Cervical Root Enlargement in Segmental Zoster Paresis: A Study with Magnetic Resonance Imaging and Nerve Ultrasound

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Abstract:
A 72-year-old woman presented with acute-progressive muscle weakness after a rash in the left upper limb. Muscle weakness was restricted to the left C5 innervated muscles. Short inversion time inversion recovery magnetic resonance imaging (MRI) showed a high-intensity signal in the left C5 nerve root, and nerve ultrasound showed its enlargement. She was diagnosed with segmental zoster paralysis (SZP) and treated with acyclovir and methylprednisolone. Her muscle strength gradually recovered, and the abnormal signal and enlargement in the left C5 nerve root improved. This is the first SZP case of confirmed improvement of abnormal findings on MRI and nerve ultrasound in association with muscle power recovery.

Key words: segmental zoster paresis, MRI, nerve ultrasound, follow-up, cervical nerve root, Sjögren syndrome

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Introduction

The frequency of varicella zoster virus (VZV) reactivation in the general population during their lifetime ranges from 20-30% (1). VZV reactivation induces various neurological diseases, such as vasculopathy, myelopathy, postherpetic neuralgia, retinal necrosis, and cerebellitis (2). Segmental zoster paresis (SZP) is an infrequent VZV-related complication characterized by focal muscle weakness, occurring in 0.5-5% of patients with VZV reactivation (3). Therefore, its characteristics, including imaging changes in SZP patients, have not been yet fully elucidated.

We herein report a case of SZP with confirmed improvement of abnormal findings on magnetic resonance imaging (MRI) and nerve ultrasound (US).

Case Report

A 72-year-old woman was admitted to our hospital with acute-progressive shooting pain and muscle weakness in the left upper limb a week prior to admission (day 1). In addition, she had developed a rash on the left upper limb at the C5 and C6 dermatomes two weeks earlier and been diagnosed with VZV infection at another hospital (Fig. 1). Her medical history showed that she had undergone surgery for right-sided breast cancer four years prior and was taking tamoxifen. She had also experienced dry eyes and mouth for three years.

A neurological examination on admission showed muscle weakness in the left deltoid, infraspinatus [Medical Research Council (MRC) Scale for Muscle Strength: 2], and biceps brachii muscles (MRC scale for muscle strength: 4). A detailed sensory evaluation was difficult because of severe pain, but we found an absent deep tendon reflex of the left
biceps brachii muscle. Findings from a complete blood count and biochemical testing, including the liver and kidney function, were normal. She tested positive for VZV EIA-IgG antibodies (antibody index: 115) and VZV EIA-IgM antibodies (antibody index: 4.25). In addition, anti-Ro/SS-A antibodies (>1,200 U/mL) and anti-La/SS-B antibodies (713 U/mL) were detected in the blood, and a salivary gland biopsy from the lip revealed lymphocytic infiltration, indicating Sjögren syndrome. A cerebrospinal fluid (CSF) analysis on admission showed a mildly increased cell count (total 13/μL: monocytes, 12; segmented cells, 1) and normal protein levels (39.3 mg/dL) (Fig. 2). A polymerase chain reaction test for VZV-DNA in the CSF was negative.

Nerve conduction studies showed no obvious abnormalities in the left median or ulnar nerves. Electromyography revealed positive sharp waves and fibrillation potentials in the left deltoid muscle. No abnormality was found on brain MRI; however, enlargement and a high-intensity signal in the left C5 nerve root were detected on spinal short inversion time inversion recovery (STIR)-MRI (Fig. 3). Contrast-enhanced spinal MRI indicated enhancement in the left C5 nerve root, including the dorsal root, on the coronal and axial views (Fig. 3). No abnormal signals were detected in other nerve roots or in the spinal cord. US also showed left C5 nerve root enlargement [cross-sectional area (CSA): 15 mm², normal range: 5.66±1.02 (mean±standard deviation) mm²] (Fig. 4) (4). No obvious enlargement was detected in the left C6 nerve root (CSA: 11 mm², normal range: 8.98±1.65 mm²) (4).

Given the above, she was diagnosed with SZP and Sjögren syndrome and treated with acyclovir 1,500 mg/day for 11 days and methylprednisolone 1,000 mg/day for 3 days twice on days 4 and 11. CSF results on day 9 showed a decreased cell count (4/μL). With continuous rehabilitation, muscle weakness in the left biceps brachii muscle improved completely by day 64, and she was discharged on day 89. After discharge, shooting pain persisted. The strength of the deltoid and infraspinatus muscles recovered fully by day 146. In addition, the enlargement and high-intensity signal of the left C5 nerve root on STIR-MRI and the enlargement on US improved by day 202 (CSA: 8 mm²) (Fig. 3, 4).
SZP is characterized by focal muscle weakness in a spinal segment with skin rash, neuralgia, and sensory symptoms due to VZV reactivation (5). Sjögren syndrome often causes different neuropathy forms, such as sensory neuropathy, multiple mononeuropathy, cranial neuropathy, and radiculopathy (6). However, radiculopathy due to Sjögren syndrome is reported to present with slow-progressive polyradiculopathy, instead of monoradiculopathy, and elevated CSF protein levels (6). These characteristics are not consistent with our case. In our patient, muscle weakness was restricted to the C5 segment, where rash and shooting pain were observed. Accordingly, we diagnosed the patient with SZP.

The pathogenic background of patients who develop SZP is not fully known. Apart from aging, an immunosuppressive state with cell-mediated immune abnormalities leading to VZV reactivation may be involved in the development of SZP (7). For example, there are reports of SZP patients with diabetes mellitus and immunocompromised breast cancer being treated with chemotherapy and radiotherapy (3, 8). In our case, tamoxifen use may have been one of the factors associated with the development of SZP (9). To date, there have been no reports concerning the direct relationship between SZP and Sjögren syndrome. However, Sjögren syndrome may have also contributed to the development of SZP in our case, as it was suggested that Sjögren syndrome is a risk for VZV reactivation (10).

Although antiviral agents and corticosteroids are mainly administered for SZP treatment, their adequate dosage and duration of administration have not been determined (11, 12). Furthermore, the prognosis of SZP has not been fully clarified, as some patients recover within several months, whereas other patients do not (3). In our case, the muscle weakness in the left upper limb recovered completely within six months, indicating a favorable outcome.

While enlargement and high-intensity signals of a single nerve root on T2-weighted MRI were detected in some SZP cases at the diagnosis, no report followed the clinical course over a long term (3, 5, 13-17). In SZP, inflammation is supposedly induced by virus reactivation or virally mediated vasculitis (13, 14). Although abnormal imaging signals may reflect inflammation, other infectious or inflammatory diseases also present similar findings that are not specific for SZP (14). This is the first case to indicate a change in US
Figure 4. Nerve ultrasound (US) findings. (A) Short-axis views of the left C5 nerve root on US. Cross-sectional area (CSA) traced by blue line. (B) The CSA of the left C5 nerve root gradually decreases [normal range: 5.66±1.02 (mean±standard deviation (SD)) mm²] (4). The CSA of the left C6 nerve root shows no obvious enlargement (normal range: 8.98±1.65 mm²) (4). The CSA of the cervical root is measured around the point where the nerve root exits over the transverse process or just distal (20, 21), and the maximum CSA around the point is defined as the CSA of the cervical root. US was performed by well-trained examiners (H.E. and K.O.) and registered neurosonographers of the Japan Academy of Neurosonography (S.W. and N.T).

We confirmed the improvement of MRI and US findings in our case. Because improvements in abnormalities on MRI and US occurred later than those in muscle weakness, the usefulness of US or MRI for predicting the outcome was not proven. The accumulation of more cases is needed to assess their usefulness, including the timing of improvements.

The authors state that they have no Conflict of Interest (COI).

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