Sarcoidosis Presenting as Interstitial Pneumonitis

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Citation

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Abstract

Sarcoidosis is characterized by the presence of non-caseating granulomas. We report the case of a young woman presenting with chronic dyspnea, hypoxemia and bilateral interstitial infiltrates on chest radiographs. Open lung biopsy was consistent with usual interstitial pneumonitis. Treatment with steroids produced a marked improvement in symptoms. Five years later the patient presented with fever, bilateral hiliar and mediastinal lymphadenopathy. Work up for infectious diseases was negative; a mediastinal biopsy was consistent with Sarcoidosis with non-necrotizing granulomas. She was started again in steroids with complete resolution of symptoms and partial improvement in lung infiltrates.

Sarcoidosis can mimic many diseases and maybe idiopathic pulmonary fibrosis (IPF) is not an exemption. Those patients with an "atypical" clinico-radiologic-pathologic diagnosis of IPF should be followed closely in order to identify any evidence of Sarcoidosis.

INTRODUCTION

The characteristic lesion in sarcoidosis is the presence of noncaseating epithelioid cell granuloma[1] and this finding is considered essential in establishing a pathologic diagnosis of sarcoidosis. We present a rare case of sarcoidosis presenting with pathologic features of Usual Instertitial Pneumonitis (UIP) without granuloma.

CASE REPORT

In 1997 a healthy 34 year-old African-American woman presented with a 6-month history of nonproductive cough and dyspnea on exertion. Bilateral dry crackles were heard at lung bases, remaining of examination was normal, no clubbing was appreciated.

She was hypoxemic and the chest radiograph (CXR) and chest tomogram (CT) showed bibasilar interstitial infiltrates with no lymphadenopathy. Pulmonary function test (PFT) revealed restrictive disease with DLCO of 23%. (Table 1)

Table 1: Serial Pulmonary Function tests and Oxygenation

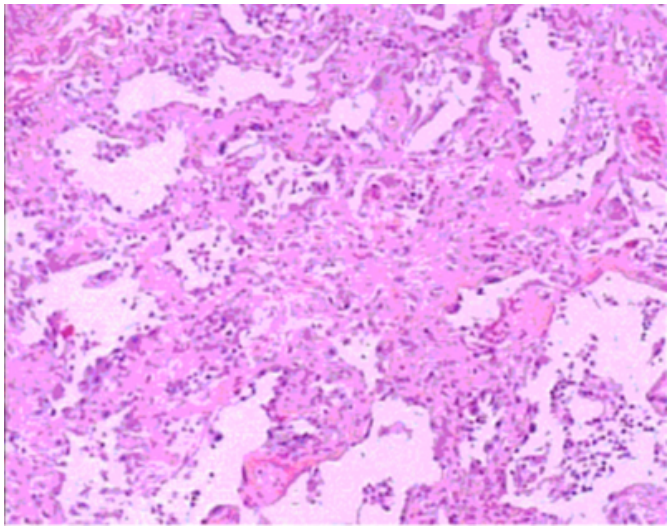
Figure 1

	June 1997	March 1998	July 2001	March 2002	July 2002
FEV1	44%	46%	68%	71%	66%
FEV1/FVC	111%	83%	87%	87%	85%
VC	42%	43%	59%	63%	60%
TLC	49%	45%	61%	68%	54%
RV	60%	44%	58%	72%	37%
DLCO	23%	31%	46%	40%	42%
PaO2	56 mmHg	70 mmHg	75 mmHg	54 mmHg	110 mmHg

Laboratory including connective tissue disease markers, HIV antibody, echocardiogram and ventilation/perfusion test were negative. Open lung biopsy showed active interstitial pneumonitis with focal fibrosis and pulmonary vascular hypertensive changes consistent with UIP (Fig 1).

Figure 2

Figure 1: High magnification of surgical lung biopsy showing moderate interstitial fibrosis, chronic inflammatory cell infiltrate and hyperplasia of alveolar lining cells. (Hematoxylin-eosin; 10x10)

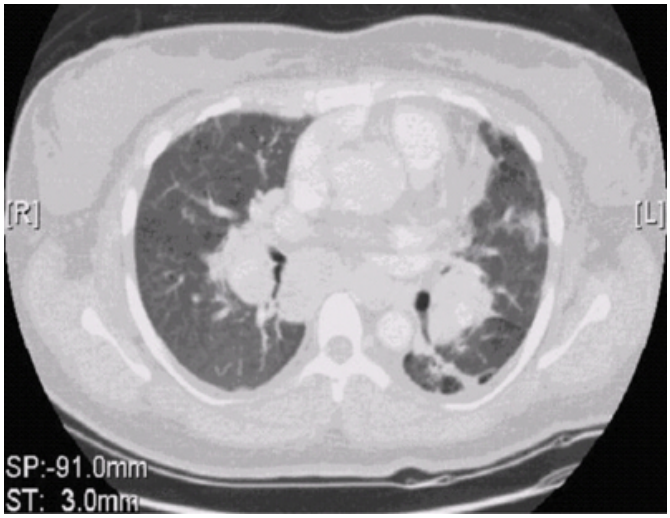


Special stains were negative for infectious microorganisms. No granuloma or giant cells were seen. The patient was discharged on 60 mg of prednisone daily and oxygen. Slowly her dyspnea improved and she was able to stop the oxygen and steroids.

In March 2002, she returned with dyspnea on exertion, dry cough and non-specific joint pain in knees and ankles. She was febrile (39 C), tachypneic with basilar lung crackles and swelling of ankles and knees. Her PaO_2 was 54 mm Hg. New bilateral hilar and mediastinal lymphadenopathy with unchanged infiltrates were seen on CXR and CT (Fig 2).

Figure 3

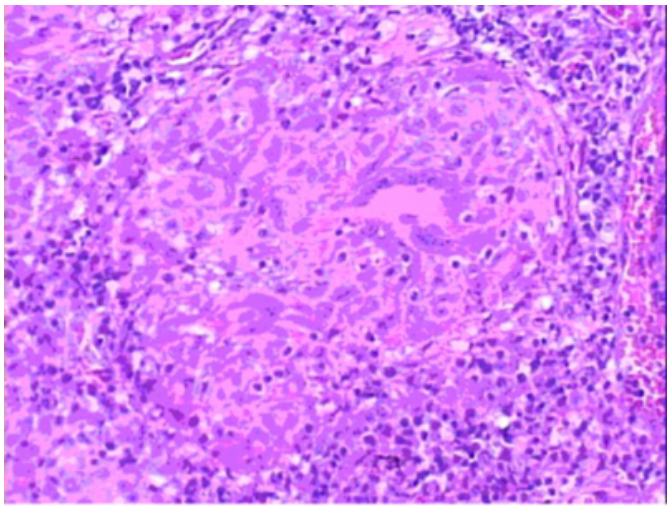
Figure 2: Chest CT scan demonstrating extensive bilateral hilar and mediastinal lymphadenopathy



Collagen disease markers were negative; serum angiotensin-converting enzyme level was elevated. Differential diagnosis included lymphoma or other malignancy, mycobacterium or fungal infection and sarcoidosis. Mediastinal lymph node biopsy revealed non-necrotizing granuloma (Fig3).

Figure 4

Figure 3: High magnification of tracheal lymph node with sarcoid granuloma comprised of epitheliod cells, well defined giant cells and lymphocytes. (Hematoxylin-eosin; 10x10)



Cultures and stains were negative. The clinico-pathologic findings were consistent with sarcoidosis. She resumed the prednisone with marked improvement in dyspnea, hypoxemia, and complete resolution of the cough, fever and joint symptoms. As of January 2004 she remains stable, with mild dyspnea of exertion

DISCUSSION

This case highlights the dilemma that clinicians encounter when confronted with an unexpected pathologic diagnosis. The diagnosis of interstitial lung diseases (ILD) depends on epidemiologic, clinical, radiological and pathological findings. The most frequent ILD are sarcoidosis, idiopathic pulmonary fibrosis (IPF) and connective disease ILD. The clinical course of IPF is invariably one of gradual deterioration with minimal response to steroid therapy[2]. End-stage sarcoidosis can be associated with fibrosis and few granuloma.[3] The natural history and prognosis is highly variable, with a tendency to wax and wane, spontaneous remission in stage III are seen in 10 to 20% of cases.[4] Nonspecific interstitial pneumonitis was seen in 62% of 128 granuloma-containing specimens from patients with sarcoidosis[5]. The age and clinical response in our patient was atypical of IPF and more consistent with the diagnosis of sarcoidosis, although the initial CT, PFT and lung biopsy were compatible with UIP. Connective disorder with sole involvement of the lung was unlikely as well. Based on this case we suggest that interstitial pneumonitis and fibrosis suggestive of IPF can be seen in sarcoidosis without granuloma formation. Sarcoidosis can mimic many diseases and maybe IPF is not an exemption. Those patients with an "atypical" clinico-radiologic-pathologic diagnosis of IPF should be followed closely in order to identify any evidence of sarcoidosis.

References

1. Rosen, Y 1994. Sarcoidosis. In D. H. Dail and S. P. Hammer, editors. Pulmonary Pathology, 2nd ed. Springer-Verlag, New York. 13-645
2. American Thoracic Society/European Respiratory Society International Multidisciplinary Consensus Classification of the Idiopathic Interstitial Pneumonias. Am J Respir Crit Care Med 2002; 165: 277-304
3. Sheffield E., Pathology of Sarcoidosis Clin. Chest Med.1997; 18:741-755
4. American thoracic society: Statement on Sarcoidosis. Am J Respir Crit Care Med 1999; 160: 736-755
5. Rosen Y, Athanassiades TJ, Moon S et al. Nongranulomatous interstitial pneumonitis in sarcoidosis. Relationship to development of epithelioid granulomas. Chest 1978; 74:122-125

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