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Effects of Corticosteroid and Chlorambucil on Multiple Pulmonary Artery Aneurysms in Behcet's Syndrome
— A Case Report —

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A patient with Behcet's syndrome in whom multiple pulmonary artery aneurysms were completely resolved after a combined treatment with corticosteroid and chlorambucil is reported.

Key Words : *Behcet's syndrome, Pulmonary artery, Aneurysm*

INTRODUCTION

Behcet's syndrome is a multisystem disorder, resulting from systemic vasculitis, which is characterized by recurrent episodes of oral and genital ulcers, iritis, and cutaneous lesions (Efthimiou et al., 1986 ; Raz et al., 1989 ; Erkan and Cavdar, 1992). Vascular involvement in Behcet's syndrome occurs in up to 25—35 % of patients, manifestating mainly as venous thrombosis (Grenier et al., 1981 ; Koc et al., 1992 ; Lie, 1992). Although infrequent, pulmonary artery aneurysm due to pulmonary vasculitis is one of the worst prognostic manifestations of the syndrome (Efthimiou et al., 1986 ; Raz et al., 1989 ; Erkan and Cavdar, 1992 ; Lie, 1992). Corticosteroid alone is not enough to prevent a fatal outcome of pulmonary artery aneurysm in most cases (Gibson et al., 1985 ; Efthimiou et al., 1986 ; Kohno et al., 1986).

In Korea, one case of Behcet's syndrome with a single pulmonary artery aneurysm was reported by Uh et al.(1994). However in their case, the aneurysm was not reponsive to medical therapy and was treated by surgery.

We report a patient with Behcet's syndrome with multiple pulmonary artery aneurysms which have been completely resolved after a combined treatment with corticosteroid and chlorambucil.

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CASE REPORT

A 26-year-old Korean man was admitted with hemoptysis of about 150 ml. Two years prior to the admission, he had developed recurrent painful oral and genital ulcers, 3—10mm oval or round, which had been sharply circumscribed and had had a gray-yellow center with a red rim. He also had developed erythema nodosum on the lower extremities. He had not taken any medication. Ten months before the admission, he noted swelling and pain in both legs, and a clinical diagnosis of Behcet's syndrome with deep vein thrombosis was made. The chest roentgenograms were normal. He was treated with prednisone and anticoagulant, and responded well. Two months prior to the admission, while still taking 15 mg of prednisone every day, he developed a dry cough, which became progressively aggravated. On the admission day, hemoptysis was developed but was not accompanied by dyspnea, fever, or pleuritic chest pain.

On examination, the patient appeared acutely ill. Blood pressure was 120/70 mmHg, pulse rate 90/min, respiratory rate 15/min, and body temperature 36.8℃. There were no oral or genital ulcers but multiple erythematous nodules were present on both legs. The remaining physical examination was normal. Laboratory data included a hemoglobin of 13.8 g %, hematocrit of 40 %, white blood cell count of 7,640/mm^3 with a normal differential count, and platelet count of 227 × 10^9/L. The erythrocyte sedimentation rate was 85 mm/hr. The prothrombin and partial thromboplastin times were normal. The anticardiolipin antibody and the lupus anticoagulant were negative. The results of the following tests were negative; antinuclear antibody, rheumatoid factor, LE cell, and VDRL. A chest roentgenogram showed two perihilar round densities and one round density in the right upper lobe. A computed tomographic scan with contrast showed a partially thrombosed aneurysm of the left lower lobe artery with a diameter of 2.5 cm, and two smaller aneurysms of the left upper lobe artery and the right lower lobe artery (Fig. 1). Bronchoscopic examination did not show abnormalities. Ventilation-perfusion radionuclide lung scans showed multiple perfusion defects in both lungs without corresponding ventilation defects (Fig. 2). Pulmonary angiography was attempted for confirmation and, more importantly, for embolization of the pulmonary artery aneurysms. However, the attempt failed as we could not introduce the catheter to the pulmonary artery because of the thrombosis in the inferior vena cava below the renal veins (Fig. 3). He was treated with a daily regimen of 1mg/kg of prednisone and 0.1mg/kg of chlorambucil. After two-months of treatment, a CT scan showed complete resolution of the pulmonary artery aneurysms (Fig. 4). The dosage of prednisone was tapered to 15mg/day over 2 months without recurrence of hemoptysis. He was still in complete remission 12 months after the treatment was started.

DISCUSSION

The primary pulmonary pathologic lesion in Behcet's syndrome is a lymphocytic and necrotizing vasculitis, involving all sizes of pulmonary arteries, veins, and septal capillaries, that may be complicated by vascular thrombosis, pulmonary infarction, and pulmonary artery aneurysm (Slavin and de Groot, 1981). Clinically overt pulmonary involvement occurs in 5 to 10 % of patients (Efthimiou et al., 1986; Raz et al., 1989). Lung involvement is characteristically associated with active disease at other sites (Efthimiou et al., 1986; Raz et al., 1989; Erkan and Cavdar,

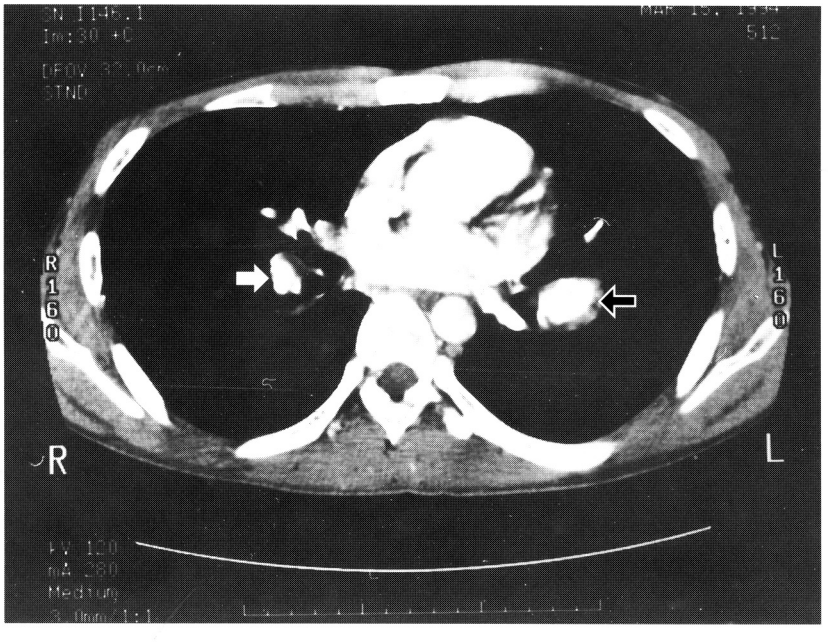

Fig. 1. CT scan of the chest with contrast showing partially thrombosed aneurysm of the left lower lobe artery with a diameter of 2.5cm(open arrow) and small aneurysm of the right lower lobe artery(white arrow).

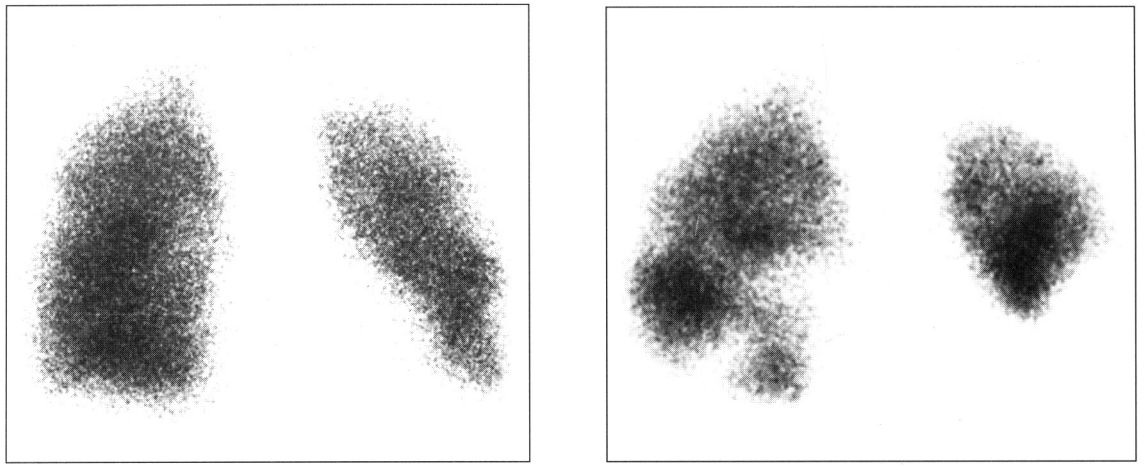

Fig. 2. Ventilation(A)-perfusion(B) lung scan of the posteroanterior view showing multiple perfusion defects in both lungs without corresponding ventilation defects.

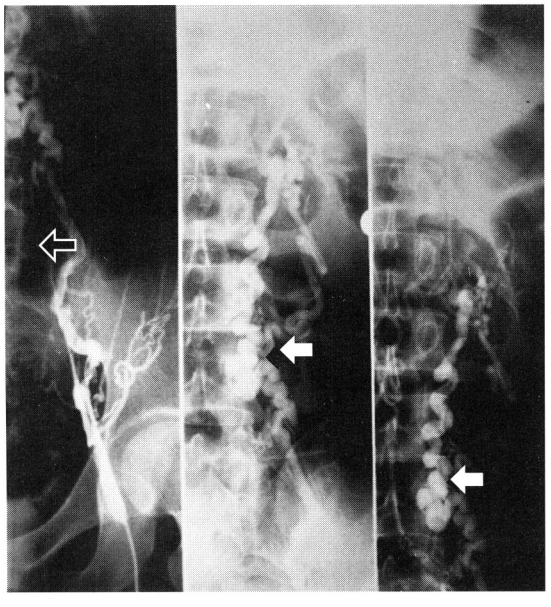

Fig. 3. Venocavagram showing occlusion of the inferior vena cava(open arrow) and rich collaterals through the vertebral plexus and lumbar veins(white arrows).

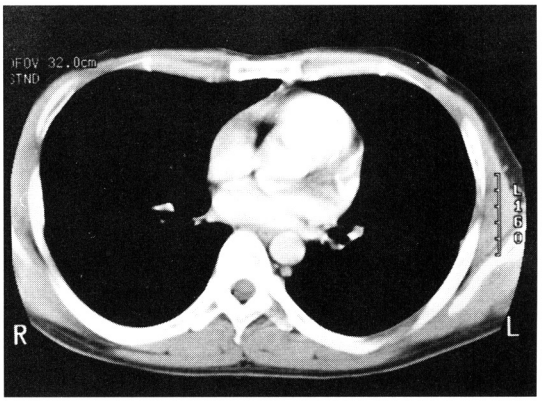

Fig. 4. CT scan of the chest 2 months after a combination treatment with corticosteroid and chlorambucil showing complete resolution of preexisting aneurysms.

1992) and generally appears after extrapulmonary manifestations (Raz et al., 1989 ; Erkan and Cavdar, 1992). Compared with the cases without lung involvement, the thrombophlebitis of the legs or the vena cava is more common in patients with lung involvement, but the ocular and skin lesions are less common (Efithimiou et al., 1986 ; Raz et al., 1989 ; Erkan and Cavdar, 1992).

Although hemoptysis, dyspnea, pleuritic chest pain, and cough are common, hemoptysis is the dominant clinical manifestation (Efthimiou et al., 1986 ; Kohno et al., 1986 ; Raz et al., 1989 ; Erkan and Cavdar, 1992) and one of the main causes of death in Behcet's syndrome (Efthimiou et al., 1986 ; Raz et al., 1989). Hemoptysis may result from bronchial erosion by aneurysm, pulmonary infarcts, extensive inflammatory disruption of alveolar capillaries, or rupture of a bronc-

hial vein caused by superior vena cava thrombi (Kohno et al., 1986). However, the massive hemoptysis resulting in death is usually caused by the rupture of pulmonary artery aneurysm (Durieux et al., 1981 ; Grenier et al., 1981 ; Gibson et al., 1985 ; Kohno et al., 1986). Therefore, in the presence of hemoptysis in patient with Behcet's syndrome, the possibility of pulmonary artery aneurysm should be considered, and a pulmonary angiography or preferably contrast enhanced CT scan of the chest should be performed (Almong et al., 1993 ; O'Duffy, 1993).

Hughes-Stovin syndrome, defined as the association of multiple pulmonary aneurysms with deep vein thrombosis, is indistinguishable from pulmonary manifestations of Behcet's syndrome and may be an incomplete form of Behcet's syndrome (Durieux et al., 1981 ; Slavin and de Groot, 1981 ; Almong et al., 1993 ; O'Duffy, 1993).

Corticosteroid has been used in the first line of therapy in Behcet's syndrome with pulmonary involvement and was relatively successful in some, especially patients without pulmonary artery aneurysm, i.e. patients in the early stage, before irreversible damage to the arterial wall develops (Cadman et al., 1976 ; Efthimiou et al., 1986 ; Raz et al., 1989). However, complete remission of pulmonary artery aneurysm following corticosteroid treatment alone is quite rare. Severe hemoptysis, although responsive initially, may recur during the therapy (Durieux et al., 1981 ; Grenier et al., 1981 ; Gibson et al., 1985 ; Kohno et al., 1986). Surgical resection or transcatheter embolization of a pulmonary artery aneurysm is indicated only in selective cases (Efthimiou et al., 1986 ; Kohno et al., 1986 ; Raz et al., 1989). Recently, O'Duffy (1993) proposed a combination therapy consisting of corticosteroid and chlorambucil in patients with pulmonary involvement based on its successful use in patients with ocular or cerebral diseases.

Our case was the first case with Behcet's syndrome in which multiple pulmonary artery aneurysms were completely resolved after a combination therapy with corticosteroid and chlorambucil. The regression of pulmonary artery aneurysms after a combination therapy in our case may indicate that this combination therapy is more effective than corticosteroid alone or at least it can give a steroid sparing effect.

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