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Polymyalgia Rheumatica or Late Onset Lupus? A Case Report

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Abstract

Systemic lupus erythematosus (SLE) is a multisystem autoimmune disease with a peak age of presentation between the 15 and 40 years with a wide variety of disease manifestations. Although there is no formal definition, late onset SLE is generally defined in the literature as onset after the age of 50. It is estimated that 2% to 20% of patients with SLE overall fall into this category. It is important for the clinician to recognize this less-common entity because arthralgia, myalgia, fatigue, and sicca symptoms in the elderly can so easily be attributed as symptoms of normal aging or other common degenerative processes rather than a systemic disease similar to SLE or Sjogren's syndrome. The following report outlines a case of late onset SLE which initially was suspected to be polymyalgia rheumatica (PMR).

Keywords

rheumatology, immunology

Introduction

Systemic lupus erythematosus (SLE) is a multisystem autoimmune disease with a peak age of presentation between the 15 and 40 years with a wide variety of disease manifestations. Although there is no formal definition, late-onset SLE is generally defined in the literature as an onset after the age of 50.[1] It is estimated that 2% to 20% of patients with SLE fall into this category.[2] This patient population tends to have a more insidious presentation with lower overall disease activity compared to the general SLE population.[3] Patients are less likely to have malar rash, photosensitivity, alopecia, Raynaud phenomenon, neuropsychiatric symptoms, renal involvement and more likely to have arthritis, sicca symptoms, lung involvement including pleuritis and interstitial lung disease.[2-5] It is important for the clinician to recognize this less-common entity because arthralgia, myalgia, fatigue, and sicca symptoms in the elderly can so easily be attributed as symptoms of normal aging or other common degenerative processes rather than a systemic disease similar to SLE or Sjogren's syndrome. The following report outlines a case of late-onset SLE which initially was suspected to be polymyalgia rheumatica (PMR).

Case History

Patient is a 64-year-old female with no significant past medical history who presented initially to an outside clinic with erythematous raised papules on her upper extremities that resolved with topical steroids and was thought to be due to insect bites. Two weeks later, the patient developed fatigue, malaise, and nasal congestion thought to be due to an upper respiratory tract infection (URI) that was treated conservatively. A month after her presumed URI, the patient presented with recurrent raised papules on her extremities followed by a polyarticular arthritis involving her metacarpophalangeal (MCP) joints and shoulders bilaterally. She was noted to have recently explored a wooded area in Virginia and was treated empirically for Lyme disease. Of note, her follow-up serologies for Lyme disease were negative, and she was

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noted to have a mildly positive antinuclear antibody (ANA) of 1:160 that was thought to be due to her family history of rheumatoid arthritis.

One year after her initial presentation, she presented to our clinic with recurrent URI symptoms, fevers, diffuse erythematous rash on her arms, back, and chest in a photosensitive distribution, and arthritis in her bilateral MCP joints and shoulders. She was treated with prednisone 20 mg with immediate resolution of her symptoms. Work up revealed negative rheumatoid factor (RF) and cyclic citrullinated peptide as well as normal erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP). Of note, these levels were determined after initiation of steroids, which likely lowered the levels from her presteroid state. Given her clinical presentation and profound response to steroids, the diagnosis of PMR was proposed as most likely.

She continued to have intermittent photosensitive rashes, fevers, and arthritis with each prednisone taper. Several months after starting treatment, while she was still on prednisone, she developed a left-sided retro-orbital headache in the setting of fever and elevated inflammatory markers. Giant cell arteritis (GCA) was suspected, so a left temporal artery biopsy was performed and did not show findings suggestive of GCA. A full-body positron emission tomography-computed tomography (PET-CT) was then performed to evaluate for large vessel vasculitis and was unrevealing. She remained on prednisone during the temporal artery biopsy and PET-CT, so she was weaned from prednisone with the recurrence of the symptoms. Another full-body PET-CT was performed and revealed diffuse, metabolically active lymphadenopathy concerning for lymphoma. She was referred to oncology, and the lymph node biopsy was not consistent with malignancy. A repeat ANA resulted at 1:640 (normal negative) with a homogenous pattern. An extractable nuclear antigen (ENA) panel resulted with positive anti-double-stranded DNA (anti-dsDNA) 77.0 IU/mL (normal $<$ 9.9 IU/mL), anti-Smith 2.7 AI (normal $<$ 1 AI), anti-SmRNP 2.5 AI (normal $<$ 1 AI) antibodies. C3 and C4 were found to be 33.4 mg/dL (normal 82.0-193.0 mg/dL) and C4 9.2 mg/dL (normal 15.0-57.0 mg/dL), respectively. Antiphospholipid antibodies, including anticardiolipin IgG and beta-2 glycoprotein IgG, were negative. A complete blood count showed a leukopenia of 3.1 K/CUMM, a normal hemoglobin of 12.6 gms/dL and a normal platelet count of 167 K/CUMM. Her urine protein to creatinine ratio was normal. Given her clinical symptoms and laboratory data, she was diagnosed with SLE approximately 1 year after presentation to our rheumatology clinic.

Discussion

Our patient presented with several nonspecific symptoms, the most predominant being pain and stiffness in her shoulder girdle that was responsive to steroids, which led to an initial diagnosis of PMR as depicted in the timeline shown in Figure 1. The diagnosis of PMR is typically a step-wise process that involves the assessing a patient's risk for the disease and their response to steroids as noted in Figure 2.[6] PMR has a strong association with GCA, which highlights the importance in screening for the development of a new headache, jaw claudication, vision loss, diplopia, and scalp tenderness over the temple which was a concern in our patient when she developed a new onset headache.[7] Despite the strong clinical evidence pointing toward PMR, our patient was actually showing early signs of SLE. SLE is a pleomorphic autoimmune disease with clinical presentations that can vary with age of presentation. After discovering that our patient had diffuse lymphadenopathy, autoimmune serologies were repeated and were overwhelmingly positive for SLE, thus securing the diagnosis. The Systemic Lupus International Collaborating Clinics diagnostic criteria, outlined in Table 1, states that the diagnosis of SLE requires 4/17 of the criteria, including at least 1 clinical and 1 immunologic criterion.[8] Late-onset SLE is a well-described syndrome that varies from SLE with an onset in younger adults. This population tends to have a lower overall disease activity and a more benign clinical course.[9] The relationship between low disease activity and the postmenopausal state in patients with late-onset SLE has implicated estrogen in the pathophysiology of SLE.[10] Hutton et al reported a case series of 3 patients in 1986 who presented with symptoms initially thought to be due to PMR or a disease along the myositis spectrum.[11] Interestingly, one patient described in this case series was a 76-year-old male who initially presented with proximal muscle pain and weight loss. An initial work up revealed a negative ANA, and patient was trialed on steroids with no improvement. Subsequent evaluation revealed a positive ANA and CK, and he was ultimately diagnosed with an SLE myositis overlap. The delay in diagnosis was similar to our patient, as her initial ANA was negative and later positive. This is a peculiar finding, as several studies have suggested that autoantibody formation may predate the development of clinical symptoms of SLE.[12,13] In addition, our patient presented with a history of several nonspecific findings, such as rash, malaise, and fevers, which were thought to be easily attributed to frequent URI's, something common in the elderly, rather than a multisystem rheumatological disease. In addition, our patient strays somewhat from the classic late-onset SLE presentation, as she presented with a rash, which is not a common symptom in this population.[14] Overall, this report highlights the importance of recognizing the entity of late-onset SLE and the multitude of ways it can present.

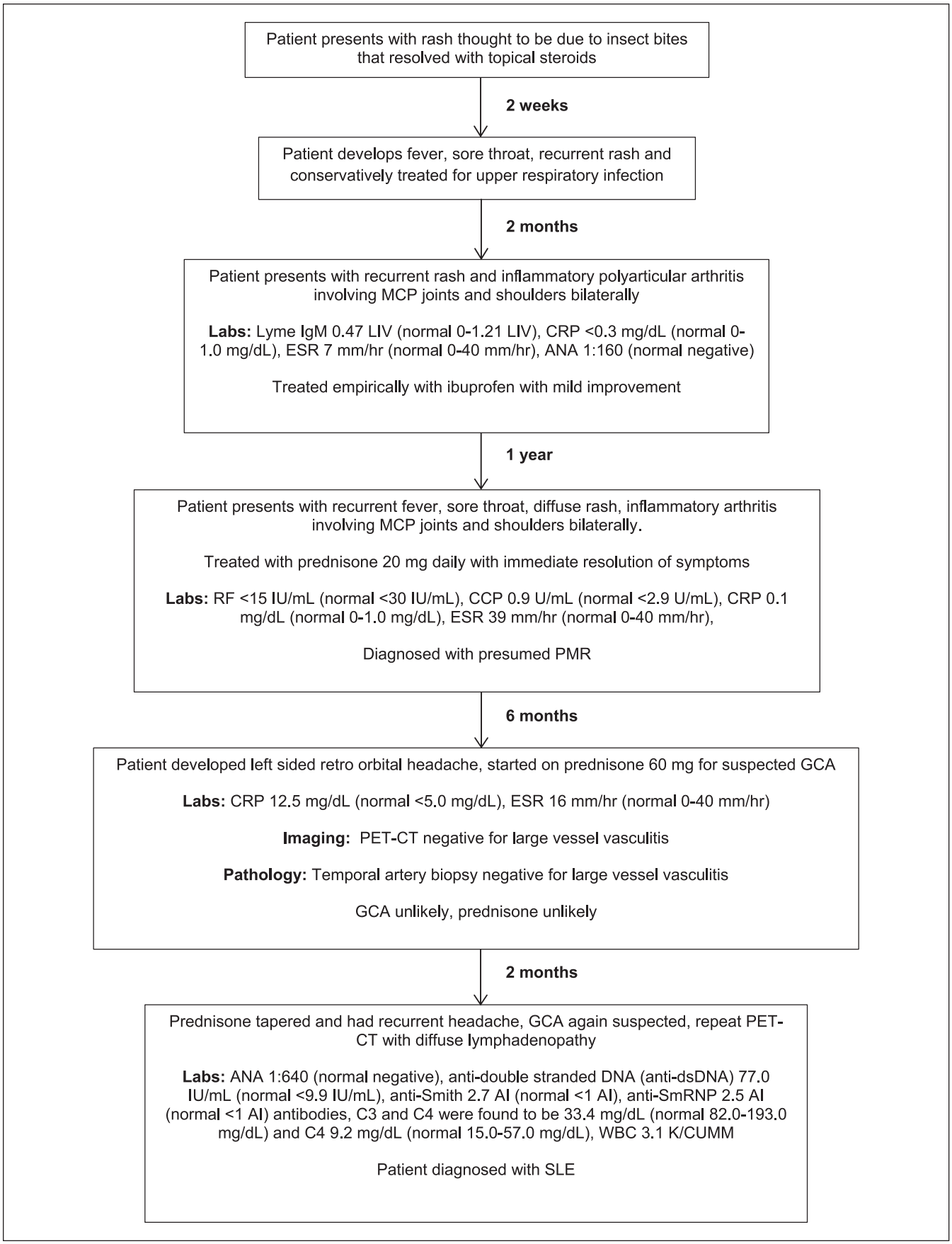

Figure 1. Timeline of events leading to patient's diagnosis.

Table 1.

SLICC criteria	
Four of 17 criteria, including at least 1 clinical criterion and 1 immunologic criterion; OR biopsy-proven lupus nephritis	
Criterion	Definition
Clinical criteria	
Acute cutaneous lupus	Lupus malar rash (not malar discoid); bullous lupus; toxic epidermal necrolysis variant of SLE; maculopapular lupus rash; photosensitive lupus rash (in the absence of dermatomyositis); or subacute cutaneous lupus
Long-term cutaneous lupus	Classic discoid rash; localized (above the neck); generalized (above and below the neck); hypertrophic (verrucous) lupus; lupus panniculitis (profundus); mucosal lupus; lupus erythematous tumidus; chilblains lupus; OR discoid lupus/lichen planus overlap
Nonscarring alopecia	Diffuse thinning or hair fragility with visible broken hairs in the absence of other causes
Oral or nasal ulcers	Palate, buccal, tongue, or nasal ulcers in the absence of other causes
Joint disease	Synovitis involving 2 or more joints and at least 30 minutes of morning stiffness OR tenderness in 2 or more joints and at least 30 minutes of morning stiffness
Serositis	Typical pleurisy for more than 1 day; pleural effusions; pleural rub OR typical pericardial pain for more than 1 day; pericardial effusion, pericardial rub; pericarditis by electrocardiography in absence of other causes
Renal	Urne protein-to-creatinine ratio/24-hour urine protein representing 500 mg protein/24 hours OR red blood cell casts
Neurologic	Seizures; psychosis; mononeuritis multiplex in the absence of other causes; myelitis; peripheral/cranial neuropathy in the absence of other causes; OR acute confusional state in the absence of other causes
Hemolytic anemia	Hemolytic anemia
Leukopenia or lymphopenia	Leukopenia ($<$4,000/mm^3 at least once) OR lymphopenia ($<$1,000 at least once) both in absence of other causes
Thrombocytopenia	Thrombocytopenia ($<$100,000/mm^3) at least once in the absence of other causes
Immunologic criteria	
ANA	ANA level above laboratory reference range
Anti-dsDNA	Anti-dsDNA antibody level above laboratory reference range (or $>$2-fold the referenc range if tested by ELISA)
Anti-Sm	Presence of antibody to Sm nuclear antigen
Antiphospholipid	Antiphospholipid antibody positivity as determined by any of the following: positive test result for lupus anticoagulant; false-positive test result for rapid plasma regain; medium- or high-titer anticardiolipin antibody level (IgA, IgG, or IgM); or positive test result for anti-beta-glycoprotein 1 (IgA, IgG, or IgM)
Low complement	Low C3; low C4; OR low CH50
Direct Coombs test	Direct Coombs test in the absence of hemolytic anemia

Abbreviation: SLICC, Systemic Lupus International Collaborating Clinics; SLE, systemic lupus erythematosus; ELISA, enzyme-limked immunosorbant assay.

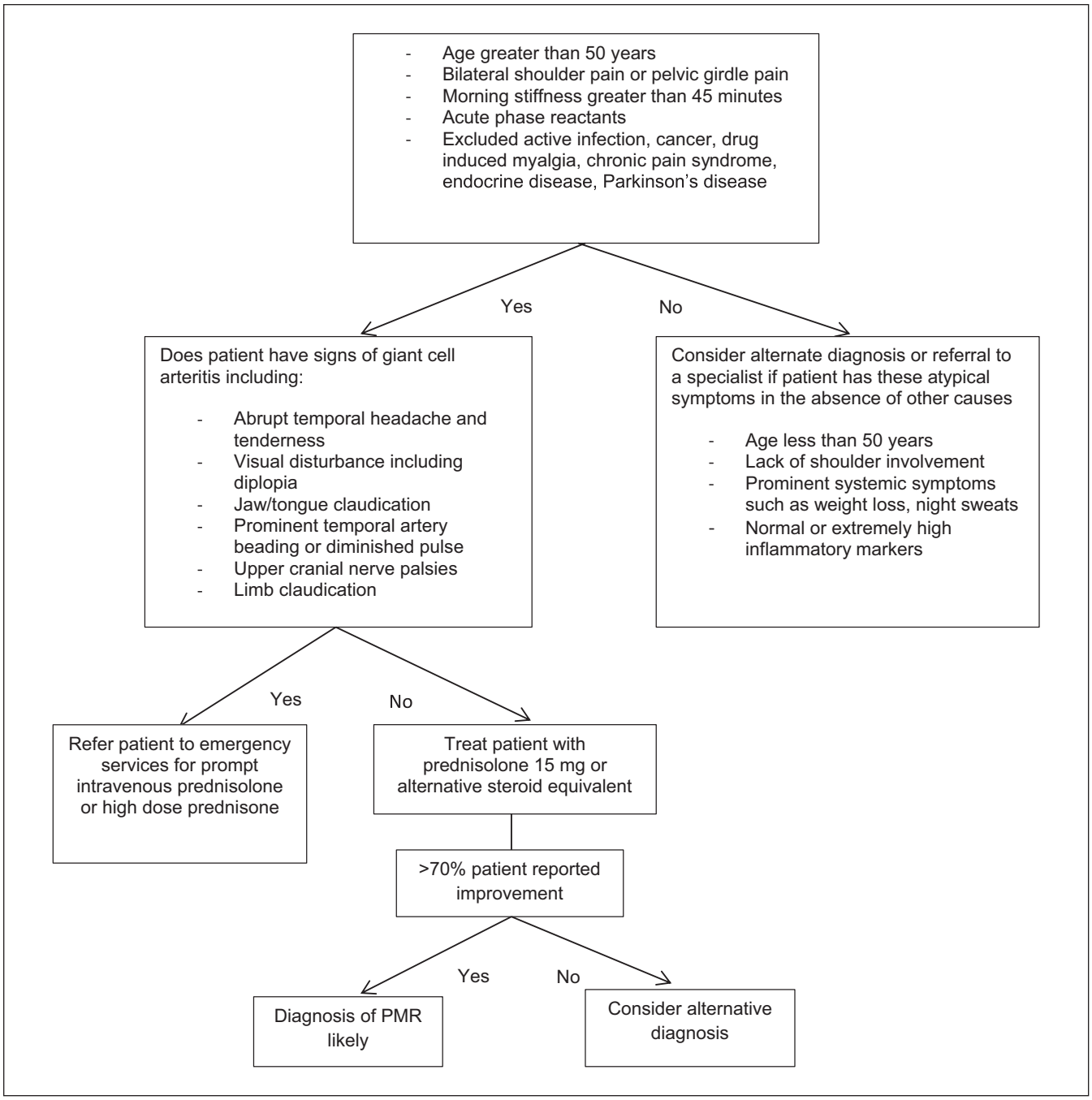

Figure 2. Diagnostic algorithm for polymyalgia rheumatica.

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Ethics Approval

Our institution does not require ethical approval for reporting individual cases or case series.

Informed Consent

Verbal informed consent was obtained from the patient(s) for their anonymized information to be published in this article.

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