Case report

A case of seronegative longitudinally extensive transverse myelitis with possible neuro sweet disease

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Dear Editor

Neuro sweet disease (NSD) is frequently described in Asian patients and is characterized by several neurologic manifestations. High levels of human leukocyte antigens (HLA) B54 and Cw1 have been strongly associated with NSD. Radiological findings are seen mainly in the basal ganglia, thalamus, brainstem and very rarely in the spinal cord.

Here, we report a 40-year-old man with subacute progressive paresthesia and gait disturbance. Spinal magnetic resonance imaging (MRI) showed longitudinally extensive transverse myelitis (LETM). Gadolinium (Gd)-enhanced lesions were located ventrodorsally in the spinal cord on the middle line and suggest inflammation or vasoedema around the spinal sulcal vein. Although various antibodies were negative, the HLA type was positive for B54 and Cw1. Steroid treatment was effective for the neurological symptoms and resulted in improved radiological findings, which supported the clinical diagnosis of NSD.

1. Case report

A 40-year-old man with a history of bronchial asthma and mood disorder presented to our hospital with paresthesia in his toes and gradually progressing gait disturbance. He was admitted three months after the initial neurological symptoms appeared. The patient was obese (body mass index: 43.3 kg/m²), but showed no other general physical problems, such as oral or genital ulcerations, skin lesions, or arthralgia. He was alert, and his body temperature was normal. He presented with bladder and rectal disturbance, paresthesia, and loss of vibrations in both ankles. He had gait disturbance due to sensory ataxia without muscle weakness.

Laboratory data was as follows: elevated white blood cell count (WBC, 13030/μl) with a slight increase in neutrophils (76.0%) and mildly elevated C-reactive protein (CRP 1.6 g/dL) and erythrocyte sedimentation rate (ESR 13 mm, 1 h). The HLA-B and C haplotypes were B54/B62 and Cw1/Cw9, respectively. Cerebrospinal fluid examination showed pleocytosis (113/μl: 100% monocyte) and an elevated protein level (259 mg/dL). Other data are shown in Supplemental data. A brain MRI showed no abnormalities. A spinal MRI showed longitudinally extensive intramedullary T2-hyperintense signals from cervical segment (C) 1 to thoracic segment (Th) 11. On axial images, hyperintensity had spread to both gray and white matter. Gd-enhanced lesions were located dorsoventrally in the spinal cord on the middle line (Fig. 1A).

The clinical and radiological course are shown in Fig. 1. The patient was treated with intravenous methylprednisolone (mPSL) pulse therapy (1 g/day for 3 days). His symptoms and radiological findings had improved remarkably. However, two months after first admission, the patient’s symptoms worsened. We used mPSL pulse treatment and started oral administration of steroids (prednisone, 0.5 mg/kg/day). Considering the laboratory findings (positive HLA B54 and Cw1, negative autoantibodies), good reactivity to systemic glucocorticoids, and no evidence for other diseases, the patient met the criteria for possible NSD, neuro-neutrophilic disease in a broad sense [1,2].

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2. Discussion

Sweet disease, also known as acute febrile neutrophilic dermatosis, is characterized by malaise, fever, leukocytosis and cutaneous edematous erythematous plaques that heal without scarring [3,4]. Characteristics include increases in peripheral blood neutrophils, CRP, and ESR [3]. Aseptic neutrophilic inflammation occurs in the dermis and in other organs. Neurologic manifestation of this condition is called NSD and can occur without skin lesions. HLA B54 and Cw1 are frequently positive in NSD patients and can be useful for diagnosis [2]. The most common associations in NSD are encephalitis and meningitis. Spinal cord lesions are very rare. One study reported 1 case of spinal lesions in 29 patients with NSD [1].

The pathogenesis of NSD is not fully clear. Skin biopsy is a valuable tool for diagnosis, with positive indications being deep dermal infiltration of mature neutrophils and the absence of vasculitis [5]. Autopsies in cases of NSD reported neuropathological findings of peri-vascular cuffing around particularly small veins, information which may be useful for the diagnosis of NSD in patients with myelitis [6]. In the present case, a skin biopsy was not possible, nor was a spinal biopsy.

Although there was no pathological evidence of NSD, pericapillary inflammation may cause reversible vasogenic edema, which we speculated was the cause of the spinal lesions because they were presumably along the vessels, and the symptoms were relatively light. These findings may be useful for diagnosis of NSD in patients with myelitis. On the other hand, it is sometimes difficult to distinguish NSD from neuro-Beçet disease because those are placed in the similar pathogenesis of neuro-neutrophilic disease [2,7]. Therefore, careful monitoring for dermatologic signs should be continued in patients with positive HLA B54/Cw1. Further cases are required to evaluate the association between HLA typing and seronegative LETM.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying data.

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Declaration of Competing Interest

Hirofumi Maruyama reports receiving grants from Daiichi Sankyo Co., Ltd., which is unrelated to the submitted work.
All other authors declare that they have no conflicts of interest.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.ensci.2020.100227.

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