Neuro-Behçet’s/Neuro-Sweet’s Disease Presents Simultaneously with Severe Tonsillitis, and Features Mimicking Bacterial Meningitis with Skin Lesions

Taiji Tsunemi¹, Yuki Sakai², Koichi Tsunoda³, Yasuhiro Irie³ and Yoshiaki Wada¹,⁴

Abstract

The patient was hospitalized due to rapidly undulant fever and sore throat. Empirical antibiotic therapy was started, however, headache also occurred. Lumbar puncture disclosed polynuclear leukocyte-predominant pleocytosis indicating that the patient suffered from bacterial meningitis. The antibiotics therapy was increased, however, consciousness became impaired and erythema multiforme-like skin lesions appeared. T2-weighted brain magnetic resonance imaging showed high signal intensity in the brainstem. HLA testing revealed B54 and Cw1. The patient presented futures of Behçet’s disease at the same time as those of Sweet’s syndrome and it was difficult to distinguish between the two diseases. Administration of prednisolone showed remarkable effect.

Key words: neuro-sweet’s disease, neuro-Behçet’s disease, pleocytosis, HLA typing

(Introduction) Behçet’s disease is a multi-system disorder with main clinical symptoms of recurrent oral ulceration, recurrent genital ulceration, and eye and skin lesions (1). Sometimes Behçet’s disease involves the central nervous system and is then called neuro-Behçet’s disease (NBD) (2). Sweet’s syndrome, also referred to as acute febrile neutrophilic dermatitis, is a related disorder (3). It is characterized by inflammatory features including malaise, fever, leukocytosis, and distinctive skin lesions, which heal without scarring, typically show symmetrical, painful, dull-red edematous plaques on the face, neck, upper part of the trunk, and limbs (4). Sometimes inflammation spreads to the other organs. Sweet’s syndrome with central nervous system is termed neuro-Sweet’s disease (NSD) (5, 6). Here we report a patient with simultaneous presentation of features of Behçet’s disease and Sweet’s syndrome. The patient abruptly presented as combined symptoms including severe tonsillitis, and bacterial meningitis mimicking features with skin lesions.

Case Report

The patient was 40-year-old Japanese man. He was admitted to our hospital because of rapidly undulant fever and sore throat. Physical examination revealed severe pharyngitis. Laboratory studies showed the white blood cells to be 17,400/mm³ with a normal differential count. The erythrocyte sedimentation rate was 107 mm in 1 h. Blood chemistry findings were normal. The concentration of C-reactive protein was 4.9 mg/dl. His HLA type was B54 and Cw1. Although pharyngeal and blood cultures were negative, the patient was diagnosed as having tonsillitis, and antibiotic therapy (piperacillin 4 g/day) was started. However intermittent fever persisted and on the eighth day of hospitalization, then he complained of severe headache and was referred to the neurology staff. He was alert with no papilloedema. He had nuchal rigidity and was hyperreflexive, with pathologi-
Figure 1. Brain magnetic resonance imaging findings. T2-weighted (a, b) and fluid-attenuated inversion recovery images (c, d) demonstrated a high-intensity lesion throughout almost the entire brain stem, extending symmetrically from the pons (a, c) up to the midbrain (b, d).

cal reflexes. Examination of cerebrospinal fluid (CSF) revealed a total nucleated cell count of 1331/mm$^3$, with 94% polynuclear cells. The total protein value was 90 mg/dl, and the glucose level was 52 mg/dl. Although repeated CSF cultures failed to yield any bacterial growth, the patient was diagnosed as having bacterial meningitis in light of the polynuclear cell-predominant pleocytosis.

Antibiotic therapy was changed to cefotaxime (12 g/day) and vancomycin (2 g/day). However, his consciousness became impaired, and bilateral abductor nerve palsy occurred. Skin lesions, including painful erythema multiforme-like lesions appeared on his face and on all extremities. Brain magnetic resonance imaging (MRI) showed high signal intensity symmetrically extending from the pons to the midbrain on T2-weighted and fluid-attenuated inversion recovery images (Figs. 1a, through 1d). Occlusion of the dural sinuses was not present. In light of the clinical features, the results of HLA typing, CSF results, and brain MRI, the patient was diagnosed as having NBD/NSD. Administration of intravenous methylprednisolone (1000 mg/day) for 3 days followed by oral prednisolone (60 mg/day) markedly ameliorated the clinical symptoms, laboratory data, and MRI abnormalities. The high signal-intensity lesion gradually decreased, and the patient was discharged after 6 months of hospitalization. Although he experienced grouped small oral ulceration in buccal mucosa and on his tongue a few times per year, no exacerbation of meningitis has occurred.

**Discussion**

Behçet’s disease is a multisystem inflammatory disorder. Since the first report of CNS symptoms associated with
Behçet’s disease (NBD) in 1941, CNS involvement has been considered one of the major and most serious complications, because it impairs the patient’s quality of life and increases the mortality rate (2).

Sweet’s syndrome is considered to be a related inflammatory disorder and was initially characterized by distinctive skin lesions. Later, Sweet’s syndrome was reported to involve multiple organs, but CNS involvement has been less common than that in Behçet’s disease. In 1999, Hisanaga et al reported a Japanese man with Sweet’s syndrome who suffered relapsing and remittent meningoencephalitis and proposed a new entity NSD (5). Since then, over 40 cases has been reported in English and Japanese literature (6).

NBD and NSD share many clinical manifestations (2, 6). Usually neurological symptoms present after the general symptoms. Common features are convulsion, headache, disturbance of consciousness, and motor palsy. The abnormal CSF findings are pleocytosis with increased protein content. Different features are cited as follows. Abnormal lesions in brain MRI are found in mainly in the brainstem in NBD whereas in the cerebrum, basal ganglia, and brainstem in NSD. The outcome is usually good and spontaneous remission is typical in NSD. In contrast, the prognosis is not good in NBD. In general manifestations, the ocular and mouth lesions are less common and less severe in Sweet’s syndrome than in Behçet’s disease. Small and clustered oral ulceration tends to present in Sweet’s syndrome (7). The frequency of HLA B51 is significantly higher in Behçet’s disease. On the other hand, those of HLA B54 and Cw1 are significantly higher in Sweet’s syndrome (8). In addition, the frequency of HLA Cw1 becomes much higher in NSD (93.8%) suggesting that Cw1 may play a critical role in NSD (6).

According to the criteria for NSD, the present case was classified as possible NSD (6) and at the same time, fulfilled the criteria for BD (1). The skin lesions, and location of the MRI lesions were typical for Behçet’s disease but the lack of eye lesions, characteristics of mouth lesions, HLA typing (B54 and Cw1) and steroid-responsiveness were compatible with NSD. Thus, the current case had overlapping features, therefore we diagnosed him as having NBD/NSD and chose steroid treatment which is effective for both diseases. Previously similar cases have been reported. There were cases of Behçet’s disease who presented typical skin lesions of Sweet’s syndrome and cases of Sweet’s syndrome who presented major symptoms of Behçet’s disease (7, 9).

Because the inflammatory symptoms are severe in some cases of Behçet’s disease /Sweet’s syndrome, the main differential diagnosis is infectious diseases (2, 4). When neurological manifestations precede or coincide with general manifestations, the diagnosis can be difficult. NBD/NSD should be considered among the differential diagnoses in patients with inflammatory neurological symptoms of unknown etiology. HLA typing, not only B51 but also B54 and Cw1, is important for diagnosis.

References


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