Pericardial Involvement as an Atypical Manifestation of Giant Cell Arteritis: Report of a Clinical Case and Literature Review

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ABSTRACT: *Purpose:* Pericardial effusion has been known to be a rare manifestation of giant cell arteritis. During the last six decades, only 24 cases have been cited in the literature. In this report, we describe the case of a patient presenting with nonspecific symptoms and development of pericardial effusion. *Procedures and Findings:* A 71-year-old woman was admitted to the hospital with low-grade fever, exertion breathlessness, atypical diffuse muscular pain, and weight loss over a period of about 5 weeks. Pericardial effusion and giant cell arteritis were diagnosed by echocardiography and left temporal artery biopsy, respectively. Treatment with corticosteroids resulted in remarkable improvement of symptoms and complete remission of pericardial effusion. One year after admission, the patient remained in a stable good condition, under low steroid maintenance dosage. *Conclusions:* The diversity of clinical manifestations (such as pericardial effusion) in such a potentially severe disease should alert the physician to prompt diagnosis and treatment in view of impending irreparable vascular damages, even in cases in which the initial presentation is quite uncommon. **KEY INDEXING TERMS:** Pericarditis; Effusion; Giant cell; Arteritis. **[Am J Med Sci 2006;332(4):198–204.]**

Giant cell arteritis (GCA) is a chronic systemic inflammatory vasculitis particularly affecting the external carotid branches.[1] Aortic dissection due to mural weakness induced by GCA has also been reported.[2]

Degeneration of large and medium-sized arteries due to GCA results in luminal narrowing associated with end-organ ischemia.[3] Atrophy of media and calcification in the internal elastic membrane of the arterial wall are considered preconditions for the development of the disease.[4] Immunologic disorders have also been thought to participate.[5]

Cardiac manifestations of GCA are less common and include angina pectoris, myocardial infarction, granulomatous myocarditis, and aortic regurgitation.[6,7] Pericardial involvement is an extremely rare manifestation of GCA. To the best of our knowledge, since 1949 only 20 cases of GCA[8–14] and 4 cases of polymyalgia rheumatica[15–18] (PMR) (the latter being a clinical subtype of GCA[19]) manifesting with pericardial effusion have been reported in literature.

We describe a case of a patient with giant cell arteritis atypically presenting with pericardial effusion. The relevant literature concerning this particular manifestation, which may disorientate the clinician from proper diagnosis and treatment, along with current knowledge for cause, pathogenesis, diagnosis, and treatment of the disease, are also briefly reviewed.

Case Report

A 71-year-old woman was admitted in the "Red Cross" General Hospital* with low-grade fever, exertion breathlessness, diffuse muscular pain, and a weight loss of 4 kg developing over a period of about 5 weeks. Headaches, visual disturbances, and jaw and tongue claudication were not reported. Her past medical history was free from any disease or use of any regular medication. On admission, her temperature was 37.6° C.

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* *"Red Cross" General Hospital is one of the biggest General Hospitals in the city of Athens. The American Journal of the Medical Sciences.* **Revision (Highlighted form).**

Physical examination revealed a pulse rate of 78 beats per minute, a respiratory rate of 14 breaths per minute, and a blood pressure of 135/85 mm Hg. Temporal arteries looked normal, without tenderness on palpation. No other abnormal findings were detected, either on physical examination or chest radiograph.

Results of laboratory tests included a hemoglobin value of 8.6 g/dL, a white blood cell count of 7.3 $\times$ $10^3/\mu$L (with a normal leukocyte differential), and a platelet count of 238 $\times$ $10^3/\mu$L. Erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) values were found to be 115 mm/h and 82 mg/dL, respectively. Results of liver function tests were as follows: SGOT, 35 U/L; SGPT, 30 U/L (reference values <37 U/L and <41 U/L, respectively); γGT, 52 U/L (reference values, 11-49 U/L); ALP, 352 U/L (reference values, 45–122 U/L).

Serum levels for creatinine (Cr), calcium (Ca), magnesium (Mg), and uric acid (UA) were found to be 1.3 mg/dL, 8.9 mg/dL, 1.8 mg/dL, and 7.4 mg/dL, respectively. Serologic tests performed for infection with cytomegalovirus (CMV IgG [IFA], CMV IgM [IFA]), Epstein-Barr virus (EBV-VCA IgG [IFA], EBV-VCA IgM [IFA]), *Brucella* (Rose Bengal, *Brucella* capture [immuno-capture assay]), *Salmonella* (Widal [group D]), hepatitis A (IgG, IgM/ELISA/Kit Abbott), hepatitis B (HBsAg: IgG, IgM total/ELISA/Kit Abbott), hepatitis C (IgG, IgM total/Micro-ELISA/Kit Abbott), $HIV_{1,2,0}$ (Microelisa/Abbott), and *Pneumococcus*, *Legionella*, *Mycoplasma pneumoniae*, and *Chlamydia pneumoniae* were all negative. Sputum cultures for infection with mycobacterial tuberculosis were negative too. Rheumatoid factor was negative and anti-DNA antibodies were not detected. Anti-nuclear (ANA) antibodies were found positive within normal limits (10–12%) at a titer of 1/80. Blood and urine cultures showed no pathologic findings.

A mild pericardial effusion, with no signs of cardiac tamponade or hemodynamic instability, was detected by two-dimensional echocardiography (Figure 1, Figure 2) performed 3 days after admission, and no pericardial drainage was warranted. Ejection fraction was calculated at 57%. The elevated values of ESR and CRP, along with the clinical presentation, raised a high degree of clinical suspicion for the presence of a systemic inflammatory

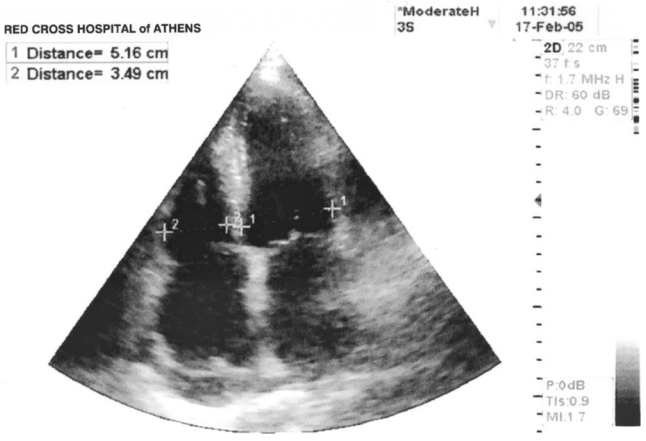

Figure 1. Isovolumic phase of systole (two-dimensional echocardiography). Tricuspid and mitral valves are closed. A marginal distention of the left ventricle is detected. A mild pericardial effusion is present. The absence of right atrial and/or ventricular indentation or collapse indicates lack of constriction. The specific two-dimensional echocardiographic criteria for pericardial tamponade and constriction are as follows: a) during diastole, indentation or collapse of the right ventricular free wall is shown. This collapse is best noted in the right ventricular outflow tract, where the free wall is most compressible.[61] The right ventricular collapse is a reliable echocardiographic sign in diagnosis of hemodynamic compromise with pericardial effusion.[62–64] b) collapse of the right atrium in diastole.[64–67]

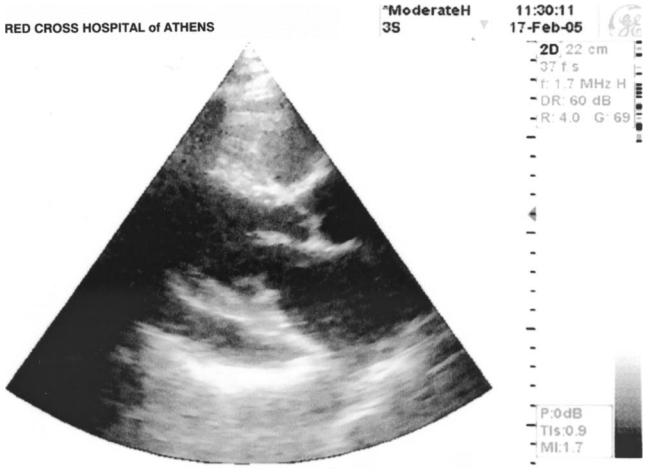

Figure 2. Filling phase of diastole (two-dimensional echocardiography). Open mitral valve, closed aortic valve, left atrium, and left ventricle are presented. Pericardial effusion is seen to surround both left atrium and left ventricle.

disease. One week after admission, a left temporal artery biopsy was performed (length of fragment, 3 cm), showing histopathologic findings compatible with those of giant cell arteritis (Figure 3, Figure 4). Specifically, intense fibromuscular thickening in the artery's intima associated with either nonspecific chronic inflammatory infiltration or with giant cells surrounding the destroyed internal elastic membrane were revealed. In addition, minimal chronic inflammation was found throughout the arterial wall up to the external elastic membrane including perivascular chronic inflammation of vasa vasorum.

Eleven days after admission, treatment was started by oral administration of methylprednisolone (Medrol tablets, 16 mg) at a dosage of 42 mg/day for more than 2 months. Two tablets were administered at 9 AM and one tablet at 6 PM. The patient's clinical condition rapidly improved within a few days. She was discharged from the hospital after completion of 3 weeks' treatment and significant improvement of her clinical condition. The rationale for this relatively long hospital stay involves the relatively long period between onset of the disease and initiation of treatment (approximately 7 weeks), the presence of pericardial involvement, and the fact that this was the time required for complete remission of symptoms.

Six weeks after discharge, the patient's ESR and CRP values were normalized (25 mm/h and <0.5 mg/dL, respectively) and a new two-dimensional echocardiogram showed no detectable pericardial effusion. At this time, a tapering dosage regimen was started by 2 mg reduction of methylprednisolone every 15 days.

Eight and a half months after the start of curative treatment, a thoracic computed tomographic scan was normal with no signs of pericardial effusion, hemoglobin and liver function test values had been normalized (Hb, 12.4 g/dL; γGT, 47 U/L; ALP, 170 U/L), and the dosage was finally fixed at 16 mg daily by mouth.

One year after admission, the patient was in a very good clinical condition, having a normal ESR value (20 mm/h) under the maintenance dosage (16 mg) of methylprednisolone.

Discussion

Giant cell arteritis, which is included among vasculitides, is rarely manifested as a systemic panarteritis associated with ischemic symptoms in the extremities, the heart, the central nervous system, the peripheral nervous system, and the abdominal and pelvic viscera.[20]