

## A Case of Behcet's Disease Complicated with a Pulmonary Artery Aneurysm and Deep Vein Thrombosis, Separately

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Behcet's disease is a chronic inflammatory disease characterized by oral ulcers, genital ulcers, uveitis, and skin lesions. Furthermore, Behcet's disease can manifest as vascular lesions, such as, those of vasculitis, venous thrombosis, or thrombophlebitis or as an arterial aneurysm. Here, the authors report the case of a pulmonary artery aneurysm and deep vein thrombosis in a 41-year-old woman with a previous diagnosis of Behcet's disease. The patient presented with hemoptysis and a cough, and was found to

have a bleeding pulmonary artery aneurysm at the right lower lung. Pulmonary arteriography was performed and the aneurysm was embolized with coils. As a result, hemoptysis did not subsequently recur. However, five years later, deep vein thrombosis occurred in the left leg. Left leg pain improved after the regional infusion of thrombolytics.

**Key Words.** Behcet's disease, Pulmonary artery aneurysm, Endovascular embolization, Deep vein thrombosis

### Introduction

Behcet's disease is a chronic, systemic, inflammatory disease of unknown origin. Its major clinical manifestations are oral ulcers, genital ulcers, uveitis, and skin lesions, and its other manifestations are multiple arthritis, gastrointestinal lesions, central nervous system involvement, and vascular lesions. Small or large vessel vasculitis in Behcet's disease leads to vessel occlusion or aneurysm formation (1), but arterial involvement occurs infrequently and presents as aortitis or a peripheral arterial aneurysm with arterial thrombosis. The pulmonary artery is the second most common site of arterial involvement in Behcet's disease (2), and pulmonary artery aneurysms can cause massive hemoptysis by rupture in bronchi. Venous thrombosis is also present in one-fourth of Behcet's disease patients, and deep venous thrombosis of the lower limb is the most frequent venous manifestation. Few cases of Behcet's disease in a female patient combined with

a pulmonary artery aneurysm and deep venous thrombosis have been reported (3). In this article, we describe a case of Behcet's disease that was complicated by a pulmonary artery aneurysm and deep venous thrombosis, separately occurred with 5 years interval.

### Case Report

A 41 year-old woman was admitted with a 2-month history of hemoptysis and sustained cough. Initially, amount of hemoptysis was small manifesting as blood-tinged sputum. Four hours before admission, she presented 30 mL of fresh hemoptysis accompanying with intractable cough. The patient had been diagnosed with Behcet's disease about 4 years previously. Her diagnosis was based on oral ulcers, genital ulcers and pathergy test. She was being treated with colchicine 1.2 mg per day and Dapsone 25 mg per day. The patient was a non-smoker and did not consume alcohol regularly. On phys-

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<Received : December 19, 2011, Revised (1st: February 24, 2012, 2nd: April 2, 2012), Accepted : April 4, 2012>  
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pISSN: 2093-940X, eISSN: 2233-4718  
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ical examination, vital signs were stable, but respiratory sounds were decreased in the right lower lung field. A peripheral blood examination revealed; hemoglobin 10.2 g/dL, hematocrit 32.6%, platelet count 545,000/ $\mu$ L, erythrocyte sedimentation rate (ESR) 47 mm/hr, and C-reactive protein (CRP) 19.7 mg/dL. Biochemical analyses of blood showed no pathological findings. Prothrombin and partial thromboplastin times were normal. But HLA-B51 test was not executed.

Chest radiography showed an ill-defined increased opacity in the right lower lung (Figure 1), and a computerized tomography (CT) scan of the thorax with intravenous contrast revealed a round enhancing lesion, measuring 2.2 $\times$ 2.0 cm, in right lower lung (Figure 2). The lesion was connected to the



**Figure 1.** Chest X-ray showing ill-defined increased opacity in the right lower lung.



**Figure 2.** Contrast computerized tomography (CT) scan of the thorax showing a round enhancing lesion, measuring 2.2 $\times$ 2.0 cm in size, in the right lower lung (arrow).

pulmonary artery, and a hypo-dense lesion was seen in the proximal pulmonary artery. Pulmonary artery angiography was performed for diagnosis and treatment. A pulmonary artery aneurysm and broncho-pulmonary shunt was demonstrated at lateral segment of right lower lung. The aneurysm was embolized with two 6 mm $\times$ 14 cm coils (NESTER, Cook, USA) and eight 4 mm $\times$ 14 cm coils (NESTER, Cook, USA) (Figure 3). After coil embolization, methylprednisolone was administered during 3 days at 1 g per day delivered by intravenous boluses. After steroid pulse therapy, intravenous methylprednisolone was changed to oral prednisolone 55 mg (1 mg/kg) per day. Post-procedural recovery was uneventful, and the patient was discharged eight days after embolization. The patient was treated with a combination of cyclophosphamide and prednisolone after discharge in an outpatient clinic. Cyclophosphamide was administered as monthly intravenous boluses of 1 g and oral prednisolone was tapered from 55 mg to 5 mg over 10 months. Cyclophosphamide was continued for 6 months and then changed to azathioprine 100 mg per day. Because of the possibility of a hemorrhagic complication, anticoagulant medication was not given.

Five years after discharge, the patient was transferred to emergency room with left leg pain presumed to be due to deep vein thrombosis in the left lower limb. Laboratory tests showed; hemoglobin 12.9 g/dL, platelets 399,000/ $\mu$ L, WBC 8,400/ $\mu$ L, prothrombin time 86%, partial thromboplastin time 38.4 seconds, ESR 36 mm/hr, CRP 15.4 mg/dL and D-dimer increased to 0.83  $\mu$ g/mL. Protein C/S activity and antigen levels was within normal range. Lupus anticoagulant was negative, but antiphospholipid antibody IgG/IgM was not tested.



**Figure 3.** Pulmonary arteriograph showing Nester coils in the pulmonary artery aneurysm.



**Figure 4.** Lower extremity venograph showing a thrombus in the left popliteal vein and the catheter used for the peripheral venous approach.

CT angiovenography revealed a thrombus in the left superficial femoral vein and popliteal vein (Figure 4). IVC filter (OPTEASE, Cordis, USA) insertion was performed for endovascular management. After filter insertion, loading dose of urokinase 100,000 IU and heparin 5,000 IU was infused through the distal portion of the left popliteal vein. After infusing regional thrombolytics, the remnant thrombus was manually removed. Two weeks later, IVC filter was removed. Leg pain and swelling improved after flow-directed regional thrombolytic therapy, and there were no evidence of a secondary pulmonary embolism or bleeding. After discharge, she was treated with azathioprine 100 mg per day and colchicine 1.2 mg per day for 3 months, and no evidence of hemoptysis recurrence was encountered after pulmonary artery embolization, warfarin was administered for 3 months, and then changed to aspirin. At 6 months after discharge, the patient remained asymptomatic with no recurrence of the leg pain.

### Discussion

Behcet's disease is a chronic, systemic, inflammatory disease. Its major manifestations are recurrent oral and genital aphthous ulcerations. Furthermore, Behcet's disease may involve neurologic, cardiovascular, pulmonary, and musculoskeletal systems, and cardiovascular manifestations have been described in 7~49% of patients. Veins are frequently affected, and their involvements result in both superficial thrombophlebitis and deep venous thrombosis in 30~40% of patients. Arterial complications occur in 1~7% of patients and pulmonary artery involvement is observed in 1% (4-6).

Men are much more likely to be affected by arterial disease.

Pulmonary artery aneurysm has a poor prognosis and is one of the leading causes of death in Behcet's disease patients. Hemoptysis, when massive and untreated, has a mortality rate of >50% (7). Medical treatments based on steroids and on immunosuppressive drugs, such as cyclophosphamide or azathioprine, have been tried, but although some have reported successful treatment results and aneurysmal regression for immunosuppressive treatments (8,9), embolization with medical therapy more widely accepted (10). Surgical treatment may be considered for refractory vessel disease.

Venous thrombosis of the lower extremities and superior or inferior vena cava occlusion frequently occur in Behcet's disease patients. Unfractionated heparin and anticoagulation historically represent the mainstay treatment for deep vein thrombosis, but anticoagulation therapy cannot be used in patients with a high risk of hemorrhagic complications. Thus, anticoagulation therapy can be administered via flow-directed regional thrombolytic therapy, which is based on the direct regional infusion of concentrated thrombolytic agent from an ipsilateral peripheral vein into the deep venous system (11). Because secondary pulmonary embolism due to small fragments can occur, IVC filtration must be performed before direct regional thrombolytic therapy.

Immunosuppressive agents are recommended for the management of acute deep vein thrombosis in Behcet's disease, but anticoagulation is not recommended by the EULAR guidelines (12). The 'vasculo-Behcet' concept has been adopted when vascular complications dominate clinical features (13).

There is some debate about use of non-soluble coil or soluble gelfoam. In this case, pulmonary artery aneurysm was treated successfully by coil embolization and deep vein thrombosis was treated by regional thrombolytics infusion. Because of co-existence bleeding risk and thrombotic complication, we must concern to determine treatment in vasculo-Behcet's disease. The early detection of vascular lesions and appropriate treatment are essential for the optimal care of these patients.

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