Polyarteritis Nodosa Presenting as Polymyositis

Spencer G. Plumley, Ronald Rubio, Said Alasfar, and Hugo E. Jasin

Background: **Skeletal muscle involvement has been well documented in patients with polyarteritis nodosa (PAN), and symptoms referable to skeletal muscle are not uncommon. However, polymyositis as a mode of presentation of PAN is uncommon. This unusual presentation of PAN has been reported only once previously in the English literature.**

Objective: **This study describes a patient who had diffuse weakness, myalgias, and markedly elevated serum creatinine phosphokinase, mimicking polymyositis. The literature dealing with the clinical aspects of muscle involvement in PAN is reviewed.**

Results: **A 24-year-old man was admitted to the hospital with a 1-month history of fever, myalgia, and muscle weakness. Necrotizing vasculitis was shown on subsequent muscle biopsy, consistent with PAN. Literature review indicated that muscle involvement is common in PAN, as has been shown by the frequency of muscular symptoms and by histologic evidence obtained from both clinical and autopsy studies. Nineteen percent of patients with PAN had documented myopathy, and autopsy series have shown skeletal muscle involvement in 30% to 48% of cases. However, polymyositis as a mode of presentation of PAN is rare. We found only 1 other patient with PAN who had elevated creatinine phosphokinase and diffuse myopathy suggestive of polymyositis.**

Conclusions: **PAN should be suspected in cases of focal or diffuse myopathy, especially in the context of a systemic disease. Biopsy of symptomatic muscles or EMG-directed biopsies can be helpful in establishing a diagnosis of PAN to allow the physician to provide early treatment.**

INDEX WORDS: Polyarteritis nodosa; polymyositis; myopathy.

POLYARTERITIS NODOSA (PAN) is a systemic vasculitis that, in its diffuse form, can affect multiple organs and cause a high degree of morbidity and mortality. There are case reports of a limited form in which only 1 organ is involved, with few or no systemic features. Because of the protean manifestations of PAN, it can mimic more common diseases and cause confusion or delays in diagnosis. This review describes the second case report of PAN to date that initially involved clinical and laboratory features consistent with polymyositis (1).

CASE REPORT

A 24-year-old healthy white man began having myalgias, fever, sore throat, and muscular weakness 1 month before admission to the hospital. He

From the Division of Rheumatology and Clinical Immunology, Department of Internal Medicine, University of Arkansas for Medical Sciences and Veterans Administration Hospital, Little Rock, AR.

Spencer G. Plumley, MD, Ronald Rubio, Said Alasfar: *Rheumatology Fellows*; Hugo E. Jasin, MD: *Director; Division of Rheumatology and Clinical Immunology, University of Arkansas for Medical Sciences and Veterans Administration Hospital, Little Rock, AR.*

Address reprint requests to Hugo E. Jasin, MD, Director, Division of Rheumatology and Clinical Immunology, University of Arkansas for Medical Sciences, Mail Slot 509, 4301 W. Markham, Little Rock, AR 72205. E-mail: jasinhugoe@uams.edu

0049-0172/02/3106-0004$35.00/0
doi:10.1053/sarh.2002.32549

received a course of oral antibiotics but showed no improvement. Thereafter, his symptoms progressed to include night sweats, weight loss of 12 pounds, and inability to rise from a chair or walk.

The patient's initial temperature was 100.1°F, heart rate 120 beats/min, blood pressure 118/68 mm Hg, and respiratory rate 26/min. He had an erythematous posterior pharynx, and a faint erythematous macular rash covered his upper torso. All peripheral pulses were palpable and cardiac examination was normal, except for tachycardia. Chest and abdominal examinations were normal. There were no focal neurologic deficits, and mental status exam was normal. The patient had diffuse muscle weakness and was able to move only against minimal resistance.

Initial laboratory test results were as follows: white blood cell count 38 K/uL, hemoglobin 19.2g/dL, hematocrit 59%, platelets 249,000/uL (normal, 150,000 to 500,000/uL), creatine phosphokinase (CPK) 41,677 IU/l (normal, 30 to 235 IU/L), total bilirubin 0.6 mg/dL (normal, 0.3 to 1.1 mg/dL), serum glutamic oxalacetic transaminase (SGOT) 1,092 IU/L (normal, 10 to 40 IU/L), serum glutamic oxalacetic transaminase (SGPT) 372 IU/L (normal, 20-80 IU/L), gamma-GTP 90 IU/L (normal, 5 to 70 IU/L), alkaline phosphatase 62 IU/L (normal, 4 to 120 IU/L), creatinine 1.2 mg/dL (normal, 0.5 to 1.1 mg/dL). Urinalysis showed a red blood cell count of 6 to 10/HPF, 26 to 50 white blood cell count/HPF, trace protein, and no myoglobin or red blood cell casts. Results from blood, urine, and cerebrospinal fluid cultures, studies for hepatitis serology, and human immunodeficiency virus were all negative. Findings from initial chest radiography were normal. The patient was hydrated and started on empiric broad spectrum antibiotics. He developed pulmonary edema and required mechanical ventilation for respiratory failure. His renal function soon deteriorated with anuria and a peak serum creatinine of 4.1 mg/dL. He required temporary hemodialysis for removal of both volume and solutes. The acute renal failure was believed to be multifactorial and consistent with acute tubular necrosis.

The patient's serum CPK had risen to 82,194 IU/L, and he developed purpuric lesions on both ankles. The results of biopsy of these lesions were consistent with leukocytoclastic vasculitis. Results from serologic studies, including antinuclear antibodies, anti-neutrophil cytoplasmic antibodies (ANCA), anti–double-stranded DNA antibodies, anti-Smith antibodies, anti-ribonucleoprotein antibodies, cryoglobulins, and rheumatoid factor, were all negative. Erythrocyte sedimentation rate (ESR) (Westergren) was 55 mm/h and C-reactive protein was 2.85 mg/dL.

Diagnostic biopsy specimens were taken from both the quadriceps and deltoid muscle groups. Focal muscle fiber necrosis, fibrinoid necrosis of medium arteries, and infiltration of the vessel walls by neutrophils was seen histologically. In conjunction with the overall clinical picture, a diagnosis of systemic necrotizing vasculitis was made. The pattern was believed to be most consistent with PAN.

The patient was given a bolus of 1 g cyclophosphamide intravenously and pulse methylprednisolone at a dose of 1 g/d for 3 days. Thereafter, he received a maintenance dose of 1 mg/kg/d methylprednisolone. Antibiotics were discontinued, and 2 weeks later, he was started on a maintenance dose of cyclophosphamide intravenously at 200 mg/d. In the third week of hospitalization, he developed gastrointestinal bleeding that persisted intermittently during the next several weeks. Endoscopy demonstrated numerous ischemic-appearing, actively bleeding ulcers along the lesser curvature of the stomach and the first portion of the duodenum. Mesenteric angiography showed focal stenotic lesions, areas of punctate hemorrhage, and an infarct within the right kidney, suggestive of vasculitis.

Renal and respiratory function improved slowly. Hemodialysis and mechanical ventilation were discontinued. He was discharged to a rehabilitation facility 10 weeks after admission on 150 mg oral cyclophosphamide and 60 mg prednisone daily. During the ensuing months, his muscle strength slowly returned to normal.

Currently, remission of this patient's vasculitis is being maintained with 15 mg/wk methotrexate and 20 mg/d prednisone. He has normal hepatic and muscle enzymes, normal renal function, and no clinical evidence of PAN.

DISCUSSION

PAN is the prototypical necrotizing vasculitis, which involves small and medium muscular arteries. Kussmaul and Maier (2) first described the classic form of PAN in 1866. The pathologic hallmark of PAN is segmental necrotizing vasculitis, which leads to vessel wall damage, microan-