A Behçet’s disease patient with intracardiac thrombus, pulmonary artery aneurysms complicating recurrent pulmonary thromboembolism

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ÖZET

Tekrarlayan pulmoner tromboemboli ile komplike pulmoner arter anevrizması ve intrakardiyak trombüs ile seyreden Behçet hastalığı olgusu


Anahtar Kelimeler: Behçet hastalığı, pulmoner arter anevrizması, pulmoner tromboemboli, intrakardiyak trombüs.

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Behçet’s disease is a multisystem disorder presenting with recurrent buccal aphthosis, genital ulcer and uveitis with hypopion (1). Pulmonary involvement in Behçet’s disease is rare, occurring 1 to 7.7% of patients (2,3). Pulmonary artery aneurysms, arterial and venous thrombosis, pulmonary infarction, recurrent pneumonia, bronchiolitis obliterans organised pneumonia, and pleurisy are the main features of pulmonary involvement in Behçet’s disease (4). There is frequently coexistence of pulmonary and cardiac complications. Cardiac involvement includes: coronary artery disease, recurrent pericarditis, myocardiopathy and endocardic involvement. Intracardiac thrombus formation is very uncommon (5).

We present a Behçet’s disease patient with intracardiac thrombus, pulmonary artery aneurysms complicating recurrent pulmonary embolism.

CASE REPORT

A twenty-years-old man was admitted to hospital due to dyspnea, haemoptysis, fever and partially loss of vision. On examination, there was no pathological finding except high temperature with 38.5°C. The chest radiograph showed bilateral, peripheral localized patchy infiltration. Considering pulmonary thromboembolism (PE), lung perfusion scintigraphy was done. Considerable perfusion defects were found in both lungs. On dynamic thorax computed tomography (CT), there was aneurysmatic dilatation and thrombus in bilateral pulmonary artery segments and also findings of pulmonary thromboembolism. A diagnosis of Behçet’s disease was made based on his clinical course and radiological findings. During treatment, the patient was admitted two times to the hospital because of recurrent pulmonary thromboembolism. At the 10th months of follow up, partially dissolution of the thrombi and pulmonary defects were observed and right ventricular thrombus was revealed by dynamic thorax CT. On a follow up period of 16 months the patient is still under treatment and doing well. We present this case because Behçet’s disease is a rarely considered cause of recurrent pulmonary embolism and intracardiac thrombus which is seen under treatment.

SUMMARY

A Behçet’s disease patient with intracardiac thrombus, pulmonary artery aneurysms complicating recurrent pulmonary thromboembolism

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Intracardiac thrombus and pulmonary embolism is a very rare manifestation of Behçet’s disease. A twenty-years-old man was admitted to hospital due to dyspnea, haemoptysis, fever and partially loss of vision. On dynamic thorax computed tomography (CT), there was aneurysmatic dilatation and thrombus in bilateral pulmonary artery segments and also findings of pulmonary thromboembolism. A diagnosis of Behçet’s disease was made based on his clinical course and radiological findings. During treatment, the patient was admitted two times to the hospital because of recurrent pulmonary thromboembolism. At the 10th months of follow up, partially dissolution of the thrombi and pulmonary defects were observed and right ventricular thrombus was revealed by dynamic thorax CT. On a follow up period of 16 months the patient is still under treatment and doing well. We present this case because Behçet’s disease is a rarely considered cause of recurrent pulmonary embolism and intracardiac thrombus which is seen under treatment.

Key Words: Behçet’s disease, pulmonary artery aneurysms, pulmonary thromboembolism, intracardiac thrombus.
Dynamic thorax CT was repeated at the end of the 10th month of the treatment. Partially dissolution of the thrombi and pulmonary defects were observed and irregular low dense area, measured 2 x 1 cm, was seen in the right ventricle (Figure 3). On echocardiography, it was revealed right ventricular thrombus. Because of intracardiac thrombus formation under treatment and repetitive attacks of PE, we added warfarin to combination therapy. On a follow up period of 16 months, the patient is still under treatment and doing well.

**DISCUSSION**

Intrathoracic manifestations of Behçet’s disease consist mainly of thromboembolism of the superior vena cava and/or other mediastinal veins; aneurysms of the aorta and pulmonary arteries; pulmonary infarct and hemorrhage; pleural effusion; and rarely, myocardial, pericardial involvement, cor pulmonale, and mediastinal or hilar lymphadenopathy (6). Pulmonary infarction is a step in the natural course of the disease. Pulmonary vasculitis and thromboses of pulmonary vessels may cause infarctions, focal or diffuse hemorrhages and focal areas of atelectasis (6-8). For this reason, thromboembolism from a cardiac cavity has previously been deemed to be relatively uncommon (9). A review by Moğulkoç et al. regarding intracardiac thrombus in 25 patients with Behçet’s disease previously published in 21 reports (5). It is also noted that PE was seen in 13 patients (52%). In seven of these 13, thrombophlebitis was observed in the major vessels and may have been the source of the embolism. Although deep venous thrombosis of the lower extremities frequently accompanies pulmonary artery aneurysms, pulmonary thromboembolism is very rare in Behçet’s disease because the thrombi in inflamed veins are strongly adherent (10).

Another review consisting reports of Turkish authors revealed one intracardiac thrombus out of 56 (1.78%) Behçet’s disease patients (11). Recently, two Behçet’s disease patients with intracardiac thrombus and pulmonary artery aneurysms have been reported (12,13). We present a Behçet’s disease patient with intracardiac thrombus, pulmonary artery aneurysms complicating recurrent pulmonary embolism. Intracardiac thrombus can give rise to pulmonary embolic phenomena and/or pulmonary embolic
phenomena was occurred from thromboses of pulmonary vessels in our patient. Is the rate of cardiac thrombus really low as seen in these reports or cardiac thrombus occurred in any time during the course of disease as seen in our patient? The frequency of this complication may be underestimated because of the clinical presentation of intracardiac thrombus is nonspecific in the majority of patients.

By the help of new imaging techniques; especially, dynamic thorax CT can be helpful including thrombus of the systemic veins, heart and pulmonary arteries (8). On dynamic thorax CT, it was revealed right ventricular thrombus in our patient. The thrombus was confirmed by echocardiography. We used combination therapy with methylprednisolone and cyclophosphamide, which is the treatment of choice in other severe systemic vasculitis (3,14). Anticoagulation carries significant risks for patients with pulmonary artery aneurysms and must be used cautiously and only after systemic immunosuppressant treatment has been given (4). In our case, pulmonary thromboembolism recurred two times in the 5th and 11th month of treatment and intracardiac thrombus was found. Therefore we added warfarin to the combination therapy because of recurrent thromboembolism under treatment.

As a result, when seeking an aetiology of pulmonary thromboembolism in a patient, Behçet’s disease should be kept in mind. Pulmonary embolism in Behçet’s disease might be related to intracardiac thrombus. The intracardiac thrombus may be a step in the natural course of the disease like in our patient. Therefore echocardiographic examination can be useful in detecting them.

REFERENCES