CASE REPORT

DOI: 10.5336/caserep.2020-77015

A Case of Polymyalgia Rheumatica with Pericardial and Pleural Effusion

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ABSTRACT Polimyalgia rheumatica is a chronic systemic inflammatory disease that affects people over 50 years old. The disease is characterized by pain and morning stiffness in the shoulders, neck, and pelvic girdle that occur within weeks. Elevated inflammatory markers and dramatic response to corticosteroid therapy are characteristic. It may coexist with giant cell arteritis in some patients. In this presentation, a patient with pleural and pericardial effusion, which is extremely rare during the course of the disease, is reported. The patient responded quickly and very well to corticosteroid therapy.

Keywords: Polymyalgia romatica; pleuro-pericardial effusion

Polymyalgia rheumatica (PR) is a systemic inflammatory disease that occurs in people aged 50 and over and is most common in the 70-80 years old.[1] The disease occurs within weeks and characteristically causes pain and stiffness in the neck, shoulders, upper arms and pelvic girdle.[2] The structures that are mainly affected in PR are not the muscles, but the proximal joints, and especially periarticular structures such as the bursa and tendons.[3] Its pathogenesis is unknown and there is no specific test for diagnosis.[4] The diagnosis is made with clinical findings. A dramatic response to corticosteroids in a short time and eliminating other diseases support the diagnosis.[4] To date, pericardial and pleural effusion has been reported in very few patients, the first of which was published in 2005.[5,6] We report a patient who presented with pleural and pericardial effusion in addition to the classical findings of PR.

CASE REPORT

A 69-year-old female patient admitted to the hospital complaining of pain in the shoulders and neck which was more prominent on the left, and pain radiating to the leg in the left hip. She also complained of pain in the chest and back, weakness and shortness of breath. She stated that she had difficulty lying on her left side. She further stated that these complaints started and progressed in the last few weeks and that she did not benefit from the NSAIDs she used during this period. With the exception of receiving levothyroxin sodium, her medical history were unremarkable.

On physical examination, the patient was alert but appeared restless and painful. On physical examination, she was afebrile, with a pulse rate of 75 beats/min, blood pressure of 120/70 mm Hg, respiratory rate of 22 breaths/min, and oxygen saturation of 98% while breathing room air. Cardiac examination revealed a regular heart rate without rubs or murmur. Breathing sounds were normal without rale or wheezing. Abdominal examination revealed no abnormalities. There was no evidence of clubbing, cyanosis, or edema of the extremities. No arthritis or skin rash were present and no lymphadenopathy was detected. Neurological examination was normal in the patient whose shoulder and neck movements were painful.

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Peer review under responsibility of Turkiye Klinikleri Journal of Case Reports.

Received: 01 Jun 2020 ***Received in revised form:*** 13 Jul 2020 ***Accepted:*** 16 Jul 2020 ***Available online:*** 26 Nov 2020

TABLE 1: Laboratory data.

Variable	Reference range	On admission
White-cell count (per mm^3)	4500-12600	9810
Hemoglobin (g/dl)	11.9-14.6	12.2
Platelet count (per mm^3)	150000-400000	616
Alanine aminotransferase (U/liter)	5-30	21
Creatinine (mg/dl)	0.45-1.20	0.83
Glucose (mg/dl)	70-100	116
TSH (µIU/ml)	0.3-4	1.628
C-reactive protein (mg/dl)	0-0.5	24.22
Erythrocyte sedimentation rate(mm/hr)	0-24	90
Rheumatoid factor (IU/ml)	<8	0-30
Anti-CCP (U/mL)	0-17	<7
ANA	<1/40	1/100
Anti ds-DNA (IU/mL)	<100	<10
Anti nRNP/Sm anticor	Negative	Negative
Anti-Sm	Negative	Negative
Anti SSA	Negative	Negative
Anti SSB	Negative	Negative
Anti- Scl 70	Negative	Negative
Anti Jo-1	Negative	Negative

The laboratory test results on admission are shown in Table 1. Briefly, thrombocytosis and elevated C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR) were detected. The other laboratory data revealed normal levels of creatinine, electrolytes, liver function tests, cardiac enzymes and thyroid stimulating hormone. Urine results were also normal. Serologic tests for rheumatologic disease were also normal except for weak positivity of antinuclear antibodies (ANAs) at 1:100 (Table 1). Electrocardiography showed normal sinus rhythm with no other abnormalities. Chest X-ray revealed left-sided pleural effusion (Figure 1). Computed tomography of the chest showed pericardial and pleural effusion, but no abnormalities in the lung fields (Figure 2). There was also no evidence of filling defects in the pulmonary arteries. An echocardiogram showed pericardial effusion with mild pericardial thickening. Abdominal ultrasonography was reported as normal except for grade II hepatosteatosis. Mammography, breast ultrasonography and thyroid ultrasonography were reported as normal.

These findings suggested PR and 20 mg prednisone was started daily. Her complaints improved dramatically within a few days. Follow-up chest X ray and transthoracic echocardiogram obtained seven days later revealed total regression of both pleural and pericardial effusion (Figure 3). CRP level decreased to 0.12 mg/dL and ESR decreased to 20 mm/h.

The patient was informed about her process and her written approval was received for a case report presentation.

DISCUSSION

Polymyalgia rheumatica is an inflammatory disease with unknown etiology that occurs in people over 50 years of age.[1,2,4] It is more common in women

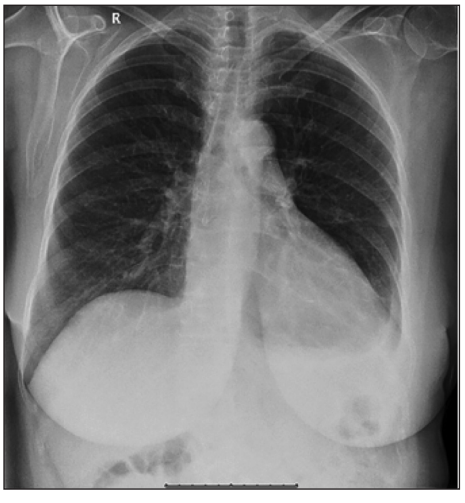

FIGURE 1: Chest X-ray revealed left-sided pleural effusion.

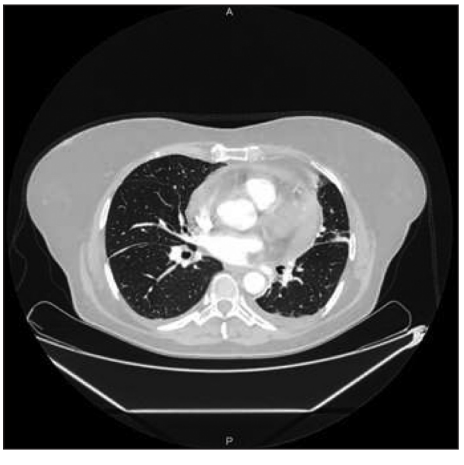

FIGURE 2: Computed tomography of the chest showed pericardial and pleural effusion, but no abnormalities in the lung fields.

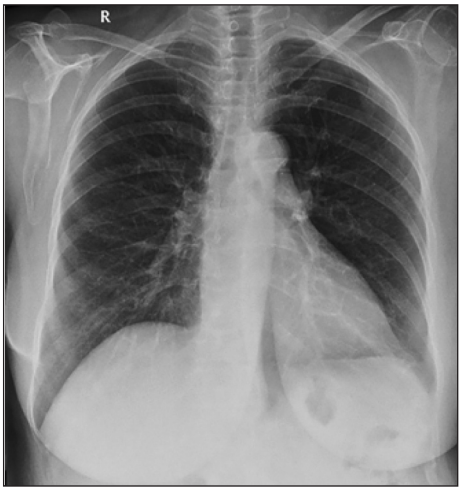

FIGURE 3: Chest X-ray on the seventh day of steroid treatment, revealed total regression of both pleural and pericardial effusion.

and in some geographic areas and is sometimes accompanied by giant cell arteritis.[7,8] They can be seen together or can occur sequentially. Giant cell arteritis has been reported in 16-21% of patients with PR.[9] Therefore, the presence of giant cell arteritis in PR cases must be investigated. The disease characteristically causes pain and stiffness in the neck, shoulders, upper arms and pelvic girdle, sometimes it can start unilaterally, but later becomes symmetrical. Pain is felt more intensely at night and in the first part of the day. Some patients may have systemic complaints such as weakness, fever and weight loss.[8]

The pathogenesis of PR is not known. Some studies have suggested that seasonal factors and infections might play a trigger role. Since it is more common in some regions, the effects of genetic factors have been investigated and it has been stated that the HLA-DRB1 * 04 allele may be related to sensitivity to PR.[8,10] Various studies have suggested that a large number of genes can contribute to PR sensitivity and their relationship with polymorphisms can vary in different populations.

There is no specific test for the diagnosis of PR. The diagnosis is based on clinical findings. In patients over 50 years of age who have classic PR complaints for at least two weeks, diagnosis can be made by the presence of high acute phase reactants, rapid recovery of symptoms with low-dose steroid therapy, and the absence of some other diseases (cancer, chronic infection or other rheumatic disorders).[3] Various criteria have been proposed for diagnosis.[1,11] According to these criteria and the criteria proposed by a Joint Task Force from the American College of Rheumatology (ACR) and the European League Against Rheumatism (EULAR) published in 2012, our patient was found to be compatible with PR.

Pleural and pericardial effusion has been reported very rarely in PM. In a systematic literature review published in 2019 investigating pericardial involvement in GCA and PR, it was reported that a total of 44 cases in GCA and 8 cases in PMR were detected as having pericardial effusion.[6] In this study, it was stated that the probability of pericardial tamponade was higher in PR cases compared to GCA. Pleural effusion is not mentioned in two of 8 cases reported pericardial effusion. Therefore, we can state that our case was the 7th case with pleuro-pericardial effusion in PR. However, it should be borne in mind that there may be cases of pericardial and pleural effusion without clinical findings, and therefore they have not been investigated, so the actual number may be even higher. The pathophysiology of pleuro-pericardial effusion developing in PMR is not clear. It has been suggested that pericardial effusion may develop secondary to vasculitis in small pericardial vessels or to existing inflammation in patients. In a study with PET in isolated PMR cases, it was reported that mild arteritis was detected in 1/3 patients.[12] It is stated that subclinical vasculitis can be found in these patients and should be followed with this respect.

The absence of headache, temporal artery abnormality, jaw claudication, scalp tenderness, visual impairment and ischemic complications, and good clinical response to low dose steroid administration in a short time eliminated the possibility of GCA association at the time of diagnosis. Since serositis can occur in various auto-inflammatory diseases, these disorders should be taken into account in the differential diagnosis. The normal serological tests and the absence of signs and symptoms suggestive of autoinflammatory diseases excludes other rheumatological disorders. (Table 1). Another disease that should be considered in the differential diagnosis is tuberculosis. Unfortunately, investigation of pleural effusion could not technically

possible as amount of fluid was low. The disappearance of pleuro-pericardial effusion with low-dose steroid and no recurrence in the absence of anti-tuberculosis therapy suggested that the effusion was not due to tuberculosis. Additionally, since the sensitivity and specificity of the Quantiferon test is particularly low in high incidence environments and is not recommended for the diagnosis of active tuberculosis, this test has not been performed. The absence of arthritis and negative of both rheumatoid factor and anti-citrulline antibodies eliminated the diagnosis of rheumatoid arthritis. Also, absence of pitting edema in hands and distal extremities, excludes the RS3PE (Remitting Seronegative Symmetrical Synovitis with Pitting Edema) syndrome. Furthermore; imaging methods, serological tests and biochemical tests have exclude malignancy, thyroid disease and other rheumatologic diseases. For these reasons, and because of the good response to low dose steroid therapy in a short time, we think that pleuro-pericardial effusion is caused by PR. Although it is extremely rare to have pleuro-pericardial effusion in PR patients, clinicians should keep this complication in mind and conduct further investigations as needed.

Source of Finance

During this study, no financial or spiritual support was received neither from any pharmaceutical company that has a direct connection with the research subject, nor from a company that provides or produces medical instruments and materials which may negatively affect the evaluation process of this study.

Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

This study is entirely author's own work and no other author contribution.

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