

Pulmonary multiple cystic lesions in a patient with Behçet's syndrome

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SUMMARY

Pulmonary involvement, mainly originating from vasculitis, is one of the features of Behçet's syndrome (BS). We describe, for the first time in literature, computerised tomography images of a male BS patient with multiple pulmonary cystic lesions possibly originated from vasculitis or bronchiolar stenosis.

Key words: Behçet's syndrome; pulmonary cysts; pulmonary artery aneurysm.

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■ CASE REPORT

Behçet's syndrome (BS) is a multisystem vasculitis with unknown etiology (1). Pulmonary artery aneurysms, arterial and venous thrombosis, pulmonary infarction, recurrent pneumonia, bronchiolitis obliterans organised pneumonia, multifocal cavitary lesions and pleurisy are the main features of pulmonary involvement (2, 3). Nevertheless, pulmonary cystic lesions have not been reported previously in patients with BS. For the first time in literature, chest computerised tomography (CT) images were presented of a BS patient with multiple pulmonary cystic lesions.

A 38-year-old male patient, diagnosed as BS according to ISG diagnostic criteria for BS (4), was referred to the emergency unit with complaints of haemoptysis. He had a history of oral aphthous and genital ulcers and erythema nodosum such as skin lesions. Oral aphthous ulcers, scars of genital ulcers and hyperpigmentations related with skin lesions were detected in his first physical examination. Moreover, pathergy reactions were seen in phlebotomy sites during following evaluation. Also, the pathergy test was found positive at a later visit. We ordered a chest CT scan to investigate the etiology for haemoptysis. Pulmonary artery aneurysm (Figure 1) and nonspecific

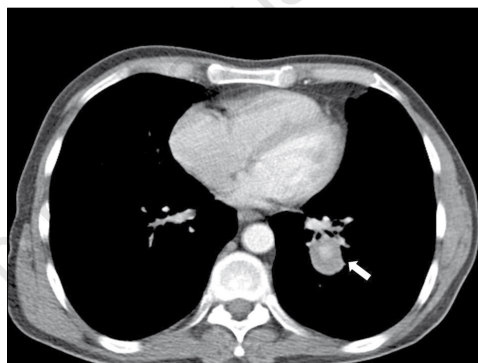


Figure 1 - Image of the former chest CT (mediastinal window). The arrow shows pulmonary artery aneurysm.

multiple nodules (Figure 2A) were found in his first chest CT scan. Furthermore, he was also concomitantly diagnosed with uveitis. Oral steroid, interferon-alfa-2a and monthly cyclophosphamide treatment was started. Then, after the third month of treatment, before evaluating radiological response to the treatment, he missed the follow up. Until this time, he was clinically stable without any symptoms or signs related to BS. One year after the diagnosis, while untreated, he was admitted to the emergency unit with the complaint of haemoptysis. In his second chest CT, pulmonary multiple cystic lesions were found in his right hemithorax (Figure 2B).

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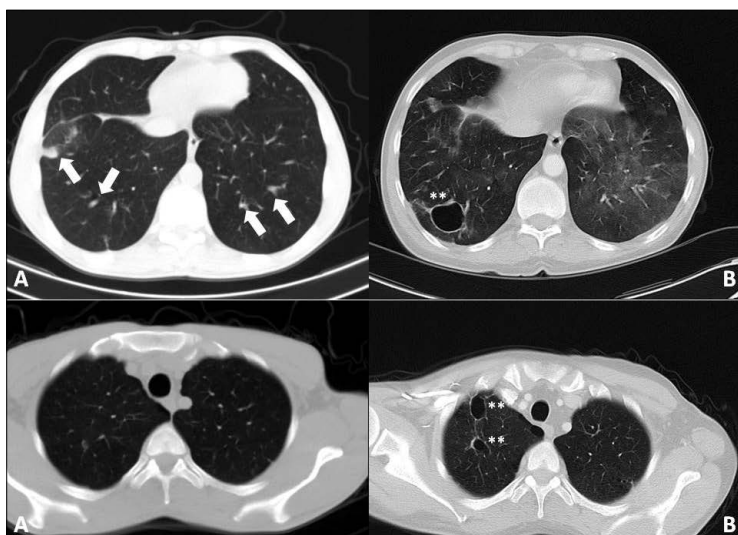


Figure 2 - A) Images of the former chest CT (parenchymal window). The arrows show non-specific pulmonary nodules. B) Images of the latter chest CT (parenchymal window). The asterisks show multiple cystic lesions.

We planned to conduct pulmonary parenchymal biopsy, but the patient refused the procedure. He had no fever and purulent expectoration on both of his applications. Also, the purified protein derivation test was smaller than five millimetres, sputum cultures for tuberculosis, fungus and other bacteria were negative. To exclude the possibility of connective tissue diseases and vasculitis, we ordered serological parameters including anti-nuclear antibody, anti-neutrophil cytoplasmic antibody, and antibodies to extractable nuclear antigens. All of the serological tests were negative. Furthermore, he had no active urine sedimentation and proteinuria. Therefore, we excluded other diseases apart from BS according to his diagnostic findings related with BS and negative serological tests. We then re-started parenteral cyclophos-

phamide treatment with pulse and oral steroids. After six months of cyclophosphamide therapy, we observed an expansion of the pulmonary aneurysm in angiography with magnetic resonance. Then, we switched the treatment to infliximab. Thereafter, we observed regression of the pulmonary aneurysm with six months of infliximab treatment.

■ DISCUSSION AND CONCLUSIONS

There were two possible explanations for this pulmonary findings related with BS. Firstly, the nonspecific nodules in the lung parenchyma might be originated from small arterial vasculitis as occurs frequently with pulmonary artery aneurysm (5). Then subsequent parenchymal infarction in connection with vasculitis might cause multiple cystic lesions. Secondly, bronchiolar stenosis/occlusion due to peribronchial inflammatory infiltration might cause cystic lesions with leading air trapping and distal airway dilatation as can be seen in lymphocytic interstitial pneumonia.

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