Sarcoidosis

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ABSTRACT

Sarcoidosis is often put forth as differential diagnosis for many clinical conditions, but incidence of sarcoidosis is very low in Indian population.[1] With availability of new diagnostic tools,more cases are being reported from India. It is also possible that some cases of sarcoidosis might be getting misdiagnosed as tuberculosis or leprosy. We present here a case of sarcoidosis in a middle aged woman, proved adequately by histopathology.

INTRODUCTION

Sarcoidosisis a multi systemic granulomatous disorder, mainly involving lungs, lymphnodes, skin and eyes but can involve many other organs. Worldwide prevalence of sarcoidosis is 20-60 per 100000. It is more common in North European countries, where the incidence is 40 per 10000 population. It is also common in North America, where it is more common and more severe in Afro American people [2] Incidence of sarcoidosis is less in China, India and Africa. This shows probable relationship with colder climate. Some evidence of familial clustering is definitely present but whether it is related to genetic factors or environmental factors is not clear. Link with atypical mycobacteria, Propionbacter acnes, Epstein Barr virus, fungi remains speculative. Non infective precipitating agents proposed are Berillium and other metals, organicantigenslike pine pollens and inorganic dust (clay).[1]

CASE REPORT

MrsB, a 62 years old female, known case of hypertension, came with c/o low grade fever intermittently since 2-3 weeks, 2-3 days of diarrheoa which subsided on treatment and general weakness. On examination, patient looked pale, she had lost about 5-6 kg weight since her last visit 4 months back. There was no icterus, no pedal oedema, no clubbing. Left inguinal lymph nodes were palpable,2-3 in number, non tender, discreet, firm and not adherent to underlying structures. No other lymph nodes were palpable.On systemic examination,CVS was normal, chest was normal except occasional rhonchi, abdominal examination showed liver enlarged 4-5 cm below costal margin, firm, non tender with smooth surface. Spleen was not palpable, there was no free fluid. Neurological examination was normal.

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Investigations

Hb10 gms%, TLC 4600 ,P82,L15,E1,M2, urine routine normal, KFT normal, F &PM sugars normal, S. bilirubin 0.8,SGPT23,SGOT40, alkaline phosphatase 208,ESR38, S.calcium8.9,TBIgG and TBIgM negative, Monteux test negative. S. Angiotensin Converting Enzyme (ACE) was elevated -152units/litre (normal range 8-65),XRC showed increased bronchovascular markings but there was no hilar lymphadenopathy.USG abdomen showed moderate hepatomegaly with increased echogenecity and mild splenomegaly.

MDCT abdominal scan showed hepatosplenomegaly. There were multiple small infiltrating lesions in both lobes of liver along with extensive retroperitoneal lymphadenopathy. Enlarged paraaortic nodes extended from superior mesenteric artery level upto aortic bifurcation, both common and external iliac and inguinal regions. Possibility of neoplastic etiology like lymphoreticular malignancy was considered as more likely cause than metastatic or infective etiology.

MDCT chest scan showed few small nodular lesions in

bilateral lung fields, more marked in upper lobes. Nodules were also located in subpleural region and peribronchovascular distribution. No hilar lymphadenopathy was seen. Impression-? sarcoidosis? tuberculosis.

Inguinal node biopsy report showed features suggestive of non caseating granulomatous lymphadenopathy with possibility of sarcoidosis. There was no evidence of malignancy.

For further confirmation of diagnosis, laparoscopic liver and abdominal lymphnode biopsy was taken with help of the surgeon. Liver parenchyma showed multiple non caseating granulomas with no evidence of malignancy. Five lymph node biopsy sections showed multiple non caseating granulomas surrounded by hyalinised collagen rings. Fiteferraco stain for AFB was negative, no fungus, no Reed Sternberg cells, no malignant cells seen. Features were highly suggestive of sarcoidosis.

A diagnosis of sarcoidosis was made and patient was put on steroid therapy, tab. wysolone 30 mgs BD .Within 15 days patient showed marked symptomatic improvement,liver size regressed from 4-5 cm to 2cm.Steroids were continued for 6 weeks and then tapered over next 2 weeks.

Repeat MDCT abdomen done after completion of treatment showed significant reduction in number and size of abdominal lymphnodes with regression of hepatic lesions also. Now 6months after treatment, patient is completely asymptomatic.

DISCUSSION

Sarcoidosis is a granulomatous disorder of uncertain etiology, seen more commonly in countries with colder climates. Commonly involved organs are lungs, lymphnodes, skin and eyes. Peak age of onset is 20-30 years, second peak of onset is noted in middle age in some countries, especially in women. Racial, geographical and ethnic differences are seen in clinical presentation. Erythema nodosum is more common in Scandinavian and British patients but is uncommon in African and Japanese patients. Cardiac involvement is commoner in Japanese patients where it is the most common cause of death[3]

Diagnosis of sarcoidosis is confirmed on histopathological evidence of widespread non caseatingepitheloid granulomas in more than one organ.

Clinically, sarcoidosis patient may be asymptomatic and hilar lymphadenopathy may be detected in x ray chest done for some other purpose.

General symptoms include fever and weight loss in 5%patients, which was present in our patient. Commonest presentation is insidious respiratory symptoms like cough and exertional breathlessness. Chest auscultation is usually unremarkable, wheeze may be heard. Upper respiratory tract may be involved. Nasal mucosal involvement causes nasal stuffiness which was present in our patient and which subsided after steroid treatment. Nasal bones and larynx may also be involved.

Lung involvement, present in almost all (>90%) patients, [4] may be of two types—bilateral hilar adenopathy (BHL) or pulmonary infiltrates. Infiltrates are more common in upper lobes than lower lobes (as was seen in our patient's x-ray). Fibrosis occurs in some patients with silent loss of lung function. Pleural involvement is rare. Complications like bronchiectasis, pneumothorax, pulmonary hypertension or corpulmonale are rare.

Lofgren's syndrome is an acute illness with erythema nodosum, arthropathy, uveitis, hilar lymphadenopathy, lethargy and mild fever seen in young females [2]

Extra pulmonary manifestations—
Skin-erythema nodosum, granulomatous involvement of old scars giving keloid like appearance, lupus pernio (chilblain like lesion on nostril), maculopapular lesions
Eyes-anterior or posterior uveitis, sicca syndrome
Heart-cardiac arrhythmias, heart blocks, sudden death
Abdomen-hepato splenomegaly, lymphnode enlargement, intrahepatic cholestasis
Neurology - mononeuritis multiplex, cranial nerve (commonly facial nerve) palsy, optic neuritis, seizures, psychosis, hypothalamo - pitutary axis involvement causing Diabetes Insipidus& hyper prolactinemia
Other-parotid and lacrimal gland enlargement, phalangeal bone cysts
Our patient had pulmonary infiltretes, abdominal and

inguinal lymphadenopathy, hepatomegaly with hepatic granulomas and nasal stuffiness.

Investigations for sarcoidosis include lymphocytopenia, mild derangement of LFTs, raised serum calcium, raised Angiotensin converting Enzyme level (ACE)

Alveolar macrophages secrete calcitriol which leads to hypercalcemia, calciuria and renal calcinosis. Alveolar macrophages also secrete ACE, thus raising S.ACE levels [4] Our patient had raised ACE levels but normal calcium levels.

XRC shows bilateral hilar lymphadenopathy, pulmonary infiltrations and fibrosis. 4 stages are described according to radiological findings [4]

Stage1-BHL, stage2-BHL plus pulmonary infiltrets, stage3-only pulmonary infiltrets, stage 4- pulmonary fibrosis. According to this classification, our patient fitted into stage3

Bronchoscopic lung biopsy and BAL may be done. Skin biopsy, lymph node biopsy and liver biopsy also can be done. Differential diagnosis of HL include tuberculosis, lymphoma, carcinoma bronchus.

Histopathology

The typical sarcoid granuloma consists of focal accumulation of epitheloid cells, macrophages and lymphocytes mainly T cells. Multinucleated giant cells are typically seen within the granuloma. Caseation is absent.

Treatment

Spontaneous remission is common. It is difficult to decide when to start treatment. In acute illness with Erythema nodosum, NSAIDs and short course of steroids may be given. Steroids are also indicated in pulmonary, renal or hepatic impairment, ocular or myocardial involvement and for hypercalcemia. [2] In severe disease, methotrexate or azathioprim is used.

Prognosisis good for majority patients. 60-70% patients experience resolution within 2 years, in 10-30 %patients there may be chronic progression.

REFRENCES

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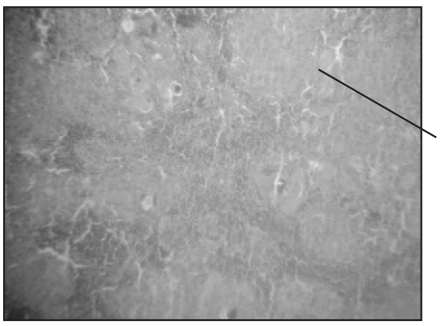
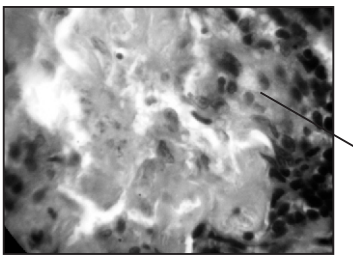
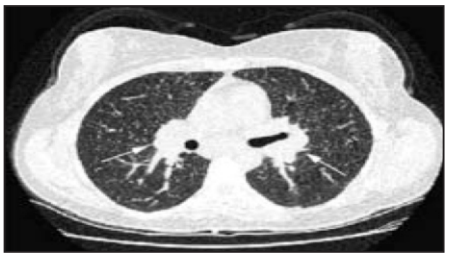
2. *Harrison's textbook of medicine, 17th edition, vol. 2: 2135-36*
3. *Lannuzzi MC, Rybicki BA, Teirstein AS.Sarcoidosis, N England J of Med 2007; 357: 2153-65*
4. *API Textbook of medicine, 9th edition, vol 2:1758-63*



Multiple granulomas in inguinal lymphnode biopsy



Wesenberg – Hamasaki body (A brown colored oval body) (W.H bodies are brown coloured inclusion bodies found in granuloma containing lymphnode in sarcoidosis)



HRCT chest showing bilateral pulmonary infiltrates with mild hilar adenopathy