

Polymyositis Presenting with Distal and Asymmetrical Weakness

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SUMMARY: *A 52 year old female with polymyositis presenting with distal and asymmetrical weakness is presented. Similar reported cases are reviewed. These patients respond favorably to steroids. Increased awareness of this unusual distal polymyositis should avoid delay in diagnosis.*

RÉSUMÉ: *Nous discutons l'histoire d'une femme de 52 ans dont la polymyosite s'est présentée avec une faiblesse distale et asymétrique. Nous faisons une revue des cas semblables publiés. Ces patients ont répondu favorablement à l'emploi des stéroïdes. La connaissance accrue de cette polymyosite distale inhabituelle devrait éviter les retards dans le diagnostic.*

INTRODUCTION

Polymyositis presenting with predominantly distal and asymmetrical weakness is rare. Only five patients have been described in detail. We describe a patient who presented with distal weakness and wasting of the left leg. The literature on this unusual form of polymyositis is reviewed.

CASE HISTORY

Mrs. M.L. aged 52 presented in 1973 with a two year history of weakness of her left foot. She noted difficulty standing on the toes of the left foot. She also found that her left leg was getting thinner. The symptoms had been slowly progressive. There was no muscle pain, backache or dysphagia. Examination showed moderate wasting of the left calf muscles, associated with moderate weakness of plantar flexion of the left foot. There was minimal weakness of the hamstring muscles on the left. Other muscles were normal. The left ankle jerk was absent. There was no sensory loss. Lumbar spine x-rays and nerve conduction velocities of the peroneal nerves were normal. No specific diagnosis was made.

The patient noted increasing weakness of her legs. She had difficulty walking on rough ground and negotiating stairs in 1975-76. During her second hospital admission in 1978, the patient also complained of difficulty standing on the toes on the right side. There were no upper limb symptoms. Her general health remained good without any weight loss. Examination showed wasting of both calf muscles, the left more than the right. There was severe weakness of ankle plantar flexion, greater on the left. Eversion of both feet was weak. Knee flexion and extension on both sides were weak. The knee and ankle jerks were unobtainable. There was no sensory loss or muscle tenderness.

Investigations showed normal hemoglobin and white cell count; ESR in the first hour was 20 mm., platelet count was 157,000/cu. mm. Electrolytes were normal. Serum proteins, calcium, phosphate, and uric acid were normal. Plasma electrophoresis was normal. Serum creatine phosphokinase was 558 IU/L (normal 0-125) and lactic dehydrogenase 285 IU/L (normal 100-270). SGOT was 33 IU/L (normal 10-40). Chest x-ray and electrocardiogram were normal. Electromyography using a concentric needle showed occasional fibrillation potentials and positive waves in the left peroneus longus muscle. The left anterior tibial muscle showed some fibrillation potentials and on voluntary contraction, slight reduction in the number of motor units. EMG of the left quadriceps, left gastrocnemius and right anterior tibial muscles showed slight reduction in the number of motor units on maximal muscle contraction. Motor conduction velocities in both peroneal nerves, left median nerve and left ulnar nerve were in the normal range. No definite conclusion was made from these results.

Open biopsy of the right quadriceps muscle showed scattered collections of lymphocytes, macrophages and occasional granular leukocytes in the interstices of the muscle. Remnants of the disintegrating muscle fibers were invaded from the periphery by the inflammatory cells (Fig. 1). Electron microscopic examination of the muscles not involved in the inflammatory process showed normal architecture. Inclusion bodies were not seen.

The patient was treated initially with glucocorticoid for about four months. Serum CPK fell to 353 IU/L; LDH was 254 (normal 100-290). It had to be discontinued because of epigastric pain and water retention. Subsequently azathioprine was started at a dose of

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Figure 1 — Histology of muscle biopsy from the right quadriceps, showing disintegrating muscle fibers and interstitial lymphocytes and macrophages. (H&E mag. x 200).

150 mgms. per day. On this, the patient's symptoms ceased to progress. CPK in January 1980 was 350 IU/L. LDH and SGOT were in the normal range.

DISCUSSION

Polymyositis is an inflammatory disease of muscle, usually affecting patients aged 40 to 70. However, patients in their eighties and children with the disease have been described. It is twice as common in females. Most large series describe patients presenting with symmetrical proximal weakness of limbs (Barwick, et al, 1963; Eaton, 1954; Pearson, 1966; Walton, 1969). The lower limbs are usually affected before the upper limbs. Dysphagia is seen in about 54% of patients. Skin lesions and Raynaud's phenomenon are also seen. In 15% of patients internal malignancy is seen (Pearson, 1966). Underlying collagen vascular disorders are seen in 25% of all patients (Barwick, et al, 1963). Steroids produce complete improvement of weakness in about 50% of patients and partial arrest in 25%. Considerable difficulty remains in 10%. The overall mortality is about 15-30% (Pearson, 1966; Rose and Walton, 1966).

The distal muscles are involved in about one-third of all patients. How-

ever, this is usually mild and follows the proximal weakness. More severe distal muscle involvement is a late feature in patients with generalized disease (Barwick et al, 1963; Eaton, 1954; Pearson, 1966; Walton, 1969). Polymyositis with onset of asymmetric weakness in the distal muscles is rare. Rose and Walton (1966) mentioned that "weakness beginning in only one extremity was encountered in some cases" among their 89 patients. No further detail was given. Rowland (1977) described one patient with predominantly distal polymyositis but gave no details. Detailed reports are available in only five patients (Eaton, 1954; Hollinrake, 1969; Bates et al, 1973; Stark, 1978; Van Kasteran, 1979). These are summarized in Figure 2. All of these patients were females aged between 45 and 65. In 3 patients, hand muscles were involved first. In 2 of these the onset was unilateral. In 3 patients, distal leg muscles were affected first. In 2 of these the weakness was asymmetrical. These patients did not have muscle pain. Dysphagia was present in 3 patients. The disease typically ran a slowly progressive course. Eventually, the more proximal muscles were involved.

Although some deep tendon reflexes were absent in our patient and in the patient described by Stark (1978), high

CPK, normal nerve conduction studies and lack of neurogenic changes in the muscle on histology would rule out any primary neurogenic disorder. Subsequent clinical course confirmed that the disorder was not neurogenic.

Recently a group of patients with inflammatory myositis with distal weakness was described by Carpenter, et al, (1978). This was named "Inclusion Body Myositis". These patients were men; the distal muscles were predominantly involved. The course of the disease was slow. The muscle histology showed massive collections of cytomembranous whorls and abnormal cytoplasmic nuclear filaments. There was no response to steroids.

We believe that the patients listed in Figure 2 did not have inclusion body myositis for various reasons. They were all female; inclusions were not seen in the muscle cells although electron microscopy was not done in all the patients. Moreover, they responded favorably to steroids.

There is a small distinct group of patients with polymyositis, where distal, and frequently asymmetrical muscle weakness is the presenting feature. The correct diagnosis is usually not made at an early stage in these patients. Their disease runs a slow course over several years yet responds well to corticosteroids. The factors determining this distribution of muscle involvement are presently unknown.

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FIGURE II

Details of Patients With Polymyositis Presenting With Predominant Distal Weakness

Author	Pt.'s age in yrs.	Sex	Predominant muscles involved	Duration of weakness when diagnosed in years	Dysphagia	Muscle Pain	CPK	Biopsy	Response to steroids
Eaton 1954	48	F	Below the knee. Upper facial muscles	3	yes	not known	not known	positive	not known
Hallinrake 1969	65	F	Initially left hand; subsequently lower limbs and right hand	6	yes	no	elevated	Left tib—ant and L. triceps positive	improved
Bates et al	62	F	Hand muscles; proximal lower limb muscles; facial muscles	2	yes	no	elevated	R. deltoid x twice positive	improved
Stark 1978	45	F	Left hand and arm — subsequently proximal leg muscles left worse than right	6	no	no	elevated	positive	arrest of the process
Van Kasteren 1979	55	F	Asymmetrical distal leg weakness; left forearm and hand. Subsequently R. forearm and hand	6	no	yes at onset	elevated	positive	improved
Our patient	52	F	Left calf muscles; subsequently proximal leg muscles	7	no	no	elevated	positive	arrest of the process

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