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European Journal of Radiology 66 (2008) 27–30

EJR
EUROPEAN JOURNAL OF RADIOLOGY

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Chest CT findings in patients with inflammatory myopathy and Jo1 antibodies

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Received 11 October 2006; received in revised form 6 May 2007; accepted 21 May 2007

Abstract

Thoracic high-resolution computed tomography scans (HRCT) of 17 patients with inflammatory muscle disorders (IMD) and positive Jo1 antibodies were retrospectively reviewed regarding presence, extension, and distribution of pathological findings. Abnormal findings were found in 14 (82.3%) patients. The predominant CT abnormality was ground glass attenuation, which was present in seven patients (41.1%), having a bilateral and diffuse distribution. In general, lesions tended to appear in the lower lobes and more specifically in the lung bases. Interlobular septal thickening was found in six patients (35.3%); it was seen in the upper and lower lobes with peripheral distribution and bilateral localization in five out of six patients. Bronchiectases, reticular opacities, and honeycombing were found in six patients (35.3%). Air space consolidation was seen in about 17% of the patients. Lung involvement is a frequent feature of IMD patients with positive Jo1 antibodies and its most common radiological pattern is that of nonspecific interstitial pneumonia.

Keywords: Inflammatory muscle disorders; Anti-synthetase syndrome; Interstitial lung disease; Lung involvement; Chest CT

1. Introduction

Anti-Jo1 myositis or anti-synthetase syndrome belongs to inflammatory muscle disorders (IMD) and is characterized by various manifestations including myositis, mechanic's hands, arthralgias, fever, Raynaud's phenomenon, and interstitial lung disease (ILD) [1,2]. Among these, ILD is the most common manifestation, found in over 70% of patients. Overall, patients with Jo1 antibodies account for about 30% of myositis patients and about 80% of myositis patients with ILD [3]. From a clinical standpoint, patients with anti-synthetase syndrome usually have a more aggressive disease course than other IMD patients, with pulmonary complications dominating the clinical picture [4]. Of the remaining anti-synthetases, anti-PL12 is found in 2–5% of patients with PM/DM and this can be associated with isolated pulmonary fibrosis [8,21].

The ILD associated with the anti-synthetase syndrome has various histopathological features including bronchiolitis obliterans organizing pneumonia (BOOP), nonspecific interstitial pneumonia (NSIP), and usual interstitial pneumonia (UIP); diffuse alveolar damage with rapidly progressive lung disease may occasionally occur [1,5–8]. On high-resolution CT imaging, these entities may have a characteristic appearance, which in conjunction with the clinical manifestations and positive Jo1 antibodies is used to make an etiologic diagnosis of the ILD without the need of a lung biopsy.

The aim of the present study was to describe the high-resolution computed tomography (HRCT) findings of the ILD in a homogeneous group of patients with positive Jo1 antibodies. Unlike similar previous studies that included a mixed cohort of IMD patients, we focused only on IMD patients with positive Jo1 antibodies.

2. Materials and methods

We retrospectively assessed 17 patients with Jo1 antibodies, referred to the Rheumatology Clinic of the Laiko University

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doi:10.1016/j.ejrad.2007.05.017

Hospital, in Athens, between 1992 and 2005. The presence of the anti-Jo1 antibodies was confirmed by counter immune electrophoresis. The patient group consisted of eight men and nine women, with a mean age at disease onset of 47.3 ± 14 years. All patients fulfilled the Bohan and Peter [9] criteria for the diagnosis of IMD. Ten out of 17 patients were classified as polymyositis (PM) and the remaining seven patients as dermatomyositis (DM); two patients in the DM subgroup had no clinical and laboratory evidence of myositis at disease onset (amyopathic dermatomyositis). Two patients had anti-Ro antibodies and one PM patient had tRNA autoantibodies. The above are known associations of the anti-synthetase syndrome and confirm the pleomorphic nature of its presentation [10–12].

All patients underwent HRCT of the lungs at various stages of their disease and the CT films were obtained and retrospectively examined by two experienced radiologists. For CT analysis, we recorded and analysed presence, location and distribution of various radiological patterns including ground glass attenuation, reticular opacity, consolidation, traction bronchiectasis and honeycombing. Two radiologists (AZ, KC) assessed independently the CT findings and in case of disagreement, final interpretation was achieved by consensus. Ground glass opacities were defined as areas with increased attenuation, which did not obscure pulmonary vessels or bronchial walls [17]. Consolidation was defined as an area of increased attenuation with obscuration of the parenchymal airways and vessels [17]. Reticular opacities were defined as linear opacities forming a net-like pattern. Honeycombing was recognized on the basis of small cystic spaces of less than 1 cm in diameter with thickened walls [17]. Traction bronchiectasis was defined as dilatation or presence of abnormal bronchial wall contour, usually within areas of honeycombing [17]. Interlobular septal thickening was defined as short lines, usually in the subpleural space, running perpendicularly to the pleural surface and representing thickened interolobular septa [17]. Mosaic pattern was considered when heterogeneous lung attenuation with a lobular or multilobular distribution was noted. Predominant distribution was assessed as being upper or lower if the lesions were primarily localized within the upper or lower lobe, respectively. Architectural distortion was considered present when interlobar fissures and hila were displaced or when bronchi or vessels were distorted. Presence of additional abnormalities such as pleural effusions was also recorded.

3. Results

The main clinical characteristics of patients with ILD and positive anti-Jo1 are shown in Table 1. Among the 17 patients whose CT scan films were evaluated, 14 patients were found to have abnormal findings (82.3%). The median time from disease onset to CT scan was approximately 2 years. The predominant CT abnormality (Table 2) was ground glass attenuation (Fig. 1, Table 2), which was present in seven patients (41.1%). These lesions were bilateral and predominantly distributed in the lower lobes. Interlobular septal thickening (Fig. 2) was found in six patients (35.3%), mainly in the upper lobes with peripheral distribution and bilateral localization in four out of five patients. The next most common abnormalities (35.3%), which were found in six patients, were bronchiectases, reticular opacity, and honeycombing (Fig. 3). In all cases, bronchiectases were bilateral with a lower lobe predilection and central distribution. Reticular opacities and honeycombing were similarly bilateral and in most cases had a diffuse distribution. Architectural distortion was noted in three patients (17.6%), with a diffuse and bilateral distribution pattern. Emphysema or air space consolidation was also detected in three patients (17.6%). Other findings that were detected in two patients (11.8%) included small nodular opacities, and pulmonary artery enlargement. Finally, thin walled parenchymal cysts, mosaic pattern, air trapping, and pleural effusion were detected in one patient (5.8%) each.

Table 1
Clinical and laboratory findings of the 17 IMD patients with positive Jo1 autoantibodies

	Patients ($n = 17$)
Sex (M/F)	8/9
Age	52.7 ± 16
Age at the disease onset	47.3 ± 14
Disease subtype, PM/DM	10/7
Disease duration since chest CT evaluation, years median	2
Arthralgias	12 (70.5%)
Raynaud's phenomenon	12 (70.5%)
Skin rash	11 (64.7%)
Mechanic's hands	7 (41%)
Muscle weakness	14 (82%)
Cough	6 (35%)
Dyspnea on exertion	8 (47%)
Elevated muscle enzymes	17 (100%)

4. Discussion

This retrospective study was carried out to describe the HRCT findings in anti-Jo1 positive patients, and, to our knowledge, is

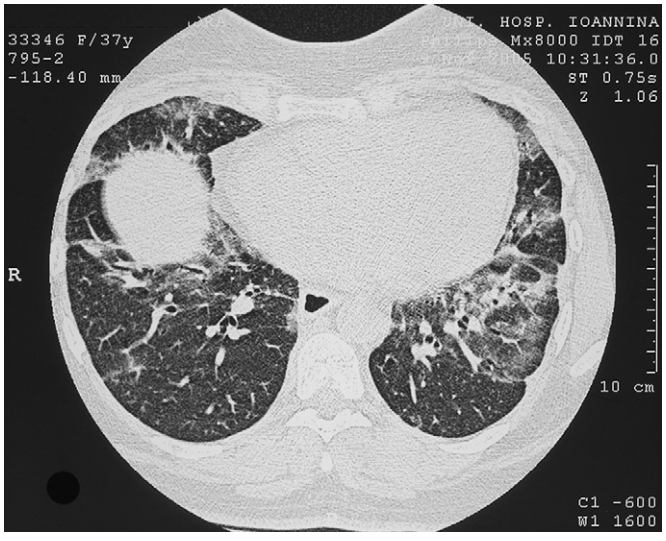

Fig. 1. Characteristic high-resolution CT in a female patient with Jo1 antibodies showing bilateral lower lobe ground glass lesions.