ISSN: 2694-1724
DOI: 10.33552/ARAR.2024.03.000556
Archives of Rheumatology & Arthritis Research
Iris Publishers

Case Report

Atypical Systemic Lupus Erythematosus Presenting as Polymyalgia Rheumatica in an Elderly Native American Male

Prashant Kaushik[1]*, Stephen Bastible[2], Megan N Tramel[3], Caleb S Alexander[3]

[1]*Chief of Rheumatology, Northeastern Health System, Tahlequah, OK; Associate Program Director, Internal Medicine Residency Program, TMG/OMECO at NHS, Tahlequah, OK; Clinical Professor of Medicine, OSU CHS Cherokee Nation, Tahlequah, Oklahoma, USA*

[2]*TMG/OMECO Internal Medicine Residency Program, Department of Internal Medicine, NHS, Tahlequah, Oklahoma, USA*

[3]*Oklahoma State University College of Osteopathic Medicine at the Cherokee Nation, Tahlequah, Oklahoma, USA*

***Corresponding author:** Prashant Kaushik Chief of Rheumatology, Northeastern Health System, Tahlequah, OK; Associate Program Director, Internal Medicine Residency Program, TMG/OMECO at NHS, Tahlequah, OK; Clinical Professor of Medicine, OSU CHS Cherokee Nation, Tahlequah, Oklahoma, USA

Received Date: March 11, 2024

Published Date: March 18, 2024

Abstract

A 78-year-old Native American male presented to the clinic for evaluation and treatment of polymyalgia rheumatica (PMR) that initially began with extreme shoulder and hip pain, later involving the neck and ankles. At the time of presentation, he had been treated with oral prednisone for a year; 5 mg in the morning and 2.5 mg in the evening. Two attempted trials without prednisone during that year resulted in a rebound of articular symptoms. Upon completing laboratory studies and evaluating this patient in the Rheumatology clinic, his presentation actually favored a diagnosis of late-onset, atypical systemic lupus erythematosus (SLE) due to his peripheral joint involvement, positive ANA (titer of 1:160 with a speckled pattern), significantly positive double stranded (ds) DNA, beta-2 glycoprotein IgG, and cardiolipin IgG antibodies. To our knowledge, this is the first of such cases to be documented.

Keywords: SLE; PMR; Native American; Elderly male

Introduction

PMR is an inflammatory autoimmune disease which predominantly affects Caucasian patients, females more than males, over the age of 70, with onset typically no younger than 50 years of age [1]. The clinical presentation is most characteristically bilateral shoulder pain, stiffness, and upper arm tenderness that may coincide with bilateral hip pain and stiffness and posterior neck muscle pain and stiffness [1]. The shoulder or hip symptoms typically have been ongoing for several weeks with the articular stiffness lasting longer than 45 minutes in the morning, with sparing of peripheral joints, and a concurrent elevation in acute phase reactants-erythrocyte sedimentation rate (ESR) or C-reactive protein (CRP) [1].First line treatment is a moderate dose of prednisone, and a dramatic, positive response in these patients can help substantiate the diagnosis [1]. PMR can lead to Giant Cell Arteritis (GCA) in 16-21% of PMR cases, thus patients must watch for sudden severe headaches, sudden severe vision changes including painless mono or bi-ocular vision loss (amaurosis fugax), jaw claudication, and scalp tenderness [2].

SLE diagnosed after age 60 often initially presents insidiously as polymyalgia rheumatica or rheumatoid disease [3]. Presentation of SLE over the age of 50 is typically considered late-onset [4]. A positive ANA of > 1:80 is required as an entry

criterion for SLE diagnosis, followed by at least 10 points from the following categories: constitutional, hematologic, neurologic, mucocutaneous, serosal, musculoskeletal, renal, antiphospholipid antibodies, complement proteins, or SLE-specific antibodies [5]. The other documented cases of late-onset SLE presenting as PMR have occurred in patients with demographics unlike this case and presented with different symptoms [6-8]. This report documents a 78-year-old Native American male presenting initially with PMR, when he in fact had late-onset SLE.

Case Report

A 78-year-old Native American male presents for evaluation and treatment of PMR. His symptoms began with joint pain in his shoulders, ankles, and hips that was severe enough to cause difficulty getting out of bed. He was prescribed prednisone (5 mg every morning and 2.5 mg every evening) for the past year, which rapidly resolved his joint pain. There were two occasions when he tried to discontinue the prednisone - once for lab work and once to see how long he could manage without the medication. His joint pains aggressively returned about one week after the first trial of discontinuation and about three weeks after the second trial. Regarding the second attempt, his pain began in his neck, causing stiffness and difficulty turning his head. Additionally, his left hand started to swell. His family history was significant for a mother with rheumatoid arthritis and osteoarthritis.

His lab work showed an elevated creatinine of 1.57 (normal 0.6-1.30), ANA positive (1:160, speckled), Beta-2 Glycoprotein IgG elevated 50.9 (normal <20), Cardiolipin IgG elevated 53.9 (normal <20), double-stranded DNA antibody (dsDNA) positive reading of 75 (normal 0-26), GFR decreased to 46 (normal > 60), anti-histone antibodies elevated to 1.1 (normal 0.0-0.9). Lupus anticoagulant was not detected. C3, C4, CRP, ESR, white blood cell counts, platelets, and hemoglobin were all within normal limits. There were no detected antibodies to rheumatoid factor, CCP, Jo-1, RNP, SSA/SSB, Smith, and Scl 70 antibodies. HLA-B27 and 14.3.3 Eta markers were negative. Further, TB QuantiFERON Gold was negative, hepatitis C virus antibodies were negative, and hepatitis B serology showed that he was non-immunized.

Through shared decision-making, the patient was started on 200 mg daily hydroxychloroquine as a disease modifying anti-rheumatic drug (DMARD) while continuing his current dose of prednisone at 5mg every morning and 2.5 mg every evening for now. He was educated on red flag symptoms concerning GCA such as sudden severe headache, jaw claudication, and sudden severe changes in vision such as blindness in one eye. He was asked to repeat labs in 3 months and return to the clinic in 4 months for follow-up. At that time, we will re-evaluate, and begin a slower tapering schedule of prednisone.

Discussion

The clinical presentation and laboratory studies on this patient point to late-onset SLE, rather than the initial diagnosis of PMR. He is a 78-year-old Native American male, which varies from the classical presentation of a young Black female under the age of 50. Although this patient's initial presentation pointed toward a diagnosis of PMR due to shoulder and hip pain at the beginning of disease course with later development of neck stiffness, the development of ankle and hand involvement put him over the required point total using SLE diagnostic criteria [5]. Further, he met the entry criterion of an ANA > 1:80 and presence of autoantibodies in each of the following categories: antiphospholipid antibodies (Beta-2 Glycoprotein antibodies and Cardiolipin antibodies-2 points), SLE-specific antibodies (Anti-dsDNA-6 points), and the aforementioned musculoskeletal symptoms (joint involvement-6 points) [5]. Overall, his SLE diagnostic score (14 points) exceeded the required total (10 points). The presence of these lab markers, in addition to his clinical presentation, met diagnostic criteria for SLE, necessitating initiation of treatment with hydroxychloroquine as a DMARD and a steroid sparing agent.

Although this presentation was atypical for SLE, and initially appeared as PMR, it shows the importance of keeping SLE on the differential list regardless of patient demographics. The prevalence of SLE is much greater among females than males with an overall prevalence of about 128.7 cases per 100,000 females and only 14.6 cases per 100,000 males [9,10]. Breaking down SLE by race/ethnicity, prevalence is much greater among the American Indian/Alaska Native (AI/AN) population compared to others with about 270.6 cases per 100,000 females and 53.8 cases per 100,000 males. Although the prevalence is significant for the AI/AN population, the total number of cases remains low, thus increasing the importance to share atypical presentations, such as the above case.

Acknowledgement

None.

Conflict of Interest

No conflict of interest.

References

1. Michet CJ, Matteson EL (2008) Polymyalgia rheumatica. BMJ 336(7647): 765-769.
2. Ramon A, Greigert H, Ornetti P, Maillefert JF, Bonnotte B, et al. (2022) Predictive factors of giant cell arteritis in polymyalgia rheumatica patients. J Clin Med 11(24): 7412.
3. Foad BSI, Sheon RP, Kirsner AB (1972) Systemic Lupus Erythematosus in the Elderly. Arch Intern Med 130(5): 743-746.
4. Altier J, Oates J, Ward C (2022) Polymyalgia Rheumatica or Late Onset Lupus? A Case Report. J Investig Med High Impact Case Rep 10: 232470962210894.
5. Aringer M, Costenbader K, Daikh D, Ralph Brinks, Marta Mosca, et al. (2019) European League Against Rheumatism/American College of Rheumatology Classification Criteria for Systemic Lupus Erythematosus. Arthritis Rheumatol 71(9): 1400-1412.
6. Maragou M, Siotsiou F, Sfondouris H, Nicolia Z, Vayopoulos G, et al. (1989) Late-onset systemic lupus erythematosus presenting as polymyalgia rheumatica. Clin Rheumatol 8(1): 91-97.
7. Hutton CW, Maddison PJ (1986) Systemic lupus erythematosus presenting as polymyalgia rheumatica in the elderly. Ann Rheum Dis 45(8): 641-644.

8. Altier J, Oates J, Ward C (2022) Polymyalgia Rheumatica or Late Onset Lupus? A Case Report. J Investig Med High Impact Case Rep 10: 23247096221089493.

9. Rees F, Doherty M, Grainge MJ, Lanyon P, Zhang W (2017) The worldwide incidence and prevalence of systemic lupus erythematosus: a systematic review of epidemiological studies. Rheumatology 56(11): 1945-1961.

10. Izmirly PM, Parton H, Wang L, W Joseph McCune, S Sam Lim, et al. (2021) Prevalence of Systemic Lupus Erythematosus in the United States: Estimates from a Meta-Analysis of the Centers for Disease Control and Prevention National Lupus Registries. Arthritis Rheumatol 73(6): 991-996.