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Original Article

Reversible basilar artery disease caused by antiphospholipid syndrome: one case report

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Abstract: Antiphospholipid syndrome (APS) is an acquired autoimmune thrombophilia characterized by venous or arterial thrombosis, and/or pregnancy loss or complications in the presence of persistently positive antiphospholipid antibodies (aPL). Thrombotic events are the hallmark of APS. The current criteria for aPL tests consist of two direct ELISAs that detect antibodies against cardiolipin or β_2-glycoprotein I (β_2GPI), and the LA assay.

Keywords: Antiphospholipid syndrome, thrombosis, treatment

Introduction

Antiphospholipid syndrome (APS) is an acquired autoimmune thrombophilia characterized by venous or arterial thrombosis, recurrent pregnancy loss, and in the presence of persistently positive antiphospholipid antibodies (aPLs), including lupus anticoagulant (LA), anticardiolipin antibodies (aCL), and anti-β_2 glycoprotein-I (β_2GPI) antibodies. APS may be presented as thrombocytopenia, livedo reticularis, stroke, transient ischemic attacks, deep vein thrombosis, pulmonary embolism, epilepsy, valve vegetations and myocardial infarction. Thrombotic events are the hallmark of APS which usually lead to stroke in younger people. According to the AntiPhospholipid Syndrome Alliance for Clinical Trials and International Networking (APS ACTION), aPL positivity is found in 17% of stroke patients <50 years of age, compared with 0.7% of control participants. Here we report a case of young patient with stroke caused by APS-related reversible basilar artery occlusion.

Case presentation

A 41-year-old woman was admitted to the hospital on December 23, 2017, presented with numbness of right limb, confusing speaking, and compulsive crying and laughing for 18 days. Twenty-three days before that, she was hospitalized in a local hospital for sudden chest distress and gasping, with 21 weeks of menopause. Ultrasound examination revealed that she had mid-term pregnancy and stillbirth. The chest CT showed a large amount of pleural effusion. She was then underwent an exploration in thoracic cavity and wedge resection of right lower lobe of lung. She was postoperatively diagnosed with severe pneumonia, right hemopneumothorax, and rupture of pulmonary artery fistula in the right lower lobe. Three days postoperation, she suddenly suffered from paralysis of right limb, confusing speaking, and compulsive crying and laughing for no reason. The brain MRI revealed abnormal signal scattered throughout pons varolii. The cerebral infarction or demyelination was suspected. There were no obvious abnormalities in routine examination, biochemistry and staining of cerebrospinal fluid. She was treated with dehydration, antivirus therapy and methylprednisolone, and then the labor induction and uterine curettage were performed. Afterwards the patient's condition was gradually improved, but the symptoms in the nervous system were not remarkably relieved. So she was transferred to our hospital on December 23, 2017. The patient had no history of cerebral infarction, hypertension, or diabetes. Childbearing history: 3 induced abortions and 1 spontaneous abortion. The examination on admission showed that she was conscious, with compulsive crying and laughing, confusing speaking, right facial paralysis and

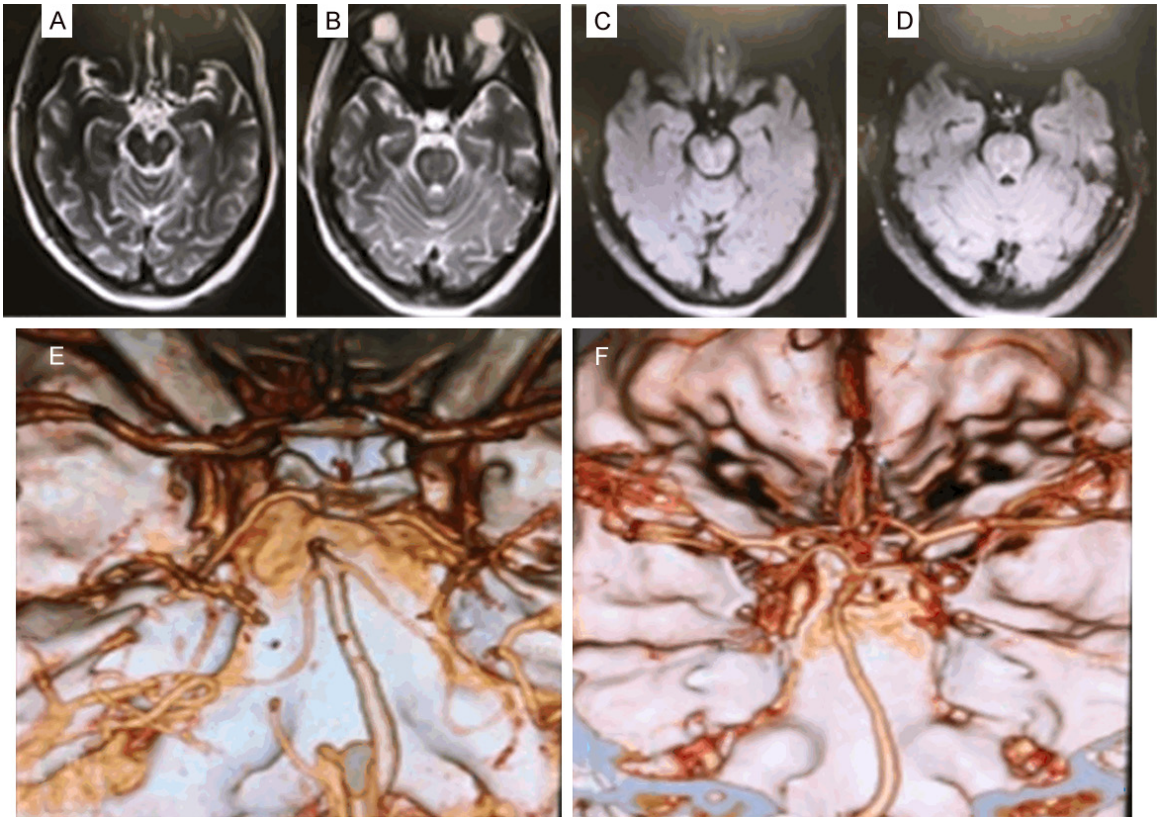

Figure 1. Reversible basilar artery disease caused by antiphospholipid syndrome. A-D: Skull MRI showed pontine, midbrain flaky long T2 signal and Flair high signal; E: CTA before head and neck treatment indicated occlusion of the basilar artery end and initial occlusion of the bilateral posterior cerebral artery; F: CTA after treatment indicated P1 segment of bilateral posterior cerebral artery and recanalization of the end of basilar artery.

right hypoglossal nerve palsy. Her muscle strength of the left extremities were level 5, while that of the right side were level 3, with the positive Babinski sign on the right side. The brain MRI showed the pons varolii, splenium of corpus callosum and bilateral brachium cerebelli **Figure 1**. Computed tomography angiography (CTA) revealed the occlusion both at the end of basilar artery and the origin of bilateral posterior cerebral artery **Figure 1**. To clarify the cause of the emboli, transesophageal echocardiography and dynamic electrocardiogram and ambulatory blood pressure were further performed, which showed normal results. She had increased white blood cells counts (11.9 × 10^9/L) and ESR level (47 mm/h). Her vasculitis-associated antibodies were negative. Of note, her anti-cardiolipin antibodies IgG and IgM were weakly positive, and anti-β_2 glycoprotein 1 IgM antibodies (β_2GP1-IgM) were 42.67 SMU (0-20). β_2GP1-IgA, β_2GP1-IgG and lupus-like anticoagulant were negative. Her hepatic, renal and thyroid functions showed no obvious abnormalities. She was diagnosed with antiphospholipid syndrome. The chloroquine of 0.2 g was taken orally twice per day, together with aspirin 100 mg and clopidogrel 75 mg once a day. After 20 days treatment, the cranio-cervical CTA showed complete revascularisation of the basilar artery and the bilateral posterior cerebral artery **Figure 1**, accompanied by significantly improved muscle strength and language function. Three months later, her anti-β_2 glycoprotein 1 IgM antibodies (β_2GP1-IgM) were 38.78 SMU.

Discussion

Diagnosis of APS includes clinical criteria of thrombosis and/or pregnancy morbidity and laboratory proof of aPLs in medium or high titers on two or more occasions at least twelve weeks apart [1]. The antiphospholipid antibodies for the diagnosis of APS are IgG/M aCL and/or IgG/M anti-β_2GPI and/or LA tests [2].

Studies provide direct evidence that aPLs cause thrombotic and obstetric APS (OAPS) manifestations. One of the main distinguishing prop-

erties of pathogenic aPLs is their binding to β_2GPI. β_2GPI is a highly glycosylated single-chain protein that is present in plasma without known physiological function. The central role of endothelial cells, monocytes, platelets, and complement has been shown in induction of thrombosis and fetal death in antiphospholipid syndrome. Endothelial cells and monocytes can be activated by antiphospholipid antibodies with anti-β_2-glycoprotein-1 activity [4].

This patient was at the childbearing age. She had the history of multiple abortions and no risk factors of vascular diseases such as hypertension, diabetes, and smoking. The onset of this disease is characterized by occlusion at the end of endobronchial artery, which may be due to the surgery and infection before the attack. The anti-β_2GP1 antibody positive in two detections confirmed the diagnosis of APS. For the treatment of APS, in order to avoid repeated thrombosis and progression of vascular lesions, it is generally recommended to use anticoagulant drugs prophylactically [2], however, in 2004, Levine et al. found that warfarin and aspirin treatment did not differ in preventing stroke recurrence through the antiphospholipid antibody stroke studies [3].

Studies have shown that the combination of aspirin plus dipyridamole and aspirin plus clopidogrel have higher efficacy than aspirin alone in patients with stroke [5] or atrial fibrillation [6].

This patient had received anti-platelet and immunosuppressive therapies. The cranio-cervical CTA 20 days after treatment showed the previously-occluded basilar artery end and posterior cerebral artery were completely recanalized. The patient's symptoms were relieved and no recurrence was observed in the follow-up till now. The internal carotid artery disease is commonly reported in literature. This patient had serious basilar artery disease and blood vessels were well-recanalized after active treatment, which is rare in previous reports. In this case, antiplatelet drugs have shown the same efficacy in blood vessels recanalization as well as in prevention of thrombosis.

APS usually causes in situ thrombosis of cerebral intracranial vessels, and is a significant cause of stroke in younger people. It is important to recognize the symptoms of APS and give appropriate therapy in order to reduce the risk of recurrence. This case suggested that APS-related thrombosis can be actively treated with anti-platelet aggregation and immunosuppressive therapy, and achieve good clinical outcome.

Disclosure of conflict of interest

None.

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