Case Report

Intra-Abdominal Sarcoidosis In 54-Year-Old Indian Lady-A Case Report

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Received Date: 28 December, 2016; Accepted Date: 20 February, 2017; Published Date: 25 February, 2017

Abstract

Commonly considered a pulmonary disease, sarcoidosis is actually a multisystemic inflammatory granulomatous disease which is not commonly found in Asians. The systemic nature of the disease can lead to diverse manifestations. We describe here the diagnostic challenge in a 54-year-old Indian lady presenting with dyspeptic symptoms and without pulmonary complaints who was later found to have disseminated sarcoidosis. Hepatoduodenal lymphadenopathy was the presenting radiological finding, with normal findings on chest radiograph. Lymph node, pulmonary and hepatic involvement was confirmed by presence of non-caseating granulomas on histological examination. Splenic involvement was diagnosed by imaging studies. This case highlighted both the variability in the presentation of the disease as well as the difficulties encountered in establishing the diagnosis. The patient was prescribed corticosteroid therapy and responded well. Strong clinical suspicion is critical in making an early diagnosis in intra-abdominal sarcoidosis in the absence of pulmonary symptoms.

Introduction

Sarcoidosis is a multisystemic inflammatory granulomatous disease of unknown aetiology [1]. Epidemiological reports of sarcoidosis vary among ethnic groups and worldwide prevalence varies between 2 to 60 per 100,000 people, with higher rates of occurrence seen in patients in northern European countries [2]. Sarcoidosis is rarely encountered in Southeast Asia and its incidence in Malaysia is unknown although it is generally thought to be rare [3]. In a report from Singapore, the overall annual incidence of sarcoidosis was 0.56 per 100,000 people [4] This multisystem disorder usually involves the lungs (>90%) and the lymphoid system (30%), and classically presents as intrathoracic hilar lymphadenopathy [5]. However, the systemic nature of the disease can lead to diverse manifestations, such as isolated retroperitoneal lymphadenopathy or intra-abdominal organ involvement, without associated pulmonary or mediastinal disease. The following is a case report describing a patient with intra-abdominal sarcoidosis who presented with retroperitoneal lymphadenopathy without pulmonary complaints.

Case

A 54-year-old Indian lady presented to the emergency department with one month history of epigastric pain associated with malaise, loss of weight and loss of appetite. Physical examination revealed painless hepatomegaly and a suprapubic mass. There was no peripheral lymphadenopathy. Laboratory tests were normal. Chest X-ray was essentially normal with no evidence of interstitial lung disease or hilar lymphadenopathy. Lung function tests and echocardiogram were normal. Ultrasonography disclosed fatty liver with hepatomegaly and a pedunculated uterine fibroid. Oesophago-gastro-duodenoscopy (OGDS) revealed pangastritis and duodenitis with thickened gastric mucosa. A biopsy of gastric mucosa showed chronic gastritis. Colonoscopy was normal. She was treated with a proton pump inhibitor but her abdominal pain persisted. Computed tomography (CT) scan of the abdomen revealed enlarged hepatoduodenal and portal lymph nodes along with a large uterine fibroid. There were changes in the lung bases which were reported as suggestive of lymphangitis carcinomatosis. Endoscopic ultrasound (EUS) was done and revealed enlarged
porta hepatis, coeliac, subhilar and mediastinal lymph nodes. She was then subjected to an open biopsy of abdominal lymph nodes as the fine needle aspiration cytology from EUS was inconclusive. At surgery, enlarged hepatoduodenal lymph nodes and a fibrotic liver were found. Histological features of the lymph node biopsy showed areas of non-caseating granulomatous involvement, highly suggestive of sarcoidosis. Stains for acid fast bacilli and fungi were negative. There was no evidence of malignancy. CT thorax was performed in view of the diagnosis and showed diffuse upper and middle lobe nodular changes with a Broncho vascular distribution and clustering, along with bronchiectatic changes and fibrosis. Transbronchial lung biopsy revealed non-caseating granulomas which were negative for acid fast bacilli, fungal and malignancy. The patient was commenced on steroid therapy. Repeat liver function tests after the diagnosis revealed elevated alkaline phosphatase levels. CT scan and ultrasonography of the abdomen showed possible liver and spleen involvement. A liver biopsy was performed under ultrasound guidance and it showed granulomatous inflammation with on-going cirrhosis, consistent with the diagnosis of sarcoidosis. Pulmonary sarcoidotic lesions showed improvement on radiological imaging two months after corticosteroid therapy. Her liver function tests normalized after steroid therapy (Figure 1-5).

Figure 1, 2 & 3: CT scan abdomenshowed enlarged hepatoduodenal, paracava and paraaortic lymphadenopathy along with uterine fibroid.

Figure 4 & 5: CT thorax showed diffuse upper and middle lobe nodular changes with bronchovascular distribution and clustering along with bronchiectasis changes and fibrosis.
Discussion

Sarcoidosis is widely considered a pulmonary disease in which 95% of patients have intrathoracic involvement. Extrathoracic involvement occurs in 40% to 50% of patients at the time of diagnosis, with isolated extrapulmonary disease occurring in less than 10% [6]. Liver involvement follows lung and lymph node in frequency (60%-90%). Other sites include spleen (40%), eyes (11-83%), skin (25%) and less commonly, the nervous system, muscles, bones and salivary glands [1,7]. Patients with pulmonary disease often present with dyspnea, cough, chest discomfort and wheezing. 30% to 50% of patients report nonspecific symptoms and are incidentally found to have findings consistent with sarcoidosis upon chest radiograph [8].

Our patient presented with a persistent epigastric pain, along with constitutional symptoms such as malaise, loss of appetite and loss of weight. There was no clinical symptom which suggested pulmonary involvement. Chest radiography and lung function test did not show any abnormalities. Based on her symptoms, gastric sarcoidosis should be considered. The most prominent symptom in gastric sarcoidosis is epigastric pain, which is typically postprandial. Other common complaints include early satiety, nausea, vomiting, and significant weight loss mimicking malignancy [9]. Diffuse mucosal involvement in gastric sarcoidosis typically results in fibrosis, which leads to widespread mucosal thickening as seen in our patient. However, biopsy of the gastric mucosa in our patient revealed only chronic gastritis. The diagnosis of sarcoidosis in this patient was made 8 months after her initial presentation. As patient was still complaining of epigastric pain despite proton pump inhibitor therapy, there should be a need to repeat the biopsy to exclude gastric involvement as sarcoidosis is a multisystemic disorder.

The diagnosis of sarcoidosis in our patient was made based on histological findings of the biopsy of the enlarged hepatoduodenal lymph nodes. Abdominal lymphadenopathy is a rare finding in sarcoidosis. In a review of 2100 abdominal CT scans by Deutch, 160 showed evidence of abdominal lymphadenopathy and only 9 were secondary to benign causes. Of these nine, only one was due to sarcoidosis [10]. A study done by Liam et al in University Hospital, 4 out of 14 patients diagnosed with sarcoidosis presented with peripheral lymphadenopathy, and none of the these was found to have abdominal lymphadenopathy [11]. Lymphoma was our initial differential diagnosis based on the clinical symptoms and CT scanfindings. However, non-Hodgkin lymphoma is more likely to cause large confluent nodes and retrocrural lymphadenopathy, which was not seen in our case [12].

Hepatomegaly was present in our patient; however, she was not jaundiced or symptomatic from her hepatic enlargement. Liver function tests at the initial presentation were normal. It is discovered that serum alkaline phosphatase levels were elevated during evaluation of the patient’s progress and hepatic sarcoidosis was confirmed on liver biopsy. In a study done by Johanna et al, liver function test abnormalities were found in 24% patients with chronic sarcoidosis among which 15% of patient were suspected to have hepatic sarcoidosis [13] Severity of the liver function test abnormalities appeared to be related to the degree of fibrosis and extensiveness of granulomatous inflammation in hepatic sarcoidosis. It was suggested in the study that ultrasonography should be considered in patients with abnormal liver function tests for more than 3 months and liver biopsy is indicated in severe liver function test abnormalities. In our case, patient’s liver function test normalized after corticosteroid therapy.

Conclusion

This is an interesting case of multisystemic involvement of extrapulmonary sarcoidosis presenting as symptomatic gastritis. Strong clinical suspicion is critical in making an early diagnosis in intra-abdominal sarcoidosis in the absence of pulmonary involvement. Patients with known sarcoidosis require careful follow up and evaluation to screen for extrapulmonary involvement, and the most life-threatening involvements include the cardiac, neurologic and renal systems. Both the physician and patient should be aware of and closely monitor the progression of symptoms, pertinent laboratory findings and initiate treatment whenever appropriate.

References


