Respir Case Rep 2012;1(2):55-58 DOI: 10.5505/respircase.2012.14633

Sarcoidosis Complicating with Bilateral Pneumothorax

Bilateral Pnömotoraks ile Komplike Olan Sarkoidoz

Funda Coşkun, Ahmet Ursavaş, Ezgi Demirdöğen Çetinoğlu, Aslı Görek Dilektaşlı, Esra Uzaslan

Abstract

Cases of sarcoidosis rarely present with pneumothorax. Case is a 57-year-old woman. She has been having complaints of cough for the past four years. The patient received corticosteroid treatment for sarcoidosis for 16 months, and visited our clinic one month after she voluntarily terminated her treatment due to an increase in complaints of cough. In her physical examination, her right hemithorax responded less to general respiration during pulmonary examination. Her lung PA X-ray demonstrated a pneumothorax line in her left hemithorax. There was an increase in dyspnea and right sided chest pain during the clinical follow-up, a day after hospitalization. The PA X-ray demonstrated a pneumotorax in the right hemithorax. Bilateral tube thoracostomy was applied on the patient and mediastinoscopy was performed. Due to the rareness of concomitant bilateral pneumothorax and sarcoidosis, what kind of information was presented in the light of literature in our case.

Key words: *pneumothorax, sarcoidosis.*

Özet

Sarkoidoz olguları nadir de olsa pnömotoraks ile prezente olabilirler. Olgumuz 57 yaşında bayan hasta. Dört yıldır öksürük şikayeti mevcut. On altı ay sarkoidoz tedavisi için sistemik kortikosteroid kullanan hasta kendi isteği ile tedavisini kestikten 1 ay sonra öksürük şikayetinde artma nedeniyle polikliniğimize başvurdu. Fizik muayenesinde sağ hemitoraks solunuma az katılıyordu. Olgunun PA Akciğer grafisinde sol hemitoraksta pnömotoraks hattı izlenmekteydi. Klinik izlemde hasta hastaneye yattıktan bir gün sonrasında nefes darlığında artış ve sağ tarafta göğüs ağrısı tarifledi. Çekilen PA Akciğer grafisinde sağ hemitoraksta da pnömotoraks geliştiği gözlendi. Hastaya iki taraflı tüp torakostomi ve mediastinoskopi uygulandı. Olgumuz bilateral pnömotoraks ve sarkoidoz birlikteliğinin nadir olması nedeniyle literatür bilgileri ışığında sunulmuştur.

Anahtar Sözcükler: *pnömotoraks, sarkoidoz.*

Department of Chest Diseases and Tuberculosis, Uludağ University Faculty of Medicine, Bursa

Uludağ Üniversitesi Tıp Fakültesi, Göğüs Hastalıkları ve Tüberküloz Anabilim Dalı, Bursa

Submitted (Başvuru tarihi): 09.05.2012 **Accepted (Kabul tarihi):** 22.07.2012

Correspondence (İletişim): Funda Coşkun, Department of Chest Diseases and Tuberculosis, Uludağ University Faculty of Medicine, Bursa, Turkey
e-mail: funda_coskun@hotmail.com

Sarcoidosis is a chronic granulomatous disease of unknown etiology. It frequently presents with bilateral hilar lymphadenomegaly and lung infiltration. Other organs and systems involved are the skin, eyes, peripheral lymph ganglions, liver, spleen, parotid glands, phalanges, the heart and nervous system. Although rare, cases of sarcoidosis may also present with pneumothorax (1). Pneumothorax occurs in 2 to 4% of patients with sarcoidosis (2). We found our case worthy of presentation because it was interesting to discover the development of bilateral pneumothorax, a rarely encountered condition, while being followed with the diagnosis of sarcoidosis.

CASE

We present a 57-year-old woman complaining of cough for the past four years. The mediastinal transbronchial needle biopsy was consistent with granulomatous inflammation and the corticosteroid treatment was started with the diagnosis of stage II sarcoidosis at the previous center she was following up. The patient received treatment for 16 months, and visited our clinic one month after she voluntarily terminated her treatment due to an increase in complaints of cough. She had cough and dyspnea, and her symptoms were worsening. She had a history of hypertension and diabetes mellitus. Nothing particular was discovered in her family history. The respiratory function tests showed FEV1:1.28 L, FVC: 1.58 L, FEV1/FVC: 81% consistent with restrictive pulmonary insufficiency. The carbon monoxide diffusion capacity was found as 72%. Arterial blood gases were normal. Her lung PA X-ray demonstrated bilateral parencyhmal lesions and hilar lymphadenopathy. A thoracic computed thomography (CT) demonstrated diffuse reticular lesions with bilateral mediastial, hilar and subcarinal lymphadenopathy and subpleural bulles. Bronchoalveolar lavage was performed who was considered having stage II sarcoidosis. CD4/CD8 was 3.3. Blood ACE level was 54 U/l. On the basis of clinical course and the radiological findings, she was diagnosed as sarcoidosis activation and 40 mg/day methylprednisolon was started and rapidly tapered to 16 mg/day after two months. The other organ systems were not involved in the course of disease.

After 3 months, she revisited our clinic with worsening dyspnea and new-onset chest pain. On physical examination, her respiratory rate was 26/min, pulse: 108/min and the blood pressure was 100/70 mmHg. Her right hemithorax responded less to respiration during pulmonary examination. No left-side breathe sounds were heard on auscultation. Other system findings were normal. In the laboratory examination, hemogram showed Hb: 13.5 gr/dl, platellets: 256,000/mm3, and hemotocrit 39%. Erythrocyte sedimentation rate was 32 mm/hour and the CRP was 3.4 mg/dl. Blood ACE level was 10.6 U/l and blood calcium was 8.6 mg/dl. The lung X-ray demonstrated a pneumothorax line in the left hemithorax (Figure 1).

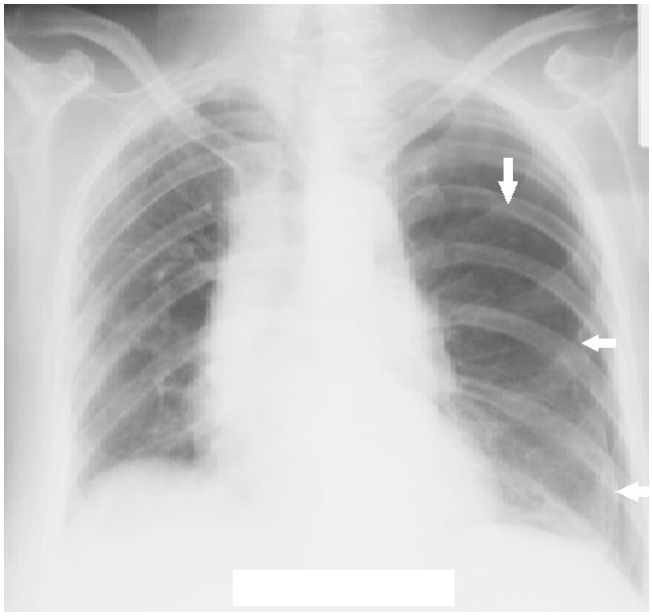

Figure 1: *PA X-ray demonstrated a pneumothorax line in her left hemithorax (see arrows)*

There was an increase in dyspnea and right sided chest pain during the clinical follow-up, a day after hospitalization. The PA X-ray demonstrated a pneumotorax line in the right and left hemithorax (Figure 2). Bilateral tube thoracostomy was applied on the patient (Figure 3) and mediastinoscopy was performed to confirm diagnosis in the patient whose lungs were both expanded after removing the thorax tubes.

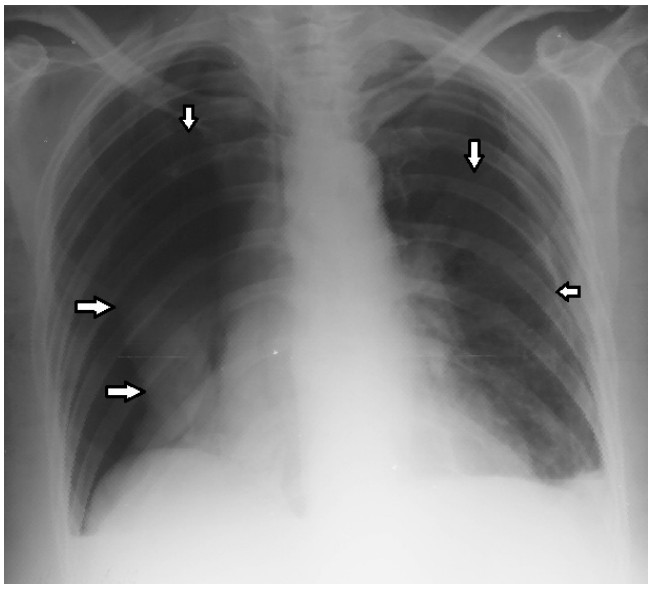

Figure 2: *PA X-ray demonstrated a pneumotorax in the right and left hemithorax (see arrows)*

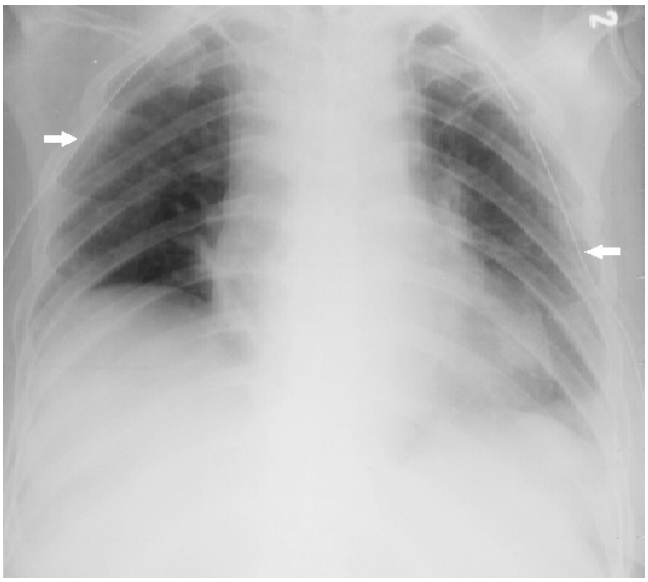

Figure 3: *PA X-ray demonstrated Lungs were both expanded, after removing the thorax tubes (see arrows)*

Following resolution of pneumothorax she underwent a thoracic CT scan. Ongoing diffuse reticular lesions with bilateral mediastial, hilar and subcarinal lymphadenopathy were observed and the corticosteroid treatment was continued.

DISCUSSION

The etiology of sarcoidosis is not well understood. It is a granulomatous inflammatory disease which involves the lungs and intrathoracic lymph nodes in more than 90% of patients, but can affect virtually every organ in the body. There is granulomatous inflammation without caseification necrosis where multinuclear giant cells and T lymphocytes (CD4>CD8 cells) are known to play role in the pathogenesis. The most commonly observed thoracic manifestations of sarcoidosis are pulmonary parenchymal involvement, lymphadenopathy (hilar, mediastinal or paratracheal) as well as airway, endobronchial, pleural, pulmonary vascular and thoracic lymph node involvement. Pulmonary parenchymal involvement is generally symmetric, diffused, reticular and nodular; and observed at the upper and middle zones. There is rarely a unilateral lesion, multiple large or solitary nodular involvements.

Cases of sarcoidosis rarely present with pneumothorax. The pathogenesis is not well understood. Many studies demonstrate that there might be development of a pneumothorax secondary to subpleural granuloma necrosis or eruption of subpleural bullae. Aleksson et al. (3) reported a 44-year-old case of sarcoidosis, presenting with bilateral pneumothorax. Another 26-years-old patient with sarcoidosis presenting with bilateral multiple cavitary lesions and a right-sided pneumothorax was reported by Yamaguchi et al. (4). In another publication, Komiya et al. (5) reported a case of 24 years old male, who visited the hospital with a left sided pneumothorax and was diagnosed with sarcoidosis through transbronchial needle biopsy. Recurrent bilateral pneumothorax in a young sarcoidosis patient was presented by Sharma and colleagues (6). Pneumothorax is known to be a rare complication of early stage sarcoidosis (7). Gomm reported another young sarcoidosis patient with hemopneumothorax in the left and pneumothorax of the apex in the right side (7). Subpleural blebs were identified in the upper left lobe of the lung in a 21 years-old male sarcoidosis patient who presented with pneumothorax by Omori and colleagues (8). They underlined the fact that, although rare, early stage sarcoidosis could present with pneumothorax in young adults (8).

In the retrospective examination of cases with bilateral pneumothorax, sarcoidosis was detected in two of 15 cases by Ayed (9). Resection of bullae or blebs was performed on all cases using video-assisted thoracic surgery, and it was suggested as a reliable surgical procedure. Nakamura et al. (10) reported another case

of a 23 year old young male. Pneumothorax was observed in the patient who showed spontaneous deterioration and presented with chest pain at the center a year after being diagnosed with sarcoidosis. Pleural effusion was observed on the same side two weeks after performing tube tracheostomy. Pleural effusion regressed following treatment with systemic steroids. Bullous changes were detected in the upper lobes and a spontaneous pneumothorax identified on the left side during more advanced tests, in a patient who visited the hospital with complaints of dry cough and dyspnea. Kanematsu et al (11) reported a diagnosis of sarcoidosis that was made after video-assisted thoracic surgery.

Apart from sarcoidosis, bilateral pneumothorax was frequently found to be concomitant with other pulmonary diseases. Some of the few pulmonary diseases observed are interstitial fibrosis, cystic fibrosis, histiocytosis X and lymphangiomatosis pneumothorax. A study conducted which showed that the rate of pneumothorax was present in variable rates depending on the diseases. They reported presence in <1% of COPD cases, 3-9% of cystic fibrosis, 1-3% of tuberculosis, 60-80% of lymphangiomyomatosis, and 4-17% of pulmonary langerhans cell histiocytosis.

Due to the rareness of concomitant bilateral pneumothorax and sarcoidosis, our patient is in the light of literature. Although rare, sarcoidosis should be kept in mind in the differential diagnoses, when encountered with a case of pneumothorax.

CONFLICTS OF INTEREST

None declared.

REFERENCES

1. Gül S, Danacı D, Evrenkaya TR, Başekim C. Sarkoidoz. T Clin Med Sci 1993; 13:296-302.
2. Froudarakis ME, Bouros D, Voloudaki A, Papiris S, Kottakis Y, Constantopoulos SH, et al. Pneumothorax as a first manifestation of sarcoidosis. Chest 1997; 112:278-80. [CrossRef]
3. Akelsson IG, Eklund A, Skold CM, Tornling G. Bilateral spontaneus pneumothorax and sarcoidosis. Sarcoidosis 1990; 7:136-8.
4. Yamaguchi M, Ohta K, Takizawa H, Kobeyashi N, Ishii A, Sugiyama H, et al. A case of sarcoidosis with right pneumothorax and multipl cavities in both lung fields. Nihon Kyobu Shikkan Gakkai Zasshi. 1995; 33:533-7.
5. Komiya T, Matsushima T, Kimura N, Tano Y. A case of sarcoidosis discovered by onset of pneumothorax. Nihon Kyobu Shikkan Gakkai Zasshi. 1995; 33:433-7.
6. Sharma SK, Pande JN, Mukhopadhay AK, Goulatia GK, Wali JP, Guleria JS. Bilateral recurrent spontaneous pneumothorax in sarcoidosis. Jpn J Med 1987; 26:69-71. [CrossRef]
7. Gomm SA. An unusual presentation of sarcoidosis spontaneous haemopneumothorax. Postgrad Med J 1984; 60:621-3. [CrossRef]
8. Omori H, Asahi H, Irinoda T, Itabashi T, Saito K. Pneumothorax as a presenting manifestation of early sarcoidosis. Jpn Thorac Cardiovasc Surg 2004; 52:33-5. [CrossRef]
9. Ayed AK. Bilateral video-assisted thorascoscopic surgery for bilateral spontaneous pneumothorax. Chest 2002; 122:2234-7. [CrossRef]
10. Nakamura H, Kitada O, Aragane K. A case of pulmonary sarcoidosis with pneumothorax and pleural effusion after improvement of pulmonary impairment. Nihon Kyobu Shikkan Gakkai Zasshi. 2002; 40:383-6.
11. Kanematsu T, Ohgushi F, Ogawa H, Nishioka Y, Shinohara T, Yanagawa H, et al. Bullous sarcoidosis a case report. Nihon Kyobu Shikkan Gakkai Zasshi. 2001; 39:117-21.