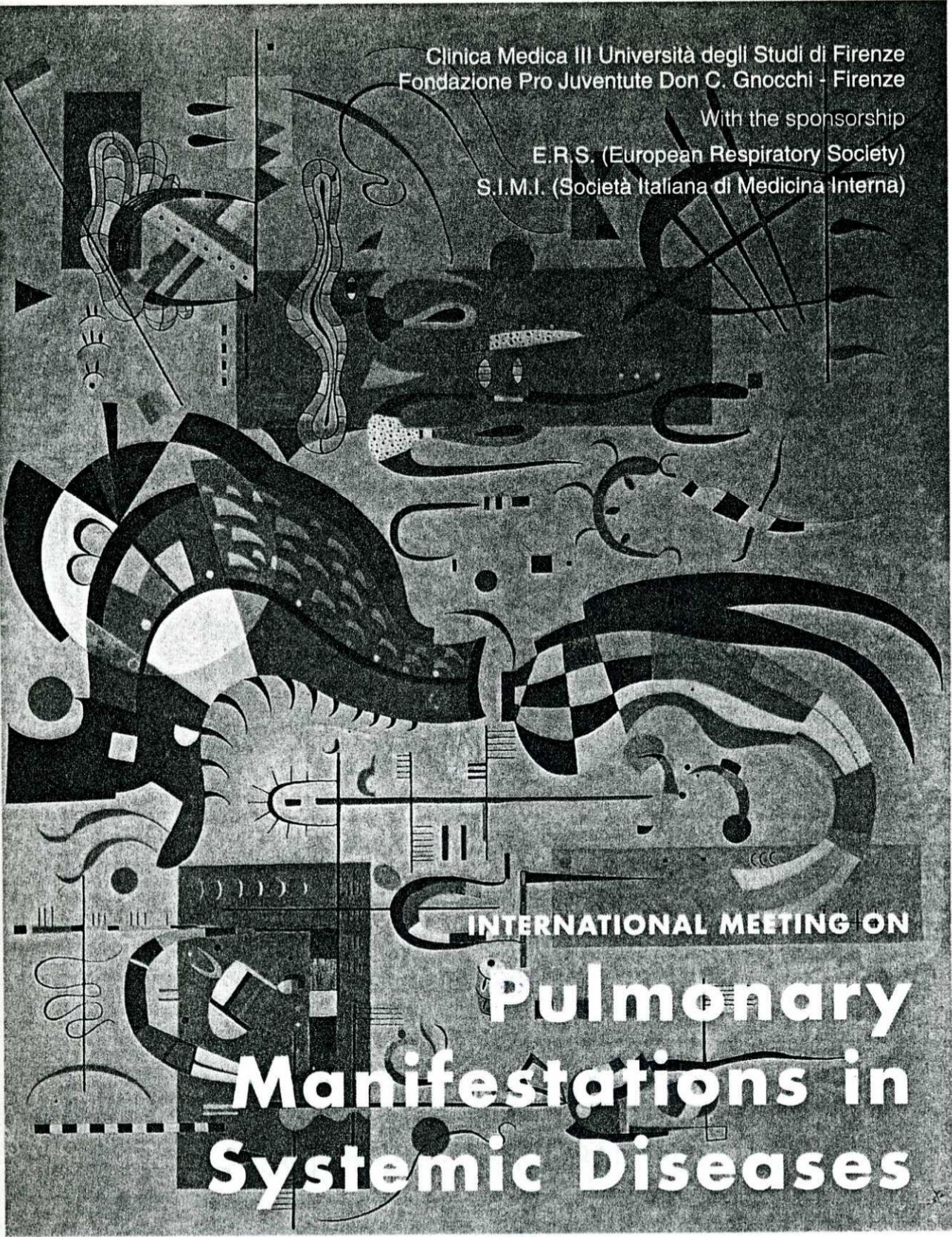


Clinica Medica III Università degli Studi di Firenze
Fondazione Pro Juventute Don C. Gnocchi - Firenze

With the sponsorship

E.R.S. (European Respiratory Society)

S.I.M.I. (Società Italiana di Medicina Interna)



INTERNATIONAL MEETING ON
**Pulmonary
Manifestations in
Systemic Diseases**

Florence (Italy) January 25th - 28th 1995

Scientific Programme

INTERNATIONAL
MEETING ON

PULMONARY MANIFESTATIONS IN SYSTEMIC DISEASES

Palazzo degli Affari,
Florence, Italy
25th-28th January, 1995

PRESIDENT
Mario Ricci (I)

ORGANIZERS
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UNDER THE PATRONAGE OF

Regione Toscana
Comune di Firenze - Assessorato alla Sanità
Università degli Studi di Firenze

SCIENTIFIC PROGRAMME

ABSTRACTS

LUNG IMAGING IN WEGENER'S GRANULOMATOSIS

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Wegener's granulomatosis is a systemic granulomatous vasculitis (small and medium-sized arteries and veins) tending to necrosis, of unknown aetiology, which may affect, singly or simultaneously a variety of organs and apparatuses (upper and lower airways, eye, kidney, skin, central nervous system, heart, lung, etc...). Pathogenesis may be traced to cell-mediated immunity or immune complex-mediated mechanism. The disease is extremely variable on onset and capricious in the evolution. A form involving only the lung, which may be asymptomatic or it may show with worsening dyspnoea, cough, haemoptysis, chest pain, fever, has been described. The radiograph anomalies (various-sized multiple nodules, solitary nodules, lobar opacities, pleural effusion and thickening, combined anomalies) are variable and fluctuating, with improvement or worsening of the involved areas. The case shown, indicative of a form of Wegener's granulomatosis, limited to the lung, lays the stress on the lung imaging that, though aspecific and capricious in the evolution, represents a fundamental moment in the diagnostic course of the disease, mainly if it is supported by specific laboratory tests (phlogosis parameters, C-ANCA, ESR, C-reactive protein, etc...). Through an aimed and precocious therapy (corticosteroid and cytotoxic drugs) these tests permit long remissions with positive reflections on the *quoad vitam* prognosis of such patients.

We observed a 34-year patient, no-smoker, with fever (39°C), chest pains and cough with haemoptysis, worsening dyspnoea and anorexy. The chest radiograph, at hospitalization, showed a modest, faded thickening in the basal left lungfield. On auscultation, harsh breathing with fine crackles on the left basal lungfield could be heard. On the assumption of an infective aetiology, an antibiotic therapy was started which didn't improve the clinical picture; on the fourth day symptomatology showed a worsening with continuous fever, severe respiratory failure ($pO_2=60\text{mmHg}$, $pCO_2=40\text{mmHg}$, $pH=7.35$), worsening of the chest radiograph with appearance of small multiple pulmonary thickenings to be found in the mid-basal right zone. Computed tomography permitted clear visualization of the lung parenchyma with disomogeneous, multiple, confluent, partly excavated infiltrates especially in the lower zones and middle lobe. The infiltrates showed several air bronchograms and a modest layer of bilateral pleural effusion together with absence of hilar and mediastinal adenopathies. The laboratory tests showed an increase in the ESR (1st hour=93), in C-reactive protein, leukocytosis, negativity of the various serological specific tests towards probable pathogens (HIV, Legionella, Mycoplasma, Rickettsia etc...), no abnormality in the urine, modest increase in transaminases. Haemoculture and urine culture were repeatedly negative. The bronchoscopy was negative as well as the search for pathogens in the broncho-aspirate fluid. Taking into account the clinical laboratory picture, we tested the level of C-ANCA (anti-neutrophils cytoplasm antibodies) in the blood. The positivity of these ones has been determining not only for Wegener's granulomatosis diagnosis and for a specific corticosteroid and cytotoxic therapy (prednisone 1mg/kg/die + cyclofosamide 2mg/kg/die) but also for following its evolution. Such therapy has determined a progressive clinical improvement attested by laboratory parameters (C-ANCA, ESR, etc...), by periodic chest radiograph checks, by lack of fever. At discharge, a month later, chest radiograph showed fibrotic and atelectatic lines with small areas of fibrotic thickening in both the lungfields which have replaced the infiltrates. The patient is being tested with periodic checks and posology adjustment in order to follow the course of the disease.