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Sarcoidosis Developing Usual Interstitial Pneumonia Successfully Treated With Lung Transplantation: A Case Report

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Abstract Sarcoidosis and usual interstitial pneumonia (UIP) may occur in the same patient, though this is rare. We report a unique case of sarcoidosis that was complicated by UIP resulting in end-stage respiratory failure. He successfully underwent lung transplantation.

Keywords: *Sarcoidosis, Usual interstitial pneumonitis (UIP), interstitial lung disease*

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1. Introduction

Although uncommon, sarcoidosis and usual interstitial pneumonia (UIP) may occur simultaneously in the same patient [1]. We report a unique case of sarcoidosis that was complicated by UIP resulting in end-stage respiratory failure that was successfully treated by lung transplantation.

2. Case Presentation

The patient is a 56-year-old Asian Indian, born in South Africa, with a history of biopsy proven sarcoidosis for the last 19 years. The original presentation,at age 37, was with coughing, dyspnoea and fatigue. Examination revealed generalised lymphadenopathy and hepatosplenomegaly and on chest radiograph, bilateral upper and midzone nodular infiltrates with hilar lymphadenopathy (Figure 1). A supraclavicular lymph node biopsy confirmed a diagnosis of sarcoidosis (Figure 2). His lung functions showed mild obstruction with normal diffusion capacity in October 2005 (Table 1) and he was treated with systemic and inhaled corticosteroids with marked improvement. He was subsequently maintained on low dose oral corticosteroids (OCS) and the chest x-ray and High Resolution Computerised Tomographic (HRCT) changes also improved with the only residual abnormalities being bilateral upper zone volume loss and mild bronchial wall thickening. The lung functions similarly stabilised for a period of approximately 5 years.

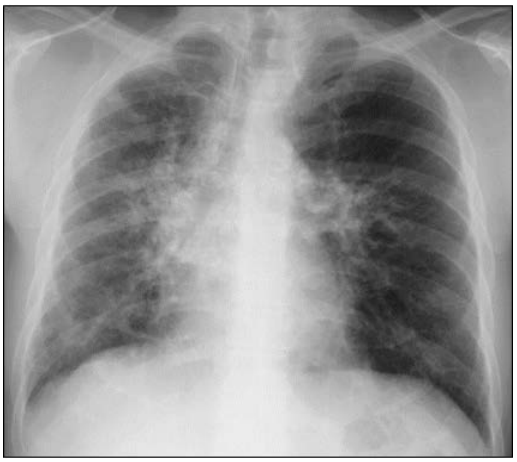

Figure 1. Chest x-ray showing bilateral hila lymphadenopathy and pulmonary infiltrate

Table 1.

Date	FEV1 (%predicted)	FVC (% predicted)	FEV1/FVC Ratio	RV (% predicted)	TLC (% predicted)	DLCO (% predicted)
2005/10	1.88 (73%)	2.64 (83%)	71	1.76 (100%)	4.55 (89%)	6.62 (90%)
2006/11	1.77 (66%)	2.46 (74%)	71	1.30 (69%)	3.98 (73%)	5.87 (76%)
2007/5	1.71 (64%)	2.27 (69%)	75	1.63 (85%)	4.04 (75%)	5.91 (77%)
2008/1	1.44 (54%)	2.09 (63%)	68	1.28 (67%)	3.34 (62%)	4.00 (52%)
2008/3	1.46 (55%)	1.97 (60%)	73	1.22 (64%)	3.26 (60%)	2.93 (38%)
2008/5	1.11 (42%)	1.54 (47%)	72	1.16 (60%)	2.83 (52%)	2.25 (29%)

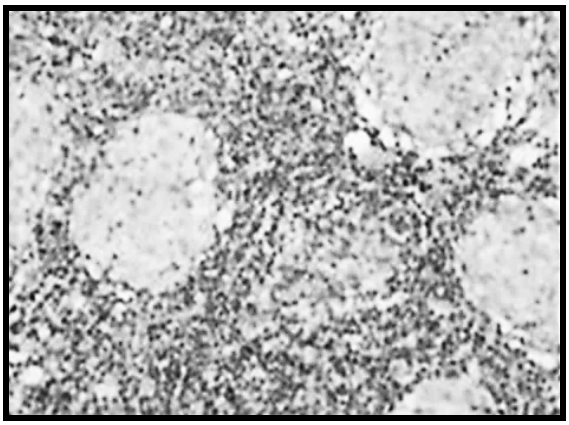

Figure 2. Peripheral lymph node biopsy showing non caseating granulomata

In the last 3 years, from late 2005 onwards, he had a series of exacerbations associated with acute deterioration in his clinical condition and lung function (Table 1).

Initially it was thought that the worsening symptoms were from a recurrence of the sarcoidosis, and he was treated with higher doses of OCS, methotrexate and bronchodilators. Despite this he continued to deteriorate with a steady decline in lung function parameters. This coincided with symptomatic deterioration with an incessant non-productive cough, dyspnoea on mild exertion and worsening malaise and fatigue. In late 2008, the chest x-ray (Figure 3) and HRCT (Figure 4 and Figure 5) showed fibrosis and honeycombing with marked volume loss especially of the right lung, with an appearance similar to that of progressive massive fibrosis. There was no ground glass infiltratenormediastinal lymphadenopathy.

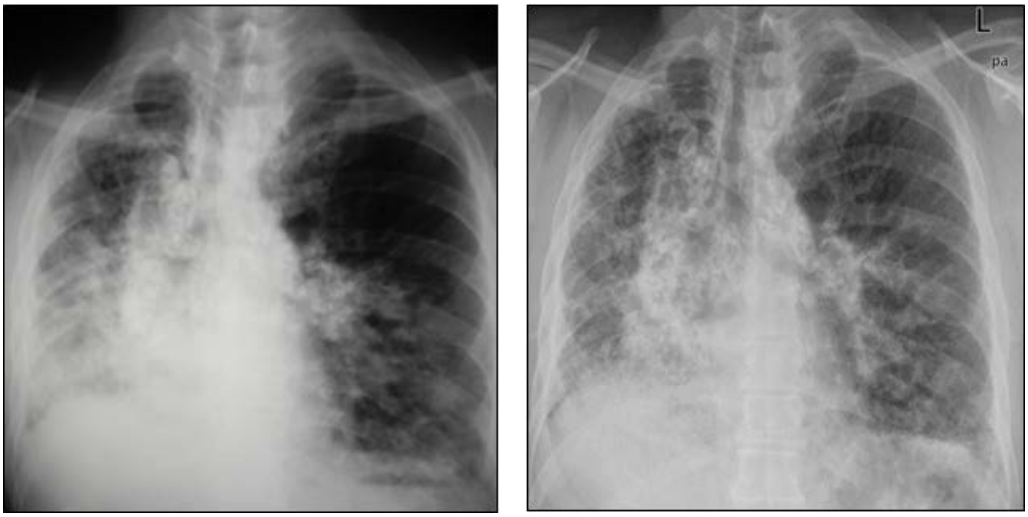

Figure 3. Chest x-ray showing progressive scarring and volume loss associated with fibrosis mainly on the right lung

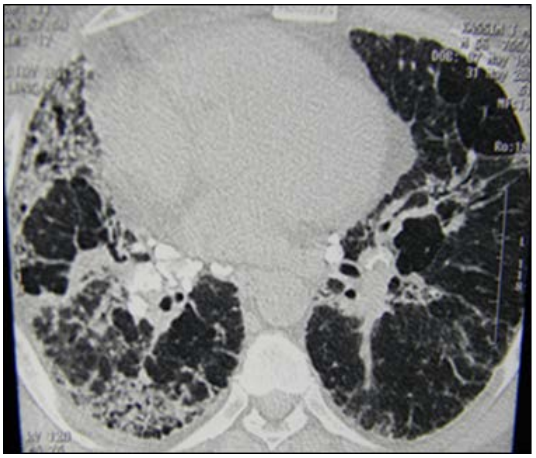

Figure 4. HRCT chest showing features of UIP with honeycombingand fibrosis

He was subjected to an open lung biopsy and a diagnosis of UIP was confirmed histologically (Figure 6). He continued to deteriorate progressively; to the point that he could barely walk a few metres and remained dyspnoeic with minimal effort. The lung function showed severe restriction with a DLCO of 29% despite azathioprine, OCS and n-acetyl cysteine (NAC). At this stage he was assessed for lung transplantation and fortunately, a single lung became available and shortly thereafter, he was successfully transplanted (Figure 7). The explanted lung confirmed the features of UIP with no active sarcoidosis granulomata.

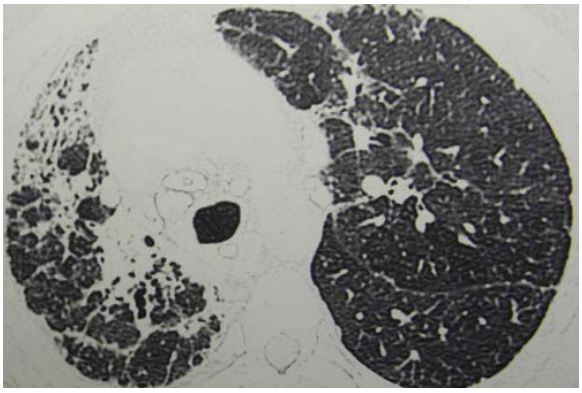

Figure 5. HRCT chest showing subpleural nodules and minimal scarring on the left and extensive fibrosis on the right mid zone

3. Discussion

Sarcoidosis is a systemic granulomatous disease of unknown aetiology. The clinical course and presentation is variable and the pulmonary changes, the most common site of involvement, generally heal without scarring, and rarely progress to significant fibrosis if managed appropriately with OCS during relapses [2]. However, a small proportion of these patients go on to develop

progressive disease associated with pulmonary fibrosis. When the disease is active however, biopsy generally shows, in addition to fibrosis, the granulomata described above.

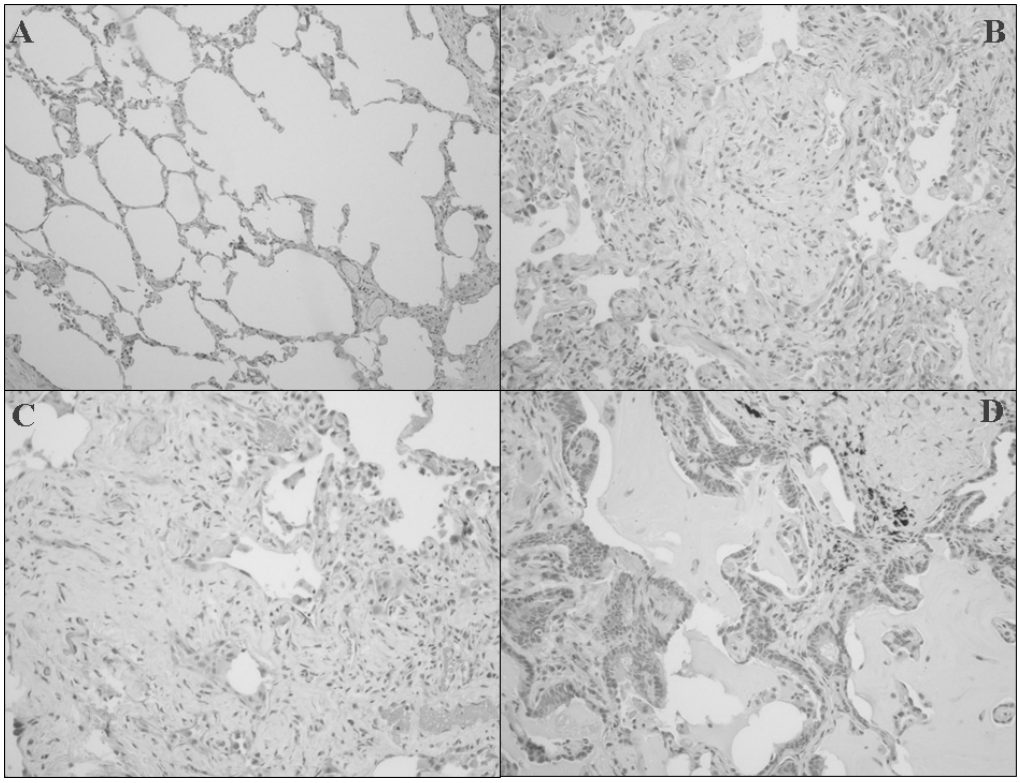

Figure 6. Histopathology (hematoxylin-eosin–stained slide, original × 20) prior to transplantation and explanted lung showing normal lung (A), fibroblastic foci (B), fibrosis (C), and cysts (D)

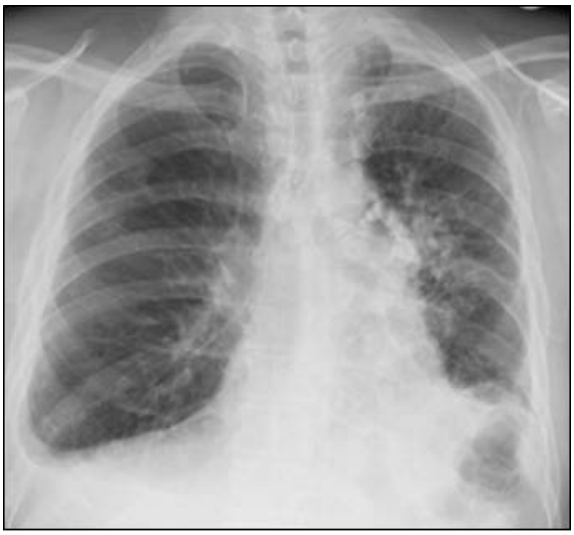

Figure 7. Chest x-ray post-transplantation

The interstitial lung diseases are a heterogeneous group of disorders characterized by inflammation and fibrosis of the pulmonary interstitium. Usual interstitial pneumonitis (UIP), the commonest of the idiopathic interstitial pneumonias, is a chronic, progressive, fatal form of fibrotic lung disease occurring primarily in older adults and characterized by shortness of breath during exertion and ultimately respiratory failure. There is currently no effective therapy available for this condition. Open lung biopsy is often necessary for diagnosis and to evaluate prognosis [3]. Both the above conditions are classified as diffuse parenchymal lung diseases and this patient had the sarcoidosis for 19 years prior to the development of UIP.

4. Conclusion

It is conceivable that the final stages of pulmonary sarcoidosis may be similar histopathologically to those of UIP. Although end stage fibrosis from sarcoidosis is considered to be distinct from UIP, there is much debate regarding the pathogenesis and morphology [4,5]. However, the relentless progression despite therapy, the absence of granulomata on biopsy and a normal serum angiotensin converting enzyme (ACE) level make the possibility that the sarcoidosis was complicated by the development of UIP more likely. The fact that one pulmonary disease is diagnosed initially does not exclude the possibility that another may occur in the same patient. It is critical to bear this in mind as the therapeutic approach may be entirely different.

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