

## **'Trigger related phenotypes in sarcoidosis: Does silica induced sarcoidosis exists?'**

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### Introduction

Sarcoidosis is a systemic disease, characterized by noncaseating granuloma. Its etiology is unknown, however suspected causes are inhaled (in)organic agents including metals and silica. Chronic beryllium disease (CBD) and sarcoidosis can be indistinguishable. Diagnosis of CBD is based on beryllium exposure, together with granulomas in lung biopsies and a positive beryllium lymphocyte proliferation test (LPT) (ATS statement, 2014). In comparisons to CBD, diagnostic criteria for silicosis are not well defined.

### Case

We present a 49 year old male plasterer with complaints of fatigue and dyspnea during exercise. The referring hospital made a previous diagnosis of pneumoconiosis. HRCT-scan showed lymphadenopathy with peripheral located calcifications, nodular abnormalities and fibrotic components. On those observations, a distinction between pneumoconiosis and sarcoidosis could not be made. Lung biopsies showed inflammatory infiltration, non-necrotizing granulomas including multiple nucleated giant cells with hyalinization and birefringent material. Energy-dispersive X-ray spectroscopy analyses resulted in the finding of aluminum, silicon and titanium. A LPT showed a positive stimulation index of 3.2 for silica. The combination of the above findings led to the working diagnosis of sarcoidosis. Since the disease was progressive, treatment with prednisone was started. As prednisone induced severe weight gain, therapy was switched to azathioprine. However, further pulmonary deterioration occurred. Therefore, infliximab treatment was started. A HRCT-scan and PET-scan were made before and after 7 months of infliximab treatment. The latter scans showed both decrease in consolidations at the HRCT-scan as decrease in activity at the PET-scan (fig. 1).

### Discussion

No effective silicosis treatment exists. Beneficial effects of corticosteroids have only been found in a few cases. This is the first case describing a patient with silicosis or, in our opinion, silica induced sarcoidosis treated with TNF- $\alpha$  antibodies. The effect of anti-TNF- $\alpha$  treatment in silicosis patients is unknown, however refractory sarcoidosis patients treated with infliximab have shown good responses. Our patient had silica exposure, granulomas and a positive silica LPT. Referring to the ATS guidelines on CBD, in our opinion diagnosis of silica induced sarcoidosis could be made here. Therefore, searching for trigger-related phenotypes in sarcoidosis could broaden therapeutic options in some patients otherwise determined as pneumoconiosis.