CLNICAL VIGNETTE

Muscle Weakness: A Case of Polymyositis

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Case Report

Approximately 4 months ago, a previously active 47-year-old man with a history of eczema, presented to his primary care provider feeling weak with diffuse myalgias and intermittent fevers up to 38.9°C. He also had a mild productive cough.

He was diagnosed with a right lower lobe pneumonia based on a chest radiograph and was treated with a 5-day course of azithromycin.

A month after completing his antibiotics he returned to his primary care provider with persistent diffuse myalgias and muscle weakness. He also noted polyarthralgias, particularly in his hands, elbows, knees and ankles.

His appetite and weight were unchanged and he was free of rashes, abdominal pain, melena, hematochezia and urinary symptoms. The patient reported no new sexual contacts or recent travel.

His past medical history included hypertension and prior cataract surgery. He was allergic to tacrolimus ointment and takes only hydrocodone as needed for joint pain.

He is married and is employed as a marketing director. He consumes about 2-3 alcoholic beverages a week, does not smoke tobacco and occasionally smokes marijuana. There is no intravenous or illicit drug use.

Vital signs included temperature of 37.2°C blood pressure of 124/68, pulse of 84, respiratory rate of 12 per minute with an oxygen saturation of 96% at room air. Exam revealed symmetrical 3/5 muscle strength in the upper and lower extremities confined to the proximal muscle groups. Mild edema was also noted on both hands and ankles. Skin exam was unremarkable.

Initial labs were remarkable for:

- Elevated CPK 1497 U/L
- Aldolase 32.6 U/L ESR 19 mm/h
- CRP 10.5 mg/L

An inflammatory myositis was considered highly likely and the patient was referred to rheumatology.

Additional rheumatologic labs ANA, Anti-Cyclic Citrullinated Peptide Antibody (Anti-CCP), Anti-Double stranded DNA, Rheumatoid Factor, SM Antibody and SM/RNP Antibody. All were unremarkable except for a mildly elevated Anti-CCP of 29.9 Units.

HIV, Hepatitis B and C, Lyme, Epstein Barr Virus, Legionella and C. Psittaci serologies were also unrevealing.

An EMG/Nerve conduction study was consistent with an inflammatory myositis. A right biceps brachii muscle biopsy revealed an inflammatory myopathy with a marked endomysial inflammatory infiltrate that primarily consisted of CD 3+ and CD 8+ T lymphocytes, consistent with polymyositis.

The patient was started on Prednisone 80 mg (1 mg/kg) tapering to 20 mg daily over the next 3 months. He was referred to physical therapy and returned to his primary care provider for cancer screening given the association of cancer with polymyositis.

Discussion

Polymyositis is an idiopathic form of inflammatory myopathy that is clinically characterized by symmetrical, proximal muscle weakness that evolves over weeks to months[1-3]. It may occur in any age but the peak incidence in adults is between 30 to 60 years.[4] It is twice as common in women.

Unlike dermatomyositis, which shares many clinical features with polymyositis, patients with polymyositis do not have any characteristic skin findings such as Heliotrope rash, Shawl sign and Gottron papules. Elevations of creatinine kinase (CK), lactate dehydrogenase (LDH), aldolase, aspartate aminotransferase (AST) and alanine aminotransferase (ALT) are commonly seen. Creatinine kinase is the most sensitive serum enzyme abnormality in polymyositis[3]. In severe cases, serum CK can be elevated 50 times the reference level.

Autoantibodies may be present in patients with polymyositis, anti-nuclear antibody assay (ANA) is seen in approximately one third of patients.

Anti-synthetase antibodies can also be present. The association of these antibodies (such as anti-Jo-1 antibodies) with interstitial lung disease, Raynaud phenomenon, fever, arthritis and mechanic's hands is known as anti-synthetase syndrome[5].

Severe muscle weakness and atrophy is often seen in patients with anti-signal recognition particle (SRP) antibodies.[6]

The electromyogram (EMG) is abnormal in 90% of cases showing motor unit action potentials (MUAPs) that are of short duration and of small amplitude or polyphasic with early recruitment patterns that are suggestive of an inflammatory process[1,7,8].

MRI of the proximal muscles, in particularly the thighs, can help assess the degree of muscle inflammation and identify a biopsy site[9].

Muscle biopsy is the gold standard in establishing the diagnosis and also to rule out other neuromuscular diseases[1-3].

The most important histologic finding is a CD8 predominant lymphocytic infiltrate localized within the muscle fascicles, which is pathognomonic for polymyositis[1,2].

Patients with polymyositis have increased risk of malignancy documented[10,11]. A population based study by Hill found the most common types of cancers linked to polymyositis were non-Hodgkin lymphoma, lung and bladder malignancies[10].

The primary treatment objective in patients with polymyositis is to restore muscle strength and to avoid further deterioration of muscle use[12]. The initial prescribed regimen includes high dose prednisone and physical therapy. Prednisone has been shown to improve muscle strength and normalize serum enzymes[10].

Methotrexate and azathioprine are commonly used as corticoid-sparing agents to prevent long-term complications of prednisone[12].

Other treatments such as hydroxycholoroquine, mycophenololate, tacrolimus, cyclosporine and intravenous gamma globulin have been shown to benefit patients who do not respond to methotrexate and azathioprine[12].

A biological therapy, Rituximab, is still being studied and has shown some promise in treating patients with refractory polymyositis[10,12,13].

Conclusion

The salient features of polymyositis are:

- Symmetrical muscle weakness, predominantly proximal
- Elevated CPK
- Abnormal EMG findings
- Pathologic muscle biopsy

The muscle biopsy is the most important diagnostic tool to distinguish polymyositis from other muscle pathologies.

Prompt diagnosis is paramount as early treatment can halt the deterioration of muscle use. Early cancer screening should also be initiated given the increased risk of malignancy in patients with polymyositis.

It is also important to consider other causes of muscle weakness such as thyroid dysfunction, neuromuscular disorders, medication toxicity and metabolic diseases as many of these are treatable and early intervention could reverse the disease process.

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