HEMOPTYSIS IN BEHÇET'S DISEASE. A CASE REPORT AND REVIEW OF THE PREVİOUS STUDİES

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INTRODUCTION

Behçet's disease is a systemic disease of unknown etiology and is thought to be leukocytoclastic vasculit (1,2,3). The major criteria of diagnosis are aphthous stomatitis, genital ulcerations, ocular lesions, cutaneous lesions, and arthritis (4). Behçet's disease is a chronic relapsing illness characterized by polymorphic clinical manifestations. Although pulmonary involvement is unusual, it is known that it is particularly more frequent in our conutry (5,6).

Behçet's disease was diagnosed in a case with hemoptysis in our department. Because the first symptom was hemoptysis, this case was interesting for us. Therefore, we approved to report the case and reviewed other cases in literature.

CASE REPORT

The patient, a 24 year old male, presented a history of high fever, cough and hemopytiysis since 3 months. First, we consulted in our department in August 1986. He had received tubercülostatics and nonspecific antibiotics before our examination. No abnormalities were found on examination. At the first examination, the sedimentation rate was 60 mm/hr. The leukocyte count was 9.500/mm^3, of which 77 percent were polymorphonuclear cells. Gamma-2 globulin was 21.2 percent in protein electrophoresis. Bronchoscopy and esophagoscopy revealed no abnormalities. Pulmonary radiography showed the bilateral alveolar infiltrates (Figure 1).

After hospitalization, genital ulcerations and aphtous stomatitis occured. Hypersansibility reactions were seen in the enjection areas. The findings on ophtal-

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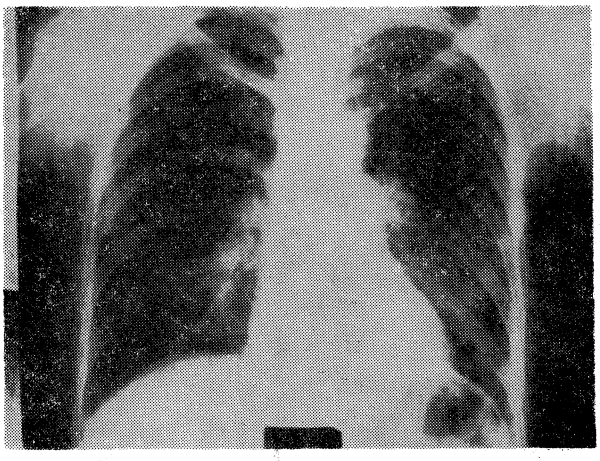

Figure 1. Bilateral alveolar infiltrates cosistant with Behçe('s disease.

mologic examination were normal. Therefore, we thought that hemoptysis may be secondary to Behçet's diseases. Pulmonary angiography wasn't able to caried out, since we were afraid of the fact that aneurysms were able to ruptured. Therapy comprising 1 mg/kg of prednisone per day was begun and lead to a relatively improvement.

DISCUSSION

According to observations of Fırat (7), pulmonary lesions were present about one percent in 245 Behçet patients (190 males and 55 females). Although pulmonary lesions were infrequent, these were fairly good defined. Recurrent pneumonies, pleural effusions, hilar adenotpathies, rounded opacities, pulmonary thrombosis, superior vena cava syndrome, and pulmonary artery aneurysms are mainly pulmonary findings in Behçet's disease (8).

The principal pathologic abnormalities is vasculitis consisting of infiltration with immuncomplexs, immunglobulin G, compleman 3 and 4 of pulmonary capillaries and venules. Indeeed, several histological studies have proved the existence of vasculitis chiefly in the venules and capillaries with infiltration of lymphocytes and plasmocytes (2,3,9).

Pulmonary arteries can also be influenced by the disease and aneurysms may occur. Multiple pulmonary arterial aneurysms are uncomon, but occur preferentially in the peripheral branches of the pulmonary arteries. In Hughes-Stovin syndrome, the multiple pulmonary aneurysms are accompanied by venous thrombosis, especially that of the vena cava (10,11,2). Hughes-Stovin sydrome may be indistinguishable from the pulmonary involvements of Behçet's disease.

The pulmonary lesions can be accompanied by thrombosis of the vena cava in Behçet's disease. Carman et al. (13) observed a such association in four of 12 patients with pulmonary lesions. Ateş et al. (8) also reported three cases with superior vena cava syndrome. Granier et al. (14) observed thrombosis of the superior or inferior vena cava in two of the five patients having three major criteria. One of the characteristics of the disease is involvement of the large vessels, mainly of the veins with a frequency of 24 percent. The venae cava are the veins most often affected. Thrombosis or aneurysms of the aorta or limb arteries have been reported rarely (4,14).

Radiological abnormalities may consist of alveolar infiltrates. as our case, has and rounded opacities. The alveolar infiltrates are probably the radiographic expression of the hemorrhage. Pleural effusion seems to be the common result of pulmonary infarction. Pulmonary angiography makes visible the severity of pulmonary arterial lesions and shows occlusions and aneurymsms (4).

From a clinical standpoint, pulmonary, manifestations always occur during exacerbation of the disease (4,13). The most frequent symptom observed in hemoptysis, which is sometimes fatal. Çobanlı (15) reported hemoptysis in 11 of 19 patients with pulmonary lesions. In our case, hemomptysis occured before the major criteria of Behçet's diseases. Since Behçet's disease is frequent in our country. It is to remembered in differantial diagnosis of hemoptysis, even major criteria are not present.

SUMMARY

In a case, hemoptysis secondary to Behçet's disease was diagnosed in our department,. Therefore, we reviewed the literature concerned with this disease.

REFERENCES

1) Hunninghake GW, Fauci, AS. Pulmonary involvement in the collagen vascular diseases. Am Rev Respir Dis 1979; 119: 471-498.

2) Leavitt RY. Fauci AS. Pulmonary vasculitis. Am Rev Respir Dis 1986; 134: 149-166.

3) Fulmer JD, Kaltreider HB. The pulmonary vasculitites. Chest 1982; 82: 615-624.

4) Grenier P, Bletry O, Cornud F, et al Pulmonary involvement in Behcet's disease. Am J Roent 1981; 137: 565-569.

5) Dilşen N, Koniçe M, Gazioğlu K, Çavdar T, Ulugay İ. Övül C. Pleuropulmonary manifestation in Belçet's diseases. Int Symp Behcet's Disease, İstanbul, 1977.

6) Akkaynak, S, Enacar N, Çobanlı B, Behçet hastalığı ve akciğer tromboembolisi. Tüberküloz ve Toraks 1973; 21: 5-6.

7) Fırat T. New concept on the cardinal manifestations of Behçet's diseases. Jpn. J. Ophtalmol 1979; 23: 46-48.

8) Ateş M, Erdin E, Uçan ES, Akkoçlu A. Behçet hastalığında vena cava superior sendıomu. Ege Üniv. Tıp Fak. Dergisi 1985; 24: 795-803.

9) Gamble CN, Weisner KB, Shapiro RF, Boyer WJ. The immune complex pathogenesis of glommerulonephritis and pulmonary vasculitis in Behçet's disease. Am J Med. 1979; 6: 1031-1039.

10) Durıeux P, Bletry O, Huchon G, Wechsler B, Chretıen J, Godeau P. Multiple pulmonary arterial aneurysms in BehJet's disease and Hughes Stovin syndrome. The Am J. Med. 1981; 71: 736-741.

11) Hughes JP, Stovin PGI. Segmental pulmonary artery aneurysms with peripheral venous thrombosis. Br J Dis. Chest 1959; 53: 19-27.

12) Teplick JG, Haskin ME, Medwick A. The Hughes-Stovin syndrome-case report. Radiology 1974; 113: 607-608.

13) Cadman EC, Lundberg WB, Mitchell MS. Pulmonay manifestations in Behçet syndrome. Case report and review of the literature. Arch Intern Med 1976; 136: 844-947.

14) Hills EA. Behcet's syndrome wih aortic aneurysms. Br Med J 1967; 4: 152-154.

15) Çobanlı B. Behçet hastalığında akciğer plevra bulguları. Türkiye Klinikleri 1985; 5: 423-424