Callosal Disconnection Syndrome Associated with Relapsing Polychondritis

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Abstract

Relapsing polychondritis (RP) is a rare inflammatory disorder of the cartilaginous structures, and it sometimes involves the central nervous system. Encephalitis associated with RP causes a wide variety of symptoms according to the affected sites. We herein report the first case of 72-year-old right-handed man who developed acute meningoencephalitis associated with RP involving the corpus callosum. After immunosuppressive therapy, his symptoms dramatically improved, but difficulty in performing bimanual movements with occasional diagnostic dyspraxia in his right hand remained. Because callosal signs are easily missed, especially in acute settings, it would be useful to know that RP can sometimes cause callosal disconnection syndrome.

Key words: relapsing polychondritis, callosal dysconnection syndrome, diagnostic dyspraxia


Introduction

Relapsing polychondritis (RP) is a rare relapsing and remitting inflammatory disorder of the cartilaginous structures (1). RP has been reported to cause central nervous system vasculitis in rare cases, thus leading to various neurological symptoms, such as meningitis, encephalitis (2), parkinsonism (3) and dementia (4), according to the site of lesions. We herein describe, for the first time, that RP may sometimes cause callosal disconnection syndrome.

Case Report

A 72-year old right-handed man with 16 years of education was found lying drowsy on the floor and thereafter was brought to the nearest hospital by ambulance. His past medical history included diabetes mellitus, hypertension, postoperative thoracic aortic aneurysm and cured gastric MALT (mucosa-associated lymphatic tissue) lymphoma. Two years before this presentation, at 70 years of age, he presented with eruptions over his body and extremities. He was given 60 mg/day oral prednisolone and the eruptions thereafter disappeared, however he developed pain and swelling in his left ear while tapering prednisolone to 5 mg/day. A biopsy of the auricular cartilage showed perichondral inflammation with lymphocytic infiltration. A diagnosis of RP was thus made and oral prednisolone was increased to 10 mg/day with a complete recovery of the symptoms. He then remained in good health until 72 years of age, when he developed a high fever and drowsiness two days before admission.

On admission, the patient was febrile and somnolent. Dysarthria and neck stiffness were evident. Blood tests revealed an elevated white blood cell count and C-reactive protein concentration. A cerebrospinal fluid (CSF) examination showed pleocytosis of 781/mm³ white blood cells with 87% polymorphonuclear leukocytes, a glucose level of 38 mg/dL (blood glucose 149 mg/dL), and a total protein of 582 mg/dL. A computed tomography scan of the brain showed no obvious abnormalities. With the presumptive diagnosis of acute bacterial meningitis, he was treated with dexametha-
in writing especially with his left hand (Fig. 2a, b). He had difficulty in repetition, and reading were preserved, but he had difficulty in his spontaneous speech. The abilities of comprehension, locutions and semantic paraphasias were sometimes noted in speech, but his articulation and prosody were adequate. Circumstantial thought was noted as to the time and place, and he achieved a score of the observed neurocognitive deficits. He was alert but disoriented. The neurological examination were not remarkable, except for the involvement of the medial frontal cortex. All CSF specimens were negative for bacteria, virus, and fungi or abnormal cells. Acute bacterial meningitis was thought to be unlikely, and alternatively meningoencephalitis associated with RP was highly suspected. All antibiotics were discontinued and oral prednisolone at a dose of 60 mg/day was started, and thereafter his condition improved gradually.

One month after admission, he was transferred to our hospital and oral prednisolone was gradually tapered to 10 mg/day and pulsed intravenous cyclophosphamide (500 mg monthly for 4 months) was administered. At that time, he complained of speech difficulty and sometimes reported that his left hand would not respond as intended. The results of a neurological examination were not remarkable, except for the observed neurocognitive deficits. He was alert but disoriented as to the time and place, and he achieved a score of 13 on the Mini-Mental State Examination (MMSE). His speech was slow and often stopped due to occasional stuttering, but his articulation and prosody were adequate. Circumlocutions and semantic paraphasias were sometimes noted in his spontaneous speech. The abilities of comprehension, repetition, and reading were preserved, but he had difficulty in writing especially with his left hand (Fig. 2a, b). He could not perform pentagon copy, cube copy and clock drawing in either hand, thus indicating a visuospatial impairment (Fig. 2e). Furthermore, mild left-sided hemispatial neglect was observed when copying line drawings with his right-hand (Fig. 2c, d). Ideomotor apraxia of both hands was observed on verbal command and on imitation. He could not identify his left hand fingers or objects placed in his left hand with his eyes closed, but he was able to do so under visual observation.

After a rehabilitation period of 4 months in our hospital, his cognitive status improved significantly and his MMSE score improved to 21. His speech became faster and more fluent except for some stuttering. He became able to write with his right hand proficiently. Visuospatial impairment also improved remarkably, but he could not copy a double-pentagon or perform clock drawing (Fig. 2f). Apraxia of both hands was markedly diminished. Despite the significant improvement, he still had difficulty in performing bimanual movements, such as tying a string, lacing up shoes and fastening a button, as the right hand would not help the left hand and at times even tried to interfere with the action of the left hand (diagonistic dyspraxia), even though he often felt a loss of control of his left hand. On repeated MRI, subcortical white matter lesions became less remarkable and the lesions in the corpus callosum persisted.

Discussion

Corpus callosum involvement associated with RP has been inferred on MRIs in a previous report, although callosal disconnection syndrome has never been previously described (5). Because callosal signs and symptoms are prone to be missed, especially in acute settings (6) which can lead to a misestimation of the disease severity, it would therefore be useful to know that RP can sometimes cause callosal disconnection syndrome accompanied by encephalitis.

It is noteworthy that the diagnostic dyspraxia in the present case was in the right hand, although it generally occurs in the left (non-dominant) hand in right-handed individuals (7, 8). This reversed lateralization can be explained by the possibility that our patient had a mixed cortical dominance. In general, dysgraphia and ideomotor apraxia emerge in one’s non-dominant hand in callosal disconnection syndrome (9, 10), but these were observed in both hands at the initial stage, which also suggests an altered cortical lateralization in this patient. Nevertheless, tactile anomia (or asterognosia) of the left hand suggests that the left hemisphere dominance of language function (10).

Another peculiar thing to be noted in this case is a discrepancy between the affected-sides of diagonistic dyspraxia and the loss of the sense of agency. It is possible that the left hemisphere accounts for the verbal expression of a feeling of the loss of control over his left hand, even if the left hand actually manifested the patient’s intention.

In the current study, we describe a patient who presented with right-sided diagonistic dyspraxia due to callosal discon-
connection secondary to RP. The present report adds an additional case of callosal disconnection syndrome in a RP patient. RP should therefore be considered as a rare cause of alien hand syndrome.

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

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References