

## Case Report

# Neuro-Behçet Disease Mistakenly Diagnosed as a Brain Abscess Because of a Ring Contrast Enhanced Lesion on Brain MRI

Hidetoshi TAKASHIMA, Yoko NAKAO, Keisuke IWANAGA, Itsuro TOMITA, Akira SATOH, Makiko SETO, Mitsuhiro TSUJIHATA

Section of Neurology, Nagasaki Kita Hospital, Nagasaki, Japan

A 27-year-old man was hospitalized for an evaluation of a high fever and left hemiparesis. Gadolinium-enhanced MRI disclosed a lesion with ring enhanced areas with marked edema extending from the right basal ganglia and thalamus to the midbrain that appeared to be a brain abscess. The lesion resolved almost completely during the 2 month period after admission. Three weeks after discharge the patient again developed a high fever, and an MRI examination disclosed a large new lesion involving the left thalamus, brainstem and upper pons. No ring enhancement was detected by gadolinium-enhanced MRI, and the new lesion again spontaneously resolved almost completely, but during the second admission the patient also developed oral aphthae and skin eruptions suggesting Behçet's disease. This is a rare case suggesting that the lesions occurring in association with neuro-Behçet disease may resolve spontaneously.

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## Introduction

Behçet disease is a multisystem vasculitis of unknown origin in which neurologic involvement has been reported to range from 4.8% to 14.3% in a large series.<sup>1,2</sup> The classical triad of oral and genital ulcerations with uveitis was originally described by the Turkish dermatologist Hulusi Behçet<sup>3</sup> in 1937. Although nonneurologic involvement generally precedes the neurologic findings, nonneurologic involvement may either go unrecognized in some cases or it may appear late in the patient's course, thus posing diagnostic difficulties.

We herein report a patient with Behçet disease in whom gadolinium-enhanced MRI disclosed a ring enhancement with marked edema mimicking a brain abscess; however, skin lesions did not appear until the second admission when a recurrence of the disease occurred.

## Case report

A 27-year-old man was hospitalized for an evaluation of a high fever and left hemiparesis on December 17, 1997 (Figure 1). He had been well until November 1994 when he noted diplopia which thereafter improved within a week. There was no history of either

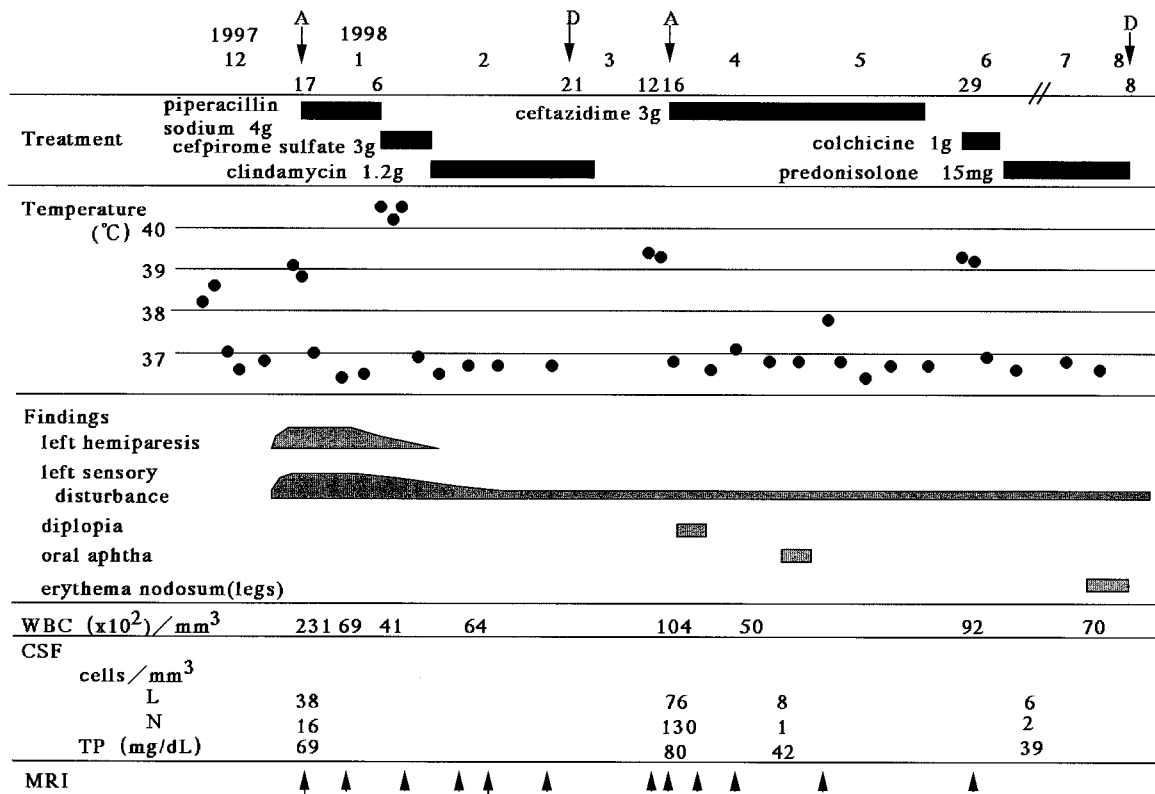
recurrent oral and genital ulcers or uveitis. Three weeks prior to admission he noted a 39°C fever, chills and headache. On December 14 he developed mild left hemiparesis.

A general examination on admission showed only tachycardia (120/min) and high fever. Neither ulcerated lesions in the mouth and genitalia or uveitis were observed. Neurological examinations disclosed very mild left hemiparesis and a sensory disturbance with facial involvement. There were no signs of either meningeal irritation or cranial nerve involvements. The patient's deep tendon reflexes were normal, and the Babinski and cerebellar signs were negative. Based on the above symptoms, acute disseminated encephalomyelitis, multiple sclerosis and Behçet's disease were all considered as possible diagnosis in this case.

The erythrocyte sedimentation rate was 85 mm/hour, leucocyte count 23,000/mm<sup>3</sup>, and the C-reactive protein level was 28.8 mg/dL. Both anti-nuclear antibodies and anti-DNA antibodies were undetectable. Brain MRI on admission revealed a high signal lesion extending from the thalamus and basal ganglia to the midbrain on the T2-weighted and FLAIR images, and a low signal lesion on the T1-weighted images on the right side (Figure 2 A). A lumbar puncture revealed 54 white blood cells/mm<sup>3</sup> (38 mononuclear and 16 polynuclear cells), and the protein level was elevated to 62 mg/dL.

**Address correspondence:** Mitsuhiro Tsujihata, M.D., Director, Nagasaki Kita Hospital, 4-61 Nameshi 5-chome, Nagasaki, 852-8061 JAPAN  
TEL: +81-(0)95-857-0001, FAX: +81-(0)95-857-5821, E-mail: kita\_m\_tsuji@mx22.tiki.ne.jp

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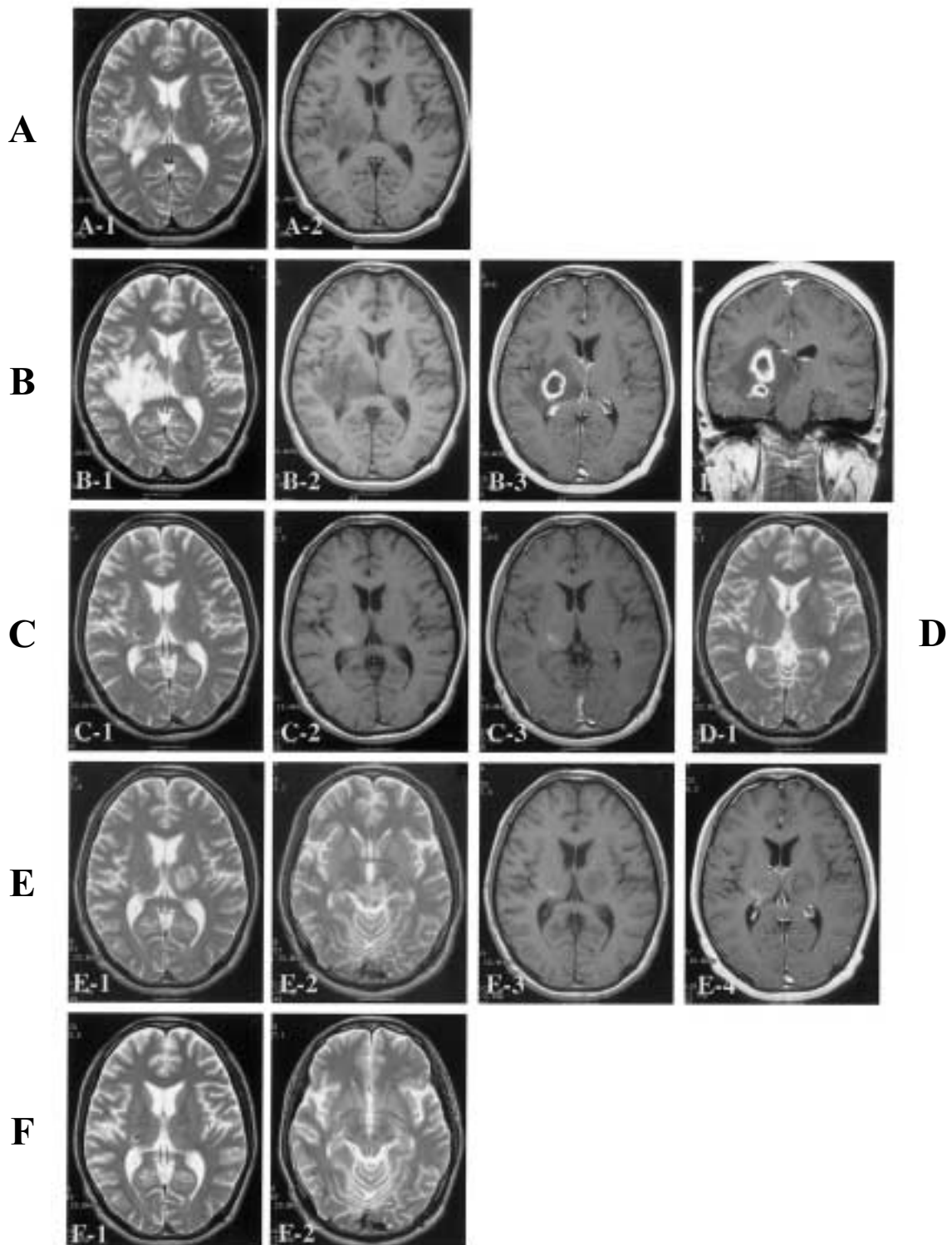
**Figure 1.** Clinical course of the patient. A=admission; D=discharge; WBC=white blood cell; CSF=cerebrospinal fluid; L=lymphocyte; N=neutrophil; TP=total protein.

The glucose level was 89 mg/dL, chloride 114 mg/dL, IgG 6.4 mg/dL, myelin basic protein 6.9 ng/mL (normal <4), and the  $\beta$ D-glucan level was less than 5.0 pg/mL. A CSF culture was also negative. MRI with gadolinium-enhancement on December 27 disclosed a ring enhancement with marked edema (Figure 2 B).

Because of the inflammatory laboratory data and the MRI findings on admission, the patient was diagnosed to have acute cerebritis (a brain abscess) and he was therefore immediately treated with piperacillin sodium (4 g/day) and glyceol (600 mL/day). The lesion almost completely resolved within 2 months after admission (Figure 2 C) and he was therefore discharged from the hospital on February 21, 1998.

On March 13, 1998 he was again admitted because of a high fever. A neurological examination on admission showed no new abnormal findings except for a mild left hemisensory disturbance which was thought to be a residual sign due to the first episode. An MRI performed on March 13 revealed no new lesions (Figure 2 D); however, MRI taken on March 16 disclosed a high signal lesion extending from the thalamus to the cerebral peduncle and upper pons on T2-weighted and FLAIR images and a low signal lesion on T1-weighted images on the left side (Figure 2 E). Gadolinium-enhancement showed only a small enhancing spot in the center of the lesion, but no ring lesion was observed. The erythrocyte sedimentation rate was 35 mm/hour, leucocyte count 10,400/mm<sup>3</sup>, and C-reactive protein 2.6 mg/dL. Cerebrospinal fluid examinations revealed pleocytosis (130

polynuclear cells and 76 mononuclear cells/mm<sup>3</sup>), and the protein level was 80 mg/dL. Antibiotics (ceftazidime 3 g/day) were thus again administered. One month after the patient's second admission aphthous lesions were noted on the patient's lips and oral mucosa, while erythematous nodules were also found on his legs. HLA typing at this point showed B51. However, the lesion on the left side has completely disappeared on both the T2- and T1-weighted images (Figure 2 F). The symptoms of high fever lasted only 2-4 days and recurred every 1 to 2 months which did not seem to be correlated with the MRI findings. We therefore changed the diagnosis of this case to neuro-Behçet disease even though his clinical findings did not completely fulfill the criteria established by the International Study Group for Behçet's disease. We changed the administration of antibiotics to oral steroids (predonisolone). Six years after the initial presentation, while being continuously administered a small amount of predonisolone, the patient continues to show a high fever lasting from 1 to 3 days every 3 to 4 months. However, such fever symptoms have not been associated with new neurological signs and symptoms or any MRI abnormalities.



**Figure 2.** Sequential changes in the MR findings. **A:** December 17, 1997. MRI shows a lesion of high signal intensity on the T2-weighted image (A-1) and a low signal intensity on the T1-weighted image (A-2) in the right basal ganglia and thalamus. **B:** December 27, 1997. The size of the lesion on the T2-weighted image (B-1) and the T1-weighted image (B-2) increased. The gadolinium-enhanced T1-weighted image (B-3 and B-4) shows a ring-enhanced lesion surrounded by marked edema. **C:** February 10, 1998. The lesion has largely resolved with some residual focus of high and low signal intensity on the T2-weighted image (C-1), a high signal intensity on the T1-weighted image (C-2) and a very slight enhancement on the T1-weighted image (C-3). **D:** March 13, 1998. No new lesion was found in the basal ganglia and thalamus (D-1). **E:** March 16, 1998. An MRI shows a new lesion of high signal intensity on the T2-weighted image (E-1 and E-2) and a low signal intensity on the T1-weighted image (E-3) in the left basal ganglia, thalamus and midbrain. A gadolinium-enhanced T1-weighted image (E-4) shows a very slightly enhanced area, but no ring lesion. **F:** April 21, 1998. The lesion on the left side has completely disappeared on both the T2-weighted image (F-1 and F-2).

## Discussion

With the introduction of MR imaging in clinical practice, numerous articles reporting the MR findings in neuro-Behçet disease have appeared. The most common imaging findings in such patients with neural parenchymal involvement are a mesodiencephalic junction lesion with edema extending along certain long tracts in the brainstem and diencephalons. The lesions seen in the acute or subacute stage tend to show asymmetric distribution, hyperintensity on the T2-weighted images, hypointensity on the T1-weighted images, and nodular enhancement within the lesions on the gadolinium-enhanced study. Such lesions tend to spontaneously resolve over time, leaving small residual lesions at the center of the original lesion. In the chronic stage the lesions frequently become atrophic and show a deposition of hemosiderin.<sup>1,2,4,5</sup>

On the other hand, such brain abscesses tend to evolve through four stages, each with different imaging features.<sup>6</sup> The initial stage of abscess formation is acute cerebritis occurring during the first 4 to 5 days of an infection which shows prolonged T1 and T2 signals on the MRI findings without a significant mass effect or contrast enhancement. By the end of the first week, the late cerebritis stage begins which also shows prolonged T1 and T2 signals on MRI. A mass effect is usually present and some minimal diffuse enhancement may also be seen. Toward the end of the second week, the capsular stage begins and MRI demonstrates a well-defined enhanced capsule of variable thickness surrounding the nonenhanced abscess. In the chronic stage, the edema decreases and it may thereafter become a small area of homogeneous enhancement with a loss of the central abscess cavity. In some cases a complete resolution can occur without residual imaging evidence of the previous infection.

Because of a lack of skin or ocular signs in this patient at onset, we initially considered the first MRI findings to be consistent with acute cerebritis and thus made a diagnosis of a brain abscess because a subsequent ring-enhanced lesion was observed on MRI.

Most articles reporting the MR findings in neuro-Behçet disease are limited to either single cases or to a relatively small series. The frequency of ring-enhanced lesions in neuro-Behçet disease has not yet been described in a relatively large series. As far as we know, only 8 cases have so far been reported in whom the MRI/CT findings showed ring-enhanced lesions according to a study by Yamada et al. in Japan.<sup>7-14</sup> In 3 of these 8 cases<sup>7,9,12</sup> the lesions were located in the basal ganglia and associated with marked edema similar to our case. However, all of these cases either had a history of Behçet disease and/or skin lesions at the onset of the disease. The case was finally diagnosed to have neuro-Behçet disease because of the lesions located extending from the basal ganglia to the midbrain and pons, which are the most common location of this disease, and also due to the later appearance of skin lesions and HLA typing of B51.

Herskovitz et al.<sup>15</sup> suggested that the CT lesions in neuro-Behçet disease may reflect a reversible breakdown in the blood-brain barrier (possibly related to inflammation) rather than gliosis or infarction. Patel et al.<sup>16</sup> reported a case in whom the MR and CT findings demonstrated abnormal signal areas in the thalamus and basal ganglia. Finally, after treatment (consisting of steroids and azathiopurine), a remission of the clinical symptoms was associated with a resolution of all abnormal signals on MR and CT. They concluded that the MR and CT abnormalities observed in neuro-Behçet disease are potentially reversible. Miyoshi et al.<sup>8</sup> reported a case in whom the CT findings showed a ring-enhancement and the lesion resolved spontaneously without any treatment using either steroids or other immunosuppressive drugs. Our case is an additional example suggesting that the lesions in neuro-Behçet disease may resolve spontaneously because it is inconceivable that the antibiotics administered to our patient under the initial diagnosis of a brain abscess could have effectively cured such lesions.

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