Granulomatous Myopathy

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Last month's case challenge (available in the video section at Practical Neurology.com) presented a case of a 65-year-old woman with a 15-year history of slowly progressive weakness of the arms and legs.

THE MOST LIKELY DIAGNOSIS IS:

1. Inclusion body myositis
2. Polymyositis
3. Granulomatous myositis
4. Vasculitic myositis
5. Parasitic myositis

The Correct Answer is Granulomatous myositis

Muscle biopsy revealed multiple non-caseating granulomata that contained many multinucleated giant cells. Patients with chronic distal hands weakness are usually given different diagnoses such as CTS, cervical radiculopathy, arthritis, etc., and it may take years before they see a neuromuscular specialist. The lack of sensory symptoms and the diffuse nature of the weakness were consistent with:

- Myopathy
- Motor neuron disease
- Hirayama disease
- Myasthenia gravis

Myopathic EMG confirmed the diagnosis of distal myopathy and raised the following possibilities:

- Distal muscular dystrophies
- Inclusion body myopathies
- Myofibrillar myopathies
- Muscle biopsy reveals unexpected diagnosis. There was several non-caseating granulomas and endomyseal inflammation.

Granulomatous myopathy is a rare form of inflammatory muscle disease. It may present as progressive proximal weakness along with MG and thymoma or as a chronic distal weakness as a manifestation of sarcoidosis. Frequently, search for sarcoidosis is not productive and the diagnosis of idiopathic granulomatous myopathy is given. CK level is normal to five times the normal level. EMG shows chronic myopathic findings (mixed short and long duration MUPs and early recruitment). Prognosis is not good and unfortunately, response to immunosupression or modulation is poor in the distal form. Unlike the proximal variant, the distal idiopathic type does not affect the heart but it may extend to the proximal muscles. ■

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1. Jasim S, Shaibani A. Nonsarcoid granulomatous myopathy: two cases and a review of literature. Int J Neurosci C. 2013 Jul;123(7):516-20.