

Case report

Pericardial tamponade in a patient with polymyalgia rheumatica

E. Calvo¹, E. Becerra¹, F.J. López-Longo¹, F.J. Cabrera², L. Carreño¹,
A. Paravisini¹, M. Cebollero³, B. Pinilla², A. Muiño²

Departments of ¹Rheumatology, ²Internal Medicine and ³Pathology, Hospital General Universitario Gregorio Marañón, Madrid, Spain.

Enrique Calvo, MD
Elena Becerra, MD
Francisco J. López-Longo, MD, PhD
Francisco J. Cabrera, MD
Luis Carreño, MD, PhD
Alexandra Paravisini, MD
María Cebollero, MD
Blanca Pinilla, MD
Antonio Muiño, MD

Please address correspondence and reprint requests to:

Dr. Francisco J. López-Longo,
Servicio de Reumatología,
Hospital General Universitario
Gregorio Marañón,
C/ Dr. Esquerdo 46,
28007 Madrid, Spain.
E-mail: fjlopezlongo@hotmail.com

Received on July 2, 2008; accepted in revised form on November 21, 2008.

Clin Exp Rheumatol 2009; 27 (Suppl. 52): S83-S85.

© Copyright CLINICAL AND EXPERIMENTAL RHEUMATOLOGY 2009.

Key words: Pericardial tamponade, polymyalgia rheumatica, giant cell arteritis

Competing interests: none declared.

ABSTRACT

We report a patient who developed pericarditis and pericardial tamponade coinciding with polymyalgia rheumatica onset. Our patient did not show any clinical sign of vasculitis; temporal artery biopsies were negative for giant cell arteritis. Pericardial biopsy in our case shows inflammatory perivascular lymphocytary infiltrates thus we believe pericardial effusion has an inflammatory-immunologic origin. Cardiac manifestations are exceptional in polymyalgia rheumatica, though it should be considered in the differential diagnosis in patients with pericarditis over 50 years. The recognition of this uncommon manifestation is very important due to the good response to corticosteroid treatment.

Introduction

Polymyalgia rheumatica (PMR) is an inflammatory disorder of unknown cause characterized by pain, stiffness and functional impairment in the scapular and pelvic girdles. It usually affects people older than 50 years, most women, showing a progressive onset (1). PMR and giant cell arteritis (GCA) are frequently overlapping conditions (2) sharing the following features: epidemiological data, clinical manifestations, and favourable response to corticosteroid treatment. Conditions mimicking PMR or presenting with polymyalgic manifestations and the possibility of a subclinical GCA should be excluded (3, 4). Visceral involvement in patients with GCA is uncommon; but in PMR is exceptional. We report a patient who developed pericarditis and pericardial tamponade coinciding with isolated PMR onset.

Case report

A 74-year-old woman presented to the Emergency Department complaining of

fever, shortness of breath with pleural pain on the left side of the thorax, a two weeks history of asthenia and weight-loss (8.8 pounds). Pain responded poorly to acetaminophen. Her medical history included bronchial asthma and hypertension. On examination, she was febrile (38°C) and pale. Blood pressure was normal. External jugular veins were slightly distended and hepatojugular reflux was negative. Cardiac auscultation was unremarkable. Lung auscultation revealed random wheezes. No adenopathies were found. Abdominal and neurological examination was normal. All arterial pulses were present.

The first data obtained were the following: elevated white blood cell count ($13.8 \times 10^9/l$), with a raised number of neutrophils ($11.3 \times 10^9/l$) and no increase in eosinophils; anemia (10.8 g/dl); normal platelet count ($402 \times 10^9/l$); C-reactive protein levels (CRP) and erythrocyte sedimentation rate (ESR) were 27.2 mg/L and 120 mm/h, respectively. Subsequent analysis showed: creatinine: 1 mg/dL; urea: 48 mg/dL; ferritin: 185 ng/mL; haptoglobin: 472 mg/dL; normal sideremia, folic acid and vitamin B values; total serum proteins: 7.7 g/dL (44% albumin, 21% alpha 2 globulin and 15.8% beta globulin); aspartate-amino-transferase: 97 UI/L; alanine-amino-transferase: 61 UI/L; gamma glutamyl-transpeptidase (108 UI/L); and normal levels of alkaline phosphatase (132 UI/L). Coagulation test showed only an elevated fibrinogen (711 mg/dL). Muscular enzymes, serum tumor markers, and urinalysis were normal.

Chest radiography showed mild cardiomegaly; blunting of both costophrenic angles; and lingula atelectasis. Computerized tomography revealed the following findings: laminar basal atel-

ectasis in the left lung; calcified right hilar; precarinal and subcarinal lymph nodes; pleural calcification; calcified right basal granuloma; and renal cysts. Pulmonary function test demonstrated a mixed pattern of severe obstructive and moderate restrictive ventilatory defects. Abdominal ultrasound was normal.

Electrocardiogram revealed the following: generalized low voltage; atrial extra-systoles; incomplete right bundle branch block; left anterior hemiblock; negative T waves in precordial leads I-VI; and Q waves in leads III and aVF. Autoantibody screening showed positive anti-nuclear antibodies (1:160) with granular pattern; but rheumatoid factor and anti-DNA, anti-ENA and antineutrophil cytoplasmic autoantibodies were negative. C3 and C4 fractions were normal. Serological markers were negative for the following: hepatitis B and C virus, Epstein-Barr virus, cytomegalovirus, herpes virus, B-19 parvovirus, rickettsias, coxiella, legionella and Mycoplasma. Mantoux test and cultured tests were also negative.

Forty-eight hours after her admission, an echocardiogram was performed disclosing the following findings: a moderate pericardial effusion (sum of anterior and posterior echo-free space was 10 mm), more accentuated in right cavities; minimum diastolic right atrial collapse; dilation of vena cava; valves and ejection fraction of the left ventricle (EFLV) were normal; pulmonary artery pressure (PAP) could not be estimated.

Four days after admission, paroxysmic atrial fibrillation appeared with hypotension. Echocardiogram showed a larger pericardial effusion (sum of anterior and posterior echo-free space was 20 mm) and signs of reduced cardiac output. Pericardiectomy was performed and a pericardial biopsy was obtained. 150 ml of serohematic fluid were yield. Laboratory data of this fluid showed the following: 5.2 g/dl proteins; 4562 UI/L lactate dehydrogenase; and 65 mg/dL glucose. Bacterial and fungi cultures were negative. Ziehl-Nielsen staining and the polymerase chain reaction technique ruled out mycobacterial infection. Histologic examination revealed inflammation of the pericardium with

fibrosis and interstitial hemorrhage areas without fibrin deposits. Immunohistochemical analysis reported perivascular lymphocytary infiltrate consisted of a mixed lymphocyte population, mostly T cells (CD45/CD3/CD5 positive) with a minority B cells population (CD79a/CD20 positive).

Three days after pericardiocentesis, chest pain and dyspnea diminished; but fever and pain on extremities movement persisted. Bilateral temporal artery biopsies were performed in order to exclude a silent GCA, demonstrating subintimal fibrosis, several elastic laminae with no infiltrate or granuloma. In both cases, the temporal artery biopsy site taken was greater than 1 cm. Eye fundus examination revealed a thinning of the neuroretinal ring in the lower temporal side, without papilledema.

Clinical and laboratory findings suggested PMR; therefore the patient was prescribed empirically prednisone (15 mg/day; patient weighted 158 lbs). Fever and polymyalgia syndrome had a rapid improvement. Liver enzymes and haemoglobin (13 g/dL) returned to normal, ESR dropped to 10mm/h and CRP to 1.1 mg/dl. The patient was released 14 days after her admittance.

Two months later, she became asymptomatic and her laboratory analyses (haemoglobin, ESR and CRP) were normal. Five months later, she underwent an echocardiogram control showing a minimum residual pericardial effusion; EFLV was 60% and PAP 40 mmHg. Six months later, corticosteroid treatment was gradually tapered and suspended. Presently, the patient continues to do well without therapy.

Discussion

PMR is a relatively common disorder. Annual incidence varies from 12.5 cases per 100,000 inhabitants to 52.5 cases (5); but increases significantly with age (19.8 cases between 50-59 years, and 112.2 between 70-79 yrs) (6). This illness has either a sudden onset or a gradual development leading to pain, stiffness and progressive disability. One-third of patients with PMR, present the following systemic features: fever; constitutional syndrome (weight loss, anorexia and malaise) (1);

and asymmetric peripheral non-erosive arthritis (7). Visceral manifestations in these patients are very uncommon in absence of GCA.

Pericardial effusion in patients with PMR without GCA is rare; and extremely singular as onset of PMR. In our report a subclinical 'silent' GCA was appropriately excluded. In the literature review made, only 4 cases of pericardial effusion in PMR have been published; 2 of them associated pleural effusion (8-11). Only one previous description of heart block attributed to a pericardial effusion associated with PMR has been made (10). On the other hand, pericardial effusion in GCA is unusual, and it has only been reported on 21 occasions over last 60 years (12-14).

Although infectious diseases may be associated with pericarditis and polymyalgia manifestations, these conditions as well as others (3) were excluded. In addition, since the presence of anemia, platelet counts greater than 400.000/mm³ and very high ESR levels have previously been reported to be more common in patients with PMR associated to GCA than in cases of isolated PMR (15), we performed bilateral temporal artery biopsies that yielded negative pathological findings for GCA. In this regard, in keeping with recent reports (16), due to the sample size of both biopsies, the possibility of a 'silent' GCA in our case was very unlucky.

This report, one of the very few cases informed of pericarditis and pericardial tamponade coinciding with PMR onset, reveals the first description of lymphocyte subpopulation from a pericardial biopsy sample. Previous reports show that patients with PMR have an oligoclonal CD8+ T cells expansion (17); although in active and untreated patients, circulating CD8+ T cells may be reduced (18). Temporal artery samples and synovial membrane of patients with isolated PMR show similar findings to that described in the temporal arteries wall of GCA, including the presence of moderate inflammatory infiltrates with predominance of macrophages and perivascular T lymphocytes (especially CD4+ T cells); but B cells, natural-killer cells, and T γ δ cells are not present (19, 20). Our pericardial

biopsy demonstrated a lymphocytic inflammatory infiltrate of perivascular localization with a predominance of T cells. Contrary to what has been previously described in temporal artery biopsies, the pericardial sample showed T cells (CD45/CD3/CD5: positive), but these cells were negative for CD4 and CD8 surface antigens. Our results are not conclusive and further research is necessary to confirm them.

Although PMR unusually shows cardiac manifestations, this disorder should be considered as a differential diagnosis in patients with pericarditis older than 50 years. The importance of identifying pericarditis in patients with PMR resides in their excellent response to corticosteroid treatment; on the contrary, in the more common 'idiopathic' or viral pericarditis steroid use should be restricted, since NSAID and colchicine are the preferred drugs. The frequency of pericarditis in PMR untreated patients remains unknown; additional studies in PMR patients that include routine echocardiographic evaluations could clarify its frequency.

References

1. SALVARANI C, CANTINI F, BOIARDI L, HUNDER GG: Polymyalgia rheumatica and giant-cell arteritis. *N Engl J Med* 2002; 347: 261-71.
2. GONZALEZ-GAY MA: Giant cell arteritis and polymyalgia rheumatica: two different but often overlapping conditions. *Semin Arthritis Rheum* 2004; 33: 289-93.
3. GONZALEZ-GAY MA, GARCIA-PORRUA C, SALVARANI C, OLIVIERI T, HUNDER GG: The spectrum of conditions mimicking polymyalgia rheumatica in Northwestern Spain. *J Rheumatol* 2000; 27: 2179-84.
4. GONZALEZ-GAY MA, GARCIA-PORRUA C, AMOR-DORADO JC, LLORCA J: Giant cell arteritis without clinically evident vascular involvement in a defined population. *Arthritis Rheum* 2004; 51: 274-7.
5. SALVARANI C, GABRIEL SE, O'FALLON WM, HUNDER GG: Epidemiology of polymyalgia rheumatica in Olmsted County, Minnesota, 1970-1991. *Arthritis Rheum* 1995; 38: 369-73.
6. NARVAEZ J, NOLLA-SOLE JM, VALVERDE-GARCIA J, ROIG-ESCOFET D: Sex differences in temporal arteritis and polymyalgia rheumatica. *J Rheumatol* 2002; 29: 321-5.
7. SALVARANI C, BOIARDI L, MACCHIONI L, OLIVIERI I, PORTIOLI I: Polymyalgia rheumatica. *Lancet* 1996; 348: 550-1.
8. TEIRA-COBOR, ZUBERO-SULIBARRIAZ, MUÑOZ-SANCHEZ J, SANTAMARÍA-JAUREGUI JM: Polymyalgia rheumatica and pericardial effusion. *An Med Interna* 1991; 8: 362-3.
9. BOHAN A: Polymyalgia rheumatica and pericardial effusion. *Ann Intern Med* 1984; 101: 147.
10. BRUCATO A, BRAMBILLA G: Polymyalgia rheumatica and pericardial tamponade. *Ann Rheum Dis* 2002; 61: 283.
11. MALONE CB, MCCARTHY GM: Polymyalgia rheumatica as an unusual cause of pleural and pericardial effusion. *J Clin Rheumatol* 2005; 11: 59-60.
12. TEIXEIRA A, CAPITAINE E, CONGY F, HERSON S, CHERIN P: [Myopericarditis during Horton disease]. *Rev Med Interne* 2003; 24: 189-94.
13. VALSTAR MH, TERPSTRA WF, DE JONG RS: Pericardial and pleural effusion in giant cell arteritis. *Am J Med* 2003; 114: 708-9.
14. BABLEKOS GD, MICHAELIDES SA, KARACHALIOS GN, NICOLAOU IN, BATISTATOU AK, CHARALABOPOULOS KA: Pericardial involvement as an atypical manifestation of giant cell arteritis: report of a clinical case and literature review. *Am J Med Sci* 2006; 332: 198-204.
15. GONZALEZ-GAY MA, GARCIA-PORRUA C, VAZQUEZ-CARUNCHO M: Polymyalgia rheumatica in biopsy proven giant cell arteritis does not constitute a different subset but differs from isolated polymyalgia rheumatica. *J Rheumatol* 1998; 25: 1750-5. Erratum in: *J Rheumatol* 1998; 25: 2483.
16. GONZALEZ-GAY MA: The diagnosis and management of patients with giant cell arteritis. *J Rheumatol* 2005; 32: 1186-8.
17. MARTINEZ-TABOADA VM, GORONZY JJ, WEYAND CM: Clonally expanded CD8 T cells in patients with polymyalgia rheumatica and giant cell arteritis. *Clin Immunol Immunopathol* 1996; 79: 263-70.
18. CORRIGALL VM, DOLAN AL, DASGUPTA B, PANAYI GS: The sequential analysis of T lymphocyte subsets and interleukin-6 in polymyalgia rheumatica patients as predictors of disease remission and steroid withdrawal. *Br J Rheumatol* 1997; 36: 976-80.
19. WEYAND CM, HICOK KC, HUNDER GG, GORONZY JJ: Tissue cytokine patterns in patients with polymyalgia rheumatica and giant cell arteritis. *Ann Intern Med* 1994; 121: 484-91.
20. MELICONI R, PULSATELLI L, UGUCCIONI M *et al.*: Leukocyte infiltration in synovial tissue from the shoulder of patients with polymyalgia rheumatica. Quantitative analysis and influence of corticosteroid treatment. *Arthritis Rheum* 1996; 39: 1199-207.
21. DASGUPTA B, MATTESON EL, MARADIT-KREMERS H: Management guidelines and outcome measures in polymyalgia rheumatic (PMR). *Clin Exp Rheumatol* 2007; 25 (Suppl. 47): S130-S136.