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Case Report

A case report of multisystemic sarcoidosis with extracardiac shunt

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ABSTRACT

A 61-year-old female patient, presented with complaints of dry cough and gradually worsening dyspnea of 3 months duration, associated with significant weight loss of >14 kg. At presentation, patient was sick, hypoxemic, and tachypneic and system examination revealed fine basal crepitations on auscultation. CT chest showed random pulmonary nodules with bronchiectatic changes. In view of respiratory distress and elevated d-dimer, CT pulmonary angiogram was done and was normal. A contrast echocardiography study hinted towards a significant right to left extracardiac shunt. There was radiological evidence of chronic liver disease with portal hypertension and numerous poorly defined nodular opacities scattered in hepatic and splenic parenchyma in post contrast images. ANA profile was positive for ribosomal-p protein along with elevated serum ACE levels. An ultrasound guided liver biopsy was ultimately performed and histopathology revealed granulomatous lesion in liver compatible with sarcoidosis.

Keywords: Sarcoidosis, Multisystemic, Extracardiac shunt, Masquerader

INTRODUCTION

Sarcoidosis is an inflammatory disease characterised by the presence of non-caseating granulomas.[1] The disease is often multisystemic and requires the presence of involvement in two or more organs for a specific diagnosis.[1] Despite multiple investigations, the cause of sarcoidosis remains unknown. The granuloma is the pathologic hallmark of sarcoidosis.[2-7] The presentation of sarcoidosis ranges from patients who are asymptomatic to those with organ failure.[1] Multisystemic sarcoidosis may pose a diagnostic challenge masquerading as a metastatic disease. Case reports of multisystemic sarcoidosis with extracardiac shunt are rare.[2]

CASE REPORT

A 61-year-old female patient, presented with complaints of dry cough and gradually worsening dyspnea of 3 months duration, with associated significant weight loss of >14 kg. No history of fever, joint pain, or skin rashes. No history of passive smoke inhalation, occupational exposure to fumes, or dust. No prior history of bronchial asthma, pulmonary tuberculosis, malignancy, prolonged immobilisation, or connective tissue disorders. No relevant past surgical, or family history. No history of travel outside state.

Physical examination

On general examination, she was moderately built and poorly nourished (BMI: 20). Conscious, oriented. No pallor, icterus, cyanosis, clubbing, lymphadenopathy, or edema. Skin, hair, nails, eyes, thyroid were normal. Pulse: rate 84/min, regular, normal character, all peripheral pulsations present, no radio-femoral delay. Blood pressure: 130/70 mmHg, sitting, recorded in both upper arms, no postural hypotension. Temperature: 98.6°F. Respiratory rate: 28/min SpO_2: 86% (room air).

System examination was otherwise normal, except for a fine basal crepitations on auscultation.

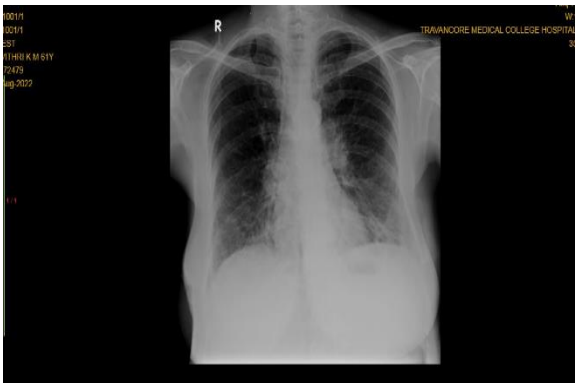

Figure 1: Chest X-ray showed reticular opacities in bilateral lower zone.

Investigations

The blood routine, urine routine, renal function tests, and liver function tests were normal. Chest X-ray showed reticular opacities in bilateral lower zone (Figure 1). CT chest showed random pulmonary nodules with bronchiectatic changes (Figure 2).

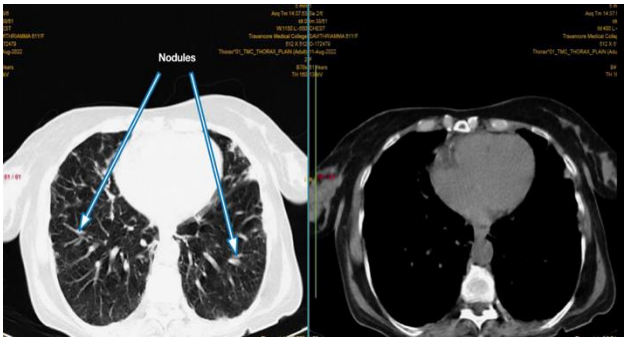

Figure 2: CT chest showed random pulmonary nodules with post covid bronchiectatic changes.

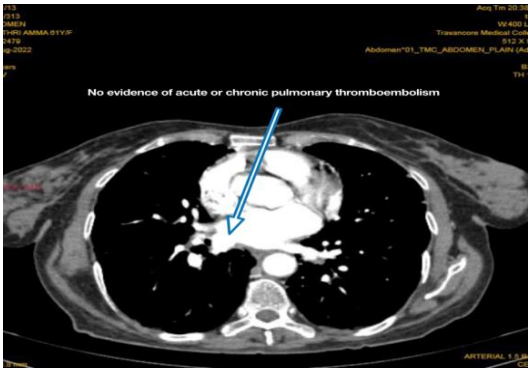

Figure 3: CT pulmonary angiogram showing no evidence of pulmonary thromboembolism.

Sputum CBNAAT was negative. In view of respiratory distress and elevated d-dimer, CT pulmonary angiogram was done and was found to be normal (Figure 3). Despite continuing antibiotics, oxygen support, nebulisation, and other anti-inflammatory medications, respiratory distress and hypoxemia persisted. Hence a 2D echocardiography was performed and was normal. A contrast echocardiography study hinted towards an significant right to left extracardiac shunt (Figure 4). There was sonographical evidence of chronic liver disease with portal hypertension which was confirmed by CT abdomen, which also revealed numerous poorly defined nodular opacities scattered in hepatic and splenic parenchyma in post contrast images (Figure 5). Upper GI endoscopy revealed Grade II esophageal varices secondary to portal hypertension (Figure 6). ANA profile was positive for ribosomal-p protein with elevated serum ACE levels. An ultrasound guided liver biopsy was ultimately performed and histopathology revealed granulomatous lesion in liver compatible with sarcoidosis (Figure 7).

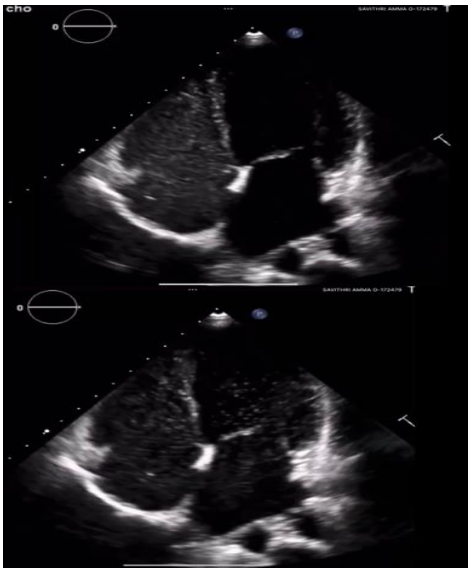

Figure 4: Contrast echocardiography study significant bubbling in LA and LV from RA after 5th cardiac cycle hinted towards an significant right to left extracardiac shunt.

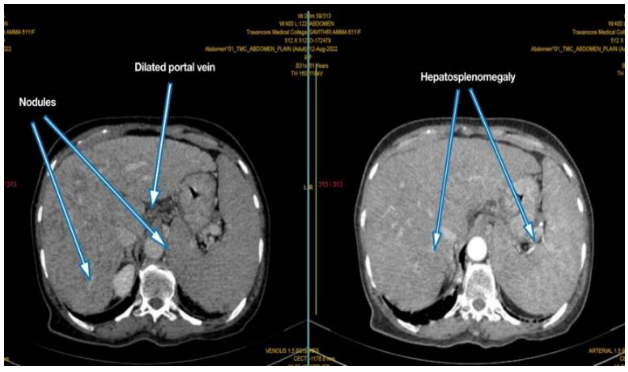

Figure 5: CT abdomen showing numerous poorly defined nodular opacities scattered in hepatic and splenic parenchyma in post contrast images.

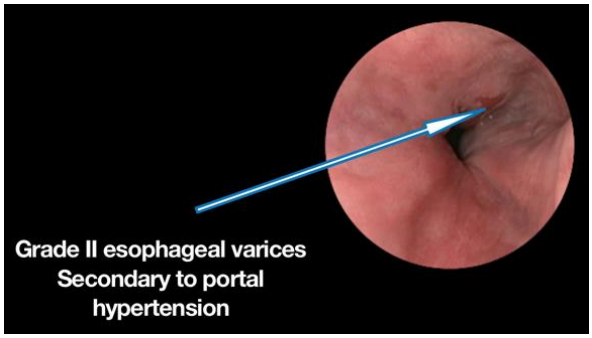

Figure 6: Upper GI endoscopy revealed Grade II esophageal varices secondary to portal hypertension.

Diagnosis

A final diagnosis of multisystemic sarcoidosis with pulmonary nodules, chronic liver disease with portal hypertension, hepato-pulmonary syndrome with extracardiac shunt was made.

Treatment and followup

The patient was started on systemic corticosteroids and immunosuppressants. Tab. Deflazacort 42 mg once daily for 3 weeks, then 36 mg once daily for 3 weeks; and Tab Mycophenolate Mofetil 1000 mg twice daily for 6 weeks. On follow-up, after 1-month significant improvement in the clinical symptoms was noted.

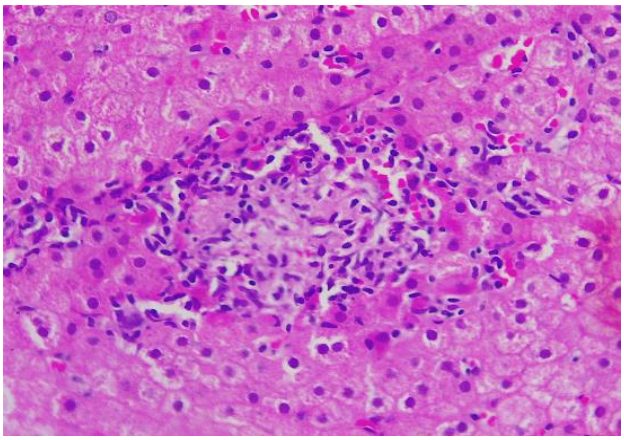

Figure 7: Non-necrotising with a tightly packed central area composed of macrophages, epithelioid cells, multinucleated giant cells, and T lymphocytes that are CD4 positive.

DISCUSSION

Sarcoidosis is best defined in histopathological terms as a disease characterised by the presence in all of several affected organs and tissues of non-caseating epithelioid-cell granulomas, proceeding either to resolution or to conversion into hyaline connective tissue.[1] The disease is often multisystemic and requires the presence of involvement in two or more organs for a specific diagnosis.[1] The granuloma is the pathologic hallmark of sarcoidosis. A distinct feature of sarcoidosis is the local accumulation of inflammatory cells.[1] The presentation of sarcoidosis ranges from patients who are asymptomatic to those with organ failure.[2] Lung involvement occurs in >90% of sarcoidosis patients.[2] 20-30% of patients will have evidence of liver involvement with intrahepatic cholestasis.[2] Skin involvement is eventually identified in over a third of patients with sarcoidosis.[2] The classic cutaneous lesions include erythema nodosum, maculopapular lesions, hyper- and hypopigmentation, keloid formation, and subcutaneous nodules.[3] A specific complex of involvement of the bridge of the nose, the area beneath the eyes, and the cheeks is referred to as lupus pernio and is diagnostic for a chronic form of sarcoidosis. In contrast, erythema nodosum is a transient rash that can be seen in association with hilar adenopathy and uveitis (Löfgren's syndrome).[3] The most common eye manifestation is anterior uveitis[3]. Arrhythmias and conduction blocks can also occur with diffuse or patchy cardiac infiltration, and is common cause of death.[4] The most common hematologic problem is lymphopenia, which is a reflection of sequestration of the lymphocytes into the areas of inflammation. Anemia occurs in 20% of patients, and leukopenia is less common. Hypercalcemia and/or hypercalciuria occur in 10% of sarcoidosis patients.[5] Neurological manifestations includes facial nerve palsy, optic neuritis, lymphocytic meningitis. The usual causes of death related to sarcoidosis are from lung, cardiac, neurologic, or liver involvement. Serum calcium should be determined as part of the initial evaluation of all sarcoidosis patients.[6] Serum levels of angiotensin-converting enzyme (ACE) can be helpful in the diagnosis of sarcoidosis. In sarcoidosis, the presence of hilar adenopathy and a nodular infiltrate is not specific for sarcoidosis[6]. The lymphocyte markers CD4 and CD8 can be used to determine the CD4/CD8 ratio of these increased lymphocytes in the BAL fluid.[7] A ratio of >3.5 is strongly supportive of sarcoidosis but is less sensitive than an increase in lymphocytes alone. The biopsy is the gold standard investigation. The diagnosis of sarcoidosis requires both compatible clinical features and pathologic findings. If the biopsy reveals non-caseating granulomas, an alternative diagnosis such as infection or malignancy must be excluded.[7] Glucocorticoids remain the drugs of choice for extensive disease.[7] Systemic therapies for sarcoidosis are usually immunosuppressive, including glucocorticoids (Prednisone), cytotoxic drugs (Methotrexate, Azathioprine) or biologics (Infliximab). Sarcoidosis is usually a self-limited, non-life-threatening disease. How-ever, organ-threatening disease can occur.[7]

CONCLUSION

Our patient presented with complaints of dry cough and gradually worsening dyspnea of 3 months duration, associated with significant weight loss, mimicking a metastatic disease. Random pulmonary and hepatosplenic nodules with significant right to left extracardiac shunt, portal hypertension further made the case a diagnostic

challenge. Ultimately histopathology report revealing granulomatous lesion supported by evidence of serum ACE levels and ANA profile narrowed the diagnosis compatible with multisystemic sarcoidosis. Clinicians may hone in on the diagnosis of sarcoidosis in the setting of 'classical' clinical syndromes, such as Löfgren's syndrome, lupus pernio, Heerfordt's syndrome. On the other hand, multisystemic sarcoidosis may pose a diagnostic challenge masquerading as a metastatic disease. This report enhances the minimal existing literature on cases of multisystemic sarcoidosis with extracardiac shunt bringing high index of suspicion, early diagnosis and timely intervention that can potentially avoid life-threatening complications and improve quality of life.

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