

Takayasu Arteritis with Intracranial Involvement Mimicking Epilepsy: Case Report and Review of the Literature

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Abstract

The symptoms of Takayasu arteritis (TA) are related to end organ ischemia. Here we present a patient with convulsions and intracranial involvement. A 15-year-old young woman was admitted with the complaint of convulsions since one and a half months previously. Her physical examination showed absent pulses and unobtainable blood pressure in both arms. Electroencephalography was normal. An arcus aorto-abdominal aortography, performed for TA prediagnosis, revealed that the subclavian artery ended as a stump at its origin on the right and was occluded by tapering on the left. Arcus aorta was normal. The right renal artery was occluded up to 80-90%. Magnetic resonance imaging of the brain revealed abnormal signal intensity in the deep white matter bilaterally. Cerebral catheter angiography showed focal stenosis of cerebral vessels; it was classified as type V according to the classification of the Takayasu Conference of 1994. A diagnosis of TA was made and 1 mg/kg steroid was given, and after a month methotrexate (15 mg/week) was added. On the tenth treatment day her pulse could be revealed. During the control period she had no convulsion. In young patients TA should be kept in mind as a rare cause in convulsion etiology. Intracranial involvement of TA must be evaluated especially if there is a headache and convulsion.

Key words: Takayasu arteritis, intracranial involvement

(Intern Med 50: 1345-1348, 2011)

(DOI: 10.2169/internalmedicine.50.4332)

Introduction

Takayasu arteritis (TA) is a chronic granulomatous inflammatory arteritis affecting large vessels, predominantly the aorta and its main branches. The symptoms of Takayasu arteritis are related to end organ ischemia. The main clinical symptoms are weakness, fever, arthralgia, skin rash, hypertension, intermittent claudication upper or lower limbs, cardiac diseases (cardiac failure, valvular or ischemic heart disease, pulmonary hypertension, pericarditis), impaired renal function and cerebrovascular manifestations (1, 2). Cerebrovascular manifestations of TA include transient ischemic attack, stroke, hypertensive encephalopathy, spastic paraple-

gia, raised intracranial pressure (2-4). Convulsion is rare and there are few studies evaluating intracranial involvement (2, 5-7). Here, we report a case presenting with convulsions and intracranial involvement.

Case Report

A 15-year-old young woman was admitted with the complaint of convulsion for 15 days. Her parents described her generalized seizures preceded by loss of consciousness which lasted for 10 minutes. Patient had post ictal fatigue and headache. Urinary and fecal incontinence were not reported. Occasionally she had headache 3 months before contractions. Her headache was not sensitive to light or sound

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Received for publication August 6, 2010; Accepted for publication February 8, 2011

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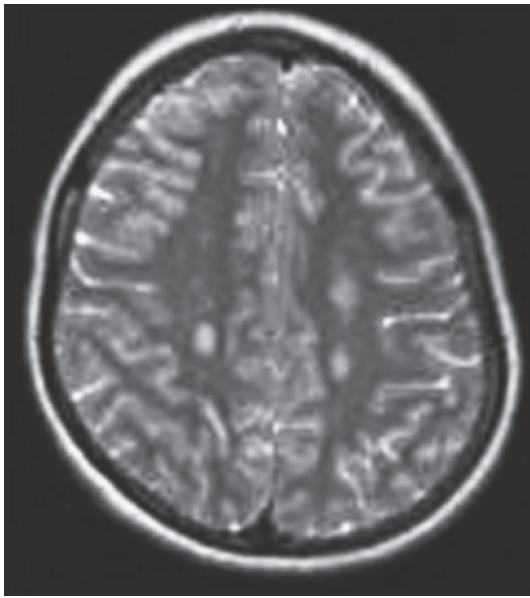


Figure 1. Magnetic rezonans imaging of brain revealed abnormal signal intensity in the deep white matter bilaterally.



Figure 2. Cerebral catheter angiography showed focal stenosis of cerebral vessels.

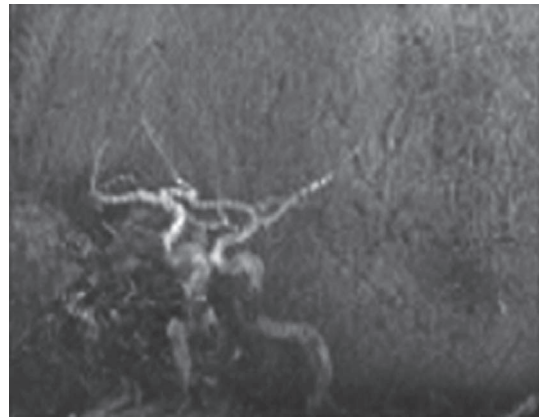


Figure 3. Post-treatment MRI angiography showed normal.

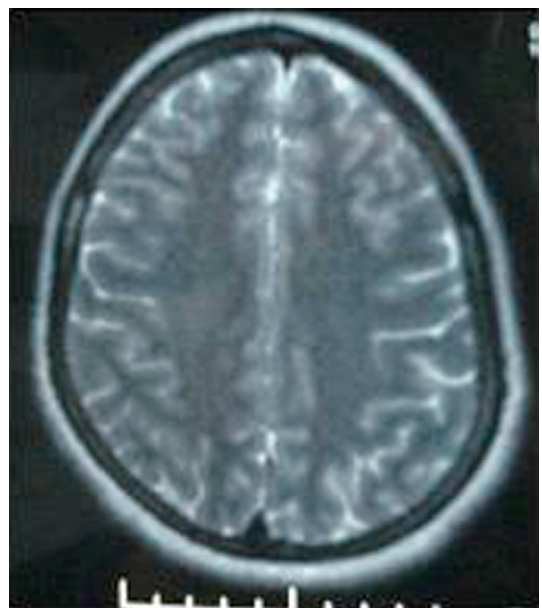


Figure 4. Post-treatment MRI of brain showed normal.

and was not associated with nausea or vomiting. Her individual and family history was uneventful, and she had no substance abuse. Physical examination showed absent pulses and unobtainable blood pressure in both arms. Skin and conjunctival pallor were noted. Cardiovascular and ophthalmoscopic examinations were normal. Laboratory findings were as follows: hemoglobin was 8.2 g/dL, hematocrit was 27%, white blood cell count was 10,300/mm³, platelet count was 516,000/mm³ mean corpuscular volume was 54 femtoliter, reticulocyte index was 1. In addition, iron was 17 mg/dL, iron-binding capacity was 173 mg/dL, iron saturation was 10%, and ferritin was 271 ng/mL. Fasting glucose level, calcium, albumin, urea and creatinin levels were within the normal range but the globulin value was 4.4 mg/dL (range between 1.8-2.6 mg/dL). Sedimentation rate was 99 mm, C-reactive protein (CRP) was 75 mg/dL (range from 0-6 mg/dL). Telecardiography and echocardiography were normal. Electroencephalography was normal. PPD test was 8 mm.

An arcus aorto-abdominal aortography showed bilateral subclavian artery occlusion and narrowing of the left common carotid artery. Arcus aorta was normal. Right renal artery stenosis was detected. TA according to the American College of Rheumatology (8) classification was diagnosed according to symptoms, signs and angiographic image. Magnetic resonance imaging (MRI) of brain revealed abnormal signal intensity in the deep white matter bilaterally (Fig. 1). Cerebral catheter angiography showed focal stenosis of cerebral vessels (Fig. 2). According to the Takayasu Conference 1994 classification it was classified as type V (9). Diagnosis of TA was made and 1 mg/kg steroid was given, and after a month methotrexate (MTX) (15 mg/week) was added. On the tenth treatment day her pulse could be revealed. During the control period she had no convulsion. Post-treatment ESR and CRP levels were measured as normal titers. Posttreatment-MRI angiography and MRI of the brain were normal (Fig. 3, 4).

Discussion

Angiography is the gold standard for the diagnosis of TA and an angiographic classification for TA was developed. According to this classification, involvement of aortic arcus indicates a higher neurologic impression (type I and type V) (3, 4, 10). The present patient was type V. Cranial MRI revealed abnormal signal intensity in the deep white matter bilaterally associated with vasculitic involvement.

Ureten and colleagues studied 45 TA patients and 6 (13%) of them had cerebrovascular events as the initial complaint (11). Wang and colleagues reported that cerebrovascular involvement may be the presenting symptoms of TA. Major neurologic events which occur are 93% dizziness, 88% headache, 67% vertigo, 63% vision loss, 51% memory loss, 43% transient ischemic attack and 34% amnesia (3). However there is not any emphasis on seizure. Another study suggested that ischemic attack, stroke, hypertensive encephalopathy, spastic paraplegia, and increased intracranial pressure occurred in 20% of the patients with TA (5). This study did not mention any seizures neither.

Neurological symptoms are associated with cerebral ischemia. The severity of ischemia is correlated with extension of vascular involvement. Generally, extracranial involvement accounts for the clinical picture (3, 5, 10). Intracranial involvement is rare (2, 6).

Intracranial stenoses in TA could be due to either vasculitic involvement or a prior embolization into the vessel. Evidence for vasculitic affection of intracranial arteries has been found in rare cases on conventional angiography (7). At least one case report has described intracranial arteritis in a patient with TA discovered at autopsy (12). In this case, thrombosis of the right internal carotid artery resulted in ischemic necrosis of the ipsilateral hemisphere.

In a prospective study Ringleb and colleagues designed, 70 TA patients were evaluated on follow-up by standardized neurological examination, sonography and MRI and intracranial pathology was found in seven patients (2). They found that the severity of vascular involvement was not correlated with neurological state. Also they did not report any convulsions.

This disease has four important complications which determine the prognosis. These are secondary hypertension, aortic regurgitation, aneurysms and retinopathy (1, 5, 10). The present patient did not have any of these complications. Half of the patients respond to steroid therapy. The present patient had a good response to steroid. In non-responsive patients MTX and Mycophenolate Mofetil can be used (5, 10). Two or more complications increase the mortality. Five-year survival is 92% and 10-years survival is 87% (1).

TA diagnosis can be delayed. Another study reported that delay is more frequent with a low sedimentation rate and in patients younger than 15 years (13). One study found that this period is on average 15.5 months (0-325 months). In

this study neurological findings were: headache, convulsion, dizziness, syncope, transient ischemic attack and stroke. These were more frequent with vertebral artery and carotis involvement. Convulsion rate was under 10%. Angiography could not show any intracranial involvement (5). The present patient also had a late diagnosis. Ruige and colleagues evaluated 16 patients and did not observe intracranial involvement (13). Also Mwipatayi and colleagues did not report any intracranial involvement in their series of 272 patients (14).

TA has morbidity and mortality by affecting cardiac, renal and cerebral tissues. Therefore, treatment must be aggressive. Clinical symptoms, inflammatory parameters, imaging methods must be used for follow-up (4-6). We used diagnostic angiography and cranial MRI for the present patient.

In young patients TA should be remembered as a rare convulsion cause. Intracranial involvement of TA must be evaluated especially if there is convulsion and headache.

The authors state that they have no Conflict of Interest (COI).

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