

## CASE REPORT

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## Circumflex-to-Bronchial Artery Fistulas in Behçet's Disease

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**ABSTRACT** A coronary-to-bronchial artery fistula is a rare entity that emerged in the case of chronic pulmonary thromboembolism (cPTE). We present a case of a 50-year-old man with coronary-to-bronchial artery fistula in Behçet's disease that had been hospitalized in consequence of pulmonary thromboembolism attacks and treated with anticoagulation therapy repeatedly. After myocardial infarction was ruled out, coronary computerized tomography angiography was performed, and demonstrated right pulmonary artery occlusion, delay of the contrast passage in the right pulmonary veins, dilated right lung bronchial arteries, and fistulas between the right lung bronchial arteries and the left circumflex artery due to cPTE. Diagnosis of coronary-to bronchial artery fistula and early treatment are very important for the prevention of fatal complications. Radiologist and clinician should be alerted for this entity.

**Keywords:** Behçet's disease; bronchial artery; coronary artery; fistula

A coronary-to-bronchial artery fistula is a rare entity that emerged in the case of chronic pulmonary thromboembolism (cPTE). If a plan of embolization exists, it is crucial to demonstrate coronary-to-bronchial artery fistulas radiologically to avoid fatal consequences such as myocardial infarction. We present a case of coronary artery-to-bronchial artery fistula in Behçet's disease. Informed consent was given.

### CASE REPORT

A 50-year-old man with Behçet's disease was admitted to our hospital for the evaluation of unstable angina pectoris. Blood test results and electrocardiogram ruled out myocardial infarction. Our patient had been suffering from Behçet's disease for 20 years. Initially, he had oral and genital painful, ulcerative lesions. For few times, he was treated with steroid therapy for panuveitis and arthritis. Pathergy test was positive. He had no neurologic or gastrointestinal involvement.

**Imaging findings:** Flash mode prospective electrocardiography-gated coronary computerized tomography (CT) angiography was performed with a dual-source CT system (Siemens Healthineers-SOMATOM Force, Erlangen, Germany), to show whether coronary artery pathology was the cause of angina pectoris. We used 70 milliliter (mL) of iohexol 350 milligram/100 mL as the contrast agent with 80 mL of 0.9% NaCl (sodium chloride) [at first, 10 mL NaCl at 5 mL/second (sec), then 60 mL contrast agent at 5 mL/sec, 10 mL contrast/40 mL NaCl mixture at 4 mL/sec, 30 mL NaCl at 4 mL/sec]. Coronary CT angiography demonstrated right pulmonary artery occlusion, delay of the contrast passage in the right pulmonary veins, dilated bronchial arteries, and fistulas between the right lung bronchial arteries and the left circumflex artery (LCx) due to the cPTE (Figure 1, Figure 2, Figure 3, Figure 4).

**Treatment:** He had been hospitalized as a consequence of PTE attacks and treated with anticoagulation therapy repeatedly. Since the patient's

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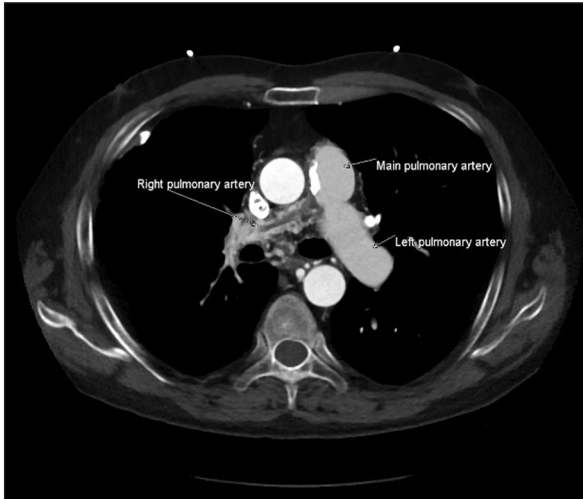
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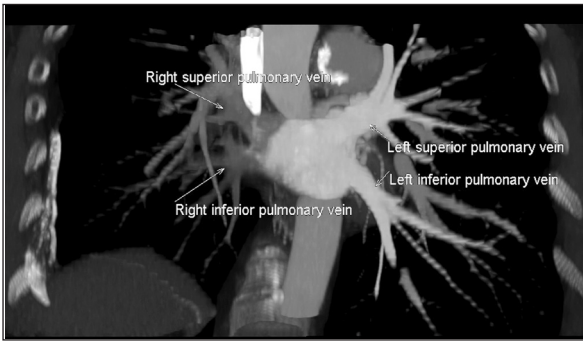
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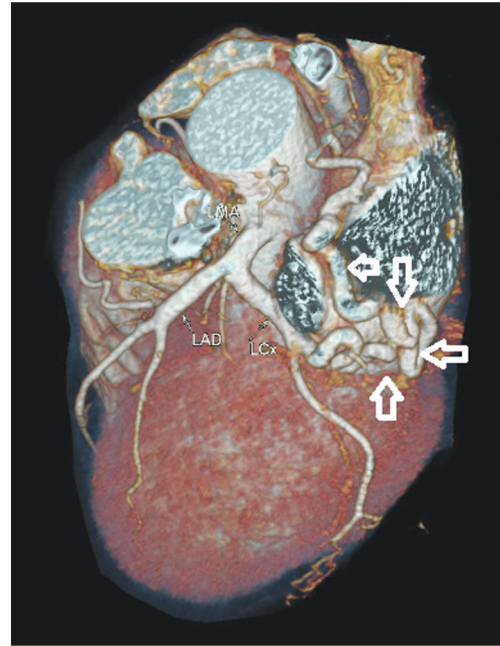
**FIGURE 1:** Axial coronary computerized tomography angiography shows occluded right pulmonary artery due to chronic thromboembolism in Behçet's disease. Also note dilated para-aortic bronchial arteries.



**FIGURE 2:** Coronal maximum intensity projection image shows delayed contrast passage in right pulmonary veins as a result of right pulmonary artery occlusion.



**FIGURE 3:** Axial oblique maximum intensity projection image shows tortuous and dilated bronchial artery-to-LCx fistulas (arrows). LCx: Left circumflex artery.



**FIGURE 4:** Volume-rendered image of coronary computerized tomography angiography shows dilated and tortuous LCx below left atrial appendage (hollow arrows). LMA: Left main coronary artery; LCx: Left circumflex artery; LAD: Left anterior descending artery.

international normalized ratio value was between 2-3, he was discharged with 5 mg warfarin per 24 hours, which he continued. Additional imaging was not performed because the patient did not come for a follow-up.

## DISCUSSION

Behçet's disease is a multisystemic, chronic recurrent inflammatory disease with non-specific vasculitis involving vessels in several organs and systems in various degrees.<sup>1</sup> Oral and genital ulcerative lesions, skin lesions like erythema nodosum, positive pathergy test, arthritis, eye lesions like uveitis, thoracic, neurologic, gastrointestinal, vascular involvement are characteristic features of Behçet's disease.<sup>2</sup> The pulmonary artery is the second most frequent vascular structure involved after aorta.<sup>1</sup> The hypercoagulability in Behçet's disease is the underlying reason for the PTE.<sup>1</sup> The highest prevalence rate of Behçet's disease is in Türkiye with 80-370/100,000.<sup>2</sup> Behçet's disease may present with pulmonary stenosis or occlusion due to the recurrent PTE or with pulmonary artery aneurysms.<sup>1</sup> cPTE leads to dilatation of the

bronchial arteries.<sup>3</sup> Fistulas between the coronary and bronchial arteries are rarely encountered and usually originated from the LCx.<sup>4,5</sup> Coronary-to-bronchial artery fistulas may occur in chronic lung diseases, vasculitis such as Takayasu, and surgical interventions, and congenitally.<sup>6-8</sup>

Compensatory dilatation of bronchial arteries and fistulas between bronchial and LCx occurred in this case as a consequence of right pulmonary artery occlusion due to the cPTE as stated in the literature.<sup>1</sup>

Coronary-to-bronchial artery fistula is a known entity.<sup>5-8</sup> To our knowledge, no case of a fistula between the coronary and bronchial artery has been reported in Behçet's disease yet although coronary-to-bronchial artery fistulas are reported in other vasculitides like Takayasu.<sup>7</sup> If a plan of embolisation exists, it is crucial to demonstrate coronary-to-bronchial artery fistulas radiologically to avoid fatal consequences such as myocardial infarction.

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### Conflict of Interest

*No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.*

### Authorship Contributions

**Idea/Concept:** Duygu İmre Yetkin, Selçuk Akkaya, Tuncay Hazırolan; **Design:** Duygu İmre Yetkin, Selçuk Akkaya; **Control/Supervision:** Duygu İmre Yetkin, Selçuk Akkaya; **Data Collection and/or Processing:** Duygu İmre Yetkin; **Analysis and/or Interpretation:** Duygu İmre Yetkin, Selçuk Akkaya; **Literature Review:** Duygu İmre Yetkin; **Writing the Article:** Duygu İmre Yetkin; **Critical Review:** Duygu İmre Yetkin; **References and Fundings:** Duygu İmre Yetkin, Selçuk Akkaya; **Materials:** Duygu İmre Yetkin, Selçuk Akkaya.

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