

Top of the Basilar Syndrome in a Young Adult Initially Presenting with a Convulsive Seizure

Koushun Matsuo¹, Chihiro Fujii¹, Ikuko Fuse², Masayuki Nakajima²,
Masahiko Takada³ and Kiyonori Miyata¹

Abstract

A 23-year-old man was admitted to our hospital due to loss of consciousness and a generalized convulsive seizure. He was diagnosed as having primary epilepsy and treated with antiepileptic drugs. Emergency CT scan of the head showed no abnormality. However, MRI scan of the head several days after admission revealed fresh infarctions caused by occlusion of the basilar artery, i.e., “top of the basilar” syndrome. This case indicates the need for precise differential diagnosis of convulsive seizure in an emergency situation. It should also be borne in mind that basilar occlusion with ‘onset seizure’ can occur even in young adults who have no risk factors for stroke.

Key words: top of the basilar syndrome, convulsive seizure, stroke in a young adult

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Introduction

Convulsive seizure is a characteristic neurological symptom of patients with stroke. The seizures associated with stroke are classified into two types: early-onset seizure, which occurs within 2 weeks after stroke, and late-onset seizure, which occurs after 2 weeks (1). Early-onset seizure is thought to be caused by ischemic or hemorrhagic lesions in the cerebral cortex (2). In particular, if a seizure occurs within 24 hours after onset of stroke, it is defined as an ‘onset seizure’ (3). In some cases, a seizure is seen as an initial symptom of stroke in the acute stage. If both seizure and hemiparesis are initially apparent, a tentative differential diagnosis (i.e., Todd’s paralysis with epilepsy or onset seizure with stroke) is difficult (4). After a diagnosis of epilepsy has been made, the possibility of stroke tends to be unlikely, especially if the patient is young and healthy.

Here, a case of basilar artery occlusion (“top of the basilar” syndrome), initially presenting as convulsive seizure, in a young adult is presented, and the difficulty involved with diagnostic work-up in this case is discussed.

Case Report

A healthy 23-year-old man was admitted because of loss of consciousness and a generalized tonic-clonic convulsive seizure. He had developed a slight upper respiratory tract infection about a week before admission. The main symptoms were sore throat and mild fever. He had no rhinorrhea, cough, or diarrhea. After the onset of the headache, he suddenly vomited several hours before admission on that day. His growth and development from birth had been normal. There were no congenital disorders or a history of epilepsy, head trauma, migraine, collagen diseases, brain tumor, drug/alcohol intoxication, psychiatric disease, arteriovenous malformation of the central nervous system, or stroke. The patient also had no family history of stroke or neuromuscular disease. On admission, the general findings were normal except for fever (37.9°C). There were neither leg edema nor skin lesions. On neurological examination, the patient was comatose, and conjugate eye deviation to the left was present. There were no meningeal signs. After resolution of the generalized convulsive seizure, right hemiparesis was noted. Bilateral Babinski signs were present. An emergency head

¹Division of Neurology, Ohmihachiman Community Medical Center, Japan, ²Division of Neurosurgery, Ohmihachiman Community Medical Center, Japan and ³Division of Radiology, Ohmihachiman Community Medical Center, Japan

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Correspondence to Dr. Koushun Matsuo, matsuo@pb4.so-net.ne.jp

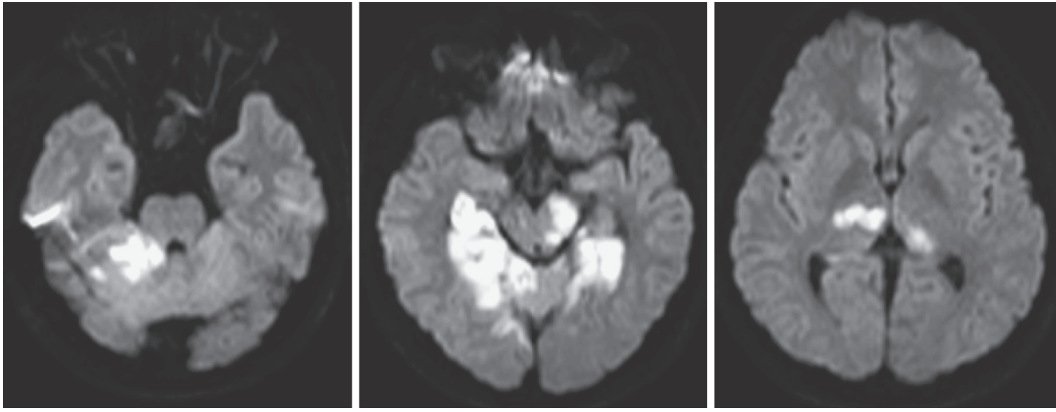


Figure 1. Head MRI scan (diffusion-weighted image) shows high intensity in the right cerebellum, left midbrain (cerebral peduncle), bilateral medial temporal lobes, and bilateral thalami. There are also no definite abnormal features in the frontal and parietal lobes and basal ganglia.

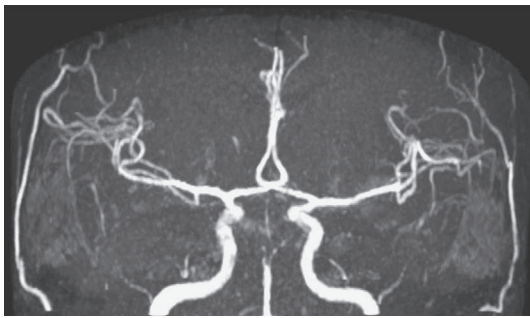


Figure 2. MR angiography (MRA) shows occlusion at the top of the basilar artery. There are no other significant abnormal findings such as an aneurysm or stenosis of the major cerebral arteries.

CT scan showed no abnormal findings. Laboratory data showed normal values except for a mild inflammatory reaction (white blood cells 12,000/ μ L, C reactive protein 0.18 mg/dL). Blood sugar and serum electrolyte levels were within normal limits. Creatine kinase increased (349 IU/L) transiently a few days after admission and immediately normalized. Cerebrospinal fluid examination showed neither pleocytosis nor any other abnormal findings.

Based on the clinical presentation and diagnostic findings, the patient was initially considered to be in the post-ictal stage of epilepsy, and thus the right hemiparesis was assumed to be Todd's paralysis. Consequently, an anti-epileptic drug (diazepam) was administered, and the patient was expected to recover consciousness. However, 4 days after admission, there was no evident improvement in his neurological signs. An electroencephalogram showed a constant diffuse slow, high voltage wave. No spike waves were detected. Therefore, it was suspected that other intracranial lesions might be present, and a head MRI scan was performed. This revealed multiple fresh infarctions in the right cerebellum, left midbrain, bilateral medial temporal lobes, and bilateral thalami (Fig. 1). MR angiography showed occlusion of the rostral segment of the basilar artery (Fig. 2).

Accordingly, therapy for cerebral infarction was started

immediately. Heparin 10,000 units/day, edaravone 60 mg/day, and glycerol 400 mL/day were administered in the acute phase. Furthermore, high dose methylprednisolone (1,000 mg/day \times 3 days) was also administered because we considered that intracranial lesions caused by vasculitis with collagen disease as a differential diagnosis immediately after revealing the result of head MRI. And citicoline was added to prevent brain edema and improve the patient's state of consciousness. Despite the delay in the correct diagnosis and initiation of appropriate therapy, the patient's neurological symptoms showed a gradual improvement.

There were no signs of atrial fibrillation or a patent foramen ovale on several examinations of the cardiovascular system, such as electrocardiogram and trans-thoracic/esophageal echocardiogram. There were no complaints of dyspnea or chest pain, and his respiratory function was essentially normal. The values of protein C/S, anti-thrombin III, platelet factor IV and other factors of the coagulation system were within normal limits. Anti-cardiolipin antibody and lupus anticoagulant were negative.

To prevent a second stroke event, antiplatelet therapy (aspirin and cilostazol) was continued. Anti-epileptic drugs were not continued because there were no further convulsive seizures. Fortunately, by 4 months, the patient showed almost full recovery of daily living activities, and he returned to his previous job 8 months after onset.

In addition, the improvements (i.e., reductions of infarct lesions and recanalization of the basilar artery) were also confirmed on the follow-up head MRI/MRA (Fig. 3). During follow-up of about 2 years after discharge, no further strokes occurred.

Discussion

Convulsive seizures are frequently observed in patients admitted for neurological emergencies, and in each case it is important to investigate the cause in order to start appropriate treatment as soon as possible. When a patient is young and healthy, the combination of loss of consciousness and

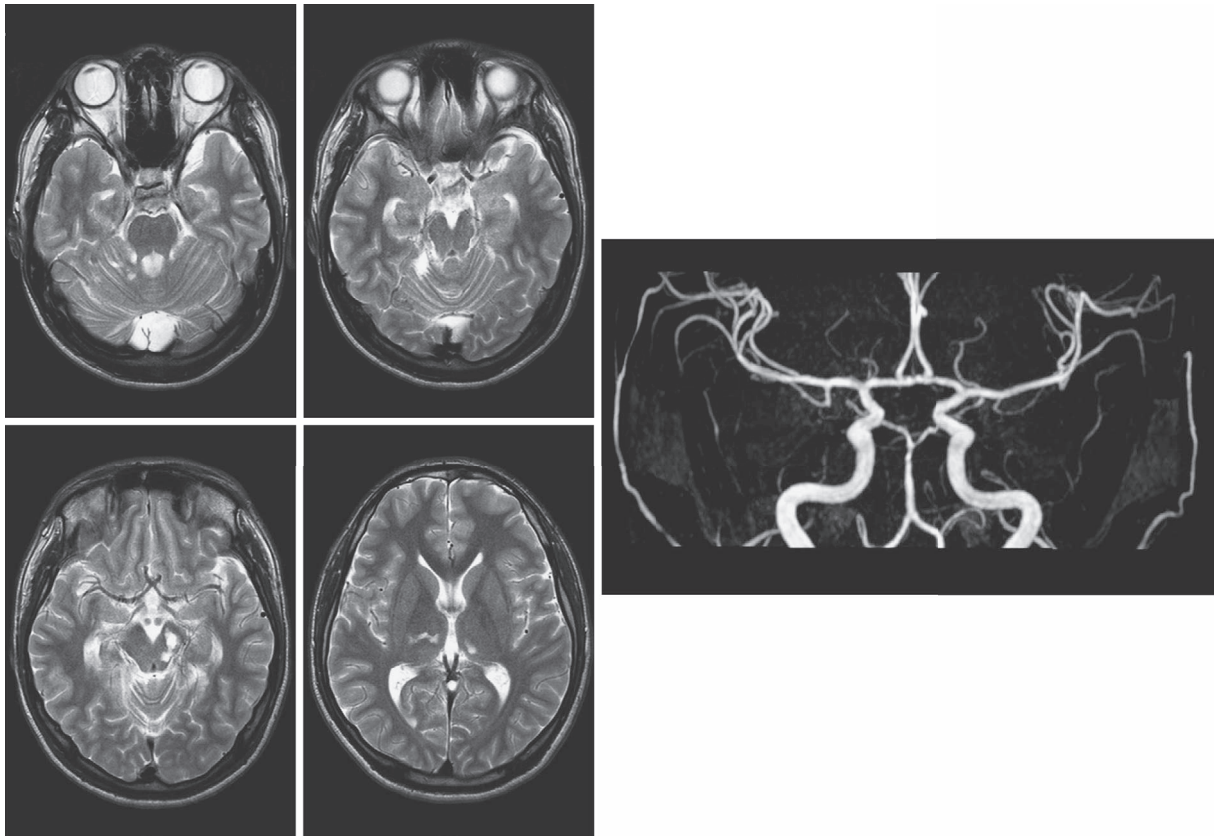


Figure 3. Follow-up images of head MRI/MRA performed 3 months after admission. In the left part, T2-weighted MRI images show the lesions of infarction that have decreased in size. In the right part, note that the MRA shows that the top of the basilar artery is clearly described, possibly reflecting the recanalization of that part.

convulsive seizure tends to be diagnosed as primary epilepsy if there is no evidence of a secondary cause such as brain tumor, meningoencephalitis, drug intoxication, hypoglycemia or electrolyte imbalance.

If the same symptoms are initially observed in an elderly patient, the possibility of stroke also needs to be considered. Burn et al reported that patients presenting with a first attack of stroke have a 2% risk of onset seizure (3). Transient conjugate eye deviation to the left in this case might reflect that a certain effect on the neural pathways of ocular movement was caused by both stroke and the complicated seizure at the same time.

Generally, early onset seizure caused by cerebral infarcts is relatively more common when the anterior circulation, rather than the posterior circulation, is affected (5). The precise mechanism whereby infarcts in the posterior circulation cause seizure is still unknown. Penfield suggested a “centrencephalic system” in which the brainstem functioned as a causative center of seizures (6). Recently, Kohsaka et al reported that brainstem auditory evoked potentials preceded the onset of cortical paroxysmal discharges in patients with absence seizure (7), thus supporting the classic theory of a centrencephalic system.

However, when encountering an emergency case of convulsive seizure, it is clinically difficult to initially suspect involvement of the brainstem, which is supplied by the verte-

brobasilar artery. Saposnick and Caplan have maintained that the convulsive-like movements in brainstem stroke are different from epileptic seizures, and that sudden onset of a decerebrate posture tends to be regarded as a convulsive seizure. They suggested that such movement is an important sign for an earlier diagnosis of pontine stroke (8). Although their incidence is low, it should be borne in mind that convulsive seizures or convulsive-like movements can be caused by brainstem lesions (included in the territory of the posterior circulation) in some cases.

If a stroke event occurs in a young adult, certain characteristic diseases in the patient’s background should be considered. These diseases may influence the onset of stroke and can include vascular anomalies (e.g., arteriovenous malformation or moyamoya disease), genetic diseases (e.g., Marfan syndrome), disorders of the coagulation system, collagen diseases, or recent trauma to the head or neck.

In a young healthy adult without any of the obvious factors mentioned above, cerebral artery dissection can also be considered as a cause of stroke (9). However, details of the background and the optimal management for cerebral artery dissection are still unknown (10).

We tried to investigate the cause of the stroke event in the present case. According to the general clinical conditions and several examinations of the cardiovascular system, paradoxical cerebral embolism and pulmonary embolism would

be ruled out. And disorder of the coagulation system would be ruled out as possible causes of the stroke event by the results of laboratory data. Because the value of platelets was normal, and also lupus anti-coagulant and anti-cardiolipin antibody were negative, anti-phospholipid antibody syndrome could be ruled out. Cerebral angiography was performed about 2 months after the onset of stroke and showed no evidence of basilar artery dissection. However, if dissection had occurred, the lesions might be repaired in this stage. In addition, headache and vomiting were present on the day of admission, which may reflect the pathogenic mechanism of basilar artery dissection. Thus, it is suspected that basilar artery dissection played an important role as a cause of stroke in this case, although no directly supportive findings were obtained.

The present case had two important clinical characteristics. One was that basilar artery occlusion presented with convulsive seizure as the initial symptom. The second was that an ischemic stroke event occurred in a young healthy adult who had no past or family history of stroke or any of the associated risk factors. These circumstances were both clinically atypical. Accordingly, ischemic stroke seemed a very unlikely possibility on admission. In fact, on the basis of the initial findings, head CT scan, and laboratory data, we mistakenly diagnosed primary epilepsy rather than stroke, and immediately administered anti-epileptic medication. As a result, several days went by before a correct diagnosis was established, thus delaying the introduction of appropriate therapy.

In patients during the acute phase of stroke, a good prognosis is dependent on a quick and accurate diagnosis. The present case illustrates that this type of ischemic stroke can occur even in a young healthy adult with no risk factors. In a differential diagnostic procedure for convulsive seizure under emergent conditions, we should consider not only pri-

mary epilepsy but also other neurological diseases. If no significant abnormality has been demonstrated by the initial head CT scan, and no recovery from consciousness disturbance is evident beyond 24 hours after onset, a follow-up CT or a further examination with a head MRI scan should be done as quickly as possible.

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

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