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Sarcoidosis with predominant hepatic involvement in a Male Patient: Case Report and Literature Review.

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Abstract: Sarcoidosis is a multisystem disorder of unknown cause. It frequently presents with bilateral hilar lymphadenopathy, pulmonary infiltration, ocular and skin lesions. Hepatic involvement is seen in relatively less number of cases. Because of diverse manifestations, patients with sarcoidosis may present to clinicians with different symptoms. We describe a case of 49 years old male who presented with typical symptoms of dry cough, weight loss and evening rise of temperature mimicking tuberculosis. No hilar lymphadenopathy was seen on CECT chest. CECT abdomen revealed liver and splenic parenchymal involvement which on further investigation by FNAC liver showed well formed epithelioid cell granulomas. Transbronchial lung biopsy was also performed which depicted noncaseous epithelioid cell granulomas suggesting systemic sarcoidosis. The diagnosis was further confirmed by ruling out other infections. In conclusion, the diagnosis of sarcoidosis has to be made through prudent and comprehensive investigations, typical imaging, histological examination and ruling out other infections.
Keywords: Hepatic granulomas, Sarcoidosis, epithelioid granuloma

INTRODUCTION

Sarcoidosis is a systemic inflammatory disorder of unknown etiology, characterized by the formation of noncaseating epithelioid cell granulomas with multisystem involvement. [1,2] Sarcoid granulomas can involve any organ,but in more than 90% of patients, clinical sarcoidosis is manifested as intrathoracic lymph node enlargement or pulmonary infiltration.[3] The eyes, skin, abdominal organs, central nervous system and bones are among the other organs involved.[4]

Significant heterogeneity in prevalence, clinical presentation and severity of sarcoidosis occurs among different ethnic and racial groups. [2]While the disease has a slight female preponderance globally, sarcoidosis is commoner in males in India. [5]

The case reported here is important because of its typical symptoms of persistant cough, history of prolonged fever with evening rise of temperature and loss of weight which is highly suggestive of tuberculosis more so in a country like India where tuberculosis accounts for one fifth (21%) of the global incidence, [6] the case could have been easily misdiagnosed and wrongly treated. Also the typical radiological finding of bilateral hilar lymphadenopathy of sarcoidosis [7] or abnormalities on chest radiographs which are detected in 85% to 95% of patients with sarcoidosis [8] is missing in our case which has made all the more diagnosis of systemic sarcoidosis with predominant hepatic involvement difficult.

Case Report

A 49 years old male, presented to a tertiary care hospital outdoor with chief complaints of dry cough for last one year associated with breathlessness which was increasing progressively for last 7-8 months. Patient complained of low grade fever of 100 Fahrenheit usually in the evening time for last two months. There was also a history of significant weight loss of around 8kgs in last 4 months. The patient got treatment at various places at his home town but with no relief. On examination pallor was noted with no significant abnormality. No added sounds were heard on auscultation of chest.Total & differential counts were in normal limits. Urine routine microscopy was normal and cultures were sterile. Peripheral blood smear showed slight degree of hypochromia, with no parasites. WIDAL test for typhoid was negative. ESR was 100mm/hr (very high). There was no past history of diabetes, Coronary Artery Disease, hypertension or tuberculosis.Digital chest x-ray done was normal.Renal function test were within normal limits. Mountex test was negative. Liver function test revealed high SGOT (AST) – 134 U/L (<50), SGPT (ALT) – 109U/L (<50) and Alkaline Phosphatase – 1170 U/L (30.00 -120) with normal serum bilirubin levels.

Initially ultrasound whole abdomen was done which showed mildly enlarged liver but with normal echotexure. Further Contrast Enhanced Computed Tomography (CECT) of whole abdomen was done which revealed hepatomegaly having longitudinal span of 15 centimeters along with splenic & liver parenchymalinvolvement showingheterogeneous attenuation with multiple ill-defined nodular & confluent hypo dense lesions. Fig. 1

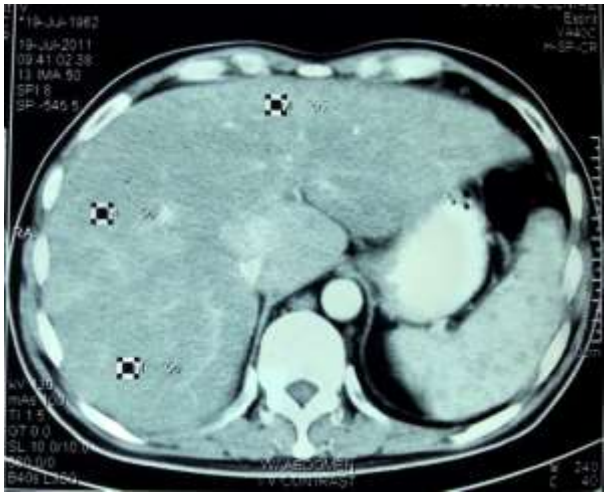

Figure 1. CECT abdomen showing heterogenous attenuation with multiple ill-defined nodular & confluent hypodense lesions in liver and spleen.

Multiple enlarged retroperitoneal (para- aortic and celiac group) and mesenteric lymph nodes (along the Superior mesenteric artery) were also seen. Rest of the abdomen was normal. CECT chest was done that showed multiple mediastinal lymphadenopathies with maximum size of 1.6 x 1.4 cm in sizein subcarinal region with subcentemeter lymph nodes in paratracheal and prevascular location. No hilar lymphadenopathy was seen. There was a nodular high density subpleural focus in the anterior basal segment of LLL (Left Lower Lung) with a wedge shaped area of atelectasis in posterior basal segment of LLL.

Fine needle aspiration (FNAC) of liver was performed after ruling out coagulopathy. FNAC yielded well formed epithelioid cell granulomas which was suggestive of granulomatous pathology. It was followed by angiotensin converting enzymes which was very high with levels of 197 U/L. Serum calcium was in normal range (9.30 mg/dl). Pulmonary function test was within normal limits. In view of inconclusive reports bronchoscopy was performed. There was significant mucosal infiltration in the left main bronchus, left upperlobe bronchus & lingula with unhealthy looking right middle lobe bronchus. Multiple bronchial and transbronchial lung biopsies were taken from left lobe. Bronchoalveolar lavage was taken andsent for AFB smear and culture (BACTEC). Biopsies showed granuloma comprising epithelioid cells, Langhan's type of giant cells & lymphocytes. Some of the giant cells showed calcified inclusions. No necrosis was seen. Stain for acid fast bacilli (AFB) and PAS was negative for microorganisms. BAL was negative for gram stain, AFB smear and pyogenic cultures. Later on AFB culture was also negative.Diagnosis of systemic sarcoidosis was made and treatment with steroids was started to which patient responded symptomatically. Liver enzymes come within normal limits in few weeks. CECT both lung & liver was repeated after 4 months of treatment that showed remarkable improvement in the form of normal liver size & normal architecture of both spleen and liver (Fig. 2) with significant reduction in size of lymph nodes . In CT chest size of lymph nodes were also reduced significantly.

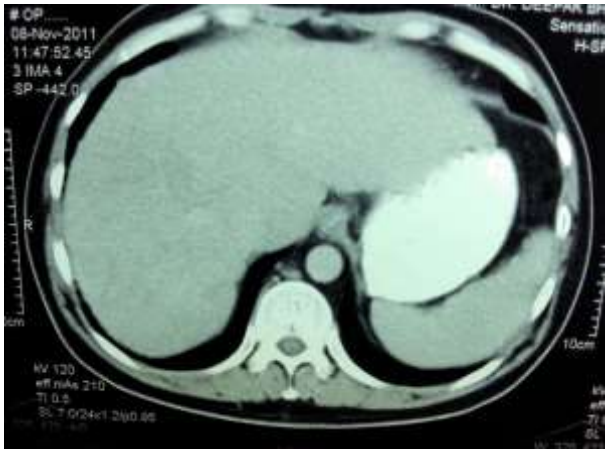

Figure 2.CECT abdomen after four months of treatment showing normal liver and spleen architecture.

DISCUSSION

The incidence of sarcoidosis varies widely throughout the world probably because of differences in environmental exposures, surveillance methods, and predisposing HLA alleles and other genetic factors.[9] In India due to resemblance to tuberculosis, compounded by lack of awareness among physicians and pathologists with lack of diagnostic facilities has all been the reason for under reporting of the sarcoidosis.The true prevalence rate of sarcoidosis in any part of India is undetermined.Among few estimates that are available,sarcoidosis constituted 10-12 cases per 1000 new registrations annually at a respiratory unit at Kolkata.[10]

Granulomatous pathology in the liver is associated with a myriad of disorders. Common causes include immunological disorders (primary biliary cirrhosis, sarcoidosis etc.), infectious agents, foreign bodies, neoplasms and drugs. Infections particularly tuberculosis are the most common cause of hepatic granulomas worldwide while in the west , non-infectious disorders like sarcoidosis and primary biliary cirrhosis are the most frequently encountered etiologies.Hepatic involvement of sarcoidosis has been reported to range between 17 and 90%.[11] In ACCESS

study, involvement of liver due to sarcoidosis was seen in 11.5% patients out of 736 patients.[12]Hepatic granulomas due to sarcoidosis are recognized most commonly as multiple and small nodules with less than 1cm in diameter. They are found in 24-94% of liver biopsies and autopsies. The classic granuloma in sarcoidosis is mainly found in the portal triads with a cluster of large epithelioid cells, often with multinucleated giant cells. [1]These granulomas may cause an asymptomatic rise in liver enzymes or chronic cholestasis. [13]In only 1% of patients, sarcoidosis resulting in cirrhosis with portal hypertension leading to liver failure is reported. [9]

Hepatomegaly due to sarcoidosis in western studies is reported to be around 12% whereas Indian studies showed 14-42% cases with liver enlargement. [5] On computed tomography detection of hepatic and splenic lesions is described in 5% and 15% of patients' respectively. [9]

Angiotensin-converting enzyme (ACE) level may be elevated in 70% of patients with sarcoidosis and is elevated in almost all patients with hepatic sarcoidosis.[14]Elevation greater than two times the upper limits of normal(8-65 U/L)are less common in other diseases.[10] In our case ACE levels were197 U/L. In general, in 20–40% of patients with sarcoidosis, ALP and γ-GTP levels are elevated, but in those with hepatic sarcoidosis the levels are usually much higher. [14]

Uncommon manifestations observed in patients with sarcoidosis from India include intraabdominal lymphadenopathy and hypodense lesions in liver and spleen.[5] Likewise splenic and liver multiple,parenchymal hypodense nodular lesions with enlarged retroperitoneal,mesenteric lymphadenopathy were demonstrated in present case.
For establishing the diagnosis specimens must be procured for histopathological examination from the most accessible site with the least invasive method [5] e.g. transbronchial lung biopsy (TBLB) from lung parenchyma,FNAC/liver biopsy in hepatic involvement or biopsies from sarcoid skin lesions.

For confirming the diagnosis, recognition of characteristic clinical findings, securing histologic evidence of noncaseating granulomas and ruling out other causes of granulomatous disease is a must.[9]No treatment is required in asymptomatic patients. Glucocorticoids are administered in patients who are symptomatic or in whom biopsy confirms granulomatous inflammation. [1]

In this report we describe an experience wherein medical history was remarkable for suspected tuberculosis and the radiological findings of bilateral hilar lymphadenopathy which are typical of sarcoidosis were missing. The treatment strategies for the two diseases (tuberculosis and sarcoidosis) are just the opposite. Immunosuppression is required in sarcoidosis whereas immunosuppression in patients with tuberculosis may lead to dissemination of the disease. We would like to emphasize that such cases are challenging for the physicians as it can mimic other infections and if wrongly treated the results can be devastating. Typical imaging, histological examination and laboratory findings are baseline to establish the diagnosis of sarcoidosis.

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