Case Report

Pleural Effusion as a Presenting Manifestation of Giant Cell Arteritis

H. GUR***, M. EHRENFELD***, E. IZSAK*

Summary
Pulmonary involvement is rare in giant cell arteritis (GCA). Only a few cases have been reported, manifested by interstitial infiltration, pulmonary nodules, pulmonary artery vasculitis, and granuloma formation. Moreover, only 3 previous cases of pleural effusion associated with GCA have been described. Herein we report a 67-year-old woman with biopsy-proven temporal arteritis, presented with prolonged fever, weight loss, cough and pleural effusion. ELISA test for the presence of anti-proteinase-3 antibodies was negative. The importance of the anti-neutrophil cytoplasmic-antibodies (ANCA) examination in the differential diagnosis from other vasculitides with pulmonary involvement is discussed.

Key words Temporal Arteritis, Pulmonary Involvement.

INTRODUCTION

Giant cell arteritis (temporal arteritis; GCA) is a systemic vasculitis of medium and large arteries, affecting predominantly the aortic branches to the head and neck (1). Temporal arteritis was first described by Horton in 1932, and is classically presented with the combination of polymyalgia rheumatica, headache, and manifestations of systemic illness (fever, anaemia, anorexia, malaise, weight loss), in an elderly patient (2). In addition, involvement of cranial arteries has resulted in other manifestations such as scalp tenderness and necrosis (3), blindness, diplopia, amaurosis fugax, jaw claudication (4), acute lingual ischaemia (5), and aortic arch syndrome (6).

In about 8% of patients, however, nonclassical extracranial organ involvement has been described including coronary vasculitis, pericarditis and myocarditis (7), peripheral neuropathy and sensorineural hearing loss (8-10), abnormal liver function tests and histology (11), giant lymph node hyperplasia (12), renal arteritis (13), and presentation with pyrexia of unknown origin (14).

Pulmonary involvement in GCA has been rarely described (7, 15-18). In the few reported cases clinical findings included cough, hemoptysis, pulmonary or interstitial infiltrates and pulmonary effusions. In some cases histology was available, revealing peribronchial and alveolar interstitial granulomata formation (15, 19), and pulmonary vasculitis (20).

Recently, it has been suggested that overlapping features of GCA and Wegener's granulomatosis (WG) do occur in some patients (21-23). In fact, one can argue that some, if not all of the reported cases of GCA with lung involvement are form frustum of WG, a systemic necrotizing granulomatous vasculitis, specifically associated with the presence of autoantibodies directed against the neutrophil serine protease proteinase-3 (24).

In this report we present a case of a biopsy proven GCA presented with pulmonary effusion and fever. ELISA test for proteinase-3 was negative, and the patient promptly responded to corticosteroid administration, further confirming the diagnosis of GCA. To our knowledge, this is the first reported case of proven lung involvement in GCA, in which other vasculitides involving the lung were ruled out by using a test for anti-neutrophil cytoplasmic antibodies (ANCA).

CASE REPORT

A 67-year-old woman was admitted for fever, cough, night sweats, and weight loss which had started 2 weeks prior to admission. Her past history was unremarkable. On physical examination she looked pale and wasted. The temperature was 38.1°C, the blood pressure was 130/80 mmHg, the pulse rate was 88/min, and the respira-

From the Department of Medicine C* and the Rheumatology Unit**. The Chaim Sheba Medical Center. Tel Hashomer 52621, Israel.
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The findings were limited to dullness and reduced alveolar breathing over the left lower thorax, accompanied by few audible inspiratory rales. There were neither signs of purpura, rash, oral ulcers, ocular inflammation, arthritis, lymphadenopathy, hepatosplenomegaly, nor scalp or temporal tenderness. Laboratory investigation revealed a sedimentation rate of 130 mm per hour (Westegren), haemoglobin concentration of 9 gr/dL, and a thrombocytosis of 617,000/μL. Automated serum chemistry profile was normal except for mild transient disturbances in liver function tests. The albumin was 3 gr/dL and the globulin was 3.7 gr/dL. Protein electrophoresis showed a slight increase in the α2 and the β2 globulin fractions. Urine, blood, sputum, and gastric fluid aspirate cultures were negative. The anti-nuclear-antibody and the latex fixation test for RF were negative, as well. The serum complement levels were slightly increased (C3 - 2.1 gr/L, C4 - 0.5 gr/L), and the C reactive protein was 15.4 mg/dL (normal values: 0 - 0.8 mg/dL). Bone marrow biopsy was nonspecific, revealing a slight hypercellularity. Gastrointestinal studies, including barium enema and gastrosopy revealed only a small diaphragmatic hernia. Radiological studies of the chest demonstrated left pleural effusion (Fig. 1). Computed tomography of the chest and the abdomen confirmed the presence of a pleural effusion associated with a small pulmonary infiltrate. Technetium and gallium scans indicated only diffuse osteoarthritic changes. Echocardiogram was normal, and pulmonary scans ruled out the possibility of pulmonary thromboembolism. Aspiration of the pulmonary effusion revealed a transudate (2.7 gr/dL protein and LDH 94 IU/ml, both less than 50% of serum values) with few mononuclear cells. The tuberculin test (PPD) was borderline positive. The ELISA test for detecting anti-neutrophil-cytoplasmic-antibodies (c-ANCA), using an anti-proteinase-3 antibodies (PR3-

![Fig 1: Chest radiogram of the patient taken on admission. A left side pleural effusion with a small infiltrate is shown.](image)

ANCA) immunoassay kit (Euro-Diagnostica AB, Malmo, Sweden), as previously described (25), was negative. Therapeutical trials of parenteral broad spectrum antibiotics and a three week anti-tuberculosis treatment (isoniazide, rifampicin and pyrazinamide) were of no avail. Temporal artery biopsy revealed a macroscopically inflamed artery. Microscopical examination showed

![Fig 2: Histological findings of the temporal artery biopsy. A. Lymphoproliferative inflammatory infiltrate involving the artery wall (x 250). B. Giant cell reaction within the area of the fragmented internal elastic membrane. (H&E x 100).](image)