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2nd Department of Radiology, Skubiszewski Medical University of Lublin

MAREK PASŁAWSKI, WITOLD KRUPSKI, JANUSZ ZŁOMANIEC

Fibrotic changes in high-resolution computed tomography in patients with sarcoidosis

Sarcoidosis is a multi-organ granulomatous disease of unknown cause, that occurs in mediastinal and pulmonary sites in 90% of cases (1, 3, 5, 8). Histologically granulomas are noncaseating, contrary to tuberculosis (4, 8). Parenchymal abnormalities often resolve spontaneously, but they evolve toward pulmonary fibrosis in 20–25% of cases. The presence of pulmonary fibrosis (in stage IV sarcoidosis) on a chest radiography is generally associated with poor pulmonary function and a poor prognosis with increased morbidity and mortality (1, 6, 8). However, the chest radiographic findings of pulmonary fibrosis show poor correlation with histopathology or pulmonary function (1).

The aim of the study is presenting the typical findings of fibrotic changes in high resolution computed tomography in patients with sarcoidosis.

MATERIAL AND METHODS

The material comprises a group of 18 patients with sarcoidosis, in whom HRCT examination was performed. The scanning was performed from lung apices to the level of diaphragm, at full inspiration with patients in supine position. The presence and character of HRCT findings were noted and analyzed. Additional expiratory scans were performed to diagnose air-trapping, and in case of subpleural densities in posterior, dependent lung areas additional scans were obtaining in prone patients positions.

RESULTS

Fibrotic changes were found in the area of confluent nodular lesions in five patients. In these fibrotic areas the bronchiectasis was seen, as well as parenchymal bands and fibrotic masses (Fig. 1). In six patients the reticular pattern was the predominant pathology. Thickened lobular septa did not resolve during the remission of the diseases, suggesting the presence of irreversible, reticular fibrotic changes (Fig. 2). Intensive fibrosis led towards destruction of the lung parenchyma, with honeycombing involving large lung areas (Fig. 3). In the areas of active disease fibrotic changes with traction bronchiectasis were accompanied by the confluent nodular changes and the presence of consolidation (Fig. 4). After resolving of active reversible changes the intensive fibrosis with large bronchiectasis forming the honeycombing pattern with reticular changes were seen in five patients (Fig. 5). In three cases the most intensive changes, both fibrotic and productive (granuloma), were localized in the lower lobe, mimicking the idiopathic pulmonary fibrosis (Fig. 6).

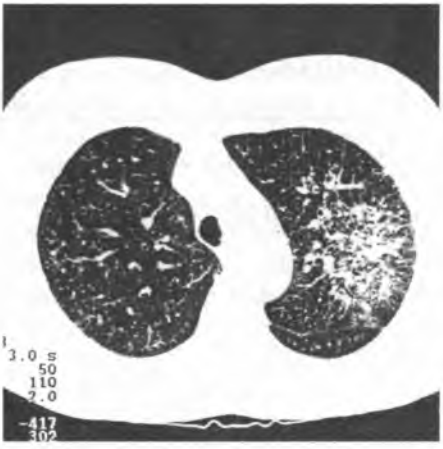

Fig. 1. Fibrotic changes in the left lung in a patient with sarcoidosis

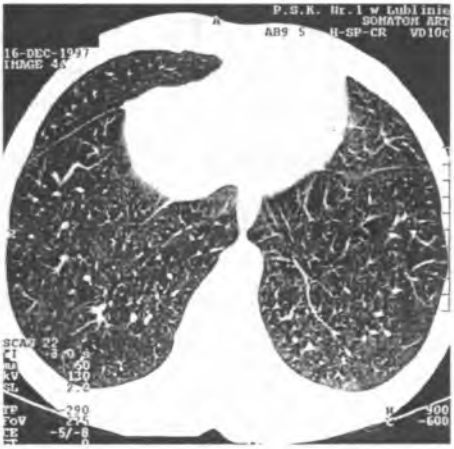

Fig. 2. Linear opacities formed by interlobular septal thickening

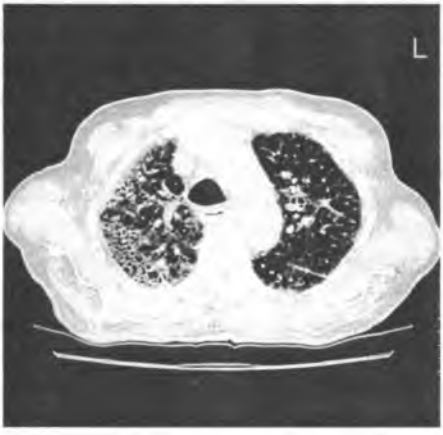

Fig. 3. Distortion of the bronchi resulting from fibrotic changes. Traction bronchiectasis

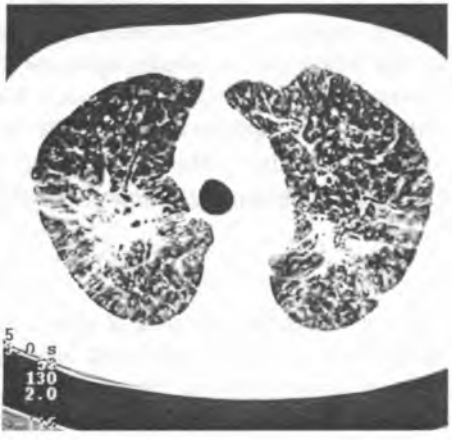

Fig. 4. Honeycombing in the areas of intense fibrosis in the right lung

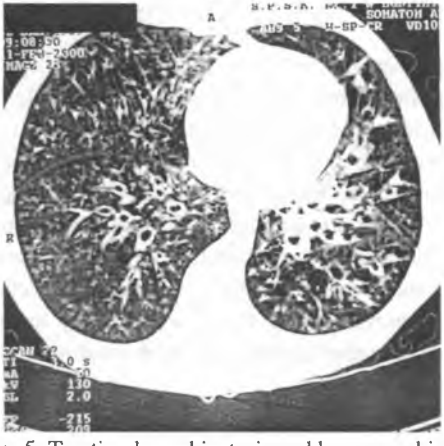

Fig. 5. Traction bronchiectasis and honeycombing as a result of fibrosis in sarcoidosis

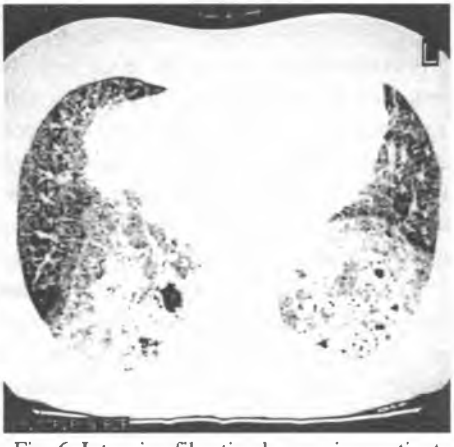

Fig. 6. Intensive fibrotic changes in a patient with sarcoidosis

DISCUSSION

Typical HRCT findings in sarcoidosis include nodules, septal and nonseptal lines, and areas of increased attenuation like ground-glass opacities, parenchymal consolidation and honeycombing areas (3, 6, 8). Nodules are the most common abnormalities detected on high resolution CT in patients with pulmonary sarcoidosis. They are usually 2 mm to 1 cm in diameter and have irregular margin. Nodules occur predominantly in a perilymphatic distribution, adjacent to interlobular septa and subpleurally (6, 7, 8).

Ground glass opacities are also often seen in sarcoidosis, defined as a hazy increase in lung density without obscuration of the underlying vessels. They correlate with the presence of numerous sarcoid granulomas below the resolution of HRCT. Although ground glass opacity usually reflect potentially treatable or reversible disease, in some patients it does not appear reversible and is likely to result from the increase in soft tissue seen in fibrosis. The presence of fibrosis is suggested by the concurrence of traction bronchiectasis and bronchiolectasis (7, 8).

Linear opacities well recognized in sarcoidosis occur less often. Septal lines are more common than polygonal structures formed by interlobular septal thickening. Long irregular linear opacities are also described. These irregular reticular opacities do not appear reversible despite diseases remission, and they are likely to represent early manifestation of fibrosis (7, 8).

Common high resolution CT findings suggesting fibrosis is the presence of distortion, usually shown by posterior displacement of the main or upper lobe bronchus. This sign as well as distortion of the interlobular fissures is likely to indicate loss of lung volume, especially in the posterior segment of the upper lobe (7, 8).

Less commonly honeycombing and formation of cysts or bullae will be evident. Fibrosis may be so marked, that parahilar opacities resembling those seen in progressive massive fibrosis may be seen. They are called conglomerate fibrosis. Severe upper lobe fibrosis may be associated with compensatory emphysema of the lower lobes. Traction bronchiectasis and bronchiolectasis are also considered to reflect fibrosis (2, 7).

Fibrosis due to sarcoidosis can be clearly assigned to one of three categories according to the main CT lesion and its topography (1).

The most frequent pattern is bronchial distortion (may be seen in nearly 50% of patients). May be divided into two categories according to the presence or absence of masses around the proximal bronchi (1, 2). The honeycombing pattern (seen in about 25% of patients) is predominantly peripheral, and restricted to the upper zones (1). The linear pattern is seen in about 25% of patients. Masses are usually not included as separate category, because it is difficult to exclude the potential reversibility in such kind of changes without long follow-up (1, 2).

As fibrosis develops over time, sarcoidosis and idiopathic pulmonary fibrosis increasingly share numerous findings. Both show irregular or nodular septal thickening, irregular interfaces, and traction bronchiectasis. In sarcoidosis, honeycombing is a less frequent feature of irreversible fibrosis as compared with idiopathic pulmonary fibrosis (6).

CONCLUSIONS

The classical HRCT findings in sarcoidosis are well known. But most of them, nodules, ground glass opacities, peribronchovascular thickening or hilar adenopathy are reversible. The irreversible fibrotic changes develop in about 25% of patients. In the early stage they are subtle and invisible on plain radiographs or even on normal CT sections. So the HRCT is necessary to reveal fibrotic changes in patients

with sarcoidosis and to monitor the progression of the diseases. Therefore, the good knowledge of HRCT signs of fibrosis is essential.

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SUMMARY

The aim of the study is presenting the typical findings of fibrotic changes in high-resolution computed tomography in patients with sarcoidosis. Material comprises a group of 18 patients with sarcoidosis, in whom HRCT examination was performed. The scanning was performed from lung apices to the level of diaphragm, at full inspiration with patients in supine position. The presence and character of HRCT findings were noted and analyzed. Additional expiratory scans were performed to diagnose air-trapping, and in case of subpleural densities in posterior, dependent lung areas additional scans were obtaining in prone patients positions. Fibrotic changes were found in the area of confluent nodular lesions in five patients. In these fibrotic areas the bronchiectasis, as well as parenchymal bands and fibrotic masses were seen. In six patients the reticular pattern was the predominant pathology. Thickened lobular septa did not resolve during the remission of the diseases, suggesting the presence of irreversible, reticular fibrotic changes. Intensive fibrosis led towards destruction of the lung parenchyma, with honeycombing involving large lung areas. In the areas of active disease fibrotic changes with traction bronchiectasis were accompanied by the confluent nodular changes and presence of consolidation. After resolving of active reversible changes the intensive fibrosis with large bronchiectasis forming the honeycombing pattern with reticular changes were seen in five patients. In three cases the most intensive changes both fibrotic and productive (granuloma) changes were localized in the lower lobe, mimicking the idiopathic pulmonary fibrosis. The classical HRCT findings in sarcoidosis are well known. But most of them, nodules, ground glass opacities, peribronchovascular thickening or hilar adenopathy are reversible. The irreversible fibrotic changes develop in about 25% of patients. In the early stage they are subtle and invisible on plain radiographs or even on normal CT sections. So the HRCT is necessary to reveal fibrotic changes in patients with sarcoidosis and to monitor the progression of the diseases. Therefore, the good knowledge of HRCT signs of fibrosis is essential.

Zmiany włókniste w tomografii komputerowej wysokiej rozdzielczości płuc u pacjentów z sarkoidozą

Celem pracy jest przedstawienie typowych obrazów zmian włóknistych tomografii komputerowej wysokiej rozdzielczości u pacjentów z sarkoidozą. Materiał stanowiła grupa 18 osób z sarkoidozą, u których wykonano badanie HRCT. Badanie TK wykonano od szczytów płuc do poziomu przepony, na pełnym wdechu u pacjentów ułożonych na wznak. Obecność i charakter zmian w HRCT była oceniana i analizowana. Dodatkowo wykonywano skany wydechowe w celu wykrycia pułapek powietrznych, a w przypadkach stwierdzenia patologii w tylnych, zależnych obszarach płuc wykonywano dodatkowo badanie u pacjentów ułożonych na brzuchu. Zmiany włókniste w obszarach zlewających się zmian guzkowatych były stwierdzone u pięciu pacjentów. W obszarach tych widoczne były rozstrzenia oskrzeli jak również pasma miąższowe i masy włókniste. U sześciu pacjentów obraz siateczki był dominujący. Pogrubiałe przegrody zrazikowe nie uległy wchłonięciu w okresie remisji choroby, sugerując obecność nieodwracalnych zmian włóknistych. Intensywne włóknienie prowadziło do destrukcji miąższu płuc, tworząc duże obszary plastra miodu. W obszarach aktywnego procesu zmianom włóknistym z rozstrzeniami oskrzeli z pociągania towarzyszyły zlewające się zmiany guzkowe i konsolidacje. Po wchłonięciu się zmian odwracalnych intensywne zwłóknienie z dużymi rozstrzeniami oskrzeli tworzącymi obrazy plastra miodu oraz zmiany pasmowate i siateczkowate były widoczne u pięciu pacjentów. W trzech przypadkach najbardziej intensywne zmiany włókniste i wytwórcze (ziarniniaki) lokalizowały się w płatach dolnych, naśladując idiopatyczne zwłóknienie płuc. Klasyczne objawy sarkoidozy w HRCT są dobrze znane. Ale większość z nich, guzki, zacienienia szkła mlecznego, zagęszczenie okołooskrzelowonaczyniowe czy adenopatia wnękowa, są odwracalne. Nieodwracalne zmiany włókniste pojawiają się u 25% pacjentów. We wczesnych stadiach są subtelne i niewidoczne na zdjęciach przeglądowych płuc, a nawet na klasycznych przekrojach TK. Badanie HRCT jest konieczne do wykrycia zmian włóknistych u pacjentów z sarkoidozą oraz do monitorowania postępu choroby. Dlatego dobra znajomość objawów zwłóknienia w HRCT jest niezwykle istotna.