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CASE REPORT

Isolated pulmonary arterial stenosis caused by Takayasu's arteritis in an elderly male

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KEYWORDS
Takayasu's arteritis;
Pulmonary hypertension;
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Summary Takayasu's arteritis has often been difficult to diagnose because of a lack of typical symptoms and other specific makers. We report here a case of Takayasu's arteritis in a 73-year-old man who was considered to exhibit isolated pulmonary artery involvement.

Pulmonary hypertension and right heart failure and severe stenosis in the main trunk and left pulmonary artery were observed. There was nothing remarkable in his routine blood-sample tests other than increased CRP and ESR. There were neither infectious nor collagen diseases. Anti-cardiolipin antibody, Antiphospholipid Syndrome, PR3-ANCA and MPO-ANCA were negative.

We diagnosed the patient as having Takayasu's arteritis based on chronic inflammation and the morphologic features of pulmonary artery lesion. However, other large vessels and the aorta were not involved. Treatment was started with glucocorticoids. The symptoms gradually improved, and pulmonary artery pressure estimated by echocardiography decreased along with inflammatiory markers. There were no remarkable changes in the stenotic lesions in the pulmonary artery but the flow limit in the left pulmonary artery was improved.

Introduction

Takayasu's arteritis is a chronic nonspecific inflammatory disease of large vessels such as the aorta and other major branches. The etiology of this disease has not yet been clarified. While it is not

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unusual for pulmonary arteries to be involved in this disease, there have been only a few reports involving isolated pulmonary arteries. Takayasu's arteritis has often been difficult to diagnose because of lack of typical symptoms and other specific markers. We report here a case of Takayasu's arteritis in a 73-year-old man who was considered to exhibit isolated pulmonary artery involvement.

Case report

A 73-year-old male had suffered from a continuous low-grade fever from September 2005 and needed to be tested in a hospital. There was nothing remarkable in his routine blood-sample tests other than increased C-reactive protein (6.5 mg/dl). Erythrocyte sedimentation rate (ESR) (60 min) 66 mm and WBC: 5100/μL were normal. Gallium scintigraphy showed increased uptake to the hilum of the lungs. Computed tomography (CT) and magnetic resonance imaging (MRI) showed artery wall thickening and a tumorous lesion in the main trunk and the left pulmonary artery (Fig. 1A and B).

The artrial dye did not enhance the pulmonary artery enough to effect the CT mean existence of flow limit of left pulmonary artery.

A biopsy through a thoracoscope was attempted to obtain detailed information. However, nothing existed outside the pulmonary artery. After a month without medication, the fever decreased and his general condition improved. At first, we did not doubt Takayasu's arteritis but a tumor which we could not judge as malgnant or benign. So we suggested to remove the tumor but the patient rejected the operation.

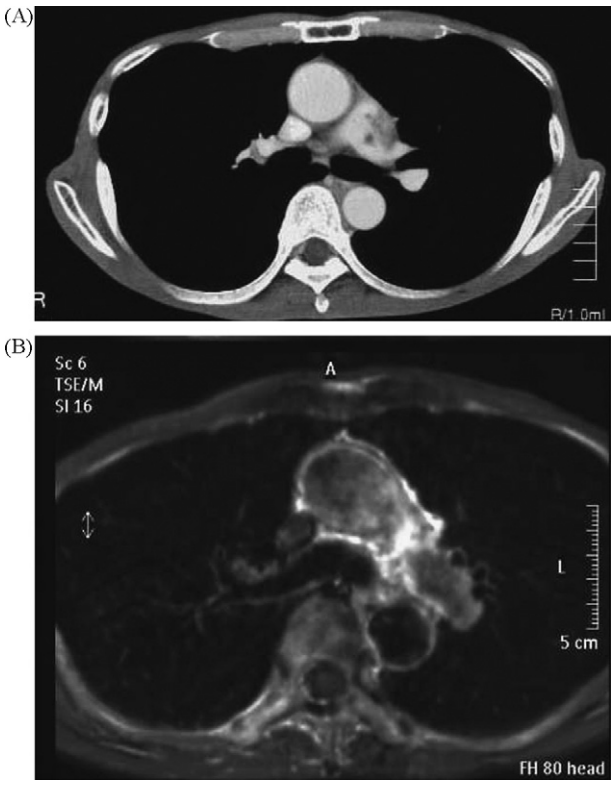

Figure 1 Computed tomography (CT, A) and magnetic resonance imaging (MRI, B) showed a tumorous lesion in the left pulmonary artery and artery wall thicking and stenotic lesion in the main trunk and the left pulmonary artery.

The patient returned to the hospital with general fatigue in June 2006. Due to the long-term duration of the rise in C-reactive protein (CRP) and the ESR, infectious and collagen diseases were ruled out. Anti-cardiolipin antibody, Antiphos-

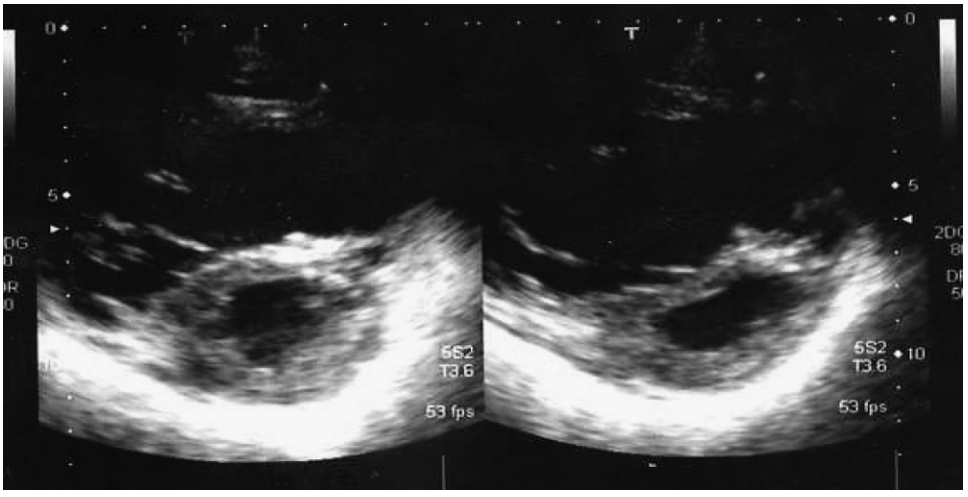

Figure 2 Echocardiography showed right heart loading.

pholipid Syndrome, PR3-ANCA, MPO-ANCA were negative.

Pulmonary hypertension and right heart failure manifesting hepatomegaly, ascites, jugular venous congestion, and pretibial pitting edema were observed. Echocardiography showed right heart loading (Fig. 2). Enhanced chest CT (Fig. 3A and B) showed a change in the pulmonary artery lesion. Tumorous lesion in the left pulmonary artery disappeared and severe stenosis in the main trunk and left pulmonary artery was observed. In fact, tumorous lesion was probably thrombus. Thrombus in the left pulmonary artery was due to the flow limit of pulmonary artery. Pulmonary perfusion scans showed reduced perfusion through most of the left lung that mismatched a pulmonary ventilation scan (Fig. 4A and B). We diagnosed the patient as having Takayasu's arteritis based on chronic inflammation and the morphologic features of pulmonary artery lesion. But other large vessels and the aorta were not involved. Treatment was started with glucocorticoids (initially 625 mg per day for 3 days and then 40 mg per day) to control Takayasu's arteritis. The symptoms gradually improved, and pulmonary artery pressure estimated by echocardiography decreased (pressure gradient 55—42 mmHg) along with the inflammation marker (CRP 10.4—0.1 mg/L) (Fig. 5).

There were no remarkable changes in the stenotic lesions in pulmonary artery but the flow limit in the left pulmonary artery was improved and pulmonary artery was enhanced enough in enhanced CT the flow limit in the left pulmonary artery was improved. But after 1 year, He developed lung abscess and pneumonia in the lower left lung because of lack of left lung perfusion due to Takayasu's arteritis.

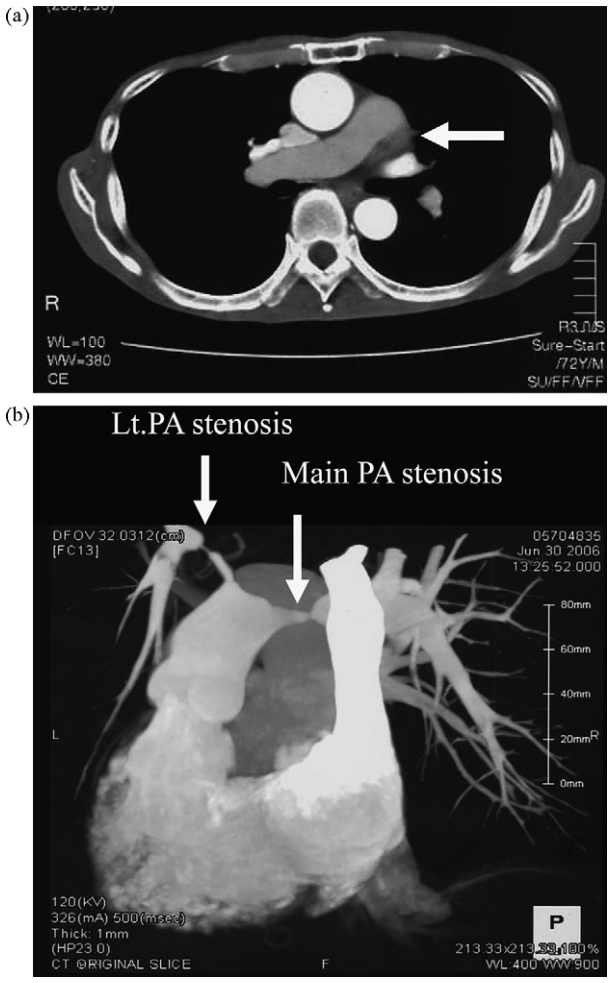

Figure 3 Enhanced chest computed tomography showed thrombus in the left pulmonary artery disappeared (A) but severe stenosis in the main trunk and left pulmonary artery (B).

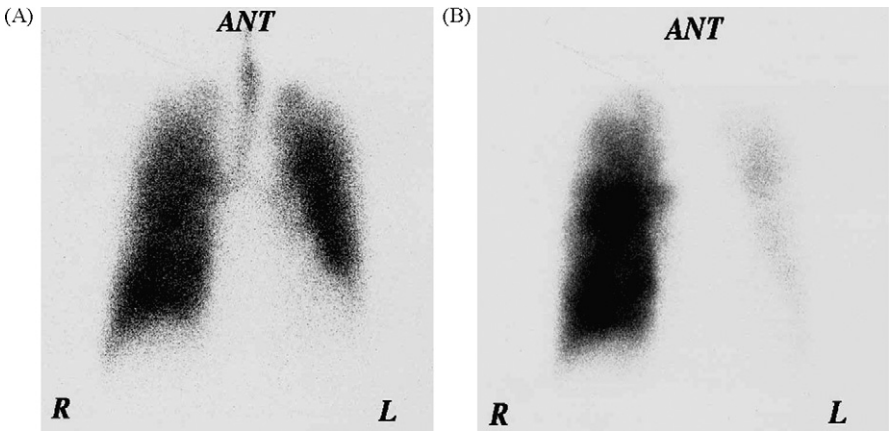

Figure 4 Pulmonary perfusion scans by scintigraphy showed a decrease in perfusion through most of the left lung that mismatched a pulmonary ventilation scan. (A) ventilation scan, (B) pulmonary perfusion scans.

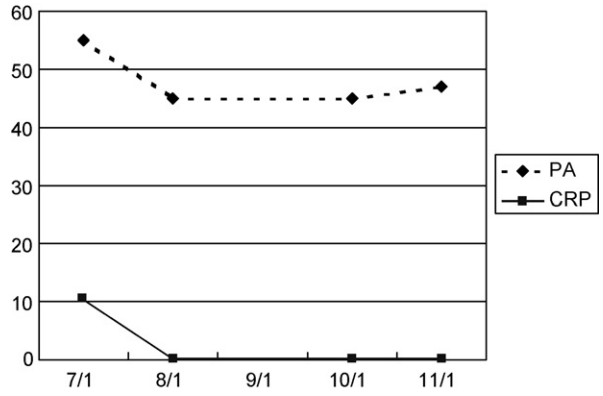

Figure 5 After the treatment with glucocorticoids, pulmonary artery pressure decreased along with the inflammation marker.

Discussion

We encountered a case of Takayasu's arteritis in a 73-year-old male who was difficult to diagnose because of lack of specific symptoms and aortic lesions. Kerr et al. [1] reported that the median delay between the onset of the first symptoms and diagnosis of the disease was 10 months in 60 cases of Takayasu's arteritis. The spectrum of clinical manifestations is thought to lead to a delay in diagnosis as was seen in our case. This disease often occurs in females (80—90%) during the second or third decade of their life.

It is reported that Takayasu's arteritis over the age of 40 is about 10—20% but male cases of Takayasu's arteritis over the age of 60 is rare [1,2].

The signs and symptoms of pulmonary Takayasu's arteritis may be subtle and some cases have been reported that mimic, for example, pulmonary thromboembolism, which has symptoms of cough, dyspnea, and hemoptysis [3—5]. In case of unexplained pulmonary vascular disease, Takayasu's arteritis should be considered.

In this case, in addition to the absence of major symptoms of Takayasu's arteritis, pulmonary hypertension and secondary right heart failure were present as the initial manifestation. Sharma et al. reported that the incidence of pulmonary artery involvement in Takayasu's arteritis was 56.1% 118 of 210 cases [6]. However, isolated pulmonary arterial stenosis caused by Takayasu's arteritis was recognized in only a few cases [7—9].

In the present case, there was no stenotic or dilated lesion of the aorta or its branches, nor was there any limb ischemia. Kerr et al. [1] reported that most cases of Takayasu's arteritis exhibit long segment lesions, which include vessel wall irregularity or post-stenotic dilatation. In this case at first, the patient showed pulmonary artery wall thickening, and thrombus which looked like a tumor with the absence of a stenotic lesion. Takayasu's arteritis was not suspected because of these morphologic features. We made the diagnosis of Takayasu's arteritis mainly based on the morphologic change in the pulmonary artery. Although we did not perform a histological examination, the final morphologic features of the pulmonary artery in this patient were equivalent to arteritis.

Glucocorticoids seem to be effective for most patients with active Takayasu's arteritis. The response to glucocorticoids during the first stage of treatment was reported to be over 50% [1,10,11]. Isolated pulmonary arterial lesions caused by Takayasu's arteritis are often diagnosed at severe progression because of the lack of symptoms. It is not clear whether glucocorticoids alone are an effective therapy in such patients. In this case, symptoms and pulmonary artery pressure showed remarkable improvement with glucocorticoid therapy alone, although the morphology of pulmonary artery stenosis on follow-up CT did not change.

Conclusion

We reported an isolated pulmonary arterial lesion caused by Takayasu's arteritis in a 73-year-old male, which appeared to be a tumor-like lesion in the early stage. Glucocorticoids alone improved the symptoms and may be effective therapy in isolated pulmonary arterial stenosis caused by Takayasu's arteritis.

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