fig. 4. Post-stenting watershed infarct due to hypoperfusion.

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