Polymyalgia Rheumatica as the First Presentation of Metastatic Lymphoma

Thatchai Kampitak

Abstract

A 48-year-old HIV-positive woman presented with progressive pain and stiffness of both shoulders and hips. She was given the diagnosis of polymyalgia rheumatica (PMR) due to high erythrocyte sedimentation rate. However, a 1-week course of prednisolone failed to improve her symptoms. She later discovered a breast lump of which histopathological tissue was consistent with a diffuse large B-cell lymphoma. Whole body bone scan revealed multiple bony metastases. The presence of atypical features of PMR and lack of dramatic response to steroids should prompt physicians to raise the probability of differential diagnoses other than PMR, and in particular, malignancy.

Key words: lymphoma, polymyalgia rheumatica, malignancy, metastasis

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Introduction

Polymyalgia rheumatica (PMR) is the most common inflammatory rheumatic disease that affects elderly patients (1). The main characteristics of PMR are limb girdle pain with stiffness and elevated inflammatory markers. This condition has become increasingly recognized since there is a progressive increase in the incidence rates of PMR, particularly in Western countries (2, 3). However, diagnosis and treatment of PMR remain problematic due to heterogeneity in clinical presentation and treatment response (1). To prevent overdiagnosis, physicians should be aware that polymyalgic symptoms may be the first manifestation of various disorders that more frequently occur in the elderly, particularly malignancy. Here, a case who presented with PMR-like symptoms secondary from bony metastasis due to malignant lymphoma is described.

Case Report

A 48-year-old woman patient presented with a 2-month history of progressive pain and stiffness involving her bilateral shoulders and pelvic girdles. She had minimal morning stiffness and her symptoms were not aggravated by active movement. She also complained of low-grade fever, fatigue, anorexia and weight loss without headache, visual loss and jaw claudication. Her past medical history included known HIV seropositive status for 2 years with CD4 T-lymphocyte counts of 235 cells/μL and HIV-RNA level of less than 50 copies/mL. Her current medications consisted of didanosine, lamivudine and efavirenz.

Physical examination was unremarkable except for mild restriction of shoulder and hip movement due to pain. All routine laboratory tests were in the normal range except for the erythrocyte sedimentation rate (ESR) of 93 mm/hr. Based on these findings, the initial diagnosis of PMR was made. However, various analgesic medications including a 1-week course of prednisolone (30 mg daily) failed to completely relieve her symptoms. She stated that the pains had developed into diffuse continuous aching and interfered with her night sleep.

One month later, she discovered a lump in the left breast which had gradually increased in size in that time. It was resected and the histopathology result revealed a diffuse large B-cell lymphoma. Computed tomography of the chest and abdomen revealed diffuse pulmonary nodules and multiple enlarged intramediastinal and intraabdominal lymph nodes. Bone scintigraphy showed multiple foci of abnormal increased radiotracer activity at left high frontal bone, proxi-
mal left humerus, distal right humerus, T10-T12 levels of the spine, left sacroiliac joint, left femoral head and distal end of right femur, which were corresponding areas to her symptoms (Fig. 1, left). After completion of chemotherapy courses, her symptoms were markedly improved along with the resolution of most radiographic features (Fig. 1, right). Unfortunately, the disease relapsed in only a few months after chemotherapy discontinuation.

Discussion

Rheumatic disorders are common manifestations in HIV-infected patients and continue to be prevalent despite the introduction of highly active anti-retroviral therapy (HAART) (4, 5). These conditions can occur as the direct result of HIV infection, a consequence of immune deficiency or a result of HIV-related treatment (6).

The main symptoms of the present patient are limb girdle pain and stiffness without arthritis and muscle weakness. Therefore, the differential diagnoses should include HIV-associated arthralgia and myalgia, musculoskeletal infection, rheumatic complications from HAART and other rheumatic disorders not directly related to HIV infection.

Arthralgia and myalgia are common symptoms in HIV-infected patients and can occur at any stage of the disease. Arthralgia is most frequently observed in the knees, shoulders and elbows. However, it is rarely severe and tends to be self-limited. Musculoskeletal infection usually develops in HIV-infected patients who have low CD4 T-lymphocyte counts. Didanosine, lamivudine and efavirenz may cause musculoskeletal symptoms such as arthralgia and myalgia but it is uncommon and usually mild and self-limited. Avascular necrosis of the bones (osteonecrosis) may occur due to either HIV infection or HAART (particularly protease inhibitor). It is also associated with a low CD4-T lymphocyte count (4-6).

Based on the above data, HIV-related rheumatic disorders seem less likely to be the responsible cause of the present patient’s symptoms due to her refractory pain and adequate CD4-T lymphocyte counts. The presence of limb girdle pain and stiffness accompanied by elevated ESR usually leads physicians to make the diagnosis of PMR. However, there are a number of disputes regarding the diagnosis of PMR in this case as follows: First, the patient’s age was less than 50 years. Second, PMR is not a common condition in Eastern countries including Thailand. Third, her ESR was rather too high. Fourth, she had many atypical features of PMR such as chronic onset of disease (more than 2 months), lack of accentuation of symptoms with movement, diffuse continuous aching symptoms, minimal morning stiffness and most importantly, poor response to glucocorticoid therapy (1, 7-9). Additionally, to the best of the author’s knowledge, there is no clear association between PMR and both HIV infection and HAART.

Various disorders can present mimicking PMR such as rheumatic diseases, SLE, vasculitis, endocrine disorders,
drugs, infective endocarditis and malignancy (10, 11). Among these, malignancy seems to be the most common entity (7). PMR-like symptoms have been reported as the first manifestation of various malignancies including renal carcinoma, gastric adenocarcinoma, colon cancer, pancreatic cancer, uterine cancer, ovarian cancer, leukemia and lymphoma (7). However, only a few cases of both Hodgkin’s and non-Hodgkin’s lymphoma initially presenting as PMR have been reported previously (12, 13). The pathogenesis of lymphoma-associated polymyalgic symptoms remains unclear. It may occur as a paraneoplastic syndrome (12, 13) or bony metastases, as in the present patient. Atypical features of PMR that are occasionally the first clinical manifestation of disseminated cancer include age younger than 50 years, involvement of only one typical site, asymmetrical involvement at typical sites, additional painful joints, an ESR of <40 or >100 mm/hr and partial or delayed (more than 48 hours) improvement after treatment with 10 mg of prednisone (8, 9). In atypical PMR, there is a significant risk for cancerous bone and joint involvement, which should be specifically searched for (8, 9).

Patients with pure PMR do not seem to have an increased risk for malignancy (11). However, the prevalence of malignancy may increase in patients who present with suspected, new-onset PMR (14). Therefore, follow-up is needed to establish the correct diagnosis. Currently, thorough investigations for malignancy in typical PMR patients are still controversial and should be reserved for patients who fail to respond to glucocorticoid therapy or have atypical presenting features (11).

In summary, this report describes a case of HIV-associated malignant lymphoma who initially presented with PMR-like symptoms. Recognition of atypical features of PMR may help physicians to determine the correct alternative diagnosis, in particular malignancy, and appropriate management.

References


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