Partially thrombosed multiple pulmonary artery aneurysm secondary to Behçet disease

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A 23-year-old man with an established diagnosis of Behçet disease (BD) for five years presented with cough and hemoptysis. The laboratory findings were normal with the exception of mild leukocytosis. An anteroposterior radiograph revealed multiple round lesions (Figure 1). He underwent multidetector computed tomography (MDCT) to determine the cause of hemoptysis. MDCT demonstrated multiple pulmonary artery aneurysms (PAAs) bilaterally (Figures 2A and 2B). Alveolar hemorrhage (Figure 3A) and subpleural fibrotic-atelectatic changes (Figure 3B) were also detected within the pulmonary parenchyma on images reconstructed using a lung algorithm.

BD is a multisystem disorder first described by Hulusi Behçet in 1937. It is a vasculitis that presents with a triad of findings including recurrent ulcers of the oral and genital mucosa with relapsing uveitis. Additional clinical manifestations were later described and include involvement of the skin, joints, large vessels, lung, brain, and gastrointestinal and genitourinary tracts (1). The underlying pathological process in BD is perivascular inflammation affecting vessels of different sizes in various organs. The inflammatory process is acute and results in destruction of the walls of the affected vessels. BD can cause aneurysms that can rupture (1). The most commonly affected arteries are the aorta, the pulmonary arteries and the femoral artery.

Vascular manifestations of BD in the thorax are observed in both the arterial and venous system. Venous involvement is, in fact, observed more frequently than arterial involvement. Occlusion with or without evidence of thrombosis of the superior vena cava or brachiocephalic vein are quite common and, less frequently, occlusion and/or stenosis

and aneurysms of the pulmonary arteries can occur. PAA is the most common lung manifestation of BD, which is the most common cause of PAAs. Additional parenchymal findings, including atelectasis, hemorrhage and infarction (focal or subpleural consolidation), have also been reported. These are commonly encountered secondary to thrombosis of the pulmonary vessels. Pneumonia, bronchitis, fibrosis, subpleural infiltrates, subpleural nodules, pleural effusion and emphysema have been described (2,3).

Clinically, patients most frequently present with hemoptysis secondary to PAA. This is more commonly observed in male patients with BD.

Figure 2) Axial contrast-enhanced computed tomography scan showing partial thrombosed pulmonary arteries in the right (A) and left (B) lung (arrows)

Figure 1) Anteroposterior chest radiograph showing multiple well-circumscribed nodular lesions (arrows)

Figure 3) Alveolar hemorrhage (A) and subpleural fibrotic-atelectatic changes (B) are also detected on parenchyma in lung window (arrows)

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KEY LEARNING POINTS

- BD is a multisystem and chronic inflammatory disorder of unknown etiology.
- The main pathological process is vasculitis and perivascular inflammatory infiltrates affecting the blood vessels.
- Vascular system involvement is observed in 20% to 40% of patients. Venous and arterial involvement are seen in BD, with venous involvement more common.
- PAA is the most frequent form of thoracic involvement, with hemoptysis being the most common clinical presentation

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