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Pseudo-dystonia in sarcoid myopathy

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Abstract

We describe a 61-year-old woman with difficulty extending her left ring finger and little finger caused by sarcoid myopathy. Since her symptom temporarily improved with carpal flexion or forearm pronation, we once misdiagnosed her as having dystonia of upper limb. Her symptom gradually worsened and muscle biopsy specimen revealed sarcoid myopathy. Muscle MRI of left forearm showed abnormal signals in the quadriceps femoris and biceps brachii muscles, and area surrounding the flexor digitorum profundus (FDP) and supinator muscles. Treatment with prednisolone was effective and stopped progression of the symptom. Adhesion of supinator muscle and FDP due to inflammation may have caused limited extension of FDP.

Key words; flexion contracture, Movement Disorders, Muscle Disease, pseudo-dystonia, sarcoid myopathy,

Introduction

Sarcoidosis is a systemic granulomatous disease of unknown etiology that affects multiple organs such as the lungs, uvea, lymphatics, heart, nervous system, and muscles(1). Symptomatic sarcoid myopathy occurs in less than 1% of patients with sarcoidosis, and, in most cases, patients have systemic symptoms prior to the development of sarcoid myopathy(2). Patients with sarcoid myopathy often manifest weakness, muscle atrophy, muscle pain, and high creatinine kinase levels, but rarely show muscle contracture. Here, we present a case with difficulty extending her fingers due to sarcoid myopathy, and who was misdiagnosed as dystonia. Sarcoid myopathy rarely causes muscle contracture.

Case Report

A 61-year-old woman presented with a 2-year history of difficulty extending her left ring finger and little finger. Before visiting our hospital, she was diagnosed with trigger finger and underwent surgery, but her symptoms did not improve. Her left ring finger and little finger were in a flexed position, and she experienced difficulty extending these fingers that improved with carpal flexion or forearm pronation (Video S1). She presented no muscle weakness, myalgia, muscle tenderness, sensory disturbance,

cerebellar ataxia or extrapyramidal sign. Her general condition was well and no visual disturbance, skin lesion or shortness of breath indicative of systemic sarcoidosis was observed. Brain, spinal, and muscle MRI and needle electromyography results were unremarkable. Though her symptoms did not show task specificity or sensory trick, upper limb dystonia was suspected because her symptom fluctuated depending on the forearm posture. Her symptoms gradually worsened and muscle MRI at 3 months after her first visit showed abnormal signals in the quadriceps femoris and biceps brachii muscles, and surrounding the flexor digitorum profundus (FDP) and supinator muscles, as well as the interosseous membrane. The patient underwent surgery for flexion contracture. During surgery, adhesion of the FDP to the ulnar periosteum and funicular structure on the surface of the FDP was observed. Examination of a biopsy specimen from the FDP showed epithelioid granuloma within the muscle. In addition, high serum angiotensin-converting enzyme levels, a negative tuberculin skin test, bilateral hilar lymphadenopathy, and F¹⁸-FDG uptake to hilar and supraclavicular lymph nodes were observed. These findings indicate systemic sarcoidosis and sarcoid myopathy. Her symptoms worsened and corticosteroid was initiated. Although the administration of prednisolone (50 mg/day orally) was effective and stopped the worsening of her symptoms, her symptoms worsened on prednisolone withdrawal. Treatment with

prednisolone was resumed, and her symptoms were stabilized with 2.5 mg/day of prednisolone.

Discussion

There are a few cases in the literature demonstrating sarcoid myopathy presenting as flexion contracture (3, 4), but this is the first report describing the extent of flexion contracture as dependent on posture. In our case, adhesion of the FDP to the ulnar periosteum and funicular structure on the FDP was observed, which limited full extension of the FDP. Improvement of the symptoms on carpal flexion can be attributed to the shortened distance between the distal interphalangeal joint and ulnar head in that posture. We observed abnormal signals on MRI in the area surrounding the FDP and supinator muscles and the interosseous membrane, suggesting the presence of inflammation and adhesions. One possible reason for the limited extension of the patient's fingers on forearm supination is that the contracted supinator muscle impinges the FDP and limits its extension.

Sarcoid myopathy may cause difficulty extending the fingers, which resembles dystonia. Changes in symptoms severity depending on posture may be helpful to distinguish sarcoid myopathy from dystonia.

Acknowledgments

The authors declare no conflict of interest.

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Figure Legend: Muscle MRI and pathology

On axial MRI images of the left forearm, (A) T1 weighted image (1.5 T; repetition time [TR] 681.0 ms; echo time [TE] 9.4 ms) and (B) Short-tau inversion recovery image ([TR] 4025.1 ms; [TE] 60.0 ms) shows high signal areas surrounding the FDP (arrow) and supinator (arrowhead) muscles. (C and D) Muscle biopsy specimens (Hematoxylin and Eosin stain) displays widespread inflammatory infiltrates, noncaseating granuloma (C, x20) and Langhans giant cells (D, x40), indicative of sarcoid myopathy.

Video S1 Legend: Pseudo-dystonia in sarcoid myopathy

The patient presents difficulty extending her left ring finger, which resembles a dystonia posture, but the difficulty improves with carpal flexion.

Carpal flexion also improves the symptom. The left little finger remains at a flexion position in every posture due to progression of the disease.

Figure. Muscle MRI and pathology

