INTRODUCTION

Conversion disorder (CD), also known as functional neurological symptom disorder, is classified as a somatic symptom and related disorder in the Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition [1]. CD may be accompanied by sensory/motor symptoms, such as weakness or partial paralysis, sensory loss, and motor deficits. Sjögren’s syndrome (SS) and systemic lupus erythematosus (SLE) are complicated by a wide spectrum of neurological disorders and present abnormalities on brain and spinal cord mag-
netic resonance imaging (MRI) and in the cerebrospinal fluid (CSF) [2-4]. The levels of interleukin-6 in CSF are also elevated in cases of central nervous system (CNS) lupus [5]. Corticosteroid therapy is a basic approach for the treatment of SLE, but treatment is intensified when severe CNS symptoms are present. In a previous study of neurological symptoms in SS, intravenous immunoglobulin (IVIG) therapy was more effective than methylprednisolone pulse therapy [6]. Approximately 30% of SS patients present with psychiatric manifestations [7], while 46% of SLE patients present with neuropsychiatric manifestations [8]. Reports of SLE and SS cases accompanied by CD are very rare, possibly because the symptoms of the two disorders are indistinguishable and can lead to an ambiguous diagnosis.

**CASE PRESENTATION**

**SS and SLE**

A 14-year-old girl was admitted to our hospital for the treatment of idiopathic thrombocytopenic purpura (ITP) because her platelet count dropped to 19,000/μL two weeks after an upper respiratory infection. The patient had experienced dry mouth and hypohidrosis for the previous two years. A butterfly rash was observed 20 days after the decrease in platelet count. There was no abnormality in the iris. Some antinuclear antibodies, such as anti-Ro/SSA and La/SSB antibodies, and rheumatoid factor were positive, while other related antibodies were negative. Urine test results were all negative. While Schirmer’s test showed normal tear production, the Saxon test and salivary scintigraphy revealed impaired salivary secretion. Other clinical findings were bilateral knee pain, an elevated serum matrix metalloproteinase-3 level, and a small amount of effusion in the knee on contrast-enhanced MRI. No abnormalities were observed on chest X-ray, echocardiography, brain perfusion single-photon emission computed tomography, or gallium scintigraphy. After SLE was diagnosed, we performed a biopsy to evaluate the patient’s stage of SLE. Kidney biopsy findings were consistent with stage 1 pathology in accordance with the International Society of Nephrology (ISN) classification. Based on the test results and symptoms, we diagnosed concomitant SS and SLE. After starting corticosteroid therapy for ITP, the patient experienced neurological symptoms starting in her lower limbs, and treatment was delayed due to the difficulty encountered in determining whether the symptoms were attributable to a neurological or mental disorder. Two cycles of methylprednisolone pulse therapy were completed one month after the onset of the neurological symptoms. Subsequently, the joint pain and erythema disappeared. Oral corticosteroids 20 mg/day and tacrolimus 3 mg/day alleviated the joint pain.

**Neurological symptoms**

Two days after the patient began corticosteroid therapy for ITP, we observed muscle weakness in her lower limbs (Fig. 1). The weakness gradually spread throughout the length of her lower limbs and to the upper limbs, leading to a loss of muscle strength in all four limbs and increased tendon reflexes. Despite mildly reduced muscle strength in her four limbs, no muscle atrophy, spastic paralysis, or bladder/rectal disorder was observed. Tests for pathological reflexes, such as Babinski reflex, were also negative, with no findings indicating pyramidal tract lesions besides her increased tendon reflexes. The patient could not lift her lower limbs but was able to hold them in a raised position. Hoover’s sign was positive. The anatomical sites of sensory impairment varied from day to day. Hypersensitivities to tactile, temperature, and pain stimuli were observed on the right side of her face. While her left lower limb and right upper limb had normal sensory perception, her right lower limb and left upper limb were hypersensitive to tactile and temperature stimuli. There were no signs of impairment in position sense, cranial neuropathy, or cerebellar ataxia. Although nociception and temperature perception returned to normal, hyperalgesia persisted in the soles of both feet on the third day of treatment; this symptom eventually decreased and disappeared completely 15 days later. Blood and CSF analysis, brain and spinal cord MRI, nerve conduction velocity, and electroencephalography revealed no abnormalities. We started physical therapy 14 days after the onset of muscle weakness. The first cycle of three-day methylprednisolone pulse therapy (500 mg) began after 7 days of corticosteroid therapy, but no improvement in the patient’s condition was observed, even after the second cycle. The first and second cycles lasted four weeks. IVIG therapy was performed because, in addition to methylprednisolone pulse therapy, IVIG therapy was found to be effective for neurological symptoms caused by SS in a previous study [9]. Therefore, five-day IVIG therapy (400 mg/kg/dose) was initiated 44 days after the onset of muscle weakness. A few days after the first five-day IVIG administration, spontaneous motor activity was observed in the lower limbs. In the upper limbs, only the fingers showed improvement. However, no clear improvement was observed after the second five-day IVIG administration (Fig. 1).
Blood and CSF analysis, brain and spinal cord MRI, and nerve conduction velocity tests were repeated but revealed no abnormalities. We made no changes to the patient’s oral medicine and started psychotherapy 85 days after the onset of muscle weakness. Spontaneous motor activity in the upper limbs resumed after hospital discharge. One month after discharge, spontaneous motor activity in all four limbs had returned to normal. However, the increased tendon reflexes in all four limbs remained.

Psychiatric manifestations

The patient had past medical history relevant to psychiatric manifestations, such as epilepsy; additionally, she had stopped attending school because of bullying and anorexia. The patient had no voluntary movement of her upper limbs but did move them upon painful stimulation, leading to a diagnosis of dissociation. After eliminating other causes of neurological symptoms, we made the diagnosis of CD with sensory and motor impairments. We recommended psychiatric counseling for her, but she refused it. We performed mental supportive care that attended to her needs without denying her wishes and gradually promoted independence.

DISCUSSION

It is of great importance to determine whether paralysis in all four limbs is caused by a neurological or mental disorder because the former urgently requires an intensive intervention, such as methylprednisolone pulse therapy or IVIG therapy [4], whereas psychotherapy and/or antidepressant drug therapy are recommended for psychiatric manifestations. Previous studies have shown that SS can accompany various types of neuropathy: ① sensory ataxic neuropathy (40% of previous cases), ② painful neuropathy without sensory ataxia (20%), ③ multiple mononeuropathy, ④ autonomic neuropathy, and ⑤ radiculopathy. In the present study, the above pathologies were ruled out for several reasons. ① Despite the presence of cutaneous sensory impairment and dyshidrosis in our patient, sensory ataxic neuropathy was ruled out because of the lack of impaired proprioception and ataxic gait, the presence of muscle weakness, and the lack of abnormalities on the spinal cord MRI or nerve conduction velocity tests. We did not measure short latency somatosensory evoked potentials (SSEP). ② Despite normal nerve conduction velocity and spinal cord MRI and the fact that the patient’s first symptom was dysesthesia in the distal portions of all four limbs, painful
neuropathy without sensory ataxia was ruled out because of the lack of aggravation of sensory impairment and the presence of muscle weakness. Hypersensitivity, a symptom of small fiber neuropathy, was observed, but the area of hypersensitivity varied from examination to examination and disappeared after a few days. Reduced thermal nociception or autonomic symptoms were absent. Multiple mononeuropathy, which involves sensory and motor disorders in the anatomical area innervated by a single nerve, was ruled out because the anatomical sites of sensory and motor disorders differed from day to day in this patient. Autonomic neuropathy, such as orthostatic hypotension and bladder rectal disorder, was unlikely because of the presence of muscle weakness and the lack of symptoms indicative of autonomic neuropathy (other than dyshidrosis) in the patient. Radiculopathy was ruled out because of normal nerve conduction velocity and no F-wave conduction delay (25.1/24.8 ms in the left/right median nerve, 23.8/23.9 ms in the left/right ulnar nerve, and 46.6/46.9 ms in the left/right tibial nerve) and because of normal MRI and CSF findings, despite the presence of muscle weakness. There were day-to-day variations in neurological symptoms, and the patient’s activities of daily living did not match the objective muscle strength findings. Moreover, the findings of sensory impairment appearing for only a few days varied from examination to examination, and the patient could not raise her lower limbs but could hold them in place. These inconsistencies in clinical findings and the positive Hoover’s sign were the reasons for suspecting CD. Similarly, in a previous study, CD was diagnosed because of a lack of abnormal findings in a patient who developed paralysis in all four limbs while undergoing steroid therapy for juvenile rheumatoid arthritis [10]. In our case, the patient also developed neurological symptoms while undergoing steroid therapy, suggesting that her pathological condition was unresponsive to steroids. Clinical testing and imaging showed no abnormality, but because it could not be completely ruled out, methylprednisolone pulse therapy was performed as a diagnostic/therapeutic regimen. The fact that psychiatric manifestations are uncommon in SS and SLE and that intensive methylprednisolone pulse therapy and IVIG therapy, which were performed to prevent any delay in treating neurological disorder, were not clearly effective in the present case led us to suspect that psychiatric manifestations were the cause of the patient’s symptoms. Because we did not believe that the neurological symptoms were due to the SLE, we did not strengthen the patient’s steroid therapy. Corticosteroid therapy with prednisolone (PSL) and tacrolimus are less effective against SLE without neurological symptoms. In our case, we started physical therapy 14 days after the appearance of symptoms and started mental supportive care 85 days after the appearance of symptoms. The patient’s condition gradually improved. We could not reduce tacrolimus, but were able to reduce PSL rapidly to 1/4 within half a year. There has been no recurrence. In a similar case, Yamaguchi et al reported that conversion symptoms of a rheumatic patient improved with physical therapy, occupational therapy, and antidepressant drug therapy [10].

CONCLUSION

We encountered a patient with no abnormal test findings whose symptoms could not be explained by medical evaluation. Based on the present findings and the patient’s history, we determined that CD was responsible for her neurological symptoms. Because early diagnosis of the cause of neurological symptoms accompanying SS and SLE is difficult, in the early stages it is often necessary to provide two lines of treatment: one for a neurological disorder and another for a mental disorder. However, when a mental disorder is suspected, treatment for neurological disorder can be minimized with early psychotherapeutic intervention.

REFERENCES