

## Analysis of clinical and imaging features of Behcet's disease-related pulmonary lesions: a series of eight cases

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*To the Editor:* Behcet's disease is a chronic vascular inflammatory disease that involves multiple systems of the body. Although the incidence of pulmonary damages is low in Behcet's disease, most of them are serious. The initial pulmonary damages of Behcet's disease are mainly manifested with mild cough, chest pain, and slight hemoptysis, which are easily misdiagnosed as pulmonary embolism, pneumonia, or other diseases. Although some studies have reported the cases of pulmonary damage in Behcet's disease, few studies have summarized the results of its specific pulmonary imaging. In this study, we described the diagnosis and treatment of eight cases of Behcet's disease-related pulmonary damage. Especially, we reviewed their clinical symptoms, especially pulmonary imaging data, to improve the understanding of this disease.

Among the eight patients, the average age was 35 years, and the average course of illness was 10 months. Hemoptysis, cough, and sputum were the most common symptoms, occurring in six, six and five patients in this study, respectively. Recurrent oral ulcers were found in six cases, vulvar ulcers were detected in one case, and iridocyclitis was found in one case. Echocardiography (Philips, GE, Wisconsin, USA) was performed on all patients, and the results suggested that there were four cases of pulmonary hypertension, of which three cases were shown with right ventricular enlargement. During the previous diagnosis and treatments, all patients had experienced misdiagnosis as pneumonia, pulmonary embolism or tuberculosis prior to admission.

Computed tomography pulmonary angiography (CTPA) (64 detector computed tomography [CT] scanner, Light-Speed VCT, GE, Waukesha, Wisconsin, USA) was performed on all patients. We found that seven cases of pulmonary arterial damage were above the lobar artery

level, and one case was at the pulmonary arteriole level. Regarding lesion imaging types, there were three types of Behcet's disease-related pulmonary arterial damage, including pulmonary arterial occlusion, aneurysmal formation, and *in situ* thrombosis formation [Figure 1]. Based on the number of pulmonary artery branches above the lobar artery level, there are 64 pulmonary artery branches in eight patients. In the pulmonary artery branch, the incidences of occlusive lesions, *in situ* thrombosis and pulmonary aneurysmal dilatation were 9.4% (6/64), 9.4% (6/64), and 6.3% (4/64), respectively. Regarding the location of lesion in all patients, there were three lesions on the right pulmonary artery trunk (4.7%, 3/64), six on the right lower lobe artery (9.4%, 6/64), four on the left lower lobe artery (6.3%, 4/64), two on the left pulmonary artery trunk (3.1%, 2/64), and one on left upper lobe artery (1.6%, 1/64). There are 48 pulmonary lobes in eight patients, and the left tongue segment was calculated as an individual lung lobe in this study. Mosaic, pulmonary infarction, consolidation, cavity, and cord shadow were five types of Behcet's disease-related pulmonary parenchymal damage imaging. In this study, the incidences of mosaic, pulmonary infarction, consolidation, cavity, cord shadow on 48 pulmonary lobes were 18.8% (9/48), 12.5% (6/48), 8.3% (4/48), 6.3% (3/48), and 2.1% (1/48), respectively. Among them, pulmonary infarction and cavity were consistent with the location of pulmonary arterial damage. One patient was misdiagnosed as mycoplasma pneumonia according to the imaging features, which showed multiple and wandering changes in bilateral pulmonary parenchyma [Figure 1F]. And the needle biopsy showed that the lesion was pulmonary small vessel wall inflammation, which was consistent with Behcet's disease-induced pulmonary small vessel inflammatory changes.

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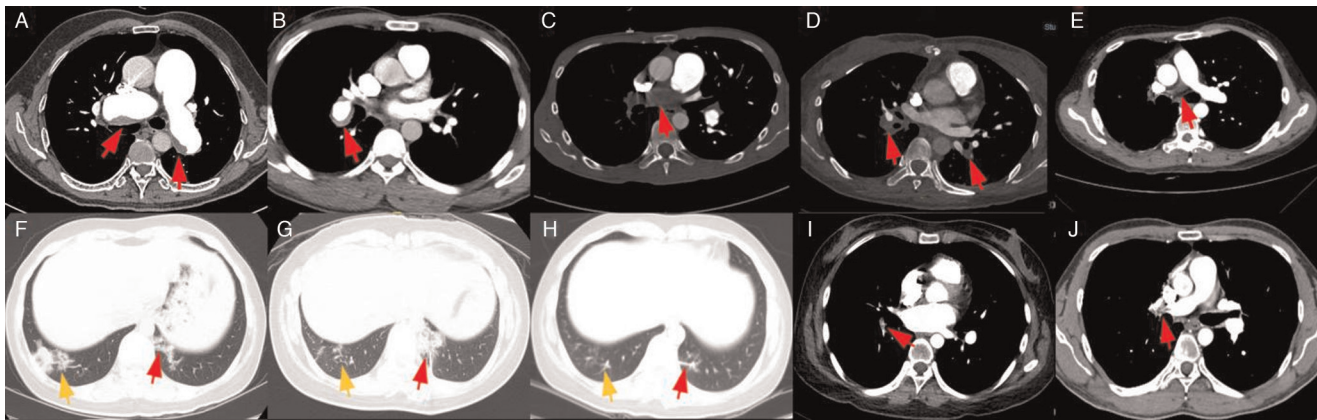
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**Figure 1:** Imaging characteristics of lung in patients with Behcet's disease-related pulmonary damage. (A) Male, 59 years old, hospitalized for chest tightness for 15 days. Behcet's disease was stable-stage. CTPA showed dilation and *in situ* thrombosis formation of the bilateral pulmonary arteries (red arrow). (B) Male, 37 years old, hospitalized for cough, sputum, and fever for more than seven months and hemoptysis for more than two months. Physical examination showed an oral ulcer. Behcet's disease was active-state. CTPA showed aneurysmal dilatation of the right pulmonary artery (red arrow). (C) Male, 32 years old, hospitalized for cough, hemoptysis, and dyspnea for more than eight months. Physical examination showed an oral ulcer. Behcet's disease was active-state. CTPA showed *in situ* thrombosis formation of the right main pulmonary artery trunk (red arrow). (D) Female, 33 years old, hospitalized for cough, sputum for 20 days, and hemoptysis for five days. Physical examination showed an oral ulcer and genital ulcer. Behcet's disease was active-state. CTPA showed right lower and left lower lobe arterial occlusion (red arrow). (E) Male, 30 years old, hospitalized for cough, sputum, fever, hemoptysis, and dyspnea for 50 days. Physical examination showed an oral ulcer. Behcet's disease was active-state. CTPA showed right main pulmonary arterial occlusion (red arrow). (F–H): Female, 23 years old, hospitalized for cough and sputum for more than six months. Physical examination showed an oral ulcer and iridocyclitis. Behcet's disease was active-state. The F and G display the pre-treatment CTPA: an exudative lesion of right lower lobe can be self absorbed before treatment (yellow arrow), while pulmonary parenchymal lesion of left lower lobe was enlarged (red arrow). The H displays the post-treatment CTPA, in which the post-absorption residual shadows were present bilaterally. (I) Female, 31 years old, hospitalized for cough, sputum and hemoptysis for more than one year. Physical examination showed an oral ulcer. Behcet's disease was active-state. CTPA showed right lower lobar arterial occlusion (red arrow). (J) Male, 32 years old, hospitalized for hemoptysis, and dyspnea for more than four years. Behcet's disease was active-state. CTPA showed right main pulmonary arterial occlusion (red arrow). CTPA: Computed tomography pulmonary angiography.

During hospital treatments, seven of the eight patients were diagnosed with active-state Behcet's disease, and one case was diagnosed with stable-stage Behcet's disease. Among them, four patients had normal erythrocyte sedimentation rate and C-reactive protein, although with evidence of recent pulmonary arterial inflammatory activity. Of the seven patients with active-state Behcet's disease, four patients received hormone, immunosuppressive, and anticoagulant therapy, two patients received hormone and anticoagulant therapy, and one patient received hormone and immunosuppressive therapy. After treatment, the disease of seven patients with active-state Behcet's disease was controlled, and the symptoms were improved. The patient diagnosed with stable-stage Behcet's disease continued to receive the previous immunosuppressive treatment.

The main findings of this study were as follows: (1) Slight hemoptysis, cough, and sputum were the main manifestations of Behcet's disease-related pulmonary damage, while the incidence of genital ulcer and ocular damage was not high; and (2) CTPA was the main method to detect Behcet's disease-related pulmonary damage, and *in situ* pulmonary arterial thrombosis formation and lobar arterial occlusion were the most common imaging features.

Behcet's disease-related pulmonary damage is relatively rare. Similar to this study, previous studies found that slight hemoptysis accompanied by low fever and dyspnea was common in patients at an early stage.<sup>[1]</sup> An insufficient understanding of pulmonary imaging of Behcet's disease-related pulmonary damage results in serious missed diagnosis and misdiagnosis of the disease. This study focused on the CTPA characteristics of Behcet's disease-related pulmonary damage. The most common

characteristics were occlusion and *in situ* thrombosis formation above the lobar artery level, which might be associated with aneurysmal formation at the corresponding level. Regarding image differentiation, Behcet's disease-related pulmonary damage needs to be distinguished from pulmonary embolism, Takayasu arteritis and pulmonary tuberculosis. Pulmonary embolism often occurs in the branches of bilateral pulmonary arteries, and the involved site is usually below the level of lobar arteries. Besides, it is often accompanied by lower extremity venous thrombosis, and anticoagulant therapy can achieve good therapeutic effects usually. Takayasu arteritis also shows stenosis and occlusion in large branches of the main pulmonary artery, however, the incidence of aneurysmal formation is low.<sup>[2]</sup> The imaging of patients with pulmonary tuberculosis usually indicates pulmonary cavities surrounded by satellite lesions, which are not be found in the case of Behcet's disease. This study suggested that CTPA is an important tool to confirm Behcet's disease-related pulmonary vascular damage. Patients suspected of Behcet's disease-related pulmonary damage should undergo this examination as soon as possible.

Generally, patients who manifested with lung damage will receive plain lung CT examination at the early stage. At this time, the characteristic damage of pulmonary parenchyma can facilitate the early screening of Behcet's disease. Similar to previous studies, the pulmonary infarction and cavity were two common types of Behcet's disease-related pulmonary parenchymal damage imaging in this study.<sup>[3]</sup> In addition, we also found that the location of pulmonary infarction and cavity were consistent with the distribution area of vascular lesions, which is an important identification point between Behcet's

disease-related pulmonary damage and pneumonia or tuberculosis. In our speculation, the vasculitic lesions in large- or medium-sized arteries are a crucial pathophysiological cause for Behçet's disease-related pulmonary parenchymal lesions, including infarcts, consolidations, cavities, and mosaics. Here, we also reported one patient with wandering consolidation, which is rare in this type of disease and easily misdiagnosed as pneumonia. We should be wary of the possibility of Behçet's disease-induced pulmonary arteritis in patients, who have no noticeable clinical signs of infection and with a poor response to antibiotics.

#### **Declaration of patient consent**

The authors certify that they have obtained consent from the patients. The consent form included permission to report clinical information in the journal. The patient understands that his/her name and initials will not be published and that due efforts will be made to conceal his/her identity, however, anonymity cannot be guaranteed.

#### **Conflicts of interest**

None.

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