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Case report

A patient with sarcoidosis who developed heterochronic involvements in different organs from initial organs during 7 years

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ABSTRACT

A 59-year-old woman, who was given a diagnosis of sarcoidosis by supraclavicular lymph node biopsy 5 years previously, was admitted for further examination following abnormal radiologic findings. Nodular pulmonary and abdominal lesions were observed by computed tomography, and liver biopsy was performed and showed epithelioid cell granulomas. She was asymptomatic and was followed up with no therapy. At 1 year follow-up, the pulmonary and abdominal lesions had nearly complete resolution. Nodular pulmonary and abdominal lesions in patients with sarcoidosis can mimic metastatic disease, lymphoma, and infection, and can reappear during disease activity. Therefore, differential diagnosis and continual follow-up are important.

1. Introduction

Sarcoidosis is a systemic granulomatous disease of unknown etiology. Although abdominal involvement is often seen, the liver, spleen, and lymph nodes are the most commonly involved sites, and usually cases are not accompanied by symptoms [1]. Lung involvement is associated with over 90% of cases, but nodular pulmonary lesions are relatively rare. Here, we present a case of sarcoidosis that developed heterochronic involvements in different organs from initial organs during 7 years with review of the relevant literature.

2. Case report

A 59-year-old woman, a current smoker, was admitted to our hospital for further examination following abnormal chest radiograph and computed tomography (CT) findings in 2011. She had no specific abdominal or thoracic symptoms. She

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was a clerical worker and had been smoking 10 cigarettes daily for 35 years.

Five years previously, she was given a diagnosis of sarcoidosis by right supraclavicular lymph node biopsy, and she had bilateral hilar lymphadenopathy (BHL), uveitis, elevated serum levels of angiotensin converting enzyme (s-ACE), elevated lymphocyte count (16.4%) and a high CD4/CD8 ratio of 9.17 upon examination of bronchoalveolar lavage fluid. There were no other distinct involvements other than in the eye, cervical lymph node, and BHL.

She was followed up with no therapy, and her eye symptoms improved gradually. Then, abnormal findings of the chest radiograph and abdominal CT were observed in 2011. A physical examination upon admission revealed a body temperature of 36.6 °C, blood pressure 136/90 mmHg, and regular pulse of 70 beats/min. The right supraclavicular was palpable. Lung and heart auscultation was normal, abdominal examination revealed no hepatosplenomegaly, and no skin rash was evident. The results of laboratory tests showed slight elevations in alkaline phosphatase (ALP), 342 IU/L; s-ACE, 26.3 IU/L (normal range, <21.3); and Krebs von den Lugen-6 (KL-6), 534 IU/L. Tests for the tumor marker, CA19-9 (48 IU/L), was elevated, but CEA (2.8 ng/mL) was not increased. Anti-nuclear, anti-mitochondrial and anti-smooth muscle antibodies, hepatitis B surface antigen, and hepatitis C virus antigen were negative. A chest radiograph at initial diagnosis showed BHL (Fig. 1A), but in 2011 it showed a reduction of BHL and multiple nodular shadows (Fig. 1B). A CT scan of the chest also showed multiple nodules, 1–2 cm in diameter, in the bilateral lung field and no hilar or mediastinal lymphadenopathy (Fig. 2A and B). The margins of the nodules were hazy and fluffy. An early contrast-enhanced CT scan of the abdomen showed diffuse parenchymal heterogeneity, irregular surface of the liver with mild hepatomegaly, and multiple low-attenuation nodules in the enlarged spleen (Fig. 2C and D). In addition, several lymph nodes around the abdominal artery were mildly enlarged. Administration of 67-gallium scintigram showed accumulation in the liver and spleen, but no accumulation in the lung and thoracic lymph nodes. Liver biopsy was performed and showed epithelioid cell granulomas in the portal area (Fig. 3). No caseous necrotic lesions were present in the granulomas, and specific staining against acid-fast bacteria and fungi was negative. Based on these findings, we concluded that the patient had developed various involvements in the lung, liver, and spleen. Since she had no symptoms and her liver function was slightly elevated (ALP was increased to 342 IU/L), we observed her without medication. At 1 year follow-up in 2012, chest and abdominal CT revealed that the pulmonary nodules and abnormal shadows of the liver and spleen had almost completely disappeared. No other involvement was seen during follow-up. The clinical course to date is shown in Fig. 4.

3. Discussion

Sarcoidosis is a non-caseating granulomatous disease of unknown etiology affecting multiple organs. Although abdominal involvements are often seen, the liver, spleen, and lymph nodes are the most commonly involved sites. Elevated s-ACE levels and abdominal findings are related, but not to chest radiographic stage [1].

Liver involvement determined by biopsy has been observed in 24–94% of patients [2,3] and was described in 11.5% of 736 patients enrolled in a case control etiologic study of sarcoidosis (ACCESS) [4]. Laboratory evidence of liver dysfunction was observed in 2–60% patients, with ALP levels being most commonly affected. Despite these observations, symptomatic liver disease occurs in less than 5% patients with sarcoidosis and it is usually not seen or manifests as homogeneous organomegaly at CT [1]. In 5–15% of cases, sarcoidosis manifests as multiple, low-attenuation nodules. By contrast-enhanced CT, the nodules appear as hypodense masses relative to adjacent normal parenchyma, and peripheral enhancement typically is not seen [5]. Similar to our case, a pattern of multiple low-density intrahepatic septa has also been described, which is suggestive of a cirrhotic liver [6,7]. Granulomatous lesions are small in size and are almost always present in liver biopsies. They typically occur in the portal and periportal zones of hepatic sinuses, are present in high numbers and are evenly distributed in the liver parenchyma. Intrahepatic cholestasis is found in up to half of all

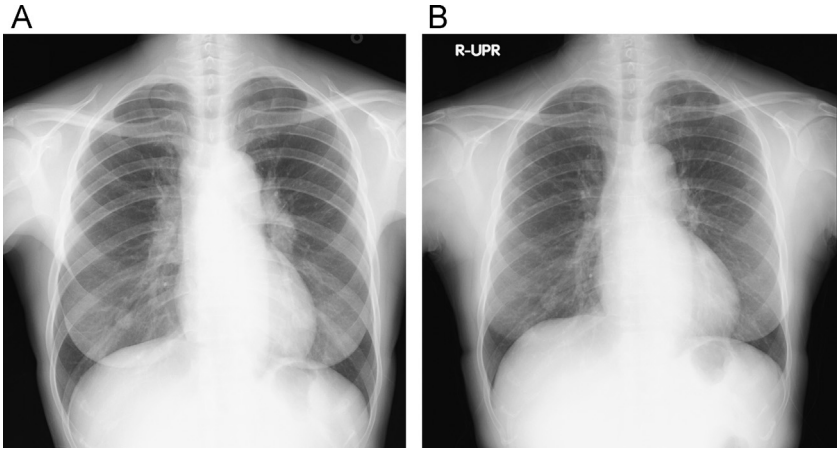

Fig. 1 – (A) Chest radiograph in 2006 showed bilateral hilar lymphadenopathy (BHL). (B)Chest radiograph in 2011 showed a reduction of BHL and multiple nodular shadows in the bilateral lung field.