Case Report

Relapsing Polychondritis with Castleman-like Lymphadenopathy: a Case Report

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Summary Relapsing Polychondritis (RP) is a systemic disorder characterized by an inflammatory process involving predominantly cartilaginous structures and the cardiovascular system. Lymphadenopathy is a very uncommon finding of RP. We report on a patient affected by RP presenting with lymphadenopathy of Castleman-like type quickly responsive to corticosteroids. The bronchial involvement and the evolution of the inflammatory process in a 3-year follow-up has been documented by computed tomography of the chest.

Key words Relapsing Polychondritis, Castleman-Like Lymphadenopathy

INTRODUCTION

Relapsing polychondritis (RP) is an infrequent episodic systemic disorder characterised by recurrent, widespread, potentially destructive inflammatory lesions involving cartilaginous structures, the cardiovascular system and organs of the special sense, such as eyes and ears (1). Lymphadenopathy is a very uncommon finding in RP which has been observed in only one patient by McAdam et al (2). Histologic changes of the lymph node similar to those found in Castleman’s disease have been associated with a variety of pathologic states including autoimmune diseases (3).

We report a case of RP with Castleman-like lymphadenopathy at the onset of the disease. A detailed description of the bronchial findings is provided since upper airway involvement represents a poor prognostic factor in patients affected by RP (2) and early recognition of the process may be useful to improve the clinical outcome.

CASE REPORT

A 50-year-old man was in good health until January 1991, when he began to complain of persistent dry cough. In April 1991, the patient developed fever, asymmetrical oligoarthritis, bilateral conjunctivitis and general malaise. At home, he was unsuccessfully treated with antibiotics. Two weeks before admission, the clinical picture was complicated by dyspnoea with retrosternal chest discomfort.

On admission, in May 1991, physical examination revealed a febrile (38°C) patient with laterocervical and supraclavicular lymphadenopathies, expiratory stridor and swelling of his left elbow and his right knee and ankle.

Laboratory investigations showed an elevation of the erythrocyte sedimentation rate (120 mm/h, Westergren), C-reactive protein (28 mg/dl) (nv < 1.2), α-2-globulins (15.1%) (nv < 11), fibrinogen (904 mg/dl) (nv < 400), ferritin (1,040 U/L) (nv < 400) and platelet count (576,000/mm3). In addition, peripheral blood leukocytosis (WBC: 13,800/mm3) with absolute neutrophilia (11,600/mm3) was present. The blood chemistry revealed raised values of alanine aminotransferase (100 U/L) (nv < 50), γ-glutamyltranspeptidase (139 U/L) (nv < 60), alkaline phosphatase (283 U/L) (nv < 200) and low serum iron (28 μg/dl). Serum electrophoresis showed a mild hyper-γ-globulinaemia (21.1%) (nv < 18%) with a polyclonal pattern and increased levels of IgG (2,100 mg/dl) (nv < 1,800) and IgA (773 mg/dl) (nv < 450). Rheumatoid factor, antinuclear, anti-native DNA, anti-mitochondrial, anti-gastric parietal cells and anti-thyroid antigens autoantibodies were negative, whereas anti-smooth muscle autoantibodies were positive at a titre of 1:160. C3c complement was elevated up to 141 mg/dl (nv < 120) and C4 was found within normal range (48 mg/dl). Thyroid function was normal. An extensive search for infectious pathogens, including hepatitis B and C viruses, Epstein-Barr
virus, human immunodeficiency virus and Toxoplasma Gondii was negative.

Chest x-ray showed a mediastinal enlargement and a narrowing of the left main bronchus without evidence of abnormal calcifications. Computed tomography (CT) of the chest revealed an enlargement of the superior mediastinal lymph nodes and the narrowing of the left main bronchus on expiration; an enlargement of pretracheal lymph nodes was also observed. CT scan of the abdomen did not reveal lymph node enlargement but showed features consistent with fatty liver. Respiratory function tests, including flow volume studies, showed a severe airflow obstruction with normal transfer factor.

Skeletal scintigraphy using 99mTc-methylendiphosphonate (99mTc-MDP) revealed intensive uptake by the cartilaginous part of the second right rib and by the larynx. A bone scintigraphy, repeated one year later, confirmed these findings showing a further increased uptake by the larynx. Total body scintigraphy with Gallium-67 citrate was negative. Electrocardiography and two-dimensional echocardiography were found to be normal and no changes were observed in two later controls in November 1993 and May 1995.

On the basis of the clinical picture and the results of radiological investigations, biopsies of the second right costal cartilage and right supraclavicular lymph node were performed. Histologic examination of the costal cartilage showed diffuse loss of basophilic staining of the matrix, sequestered islands of degenerating cells (Figure 1) as well as deposition of calcified material. Lymphocyte and plasma cell infiltrates were observed in the perichondrial tissue in which immunohistologic characterization using monoclonal antibodies revealed a prevalence of T lymphocytes carrying the CD45RO antigen. Lymph node biopsy demonstrated a normal nodal architecture with marked follicular hyperplasia (Figure 2) and prominent interfollicular plasmacytosis (Figure 2-inset) with normal ratio of $k$ and $\lambda$ light chains expression indicating a polyclonal pattern; these findings were regarded as consistent with Castleman-like lymphadenopathy (3). Bilateral percutaneous bone marrow biopsies revealed diffuse hyperplasia, reactive plasmacytosis and eosinophilia without evidence of lymphomatous involvement of the marrow.

A diagnosis of RP was made (2) and corticosteroid therapy with prednisone 50 mg daily was started which resulted in rapid defervescence, remission of the oligoarthritis, reduction of superficial lymphadenopathies and improvement of the dyspnoea and laboratory parameters of inflammation. A chest CT scan, performed in June 1991, showed reduction of both the number and volume of the mediastinal and pretracheal lymph nodes. The follow-up of the patient was characterized by several episodes of bilateral auricular chondritis and conjunctivitis.