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A Case of Endobronchial Sarcoidosis with Recurrent Pneumonia

Tekrarlayan Pnömoni ile Seyreden Endobronşiyal Sarkoidoz Olgusu

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Abstract

Sarcoidosis is a chronic, multisystem and non-caseating granulomatous disease of unknown etiology. Its diagnosis is based on a combination of clinical and radiographic findings and the demonstration of typical granulomas, and can often be made based on the exclusion of other granulomatous diseases. There is lung involvement in 90% of cases, often in the form of parenchymal disease, while involvement in the form of an endobronchial mass is a rarer condition. Our patient underwent bronchoscopy after being diagnosed with sarcoidosis around 3 years earlier, and was followed up without treatment due to a history of pneumonia that had recurred three times in the same localization in the previous year. An endobronchial lesion was identified on bronchoscopy, while a biopsy revealed granulomatous inflammation. Progressive sarcoidosis was considered based on the clinical and radiological findings and the patient was started on steroid treatment. We present this study of a case of sarcoidosis with recurrent pneumonia in the same localization and endobronchial involvement to emphasize the need to consider sarcoidosis in the differential diagnosis of endobronchial mass lesions.

Key words: *Sarcoidosis, endobronchial, treatment.*

Öz

Sarkoidoz etiyolojisi bilinmeyen, kronik, multisistemik nonkazeifiye granülomatöz bir hastalıktır. Tanısı, klinik ve radyografik bulguların birlikteliğine, tipik granülomların gösterilmesine dayanır ve çoğu zaman diğer granülomatöz hastalıkların dışlanmasıyla konulabilir. Akciğer tutulumu %90 oranındadır. Sarkoidozda akciğer tutulumu sıklıkla parankimal hastalık şeklindeyken, nadiren endobronşiyal kitle şeklinde tutulum görülebilir. Yaklaşık 3 yıl önce sarkoidoz tanısı konan ve tedavisiz izlemde olan olgumuza son bir yıl içinde aynı lokalizasyonda üç kez tekrarlayan pnömoni öyküsü olması nedeniyle bronkoskopi yapıldı. Bronkoskopide endobronşiyal lezyon görülüp alınan biyopsisinde granülomatöz inflamasyon saptanan hastada klinik, radyolojik bulgularla progresif sarkoidoz düşünüldü ve steroid tedavisi başlandı. Aynı lokalizasyonda tekrarlayan pnömoni ile gelen ve endobronşiyal tutulum gözlenen sarkoidoz olgusunu, endobronşiyal kitle lezyonların ayırıcı tanısında sarkoidozu vurgulamak için sunduk.

Anahtar Sözcükler: *Sarkoidoz, endobronşiyal, tedavi.*

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Sarcoidosis is a multisystem disease of unknown cause that is characterized by non-caseating granulomas. Although lung involvement is observed in 90% of cases, there may also be eye, skin, liver, bone, joint, heart and brain involvement (1). Lung involvement is often in the form of parenchymal disease and nodular involvement may be seen in the bronchial mucosa, while involvement in the form of endobronchial masses is very rare (2). Our patient underwent bronchoscopy, having been diagnosed with sarcoidosis around 3 years earlier, and had been followed up without treatment due to a history of pneumonia recurring three times in the same localization in the previous year. An endobronchial lesion was noted on bronchoscopy, while a biopsy revealed aggranulomatous inflammation. Progressive sarcoidosis was considered based on the clinical and radiological findings, and steroid treatment was started. We present here a case of sarcoidosis with recurrent pneumonia in the same localization and endobronchial involvement.

CASE

A 31-year-old male patient presented with complaints of fever, cough, joint pain and shortness of breath for the last 10 days. The patient had been using quinolone antibiotics for about 1 week following a diagnosis of pneumonia, and had no additional disease or smoking history. His medical history revealed that he applied to a health institution twice with the same complaints around 6 months earlier and had been treated with antibiotics twice with a diagnosis of pneumonia. It was further learned that the diagnosis of sarcoidosis had been made based on endobronchial ultrasonography and a biopsy of the mediastinal lymph nodes in 2018, and he was under follow-up without treatment. A thorax computed tomography (CT) taken in 2018 revealed bilateral hilar and paratracheal lymphadenopathy, as well as micronodular infiltrations of the parenchyma. Upon admission to our hospital, bilateral diffuse rhonchi were detected during a physical examination of the patient. Laboratory results revealed leukocytosis and C-reactive protein to be high in a complete blood count, while the patient's biochemical parameters were normal. Angiotensin converting enzyme: 32 U/L, Calcium: 9.9 mg/dl and 24-hour urine calcium normal. Bilateral hilar fullness was observed on a chest X-ray, along with a homogeneous increase in density extending from the hilus to the periphery in the upper zone of the right lung (Figure 1). On Thorax CT, on the other hand, revealed peribronchial density increments in the right hilar area adjacent to the right lung upper lobe minor fissure, and a ground glass area in the right upper lobe anterior with an appearance of peripheral consolidation (Figure 2). The patient, who had fever and was thought to have postobstructive pneumonia was started on piperacillin + tazobactam treatment, having used quinolone group antibiotics before. A fiberoptic bronchoscopy performed due to recurrent pneumonia revealed a smooth-surfaced mass that almost completely occluded the anterior segment of the right lung upper lobe and a biopsy was taken (Figure 3). The histopathological examination of the endobronchial lesion revealed a granuloma structure in the bronchial mucosa but no necrosis (Figure 4). In a microbiological examination of the bronchoalveolar lavage, no acid resistance bacillus was observed and no growth was detected in the culture. Pulmonary function tests revealed FVC: 68%, FEV1: 57% FEV1/FVC: 87% Diffusion: 66% and moderate restriction. A diagnosis of progressive sarcoidosis with clinical, radiological and physiological findings was made, and the patient was started on a 32-mg steroids treatment. The patient's complaints have since decreased, and his treatment and follow-up is continuing. Regression was observed on a chest X-ray in the 3rd week of treatment (Figure 5).

DISCUSSION

A diagnosis of sarcoidosis is based on clinical and radiographic findings, histopathological evidence, non-caseating granulomas on biopsy and the absence of alternative etiologies (3). Common complaints at admission include nonproductive cough, dyspnea on exertion, chest pain and hemoptysis (rare) (50% of cases). Systemic complaints such as fatigue, fever and anorexia may be present in up to 45% of cases, while approximately 35% of patients may present with acute fever, polyarthralgia, erythema nodosum and bilateral lymphadenopathy (Lofgren's Syndrome) (3). Cough, fever and chest pain were present in our case, and the symptoms regressed several times after antibiotics, although the patient applied again with the same complaints.

Invasive methods such as mediastinoscopy, open lung biopsy, scalene lymph node biopsy and skin biopsy are used for pathological sampling in the diagnosis of sarcoidosis, although fiberoptic bronchoscopy – a less invasive method – can also be employed (4,5). Transbronchial biopsy, transbronchial fine needle aspiration, endobronchial biopsy and Bronchoalveolar Lavage (BAL) are performed as interventional procedures in fiberoptic bronchoscopy (5).

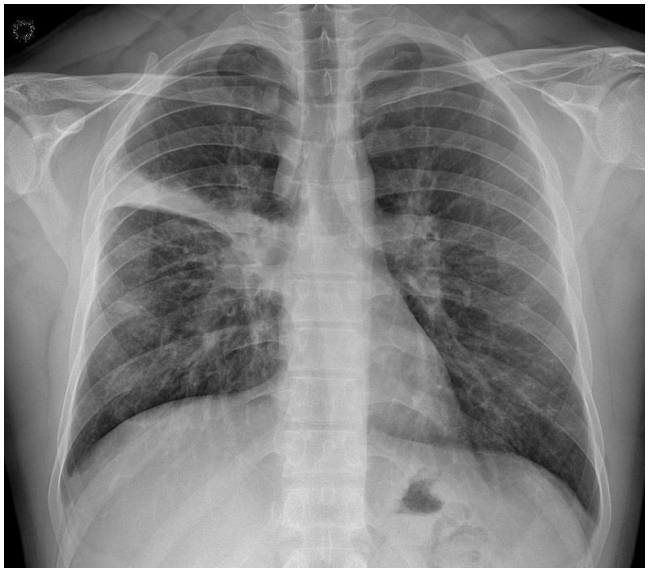

Figure 1: Bilateral hilar fullness and homogeneous increase in density extending from the hilus to the periphery in the upper zone of the right lung on Chest X-ray

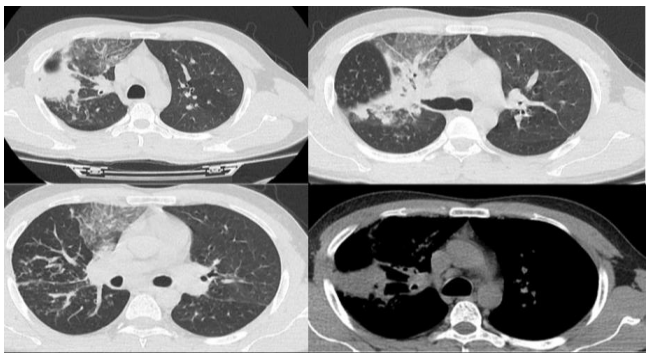

Figure 2: Peribronchial density increments notable in the right hilar area, adjacent to the right lung upper lobe minor fissure, while the ground glass area in the right upper lobe anterior has an appearance that creates peripheral consolidation on Chest CT

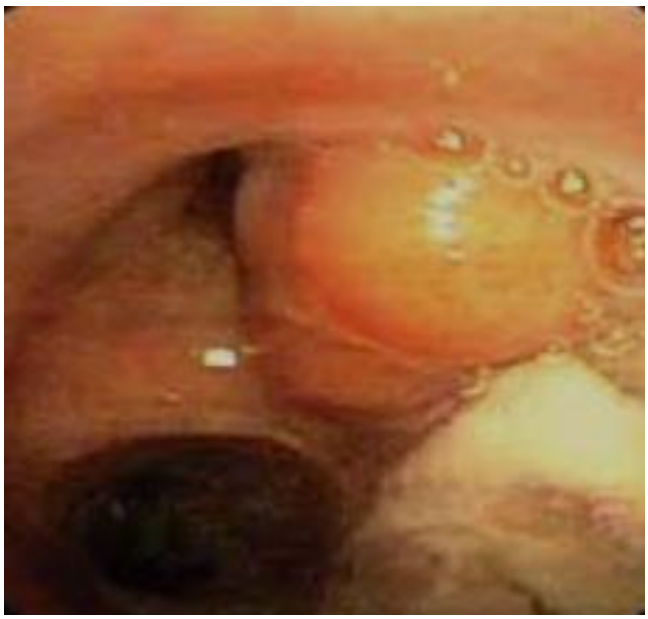

Figure 3: Flexible bronchoscopy revealed an endobronchial mass lesion at the anterior segmental bronchus of the right lung upper lobe

Endobronchial mucosal involvement is a common finding in fiberoptic bronchoscopy, and is characteristically described as a curbstone or pebble appearance. These may have areas with a nodular appearance that when biopsied take the form of granulomas (5). In cases of sarcoidosis, other abnormalities that may be detected from fiberoptic bronchoscopy include mucosal hyperemia, mucosal infiltration, bronchostenosis, or rarely, endobronchial mass lesions, as in our case (4). In a case series study by Kiter et al. (6), 0.4% endobronchial mass lesions were reported, while no endobronchial involvement was detected in other sarcoidosis series (7-9). Kumbasar et al. (10), Akpinar et al. (11) and Güngör et al. (12) all detected endobronchial mass lesions, as in our case, with granulomas observed in their biopsies of the lesions. Our case was followed-up with previous diagnosis of sarcoidosis and the absence of symptoms at the time of diagnosis, and was considered to have progressive sarcoidosis due to the presence of an endobronchial lesion, the recurrent postobstructive pneumonia and indicative symptoms.

The narrowing of the bronchial lumen in sarcoidosis can be a result of three mechanisms: the mechanical narrowing of the lumen due to compression of the enlarged lymph nodes; the submucosal invasion of sarcoid granulomas; and the presence of an endobronchial mass. Endobronchial masses have been reported only rarely, while the first two mechanisms are more common (13). Whether the endobronchial mass lesion is a form of sarcoidosis or an indicator of the extent of lung involvement of the disease has yet to be fully elucidated. Bjermer et al. (14) reported inflammatory activity in BAL to be greater in cases with granuloma detected during endobronchial biopsy than in cases without granuloma identified from an endobronchial biopsy.

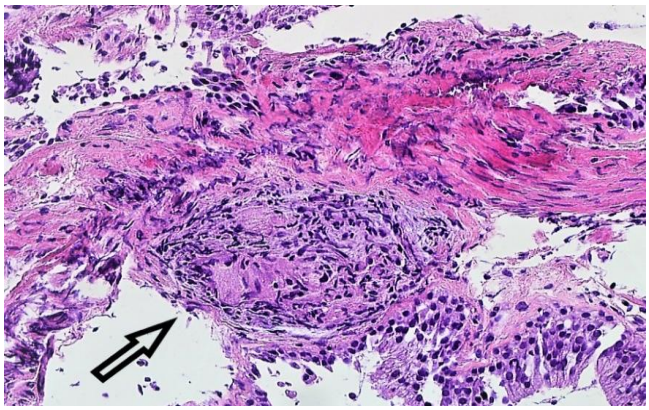

Figure 4: Granulomatous inflammation of the bronchial mucosa (H&E X200)

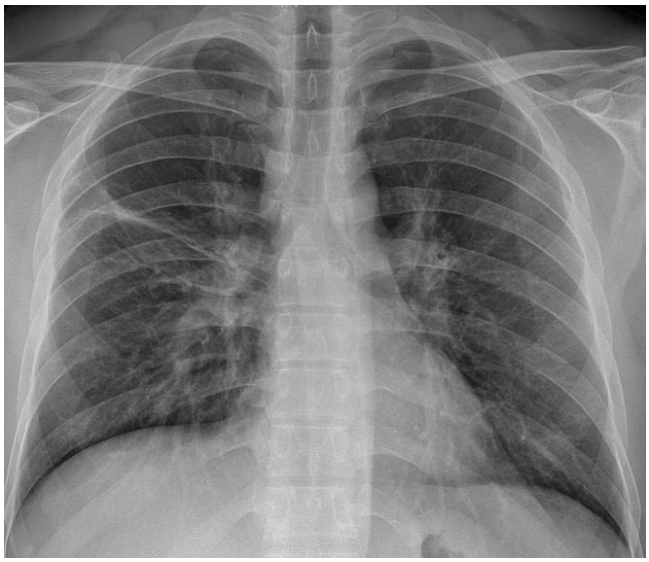

Figure 5: Regression was observed in chest X-ray in week 3 of treatment

Although the exact pathogenesis is not known yet, it is commonly assumed that sarcoidosis is an exaggerated immune response to an unidentified pathogen. Recent studies have suggested that T-cells play a central role in the development of the disease, possibly due to their excessive cellular immune response (15). A CD4:CD8 ratio of >3.5 in bronchoalveolar lavage can be detected in 50% of cases, which is most likely an outcome of the accumulation of CD4 cells and the release of interleukin-2 at disease activity sites. This leads to the development of non-caseating granulomas that are rich in epithelioid cells, which is a hallmark of sarcoidosis, on microscopic examination (although not specific) (3). In a BAL analysis, Kumbasar et al. (10) reported a CD4/CD8 ratio of 5, while Akpınar et al. (11) found it to be 3.5. No BAL analysis was performed in the present study.

It has been reported that endobronchial lesions can disappear upon corticosteroid treatment in sarcoidosis cases with endobronchial involvement (16). Akpinar et al. (11) reported the disappearance of an endobronchial mass lesion after corticosteroid treatment, and the CD4/CD8 ratio in BAL decreased to 1.18 in their case. Güngör et al. (12) reported that complete remission was achieved in both of their cases with drug-free follow-up, and emphasized that a drug-free cure could be achieved with close follow-up in cases of endobronchial sarcoidosis. Cases that have improved without treatment have been reported in the literature (17), but in our case, a radiological and clinical response was obtained with steroid treatment.

In conclusion, we describe here a case of sarcoidosis that presented as an endobronchial mass, which highlights the notion the need to include sarcoidosis in a differential diagnosis in patients presenting with an endobronchial mass, as early treatment may improve the outcome. It is recommended that a flexible bronchoscopy be performed to search for endobronchial masses in patients with suspected sarcoidosis based on CT findings and who show recurrent pneumonia at the same localization, even when CT scans reveal no such endobronchial mass lesion, as in our patient. In cases of endobronchial sarcoidosis, treatment can be provided with steroids.

CONFLICTS OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS

Concept - B.D., C.A., S.U., M.T., B.S., E.C.; Planning and Design - B.D., C.A., S.U., M.T., B.S., E.C.; Supervision - B.D., C.A., S.U., M.T., B.S., E.C.; Funding - B.D., C.A., S.U., M.T., B.S., E.C.; Materials - B.D., C.A., S.U., M.T., B.S., E.C.; Data Collection and/or Processing - B.D., C.A., S.U., M.T., B.S., E.C.; Analysis and/or Interpretation - B.D., C.A., S.U., M.T., B.S., E.C.; Literature Review - B.D., C.A., S.U., M.T., B.S., E.C.; Writing - B.D., C.A., S.U., M.T., B.S., E.C.; Critical Review - B.D., C.A., S.U., M.T., B.S., E.C.

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