The diagnosis of sarcoidosis pleurisy by medical thoracoscopy: Report of three cases

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ÖZET

Sarkoidozis plörezi tanısında medikal torakoskopi: Üç olgu bildirimi


Anahtar Kelimeler: Plevra, sarkoidozis, torakoskopi.

SUMMARY

The diagnosis of sarcoidosis pleurisy by medical thoracoscopy: Report of three cases

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Sarcoidosis is a multisystem disorder, characterized by noncaseating granulomas in the organs involved. The most affected organs in sarcoidosis are lung, lymph nodes, skin, eyes, and liver. Although the reports of pleural sarcoidosis have been increasing in the literature, the incidence of this condition is not well-known. Nevertheless, a recent article reports that the incidence of pleural effusion is in a range of 0.7-10% of all sarcoidosis cases (1). The exact diagnosis of pleural sarcoidosis can be made by showing noncaseating granulomas in pleural tissue samples. However, it is not easy to prove pleural involvement caused by sarcoidosis (2,3). The value of medical thoracoscopic intervention in the diagnosis of pleural sarcoidosis remains to be established. In this presentation, we aimed to discuss medical thoracoscopic findings in three cases of sarcoidosis pleurisy.

**CASE REPORTS**

**Case 1**

A 59-year-old man was admitted to our hospital (Spedali Civili Brescia) with a mild dyspnea under effort for several weeks. On physical examination, diminished breath sounds, and dullness to percussion at the left base were obtained. Chest X-ray on his admission showed bilateral pulmonary nodules and left moderate-sized pleural effusion. Thoracic computerized tomography (CT) scan revealed multiple round nodules in both lungs intraparenchimally, large pleural effusion on the same side, and multiple lymph nodes in precardinal and aorto-pulmonary window position.

Pulmonary function test results were consistent with a mild restrictive defect. Tuberculin skin test measurement was negative. Serum calcium and angiotensin converting enzyme levels were also within normal limits. Electrocardiogram was normal.

In fiberoptic bronchoscopy, no abnormality was seen with the exception of chronic bronchitic picture on the bilateral bronchial walls. Transbronchial biopsies were taken into the inferior segment of the lingula and were reported as a noncaseating granulomatous inflammation that is compatible with pulmonary sarcoidosis.

Thoracentesis revealed clear, serous fluid with an exudative characteristic. Pleural fluid cytologic examination showed predominately lymphocytic inflammatory cells and nonneoplastic microscopic features. On medical thoracoscopy, the parietal pleura showed multiple nodular anthracosis, more intense near the costophrenic border, and phlogistic areas with irregular pleural thickening (Figure 1). The visceral pleura showed no significant alterations, but so-
me anthracotic nodulations were present. Multiple parietal pleural biopsies taken by medical thoracoscopy confirmed the diagnosis of sarcoidosis pleurisy as histopathological. Results of mycobacterial and fungal cultures of pleural tissue were negative. Ocular and dermatological examinations showed no sarcoidosis involvement. Prednisone therapy, 40 mg daily, was started, and gradually tapered of this dosage with a clinical improvement.

Case 2
A 68-year-old woman was admitted to the hospital with complaints of right pleuritic chest pain and mild exertional dyspnea. Her physical examination findings were unremarkable. Chest X-ray on her admission revealed a blunted costophrenic angle which is considered to be a minimal pleural effusion on the right side, bilateral hilar and paratracheal lymph node enlargement as well as, a diffuse pulmonary interstitial process. Thoracic CT findings verified the chest X-ray abnormalities.
Pulmonary function test measurements revealed mild restrictive ventilatory defect and decreased diffusion capacity for carbon monoxide. Tuberculin skin test was negative.

Dermatologic examination revealed inflammatory skin lesions. Punch biopsy of skin was reported as noncaseating granulomas, and interpreted as sarcoidosis dermatitis. Ocular involvement of sarcoidosis was not observed.
The place of minimal pleurisy on the right side was established by thoracic ultrasonography. Medical thoracoscopy was performed to illuminate the etiology of pleural effusion. Pleural space was in normal appearance at first glance, and it was pointed out that some micronodularity of visceral and parietal pleural membranes were present (Figure 2). Biopsies taken from parietal pleural nodules showed noncaseating granulomas in histological examination. Pleural tissue cultures for mycobacteria and fungi were negative. Oral prednisone (40 mg daily) treatment was given and gradually tapered during the follow-up period.

Case 3
A 27-year-old woman transferred to our hospital from another center. In her medical records; bilateral pneumonia, splenomegaly and conjunctival nodules had been observed three months earlier. At her admission to our center, she was complaining of fatigue, loss of appetite, dry cough and dyspnea.
The chest X-ray on admission was similar to the one taken 3 months earlier, and revealed bilateral pleural effusion as well as, hilar and paratracheal lymph node enlargement. Thoracic CT findings verified the chest X-ray abnormalities.

Pulmonary function test measurements revealed mild restrictive ventilatory defect and decreased diffusion capacity for carbon monoxide. Tuberculin skin test was negative.

Dermatologic examination revealed inflammatory skin lesions. Punch biopsy of skin was reported as noncaseating granulomas, and interpreted as sarcoidosis dermatitis. Ocular involvement of sarcoidosis was not observed.

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heal lymph node enlargement. Thoracic CT confirmed the chest X-ray findings, showing bilateral minimal pleural effusion and, conglomerated lymphadenopathies, the largest size of which is 4 cm. In abdominal CT, hepatosplenomegaly was observed. Bone marrow biopsy was performed to exclude lymphoma, and it was in normal cellularity.

Left palpable supraclavicular lymph node excisional biopsy was carried out, and reported as noncaseating granulomatous inflammation, compatible with sarcoidosis lymphadenitis. Ga- lium-67 scintigraphy revealed bilateral supraclavicular, mediastinal, hilar abnormal hiperaccumulation. Intense lacrimal, parotid, liver and spleen involvements were also seen in the gallium scan. These scan results were consistent with active sarcoidosis.

Percutaneous needle aspiration of spleen was diagnosed as sarcoidosis splenitis. Medical thoracoscopy from the left side showed diffuse micronodulation and hyperemia of parietal pleural surfaces, and adherence with nodulation in both the lung apices (Figure 3). Parietal pleural biopsies were reported as noncaseous granuloma consistent with pleural sarcoidosis. Tuberculosis and fungal pleurisy were also ruled out by required cultures in this patient. Prednisone therapy with 40 mg daily was administered for 2 months. After her clinical condition’s improvement, therapy was briefly tapered. Due to her clinical picture of active sarcoidosis with multiorgan involvement, she underwent this therapy for two years to avoid relapse.

DISCUSSION

Since the first report by Schaumann in 1933 on pleural sarcoidosis, pleural effusion is still known as a rare manifestation of sarcoidosis in a review of published studies (1,4). It is difficult to understand why pleural disease is scarce in sarcoidosis, although the involvement of pulmonary parenchymal, hilar and mediastinal nodes is present in about 90% of all sarcoidosis cases. Our series are relatively small; however there were also pulmonary and mediastinal involvements of disease in three cases with sarcoidosis pleurisy reported here. We have suggested that pleural involvement in sarcoidosis might be found more frequently if advanced evaluation of pulmonary sarcoidosis was carried out to identify pleural abnormalities.

In a recent article published by Szwarcberg and colleagues, 25 (41%) out of the 61 patients with sarcoidosis had pleural involvement established by the thoracic CT scans. Of the 25 patients with pleural involvement, only 5 (20%) had pleural effusions (5). A drawback of these pleural sarcoidosis cases is that the diagnosis is based on CT scans’ findings without histologic evidence. Indeed, pleural sarcoidosis is not a diagnosis that can be made in the light of radiographic findings. It should be proved by the histopathologic confirmation of noncaseating granulomas in pleural biopsies. Since the CT scans show some subpleural nodules which are specific to pulmonary sarcoidosis, some radiologists believe that they can practically provide the diagnosis of sarcoidosis (5). This approach may be valid for subpleural nodules in sarcoidosis, however the presence of pleural fluid should be evaluated by interventional pleural techniques. Thoracentesis is the first step to evaluate the nature of pleural fluid. However, there are no specific findings about pleural effusion in sarcoidosis. The two most frequent features reported in sarcoidosis pleurisy are; 1. Fluid nature is exudate, 2. Lymphocytes in fluid are the predominant cells of leucocytes (6). However, similar fluid characteristics are present in tuberculous pleurisy and the practice of administering unnecessary antituberculous therapy to patients with exudative, lymphocytic pleurisy is widespread, especially in endemic areas for tuberculosis (7).

The largest case study of sarcoidosis patients (diagnosed on a biotic basis) is made of six cases and was published by Sharma et al (2). Butin’s review of thoracoscopy did not also mention sarcoid-related pleurisy (8). Until recently, a total of 43 proven sarcoidosis pleurisy patients were reported in the literature, including this report (1). In our first and second cases, pleural nodules seen in thoracoscopy were randomly dispersed in all pleural surfaces, and in only third case, diffuse pleural nodularity was obser-
Nodularity, hyperemia, and inflammatory aspects were common findings in these cases, but the distribution of nodules on pleural surfaces was not observed uniformly. Relatively large anthracotic nodules in the first case and micronodules in the second and third cases were seen in diagnostic thoracoscopy. These findings showed that nodules caused by sarcoidosis pleuritis can appear in different forms. Percutaneous pleural biopsies would not be occasionally diagnostic in sarcoidosis cases with pleural effusion similar to these reported cases as regards our thoracoscopic observation, since nodules can be seen more seldom in parietal pleural surfaces than in the appearance of tuberculous pleuritis. Panadero et al. pointed out that percutaneous biopsy needles provide a better yield in tuberculous pleurisy than in other situations, due to the diffuse involvement of the parietal pleura (9). In our patients, diagnosing all 3 patients with other procedures shows that we should only use in order to eliminate other causes for these patients.

Sarcoïd-related pleurisy can take place more frequently unilaterally, but bilateral pleural effusion has been reported in some patients as seen in our third case (3). Most effusions are small or modest in size according to our findings. Post-thoracoscopy complications were not observed in our three patients. It can be claimed that thoracoscopy can be safely carried out in patients with minimal pleurisy detected in radiographics as well.

Although most patients develop sarcoïdosis in their early adult life, patients with sarcoïd pleurisy tend to be of an older age as recognized in our first and second patients (3). It may be assumed that the disease could be present for a long time when pleural sarcoïdosis was detected. Incomplete resolution of the pleural effusion in sarcoïdosis can cause progressive pleural thickening or trapped lung according to some case reports (3,5). Treatment of sarcoïdosis should be based on the activity and the stage of the disease. Therapy with systemic corticosteroids should be considered for the symptomatic patients and if the effusion is recurrent (3). Corticosteroid therapy was applied to all patients affected by symptomatic and active diseases, and the treatment has turned out to be efficient for all patients as proved by their clinical improvements.

In summary, medical thoracoscopic interventions could be a good option to diagnose sarcoïd pleurisy. Further studies are required to evaluate the place of medical thoracoscopy in the diagnosis of sarcoïdosis pleurisy.

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