

**C39**

## **Recurrent Complex Regional Pain Syndrome Type I in a Patient with Amyotrophic Lateral Sclerosis**

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### **Introduction**

Amyotrophic lateral sclerosis (ALS) is a rapidly progressing neurodegenerative disease that involves limb, axial, bulbar, and respiratory muscles. ALS involves degeneration of the motor system combining upper and lower motor neuron signs. Although sensory or dysautonomic signs are not clinically evident, pain can occur in these patients. However, it is usually secondary to severe paresis and immobility, such as frozen shoulder resulting from severe upper limb paresis. In previous research, there have been several reports about ALS patients with complex regional pain syndrome (CRPS). To the best of our knowledge, however, there has been no report about recurrent complex regional pain syndrome in patients with ALS. Here, therefore, we report on an ALS patient with recurrent CRPS, which seemed to be caused by adhesive capsulitis.

### **Case**

A 60-year-old man with ALS was admitted to our rehabilitation clinic with a two-year history of progressive motor weakness. He also had right shoulder, wrist and hand pain with swelling of the right hand and wrist joint. The three-phase bone scan revealed increased uptakes in the right carpal, metacarpophalangeal, proximal interphalangeal joint, which is consistent with CRPS. He had recurrence of CRPS in his arm with intervals of 4 months.

### **Discussion**

CRPS type I is a syndrome with pain and signs of swelling, hyperhidrosis, vasomotor instability and trophic changes in the skin, muscle, and bone in the affected limb, and, by definition, without major nerve injury. There are several theories regarding the pathogenesis of CRPS. Some scientists emphasize the role of the sensitization of the peripheral nerve or spinal cord dorsal horn neuron, whereas some clinicians give weight to abnormalities in the structures of the central nervous system. Recently, some clinicians have emphasized the role of immobilization as a contributing factor for CRPS. It has also been reported that only immobilization without any tissue injury could cause mechanical allodynia in an animal study of a CRPS rodent model. In our case, CRPS type I developed by immobility itself as well as frozen shoulder caused by immobility of the right upper limb. In fact, the patient's right shoulder pain preceded swelling and pain of the right hand and wrist joint in the clinical history. Therefore, his uncontrolled shoulder pain due to the frozen shoulder combined with immobility seems to have progressed into CRPS type I. The second CRPS seems to have recurred as the frozen shoulder became aggravated due to immobility of the right upper limb after the first steroid pulse treatment. Collectively, in patients with ALS, frozen shoulder is more likely to occur

due to immobility of the upper limb, which may also progress to CRPS. In ALS patients with frozen shoulder, therefore, early treatment, such as an intra-articular corticosteroid injection, PROM and stretching exercises will be necessary to prevent progression to CRPS

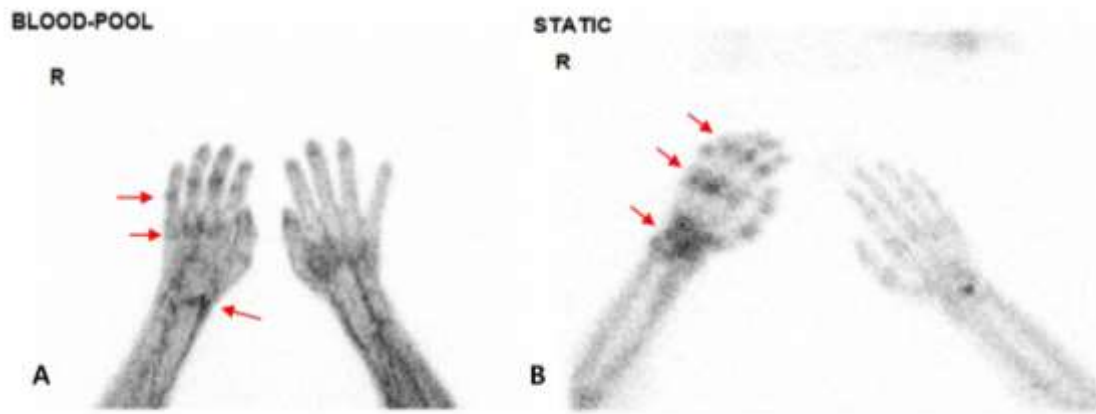


Fig 1. Three-phase bone scan of both upper limbs. (A) In the blood flow and pooling phase, increased venous blood pooling of the right hand was shown compared with the left hand. (B) In the osseous phase, increased uptake in the right carpal, metacarpophalangeal, proximal interphalangeal joint was shown compared with the left hand.