CASE REPORT

Pleuropericardial effusion: an unusual presentation of polymyalgia rheumatica

Lovely Chhabra,¹ Ramprakash Devadoss,² Karthik Gnanapandithan,³ David H Spodick⁴

¹Hartford Hospital, University of Connecticut School of Medicine, Hartford, Connecticut, USA

²Department of Internal Medicine, Saint Vincent Hospital, University of Massachusetts Medical School, Worcester, Massachusetts, USA

³Saint Vincent Hospital, University of Massachusetts Medical School, Worcester, Massachusetts, USA

⁴Department of Cardiovascular Medicine, Saint Vincent Hospital, University of Massachusetts medical School, Worcester, Massachusetts, USA

Correspondence to
Dr Lovely Chhabra,
lovids@hotmail.com

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SUMMARY

Polymyalgia rheumatica (PMR) is an idiopathic systemic inflammatory condition manifesting as aches and muscle stiffness of the proximal muscle groups. Very rarely, PMR can be associated with serositis. We report a very rare case of PMR where the patient's primary presentation was a pleuropericardial syndrome (pleuropericardial effusion).

BACKGROUND

The term 'polymyalgia rheumatica (PMR)' was coined by Barber in 1957,¹ though the first actual description of the disease was as early as 1888 by Bruce under the name 'senile rheumatic gout'.² It is an inflammatory condition of unknown cause characterised by aching and morning stiffness in the cervical region, shoulder and pelvic girdles.³ It is frequently seen in people over 50 years of age with peak incidence during 70–80 years of age.³ The reported annual incidence in Europe and USA varies between 1.3 and 11.3/10 000 individuals. Increased incidence has been noted in populations at high altitudes and of Scandinavian origin.^{4 5}

Apart from proximal muscle involvement, pericardial effusion with or without tamponade physiology has also been recognised in association with PMR.^{6–9} However, there have been only two previous reports of pleuropericardial effusion with PMR.^{10 11} We report yet another unusual case of PMR where the patient's primary presentation was a pleuropericardial effusion.

CASE PRESENTATION

An 86-year-old Caucasian woman with a medical history of hypertension, hyperlipidaemia, hypothyroidism and remote history of PMR was admitted with substernal chest pain and progressively worsening shortness of breath of approximately 1 week duration. The chest pain was described as dull-aching with no definite radiation or exacerbation with exertion, but worsened with deep inspiration. She also reported bilateral shoulder and upper back pain since a cataract surgery 2 months ago for which she was taking cyclobenzaprine with some relief. The above events were also surrogated by a 10-pound weight loss and worsening lethargy in the last month. She denied any headache or visual disturbance on presentation.

Her medications included levothyroxine, amlodipine, furosemide, omeprazole, multivitamin and calcium supplements along with cyclobenzaprine as needed for the shoulder and back pain. Physical

examination on presentation was significant for mild pallor, tachycardia and decreased breath sounds in the bilateral lower lung field. Her other systemic examination was within normal limits.

INVESTIGATIONS

The initial laboratory results were significant for anaemia (haemoglobin 9.1 g/dL) and mild neutrophilic leucocytosis. Further evaluation of anaemia revealed a normochromic and normocytic morphology with elevated ferritin, low iron and total iron binding capacity. Renal function test reported a calculated glomerular filtration rate of 32 mL/min without any notable proteinuria. Liver function test was within normal limits with total protein of 6.7 g/dL with albumin of 3.3 g/dL. Protein electrophoresis showed a pattern consistent with non-selective protein loss. Inflammatory markers on admission were elevated with erythrocyte sedimentation rate (ESR) of 90 mm/h and C reactive protein (CRP) of 167.23 mg/L. Immunological evaluation revealed positive antinuclear antibodies (ANA) and rheumatoid arthritis (RA) factors, however they were considered non-specific in the absence of distal rheumatological symptoms.

ECG showed diffuse low-voltage QRS criteria and non-specific ST-T changes. Chest radiogram showed bilateral pleural effusions and cardiomegaly (figure 1). A CT performed to elaborate the findings of radiogram revealed moderate-to-large sized pericardial effusion along with bilateral pleural effusion (left more than right) and associated mild compressive atelectasis of both lungs (figure 2). Emergent echocardiogram showed normal left ventricular systolic

Figure 1 Chest radiogram showing bilateral pleural effusions and cardiomegaly.

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Figure 2 Chest CT performed to elaborate the findings of radiogram revealed moderate-to-larger pericardial effusion along with bilateral pleural effusion (left more than right) and associated mild compressive atelectasis of both lungs.

function with mild tricuspid regurgitation and mild-to-moderate aortic insufficiency. Right ventricle was reported to be normal in size and function. There was moderate pericardial effusion without any evidence of tamponade physiology (figure 3).

DIFFERENTIAL DIAGNOSIS

The patient had symptoms compatible with the diagnosis of PMR (age >50, pain and stiffness of the proximal muscles—bilateral shoulder and upper back, elevated ESR) but there were also some atypical features—positive ANA and RA factor, pleuropericardial effusion and sparing of the pelvic girdle.

TREATMENT

As the patient did not have any distal rheumatological symptoms and solid malignancy was ruled out with CT, a trial of steroids was planned. She was started on intravenous steroids and the same was transitioned to low-dose oral prednisone with dramatic clinical improvement within 48 h.

OUTCOME AND FOLLOW-UP

A follow-up re-evaluation of inflammatory markers showed a down trending pattern with ESR at 28 mm/h within a week of

Figure 3 Echocardiogram showing moderate pericardial effusion without any evidence of tamponade physiology.

steroid initiation. CRP after 2 months of therapy was 0.16 mg/L and a follow-up echocardiogram at that time revealed significant reduction of the pericardial effusion. The patient continued to maintain good symptomatic recovery and the steroids were slowly tapered with caution to avoid exacerbation of symptoms on recovery. Repeat CT of the chest performed for an alternate indication after completion of the steroid regimen showed no evidence of pleural effusion.

DISCUSSION

Two independent diagnostic criteria sets formulated as early as the 1980s, have commonly been used in practice for the diagnosis of PMR (table 1).^{3 12 13} The only difference between the two is the inclusion of the responsiveness to steroids.

The pathogenesis of PMR still remains elusive. Increased frequency of occurrence in certain population has raised suspicion about environmental and genetic causes, however a viral cause has also been suspected.³ The shoulder has been reported to be the most common site of involvement followed by hips and neck. The shoulder and pelvic girdle pain usually radiates distally to the elbows and knees, respectively.¹¹

Retrospectively, our patient fulfilled all criteria in both the sets for the diagnosis of PMR. She remained symptom free on follow-up despite tapering off steroids which goes against any other rheumatological diagnosis. Interestingly, follow-up ANA titres conducted after complete symptomatic recovery returned negative. Apart from those criteria points, our patient also had other systemic manifestations with anorexia, weight loss and mild anaemia consistent with reported features of PMR.¹⁴ Myositis, one likely differential for proximal muscle weakness was ruled out with normal creatine phosphokinase (CPK) levels on admission. Late onset systemic lupus erythematosus (SLE) is another close differential as it has been shown to mimic PMR,¹⁵ but the same appears improbable as the patient lacked other symptoms to qualify for the diagnosis of SLE and also experienced complete resolution of symptoms with steroids alone. It is imperative to remember that steroids are the treatment of choice in patients with pericardial involvement in the setting of underlying systemic connective tissue disease.^{16 17}

PMR presenting with pericardial effusion has been identified in the past, however pleuropericardial effusion has not been a well-reported phenomenon with PMR. A thorough literature search revealed only two such reports as previously described.^{10 11}

Table 1 Diagnostic criteria for PMR

Criteria of Chuang <i>et al</i> ¹² (all criteria must be present)	▶ Age of 50 years or older ▶ Bilateral aching and stiffness for 1 month or more involving two of the following areas: neck or torso, shoulders or proximal regions of the arms and hips or proximal aspects of the thighs ▶ ESR greater than 40 mm/h ▶ Exclusion of all other diagnoses except giant-cell arteritis
Criteria of Healey ¹³ (all criteria must be present)	▶ Pain persisting for at least 1 month and involving two of the following areas: neck, shoulders and pelvic girdle ▶ Morning stiffness lasting more than 1 h ▶ Rapid response to prednisone ▶ Absence of other diseases capable of causing musculoskeletal symptoms ▶ Age of more than 50 years ▶ ESR greater than 40 mm/h

ESR, Erythrocyte sedimentation rate; PMR, polymyalgia rheumatica.

Malone and McCarthy first described a case of pleuropericardial effusion in an elderly patient who presented with a pleuritic chest pain, and shoulder girdle aching and stiffness. Their patient responded well to oral prednisolone 15 mg/day dosing.¹⁰ Similarly, Sánchez Ruiz-Granados *et al*¹¹ reported a case of an 80-year-old patient who presented with primary symptoms of PMR however a coincidental pleuropericardial effusion was noted on that visit.

Isolated pleural effusion has also not been reported with PMR. Minimal pericardial effusion may often be experienced in the setting of a pleural effusion such as in congestive heart failure; however, a moderate pleuropericardial effusion (such as in our case) indicates significant independent serositis at the level of pericardium and pleura. Surface ECG may often reveal low-voltage QRS changes, PR depression, and even electrical or vector alternans if effusion size is large.^{18–20} Whether this pleura is the one adjacent to the inflamed pericardium is a questionable theory. Serositis can occur with any inflammatory pathology, but in our patient the absence of markers towards any other diagnosis and significant recovery with prednisone and maintaining symptoms-free state even after weaning off the steroids without any other disease modifying agent, goes in favour of PMR.

This presentation with the previous examples warrants consideration of pleuropericardial effusion without any localising rheumatological symptoms as one of the clinical pictures of PMR. Physicians should be aware of this rare differential diagnosis in order to make an appropriate informed decision and selection of treatment therapy.

Learning points

- ▶ Polymyalgia rheumatica (PMR) is an idiopathic systemic inflammatory condition manifesting as aches and muscle stiffness of the proximal muscle groups. Very rarely, PMR can be associated with serositis.
- ▶ Apart from proximal muscle involvement, pericardial effusion with or without tamponade physiology can be very rarely present in association with PMR.
- ▶ Steroids are the therapy of choice of pleuropericardial effusion syndrome in the setting of PMR.

Competing interests Dr David H Spodick receives royalties from his textbook, *The Pericardium: A Comprehensive Textbook (Fundamental and Clinical Cardiology)*. New York: Marcel Dekker, 1997.

Patient consent Obtained.

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REFERENCES

- 1 Barber HS. Myalgic symptoms with constitutional effects: polymyalgia rheumatic. *Ann Rheum Dis* 1957;16:230–7.
- 2 Bruce W. Senile rheumatic gout. *BMJ* 1888;2:811–13.
- 3 Salvarani C, Cantini F, Boiardi L, *et al*. Polymyalgia rheumatica and giant cell arteritis. *N Engl J Med* 2002;347:261–77.
- 4 Van Hecke O. Polymyalgia rheumatica—diagnosis and management. *Aust Fam Physician* 2011;40:303–6.
- 5 Chuang T, Hunder G, Ilstrup D, *et al*. Polymyalgia rheumatica: a 10-year epidemiologic and clinical study. *Ann Intern Med* 1982;97:672–80.
- 6 Bohan A. Polymyalgia rheumatica and pericardial effusion. *Ann Intern Med* 1984;101:147.
- 7 Cobo T, Zubera Salibarría Z, Muñoz Sánchez J, *et al*. Polymyalgia rheumatica and pericardial effusion. *Ann Intern Med* 1991;8:362–3.
- 8 Brucato A, Brambilla G. Polymyalgia rheumatica and pericardial tamponade. *Ann Rheum Dis* 2002;61:283.
- 9 Calvo E, Becerra E, Lopez-Longo FJ, *et al*. Pericardial tamponade in a patient with polymyalgia rheumatica. *Clin Exp Rheumatol* 2009;152(Suppl):S83–5.
- 10 Malone CB, McCarthy GM. Polymyalgia rheumatica as an unusual cause of pleural and pericardial effusion. *J Clin Rheumatol* 2005;11:59–60.
- 11 Sánchez Ruiz-Granados E, Del Castillo Madrigal M, Romero Jiménez MJ. Pleural and pericardial effusion in a patient with polymyalgia rheumatica: a case presentation. *Rheumatol Clin* 2013;9:376–8.
- 12 Chuang TY, Hunder GG, Ilstrup DM, *et al*. Polymyalgia rheumatica: a 10-year epidemiologic and clinical study. *Ann Intern Med* 1982;97:672–80.
- 13 Healey LA. Long-term follow-up of polymyalgia rheumatica: evidence for synovitis. *Semin Arthritis Rheum* 1984;13:322–8.
- 14 Salvarani C, Macchioni P, Boiardi L. Polymyalgia rheumatica. *Lancet* 1997;350:43–7.
- 15 Hutton CW, Maddison PJ. Systemic lupus erythematosus presenting as polymyalgia rheumatica in the elderly. *Ann Rheum Dis* 1986;45:641–4.
- 16 Chhabra L, Spodick DH. Letter by Chhabra and Spodick regarding article, “Treatment of acute and recurrent idiopathic pericarditis”. *Circulation* 2013;128:e391.
- 17 Chhabra L, Spodick DH. Pericardial disease in the elderly. In Aronow WS, Fleg JL, Rich MW, eds *Cardiovascular disease in the elderly*. 5th edn. London, UK: CRC Press, 2013:644–68; (ISBN: 1842145436).
- 18 Chhabra L, Spodick DH. Ideal isoelectric reference segment in pericarditis: a suggested approach to a commonly prevailing clinical misconception. *Cardiology* 2012;122:210–12.
- 19 Chhabra L, Spodick DH. ECG in pericarditis and ST-elevation myocardial infarction: timing of observation is critical. *Am J Med* 2014;127:e17.
- 20 Chhabra L, Spodick DH. Swinging heart and vector alternans: signs of impending doom. *Heart* 2013;99:1545.

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