Medullary ischemia due to vertebral arteritis associated with Behçet syndrome: a case report

Takanobu Kaido,[1] Taisuke Otsuki,[1] Masafumi Ogawa,[3] Akio Takahashi,[1] Yuu Kaneko,[1] Toshiyuki Yamamoto[2] and Yasuhiro Nakata[3]

Summary

Here we report an extremely rare case of Behçet syndrome (BS) that showed acute onset of Wallenberg syndrome and was treated successfully by corticosteroids. A 51-year-old woman with BS had a sudden onset of Wallenberg syndrome. Three days after the onset, she was transferred to our institute. In the magnetic resonance imaging (MRI) study on admission, T2-weighted and fluid-attenuated inversion recovery images showed a high intensity area in the left paramedian region of the medulla oblongata. Contrast-enhanced T1-weighted images showed enhancement in the vessel wall of the left vertebral artery. We diagnosed her as having Wallenberg syndrome due to the acute vertebral arteritis associated with BS. After initiation of high-dose steroid therapy, her symptoms gradually improved. Two months after admission, she was discharged from our institute with mild hemihypesthesia. We hypothesized that vertebral arteritis due to BS had caused hypoperfusion of the medullary perforators causing Wallenberg syndrome in our patient. ***(Asian Pac J Allergy Immunol 2012;30:239-42)***

Key words: *Medullary ischemia, Wallenberg syndrome, vertebral arteritis, Behçet syndrome, steroid*

From the 1. Department of Neurosurgery
2. Department of Neurology
3. Department of Radiology, National Center Hospital, National Center of Neurology and Psychiatry, Kodaira, Tokyo, Japan
Corresponding author: Takanobu Kaido
E-mail: kaido@ncnp.go.jp
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Introduction

Lateral medullary syndrome (that is Wallenberg syndrome) is recognized if some of the characteristic features, such as nystagmus, ipsilateral Horner's syndrome, ipsilateral limb ataxia, contralateral limb and trunk spinothalamic impairment, dysphagia and dysphonia, and ipsilateral facial sensory and motor disturbance, are present. Although WS has been long thought of as the disease of the posterior inferior cerebellar artery, most of the cases are caused by occlusion or stenosis of the intracranial vertebral artery which occludes the origins of the small direct perforators feeding the lateral medulla.[1]

An ischemic change of the medulla oblongata can be detected using magnetic resonance imaging (MRI). If the symptoms are concordant with the findings of neuroimaging, therapy must be commenced immediately, according to the cause. Some causes of WS are common but less severe, such as embolism, thrombosis, and arterial dissection. Others are rare but severe, such as infection, inflammation, thrombophilia, neoplasm, and drug-induced conditions. An accurate clinical history and a competent examination are the keys to identifying which patients are likely to have had a stroke.

As a cause of stroke, Behçet syndrome (BS) is uncommon. Moreover, it is seldom reported that Wallenberg syndrome is associated with BS. Therapeutic approaches for CNS-NBS in the acute phase differ from those for ischemic stroke. Corticosteroids are necessary to treat BS with acute central nervous system (CNS) involvement.[2]

We report a rare case of BS that showed acute onset of Wallenberg syndrome and was treated successfully by corticosteroids.

Case report

Since the age of 25, a woman had oral ulcers, skin lesions, and uveitis. These symptoms fulfilled the diagnostic criteria for Behçet syndrome.[3] Before this episode, she had been administered prednisolone at

a dose of 7 mg/day, an immunosuppressant (cyclosporine), a monoclonal antibody against tumor necrosis factor (TNF)- α (infliximab), as well as antiplatelet therapy.

At the age of 52, she had a sudden onset of salivary dysphagia and a cold sensation on her right hand. She was admitted to an emergency hospital. Horner's syndrome of the left side and right hemihypesthesia were noted. Computed tomography of the brain revealed no abnormal findings. She was diagnosed with a brain infarction. Despite thrombolytic therapy, her condition did not improve. She administered 10 mg/day of prednisolone via intravenous drip infusion. Three days after the onset of this episode she was transferred to our institute.

In the magnetic resonance imaging (MRI) study on admission, T2-weighted images and fluid-attenuated inversion recovery (FLAIR) images showed a hyperintense area in the left paramedian region of the medulla oblongata (Figure 1a, b). The area showed a slightly high intensity in diffusion-weighted images (Figure 1c, d), whereas apparent diffusion coefficient (ADC) maps showed no abnormal intensity area in the medulla oblongata (Figure 1e).

Contrast-enhanced T1-weighted images showed enhancement in the vessel wall of the left vertebral artery, whereas the hyperintense area in the medulla oblongata on T2-weighted and FLAIR images did not show enhancement (Figure 1f, g). MR angiography (MRA) showed an irregular and narrowed left vertebral artery (Figure 1h). Analysis of CSF revealed normal cellularity, normal protein concentration, and negative oligoclonal IgG bands, but a significant increase of interleukin (IL)-6 (103 pg/mL; normal, <4.3 pg/mL).

We diagnosed her condition as Wallenberg syndrome induced by transient ischemia, edema,

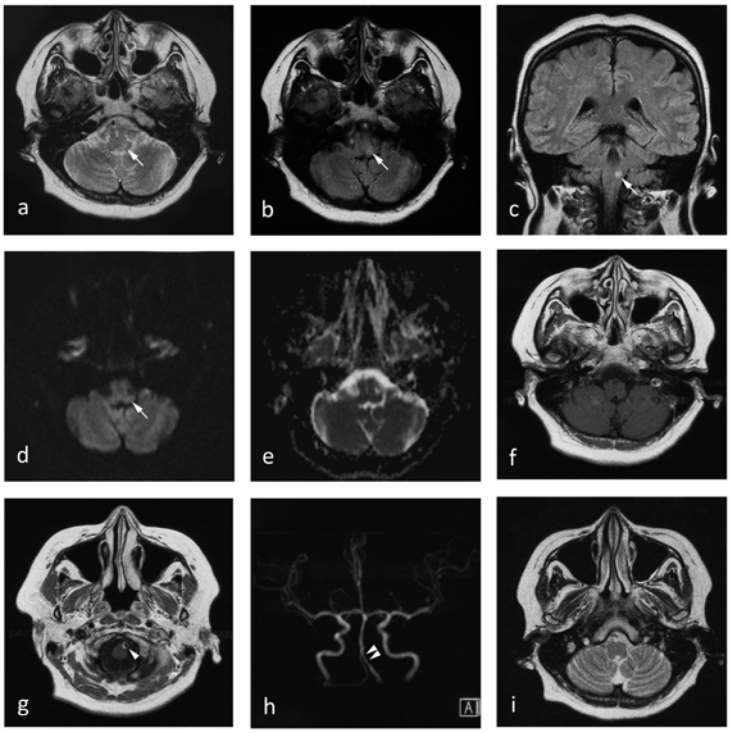

Figure 1. Magnetic resonance (MR) images at 3 (a-h) and 17 days (i) after the onset of Wallenberg syndrome. (a-d) Axial T2-weighted image (a), axial (b) and coronal (c) fluid attenuated inversion recovery images, and an axial diffusion-weighted image (d) showing a hyperintense area in the left paramedian region of the medulla oblongata (arrows). The axial apparent diffusion coefficient map (e) shows no abnormally intense area in the medulla oblongata. Contrast-enhanced axial T1-weighted images (f and g) show no enhancement of the medulla oblongata (f) but show enhancement in the vessel wall of the left vertebral artery (g) (arrowhead). Coronal maximum-intensity-projection reconstruction from the time-of-flight MR angiography (h) shows an irregular and narrowed left vertebral artery (double-arrowheads). In the MR study 17 days after onset, axial T2-weighted image (i) shows no abnormal intensity in the medulla oblongata.

and/or inflammation of the medulla oblongata due to vasculitis of the left vertebral artery caused by BS. We decided to increase the dose of prednisolone intravenously administered to 30 mg/day on the second day after admission to our institute, and 50 mg/day for 10 days on the third day. Cyclosporine was discontinued because of its possible neurotoxicity.

Her dysphagia was evaluated by swallowing with videofluoroscopy (VF), which revealed severe disturbance of swallowing, especially in the left side of the larynx. VF was performed by aspirating 10 ml of twofold diluted 110% w/v barium solution. Rotation of her head to the left side made it possible for her to swallow.

After initiation of high-dose steroid therapy and rehabilitation, her symptoms gradually improved. On the MRI study carried out 17 days after the onset, the hyperintense area disappeared on T2-weighted images (Figure 1i). Prednisolone was administered orally from the 12th day after admission and was tapered gradually as follows: 30 mg/day for 7 days, 25 mg/day for 28 days, and 20 mg/day as a maintenance dose. Two months after admission, she was discharged from our institute with mild hemihypesthesia.

Discussion

We report an extremely rare case of BS that showed acute onset of Wallenberg syndrome and was treated successfully by corticosteroids. Here we specially focus on three points: the etiology of Wallenberg syndrome in BS, the diagnosis of acute vertebral arteritis and the treatment of neuro-BS using corticosteroids.

First, we hypothesized that vertebral arteritis due to BS had resulted in hypoperfusion of the medullary perforators causing Wallenberg syndrome in our patient. BS is a chronic relapsing vascular inflammatory disease of unknown origin. Inflammation due to BS can involve vertebral arteries[2]. Moreover, occlusion of an atheromatous branch artery from the vertebral artery induces medullary infarct[4] and inflammation of the vertebral artery would occlude a branch artery and induce medullary ischemia. Wallenberg syndrome associated with BS is extremely rare and PubMed included only one case report of a 23-year-old woman with BS who had shown Wallenberg syndrome 4 years after the onset of BS and who was treated with transfusion successfully.[5] Various causes such as vasculitis, low-grade inflammation, demyelination, and degenerative changes were reported.

Next we discuss how we diagnosed the patient as having vertebral arteritis. Key clues of vertebral arteritis due to BS in our patient were the findings on MRI, CSF studies, and the past history. The vessel wall of the vertebral artery was enhanced and narrowed, providing potent evidence of arteritis. In addition, MRI clearly showed a lesion in the medulla oblongata adjacent to where the changed vertebral artery was located. We believe that the high intensity on diffusion-weighted images was a T2 shine-through effect because ADC maps showed no abnormal intensity and high intensity on T2-weighted and FLAIR images in the medulla oblongata indicated transient ischemia due to inflammation. Supporting our hypothesis, previous studies have reported that a frequent finding in neuro-BS is the resolution or the decrease in the size of the lesions when follow-up imaging studies are available.[2,6] As the general rule, IL-6 activity of CSF is significantly increased, which is supported by a previous study[7] in which all patients with progressive neuro-BS showed significant increases in CSF-IL-6 activity. These findings and past history of BS provided confirmation of the diagnosis and led to successful treatment with steroids.

Finally, administration of prednisolone dramatically alleviated the patient's condition. Therapeutic approaches for acute neuro-BS are quite different from those for ischemic stroke. Corticosteroids are used to treat acute neuro-BS[2]. Therefore, lack of a diagnosis of acute neuro-BS would lead to loss of the opportunity for appropriate treatment. Because there is no evidence for the various treatment modalities for neuro-BS at present, current guidelines in management are based on empirical observations and oral prednisolone or high-dose intravenous methylprednisolone are used to treat acute neuro-BS.[8] Even though the effects are short-lived and they do not prevent further attacks or progression, well-timed administration of corticosteroids can improve similar conditions.

Conclusions

We report a rare case of BS that showed an acute onset of Wallenberg syndrome and was treated successfully by corticosteroids. We hypothesized that vertebral arteritis due to BS had resulted in hypoperfusion via the medullary perforators to cause Wallenberg syndrome in our patient. Findings from MRI and CSF examinations and the past history

were useful in the diagnosis of vertebral arteritis. Administration of high-dose steroids can improve similar conditions.

Conflict of interest

The authors have not reported any conflicts of interest.

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