Stroke as the First Clinical Manifestation of Takayasu’s Arteritis

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Key Words
Takayasu’s arteritis · Ischemic stroke · Vasculitis of the central nervous system

Abstract
Takayasu’s arteritis is a chronic inflammatory disease, and neurological symptoms occur in 50\% of cases, most commonly including headache, dizziness, visual disturbances, convulsive crisis, transient ischemic attack, stroke and posterior reversible encephalopathy syndrome. The aim of this study was to report the case of a young Brazilian female with a focal neurological deficit. She presented with asymmetry of brachial and radial pulses, aphasia, dysarthria and right hemiplegia. Stroke was investigated extensively in this young patient. Only nonspecific inflammatory markers such as velocity of hemosedimentation and C-reactive protein were elevated. During hospitalization, clinical treatment was performed with pulse therapy showing improvement in neurological recuperation on subsequent days. In the chronic phase, the patient was submitted to medicated angioplasty of the brachiocephalic trunk with paclitaxel, with significant improvement of the stenosis. At the 6-month follow-up, the neurological exam presented mild dysarthria, faciobrachial predominant disproportionate hemiparesis, an NIHSS score of 4 and a modified Rankin Scale score of 3 (moderate incapacity). In conclusion, Takayasu’s arteritis must be recognized as a potential cause of ischemic stroke in young females.

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Introduction

Takayasu's arteritis is a chronic inflammatory disease that affects the aorta and its principal branches as well as the pulmonary artery, resulting in the formation of aneurysms and arterial stenosis [1]. In general, it occurs in women between the 2nd and 3rd decade of life [2]. The disease usually presents two phases: one systemic or ‘pre-pulseless’ phase, characterized by constitutional symptoms such as fever, weight loss, arthralgia, myalgia, lethargy and mild anemia, and a later or ‘pulseless’ phase, characterized by secondary clinical manifestations and arterial stenosis. Neurological symptoms occur in 50% of cases and most commonly include headache, dizziness, visual disturbances, convulsive crises, transient ischemic attack, stroke and posterior reversible encephalopathy syndrome [3].

Case Report

A 19-year-old Caucasian female from Brazil was admitted with focal neurological deficit 3 days after symptom onset. The patient had been diagnosed in another center with anxiety. She presented asymmetry of brachial and radial pulses, aphasia, dysarthria and right hemiplegia. A CT scan showed extensive ischemia in the left cerebral hemisphere (fig. 1). Cerebral angiography displayed occlusion of the right subclavian and left common carotid arteries, stenosis of the brachiocephalic trunk and flow inversion of the right vertebral artery and left posterior communicating artery, irrigating the territory of the left common carotid artery (fig. 2). Abdominal ultrasonography showed celiac trunk stenosis (fig. 3). Stroke was investigated extensively in this young patient. Only nonspecific inflammatory markers such as velocity of hemosedimentation and C-reactive protein were elevated. During hospitalization, clinical treatment was performed using pulse therapy with methylprednisolone 1 g/day for 3 days as well as methotrexate 15 mg/day, acetylsalicylic acid 200 mg/day and simvastatin 20 mg/day. After pulse therapy, prednisone 60 mg/day was initiated, showing progressive neurological recuperation on the subsequent days. After 4 months, the patient was submitted to medicated angioplasty of the brachiocephalic trunk with paclitaxel, with significant improvement of the stenosis. At the 6-month follow-up, the neurological exam presented mild dysarthria, faciobrachial predominant disproportionate hemiparesis, an NIHSS score of 4 and a modified Rankin Scale score of 3 (moderate incapacity).

Discussion

Stroke incidence in this disease is 10–20%, but stroke as a first manifestation of Takayasu’s arteritis in young patients is rarely found in the literature [4]. The patient in this study did not show any systemic manifestations at any moment during the clinical follow-up, which also leads to an uncommon form of presentation of the disease. The stroke mechanisms in Takayasu’s arteritis are described as embolism of stenotic or occlusive lesions of the aortic arch and its branches, hypertension, cardioembolism and cerebral hypoflow [5]. The principal objective of the clinical treatment of Takayasu’s arteritis is to control the activity of the disease, thus achieving radiological and clinical improvement. Invasive treatments including angioplasty and vascular surgery (bypass) must be considered only for stenoses or occlusions of critical arteries such as renal arteries, common carotid and internal carotid arteries. In our case, there was a delay in the diagnosis because the symptoms were
associated with a psychiatric condition. In conclusion, Takayasu’s arteritis must be recognized as a potential cause of ischemic stroke in young females.

Disclosure Statement

The authors declare that they have no conflicts of interest.

References


Fig. 1. CT scan showing extensive ischemia in the left cerebral hemisphere.
Fig. 2. Angiography showing occlusion of the right subclavian and left common carotid arteries, and stenosis of the brachiocephalic trunk.

Fig. 3. Abdominal ultrasonography showing celiac trunk stenosis.