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Case Report

A rare bilateral internal carotid artery occlusion (BICAO) with mild clinical symptoms and no risk factors-etiology, collateral circulation and clinical management: case report

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Abstract: Bilateral internal carotid artery occlusion (BICAO) is a rare disease that often results in a fatal ischemic event. So far, only a few cases of BICAO with severe clinical symptoms have been reported, most of which developed serious cerebral infarction, presented poor outcomes and carried a risk of recurrent ischemic events. We report here a case of a 46-year-old female BICAO patient with no evidence of cerebral infarction but only presenting mild clinical symptoms, such as dizziness and nausea. Interestingly, all known risk factors of atherosclerosis and artery stenosis/occlusion were negative. Digital subtraction angiography (DSA) and computed tomography angiography (CTA) demonstrated that a high-flow collateral circulation was formed through the vertebrobasilar system. Due to the mild clinical presentations, conservative therapy and timely follow-up were selected for the patient. Based on our case, three possible hypotheses of BICAO formation are discussed. As the proper treatment of BICAO is still controversial, we believe that long-term observation is necessary to obtain a better understanding of the therapeutic effects.

Keywords: Bilateral internal carotid artery occlusion, ischemic infarction, case report

Background

Most ICA occlusions occur in patients with a history of cardiovascular disease such as moyamoya disease, atherosclerosis, coronary heart disease and stroke [1, 2]. According to the study by Mead GE *et al.*, 99.6% of occlusions involve only one side of the internal carotid artery [3]. Occlusion of both sides of the internal carotid arteries is an extremely rare condition. So far, only a few studies on BICAO with severe clinical symptoms have been reported [4-8], whereas long-term survival cases with mild clinical symptoms remain non-existent. Therefore, understanding the characteristics of this BICAO case with a mild clinical presentation may help to deepen our understanding of this disease.

Case report

A 46-year-old female with a 6-year history of dizziness and nausea was admitted to our hospital. Except for mild dizzy symptoms, no disorders of consciousness or amaurosis were reported. On physical examination, neither numbness nor weakness of the limbs was found. Moreover, all pathological signs and meningeal irritation were negative. Meanwhile, she had no habit of smoking and no history of hypertension, hyperlipidemia or diabetes mellitus. Digital subtraction angiography (DSA) and computed tomography angiography (CTA) demonstrated that the bilateral internal carotid arteries (ICAs) were severely occluded and that the entire Willis circle was completely enhanced, with the vertebral-basal arteries significantly enlarged (**Figure 1**). Furthermore, no evidence of an infarction area was shown on traditional MR imaging (**Figure 2**). Considering her clinical mild symptoms and radiological good compensation, conservative therapy including weight control and antiplatelet therapy with aspirin and timely follow-up were selected. The patient was still in good neurological condition at her latest follow-up (May 8th, 2017).

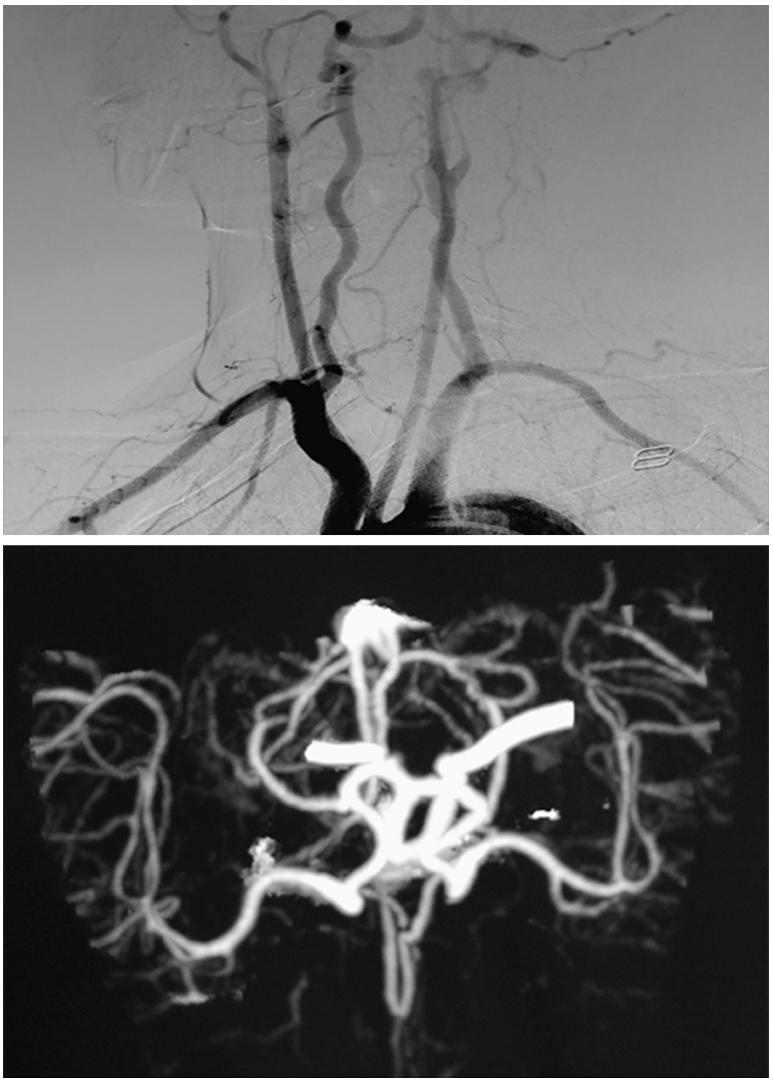

Figure 1. Digital subtraction angiography (DSA) and computed tomography angiography (CTA). Digital subtraction angiography (DSA) and computed tomography angiography (CTA) of the patient, performed on the latest follow-up on May 8th, 2017. Both internal carotid arteries were completely occluded, there was no distal flow of the dye, and the entire Willis circle was complete enhanced with the vertebral-basal arteries significantly enlarged. Single column fitting image.

Discussion

BICAO is an extreme rare disease that only accounts for 0.4% of transient ischemic patients with completed stroke [5, 6]. We reviewed another 5 BICAO cases in reports published between 2003 to 2015 (**Table 1**). Although it is rarely seen clinically, BICAO patients can be expected to suffer serious consequences due to fatal ischemic stroke [9]. It was reported that the overall mortality of 21 BICAO patients followed up for 1-11 years (average: 6 years) was 52% [4].

Although the correct mechanism of arterial stenosis/occlusion remains unanswered, atherosclerosis is considered the most significant [10, 11]. Hypertension, hyperlipidemia, diabetes mellitus and smoking are the main risk factors contributing to atherogenesis and increasing the risk of artery stenosis/occlusion [12, 13]. Interestingly, in our case, none of these abovementioned risk factors was found. We suspected the following three possible mechanisms of BICAO formation in this case: First, the congenital malformation of bilateral ICA occlusion is formed with vertebrobasilar compensation. Second, on the basis of a good blood supply through the posterior circulation, the congenital stenosis of the bilateral ICA gradually aggravated into a total occlusion during the perimenopause period by the fluctuation in hormone and hydronium levels, which might increase the blood viscosity and develop thrombosis [14]. Third, the bilateral internal carotid artery occlusion is formed by traumatic stimulation [15, 16]. Due to her mild clinical presentation and lack of definite risk factors, we considered her bilateral internal carotid artery occlusion to be less likely acquired without any stenosis; otherwise, the symptoms would be more serious when her intracranial hemodynamics changes.

According to the literature review, most BICAO cases develop into a fatal ischemic stroke. In our case, although severe occlusions formed in the bilateral ICAs, the patient presented with only a few mild clinical symptoms, such as dizziness and nausea. This might be attributed to

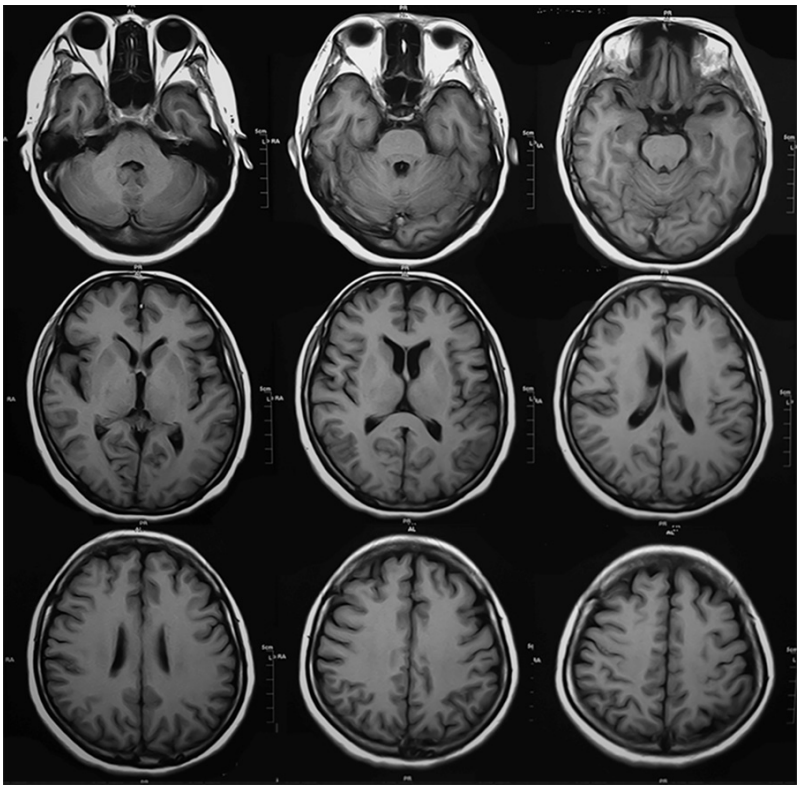

Figure 2. Traditional MR Imaging. Traditional MR imaging showed no evidence of a cerebral infarction. Single column fitting image.

her satisfactory collateral circulation formation. The compensatory circulation of BICAO mainly relies on two approaches: the vertebrobasilar system and external carotid/ophthalmic anastomosis [14]. For this patient, CTA and DSA revealed that the vertebrobasilar arteries and bilateral posterior communicating arteries were apparently enlarged. No significant extra blood supply via the external carotid artery was found as expected. Thus, the collateral circulation in this BICAO case was supplied by the vertebrobasilar system into the Willis circle via the posterior communicating arteries.

The proper treatment of BICAO remains controversial with respect to whether surgical treatment is selected. For neurosurgeons, apart from vascular bypass, external carotid artery revascularization, including carotid endarterectomy (CEA) and carotid artery stenting (CAS), is another choice. According to the report by Friedman et al., only 10% of BICAO patients who undergo external carotid artery revascularization experience a transient ischemic stroke during the follow-up period. Furthermore, they found that extracranial-intracranial bypass or medical therapy alone is not an effective way to provide a sufficient blood supply to improve patient symptoms [17]. However, a meta-analysis conducted by Mylonas SN *et al.* revealed no significant difference in therapeutic effect between medical therapy and revascularization [18]. So far, the clinical data of medical or surgical bypass management in BICAO is also limited. Persoon et al. observed that 57 patients with BICAO treated by medical therapy achieved a better prognosis than did the surgical group [19]. Although Yoshida S *et al.* reported that a BICAO patient acquired a good clinical result after undergoing a bypass, a randomized trial showed that extracranial-intracranial bypass failed to improve cognitive function within a 2-year follow-up period compared with conservative treatment [20]. For our patient, considering her mild clinical symptoms and satisfactory compensation, conservative therapy was selected by far.

BICAO is a rare and serious vascular disease and often results in a fatal ischemic event. Here, we report an extremely rare BICAO patient with only mild symptoms and the absence of radiological infarction. More interestingly, all known risk factors of atherosclerosis and artery stenosis/occlusion were negative. The collateral compensation completely relied on the vertebrobasilar system by cross-filling the Willis circle. So far, due to the limited cases, the proper treatment of BICAO remains controversial. Furthermore, long-term observation is necessary to obtain a better understanding of its therapeutic effect once surgical or conservative treatment is selected.

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Table 1. Summary of previous similar case reports of bilateral internal carotid artery occlusion

Author	Year	Patient	Image	Treatment	Clinical presence
Amin OS	2015	52-year-old man	Ischemic infarction at the region of the right middle cerebral artery and developed a new infarction at the region of the left middle cerebral artery 1 year later	Conservative treatment	Died of severe aspiration pneumonia approximately 2 months after the second stroke
Anand P	2014	39-year-old woman	Left-hemispheric stroke and developed a right-hemispheric stroke 1 year later	Conservative treatment	Expressive aphasia and right arm and leg weakness
Georgios Tsivgoulis	2013	73-year-old man	Bilateral cortical infarctions in both MCA territories	Conservative treatment	Right upper arm weakness then developed to quadriplegic and lethargic. The patient expired 1 day later
Ebru Bekircan	2009	91-year old woman	Extensive infarctions encompassing complete territory of the anterior and middle cerebral arteries bilaterally	Conservative treatment	Sudden development of coma and expired peacefully
Rabinstein AA	2003	56-year-old man	Extensive callosal infarction as well as the left frontal stroke	Conservative treatment	Rapidly progressive cognitive decline. The patient expired from aspiration pneumonia 10 days later

Disclosure of conflict of interest

None.

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