CASE REPORT

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New approach to Caplan's syndrome

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Abstract: A combination of rheumatoid arthritis and pneumoconiosis is observed in 2-6% of cases of diseases among people occupationally exposed to either the crystal or fibrous form of silica. Chest radiology of patients affected by this syndrome shows multiple, rounded opacifications, usually 0.5-5.0 cm in diameter, as well as positive serologic reactions to the presence of the rheumatoid agent. The authors analysed a comprehensive material of 187 cases with the diagnosis of pneumoconiosis in grinders/casting cleaners with an homogenous source of exposure to fibrous dust, and one observed case of Caplan's syndrome in a female who worked as a crane operator. The diagnosis was made based on occupational interview, evaluation of X-ray of the chest and bones, and serologic response typical for rheumatoid arthritis. Pneumoconiosis preceded rheumatoid arthritis by several years. Within 7 years of ambulatory follow-up, the progression of radiological changes in the lungs remained moderate, both with respect to type and density of the opacities.

Key words: Caplan's syndrome, collagen pneumoconiosis, rheumatoid arthritis

BACKGROUND

The co-occurrence of pneumoconiosis and rheumatoid arthritis is a relatively rare phenomenon. The specific character of pneumoconiosis was first described by Caplan in 1953 in a miner working in an anthracite mine. Caplan's syndrome occurs in silicosis and asbestosis and concerned 2-6% of cases [2]. A distinctive feature of this form of pneumoconiosis is the presence in the radiological image of multiple, rounded opacifictions, 0.5-5 cm in diameter, with a tendency to central necrosis and calcification. Positive serologic response to the presence of the rheumatoid agent is observed in patients [1, 3, 4, 5-7].

We analysed comprehensive material concerning pneumoconiosis diagnosed in grinders/casting cleaners, with an homogenous source of exposure to fibrous dust, and one observed case of Caplan's syndrome, based on occupational interview, evaluation of X-ray of the chest and bones, and serologic response typical of rheumatoid arthritis. Considering a specific radiological image of pneumoconiosis in the above-mentioned syndrome, which was disproportionate to the clinical course of the disease, a case is presented which is relatively rarely noted in occupational pathology and radiology.

CASE PRESENTATION

A female aged 49, employed for 22 years at the workpost of a crane operator at an iron foundry section, was exposed to the inhalation of industrial dust containing free silica in concentrations exceeding the maximum allowable standard (MAC) for industrial dust. The complaints reported covered quick fatigue, post-effort dyspnea, attacks of dry cough, chest pain, and additionally complaints concerning the joints typical of rheumatoid arthritis. X-ray of the chest and serologic tests enabled diagnosis of the disease. The patients is being cared for by the Rheumatology Outpatient Department and the Outpatient Department for Occupational Diseases.

Result of high resolution computed tomography (HRCT). cross-sections of the lungs, 2 and 3 mm thick, performed by high resolution tomography, showed the presence of diffused nodular-fascicular opacities, blending especially in the upper zones of the lungs. The cross-sections revealed changes of a type of diffused parenchymal fibrosis, multiple nodular opacities, thickened vascular-bronchial fascicles, focal emphysema, creating an image of so-called 'milk glass', hiluses of vascular properties, and a prominent pulmonary cone of the heart.

Result of rtg of the chest. Multiple micronodular and micromacular darkenings and strip-like fibrosis, most intensive in the upper and central areas, of a character of pneumoconiotic changes.

Perfusion lung scintigraphy. Perfusion disorders in both lungs, with disseminated considerable impairment of blood flow through the lungs, especially the left lobe. Decreased perfusion gradient indicated the presence of pulmonary hypertension.

Result of rtg of hand and wrist joints. Narrowing and blurring of articular spaces of the wrists, with the presence of erosions and destructive changes of joint surfaces (rheumatoid arthritis). Diffused decalcification of the bone parts of the hands and feet.

DISCUSSION

Pneumoconiosis with rheumatoid arthritis is the co-occurrence of two nosological units, rarely observed in occupational pathology.

Rheumatoid nodules showed a slightly different structure from simple silicosis. In the structure of the nodule, a focus of central necrosis was diagnosed, surrounded by a layer of

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macrophages, leukocytes and fibroblasts, and alternately, collagen fibres which presented a relatively characteristic image of the disease. Despite a considerable progression of lung changes in silicosis, in both patients the respiratory and cardiovascular efficiency was sufficient to perform daily life activities [3, 4, 6].

During the 5-7 years of ambulatory observation, the progression of radiological changes in the lungs remained moderate, with the maintenance of normal lung ventilation and perfusion parameters at rest.

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