

# Pulmonary and Thoracic Vascular Involvement in Behçet's Disease on CT

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## Abstract

Behçet's disease (BD) is characterized by recurrent oral and genital ulcerations, ocular and additional clinical manifestations. The exact cause of the disease is unknown. Thoracic manifestations of BD show a wide spectrum including vascular, mediastinal, pleural and parenchymal involvement. CT is the imaging modality that provides the assessment of the thoracic presentations of BD. The typical presentations of BD are aneurysms of the pulmonary arteries and venous occlusion.

Aneurysms of the aorta, occlusion of vena cava superior, venous aneurysms, mediastinal lymphadenopathy, mediastinal fibrosis, pneumonia, pulmonary hemorrhage, infarction, pleural and pericardial effusions, and pleural nodules are other thoracic manifestations of BD. Assessment of these thoracic involvements can be used in the diagnosis of BD.

**Keywords:** Behçet's disease; CT; Thorax

## Introduction

Behçet's disease (BD) is a multisystem inflammatory disease. Although genetic factors and microorganisms are considered as causes of the disease, the exact etiology is unknown. It is characterized by recurrent oral and genital ulcerations and uveitis. Behçet first described the clinical trial of the disease in 1937 [1]. BD may affect the joints, the skin, the gastrointestinal system, the central nervous system, the heart and vascular system, and the lungs [2]. Vascular involvement is seen approximately in 25-30% of patients [3]. The main pathologic process is vasculitis and inflammation [4]. Pulmonary manifestations are relatively infrequent and have been reported in 1-10% of patients [2,5]. The disease is common in Mediterranean countries, Middle East and Asia. The prevalence rate in Turkey is 80-370 per 100.000 [2,3]. Although chest radiography is the first imaging modality, CT is the preferred method for diagnosing thoracic involvement of BD. We review the pulmonary, pleural and thoracic vascular manifestations of BD with CT findings.

## Clinical Diagnosis

International Study Group described the diagnostic criteria of BD in 1990. The diagnosis is depend on detection of recurrent oral ulcerations, and two of the following criteria including:

- Recurrent genital ulcerations;
- Uveitis, retinal vasculitis;
- Skin lesions (erythema nodosum, folliculitis); and
- Positive skin pathergy test (pustule formation 24-48 h following skin prick) [6].

## Imaging Techniques

Patients with pulmonary symptoms are assessed with chest x-ray. Chest radiographs can show hilar or mediastinal enlargements. Pulmonary artery and thoracic aortic aneurysms can cause hilar and mediastinal enlargements. Parenchymal changes in BD are non-specific and usually seen as focal opacities [7-10].

CT is the preferred imaging method in the diagnosis and follow-up of BD. CT Angiography (CTA) maintains excellent analysis of the aorta, pulmonary arteries and veins. The spatial resolution of CT is better than Magnetic Resonance Imaging (MRI). Additionally, CT can show the parenchyma and the pleura more detailed than MRI [11].

## Thoracic Vascular System Involvement

Pulmonary and thoracic aortic aneurysms, arterial and venous thrombosis are the main thoracic vascular manifestations of BD [12,13]. Vascular involvement occurs in 25-30% of patients and the complications can cause death [13-15]. Venous thrombosis is seen more than arterial involvement and presents 85% of vascular involvement. Superior vena cava occlusion is the most serious complication of venous involvement in BD [16]. Superior vena cava occlusion causes enlarged collateral veins in the mediastinum and chest wall (Figures 1 and 2). Thrombus can occur in the superior vena cava or propagate from a distal vein [17]. Venous aneurysms are rare in BD. Venous aneurysms in BD are due to presence of a predisposing factor such as trauma or other inflammatory disorders [18]. Arterial manifestations occur less frequently comparing with venous involvement and constitute 12% of vascular complications [12,19,20]. Aneurysm of the thoracic aorta, pulmonary artery and major branches occur in 65% of patients and occlusion in 35% [20,21]. BD is one of the most common causes of pulmonary artery aneurysms (Figures 3 and 4). Pulmonary artery aneurysms are caused by trauma, infections or Behçet's disease (BD). Less common causes are pulmonary hypertension, congenital heart disease and neoplasm [22]. The inflammation of the vessel wall can cause dilatation and aneurysm formation.

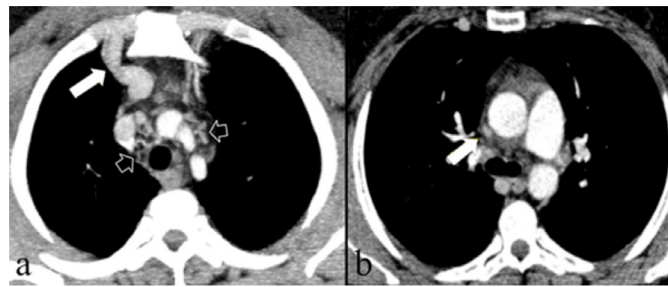
Inflammatory cellular infiltration also causes vessel wall thickening [23]. Steroid with combination of immunosuppressive agents can regress the pulmonary artery aneurysms. Early diagnosis of pulmonary artery aneurysm plays a critical role in the prognosis [24,25]. Occlusions and thrombosis of the pulmonary arteries are other complications of BD (Figure 5). The most common symptom, hemoptysis. Massive hemoptysis can cause death. Aneurysm rupture into a bronchus and pulmonary vessels thrombosis cause hemoptysis [12]. Pseudoaneurysms of the pulmonary artery, aortic arch, subclavian artery and coronary arteries have been reported in BD [26-28].

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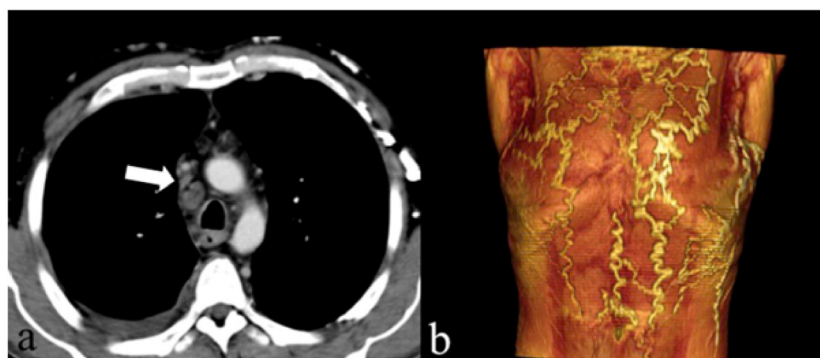
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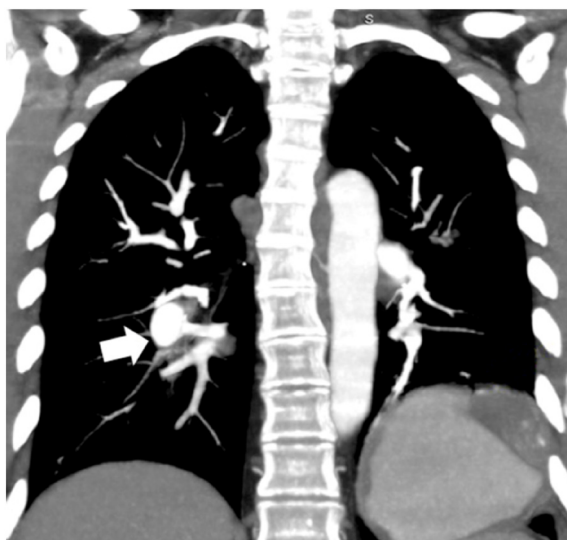
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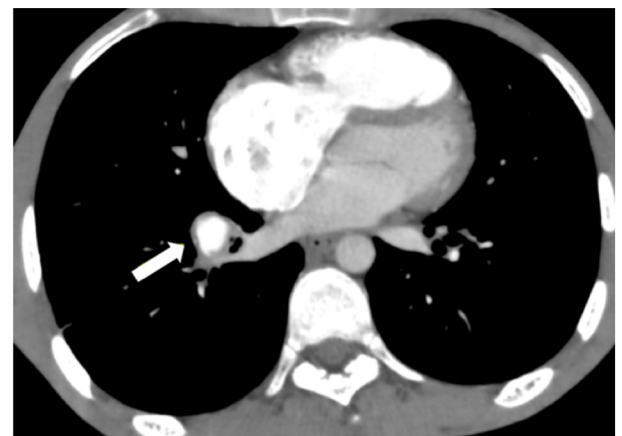
**Figure 1:** 30-year-old woman with Behçet's disease. Axial maximum intensity projection (a) and conventional (b) contrast enhanced CT images show occluded superior vena cava (short arrow in b) and enlarged right internal mammary vein (white arrow in a) and mediastinal collaterals (open arrows).



**Figure 2:** 40-year-old man with Behçet's disease. Axial contrast enhanced CT image (a) shows occluded vena cava superior and right paratracheal enlarged lymph node (arrow). Right pleural effusion is seen. (b) 3D volume rendering image shows collateral veins on the anterior chest wall.



**Figure 3:** 33-year-old woman with Behçet's disease. Coronal maximum intensity projection CT image shows right saccular (arrow) and left fusiform partially thrombosed pulmonary artery aneurysms.



**Figure 4:** 23-year-old man with Behçet's disease. Axial CT image shows the right pulmonary artery aneurysm (arrow).

## Mediastinum and Heart

Mediastinal lymphadenopathy can be seen in BD (Figure 2a) [3]. Fibrosing mediastinitis is very rare in BD. Diffuse form of fibrosing mediastinitis may be presented as a mediastinal mass [3]. One of the serious complications of BD is intracardiac thrombosis. It occurs mostly in the right atrium or ventricle. The prognosis of intracardiac thrombus is poor [29,30]. Pericardial effusion can occur because of

the inflammation of the pericardium or thrombosis of the superior vena cava [3].

## Pleura

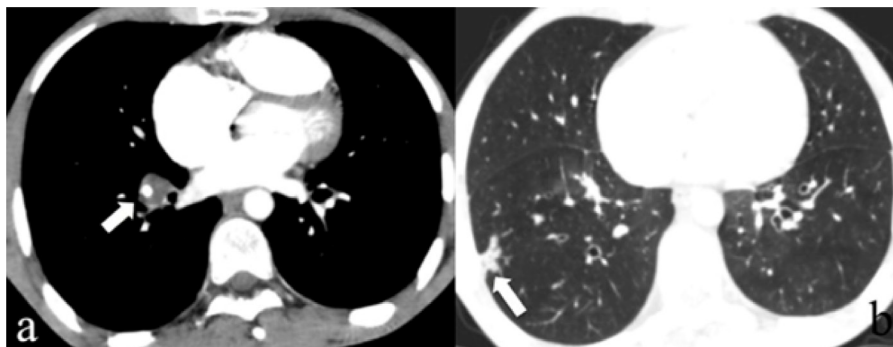
Primary pleuritic disease is not common in BD [14]. Pleural effusion and pleural nodules can be seen in BD [2,13,14]. Pulmonary infarction, pneumonia, thrombosis of superior vena cava or vasculitis can cause pleural effusion (Figure 2a).

## Lung

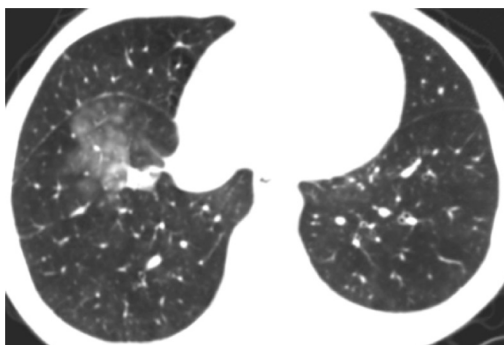
Lung parenchyma and pleural involvement occur in 1-10% of BD [31,32]. Pulmonary infarctions, pneumonia, pulmonary hemorrhage,



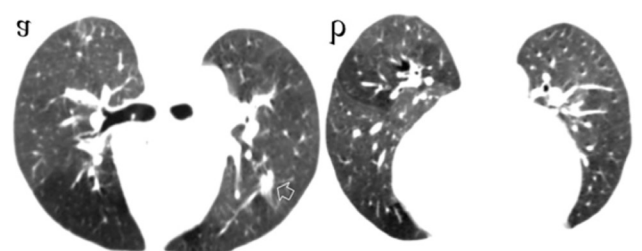
**Figure 5:** 27-year-old woman with Behçet's disease. Axial CT image demonstrates the thrombosed left pulmonary artery branch (arrow).



**Figure 6:** 28-year-old man with Behçet's disease and hemoptysis. Axial CT images show the thrombosed right pulmonary artery aneurysm (arrow in a) and peripheral consolidation in the right lower lung due to pulmonary infarction (arrow in b).



**Figure 7:** 24-year-old man with hemoptysis who had known Behçet's disease. Axial CT image shows alveolar opacities in the right lower lobe. This finding may be due to hemorrhage or pneumonia.



**Figure 8:** 32-year-old woman with Behçet's disease. Axial CT images show mosaic attenuated lung parenchyma and a nodule with irregular margins in the left superior lobe (open arrow in a). This nodule may have developed secondary to vasculitis or infarction (b).

atelectasis, bronchitis, fibrosis and emphysema are the involvement types of lung in BD. Thrombosis of pulmonary arteries may cause infarction, atelectasis, and hemorrhage (Figure 6) [12,13,18]. Focal or diffuse alveolar opacities, wedge-shaped consolidations, linear opacities, atelectasis, cavitary nodules, round opacities, reticulation and mosaic attenuation can be seen in the lung parenchyma (Figure 7). Alveolar opacities occur because of the vasculitis and hemorrhage [4,14,25]. In some patients with BD mosaic attenuation may be seen on CT scans due to small airway diseases or pulmonary artery thrombosis (Figure 8a and 8b) [14]. Pulmonary hemorrhage or pneumonia causes airspace

consolidation or ground-glass opacification. Pneumonia may be seen secondary to immunosuppressive therapy [33]. Pulmonary infarctions can cause atelectasis, wedge shaped subpleural consolidations, linear parenchymal opacities or pleural effusions. Small-vessel vasculitis can cause reticulation [12,29].

## Conclusion

Vascular involvement in BD can cause serious complications. Patients with pulmonary artery aneurysms have poor prognosis. Aortic and superior vena cava involvement may occur. Lungs, pleura, mediastinum may be involved. CT can show different thoracic manifestations of BD.

## Conflict of Interest

The authors declare no conflict of interest. None of the authors has disclosed financial interests related to the material in the manuscript.

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