Pulmonary Infarction as the Initial Manifestation of Takayasu’s Arteritis

Tomomi Nakamura1, Shinichiro Hayashi1, Mami Fukuoka1, Naoko Sueoka1 and Kohei Nagasawa2

Abstract

A 50-year-old woman reporting sudden-onset chest pain was diagnosed as having pulmonary infarction associated with Takayasu’s arteritis. She had experienced moderate malaise and cough for 3 months. Computed tomography (CT) and magnetic resonance imaging (MRI) showed wedge-shaped infiltrative shadows typical of pulmonary infarction in the right lung. Although pulmonary artery involvement in Takayasu’s arteritis is well documented, most patients show only signs of mild to moderate pulmonary hypertension. Few reports discuss patients with symptoms due to pulmonary infarction as the initial manifestation. Takayasu’s arteritis should therefore be considered a differential diagnosis in pulmonary infarction.

Key words: Takayasu’s arteritis, pulmonary artery involvement, pulmonary infarction

Introduction

Takayasu’s arteritis, a chronic nonspecific inflammatory disease of large vessels such as the aorta and its major branches, occurs most often in young women. Although more prevalent in Asians than other ethnic groups, the disease is distributed worldwide. In addition to the aortic arch and its branches, the pulmonary arteries are also involved in about 50-80% of the patients (1-7). A relatively small number of reports have referred to symptomatic events associated with pulmonary artery involvement as the initial manifestation at diagnosis, some detailing symptoms due to pulmonary artery obstruction, such as severe dyspnea, which may mimic pulmonary thromboembolism (5, 6, 8, 9). Devouassoux et al discussed a patient with pulmonary infarction associated with Takayasu’s disease (3). To expand this knowledge, we report a case of Takayasu’s arteritis with symptoms of pulmonary infarction as the initial manifestation.

Case Report

A 50-year-old woman was admitted for sudden-onset right chest pain in December 2003. She had suffered from general malaise and cough for more than 3 months and, was found to have a low-grade fever. She did not have a history of pulmonary thromboembolism. Her height and weight were 153 cm and 47 kg, respectively. She did not have histories of peripheral or deep venous thrombosis or gynecologic operation. She experienced parturition three times without any problems. Blood pressure measured supine was 98/70 mmHg in bilateral arms, 130/70 mmHg in the right leg and 126/70 mmHg in the left leg. PaO2 was 94 mmHg and PaCO2 42 mmHg breathing room air. A systolic murmur at the second intercostal space of the left sternal border and bilateral cervical bruits were noted. Peripheral blood count indicated a mildly elevated white blood cell count (8,600/μl) and a moderately elevated platelet count (64.2×10⁴/μl). Enhanced erythrocyte sedimentation at 130 mm/hour and increased C-reactive protein at 11.4 mg/dl indicated an inflammatory process. There was no sign indicating nephritis in blood and urine studies. Antinuclear antibody, PR3-antineutrophil cytoplasmic antibody, MPO-antineutrophil cytoplasmic antibody, and anticardiolipin antibody were negative. Activated partial thromboplastin time (APTT) was not prolonged. Chest radiography showed an infiltrative shadow in the lower right lung field, recognized as wedge-shaped

1 Division of Pulmonary, Department of Medicine, Saga University Medical School, Saga and 2 Division of Collagen Vascular Disease, Department of Medicine, Saga University Medical School, Saga
Received for publication December 9, 2005; Accepted for publication March 31, 2006
Correspondence to Shinichiro Hayashi, Department of Medicine, Saga University Medical School, 5-1-1 Nabeshima, Saga 849-8501
infiltrations in the subpleural area of the middle and right lower lobes on computed tomography (CT) (Fig. 1A) and magnetic resonance image (MRI) (Fig. 2). Chest CT and MRI showed abnormal wall thickening in the ascending aorta, the aortic arch and its 3 main branches, and the main pulmonary artery. Severe stenosis was notable in the distal portion of the right main pulmonary artery (Fig. 1B, Fig. 2). Pulmonary ventilation/perfusion scans showed multiple perfusion defects in the right lung (Fig. 3). No signs suggestive of pulmonary hypertension were seen in electrocardiography or echocardiography. We diagnosed the patient as having Takayasu’s arteritis. Anticoagulant therapy initiated with warfarin was followed by 40 mg/day prednisolone administration. As wall thickening of the major vessels lessened, the prednisolone dosage was gradually tapered to 7.5 mg/day and the patient remains in good condition.

**Discussion**

In our patient, the clinical findings indicated that arteritis in large vessels were affected. We diagnosed the patient as having Takayasu’s arteritis based on the following criteria: female gender, systemic inflammation, and stenosis and wall thickening of large vessels with elastic tissue in CT and MRI. Infiltrative shadows in the right lung were diagnosed as pulmonary infarction based on CT and MRI, i.e., subpleural T1-weighted hyperintense wedge-shaped infiltration on MRI. In large vessel involvement, temporal arteritis and anti-phospholipid syndrome (APS) should be considered in the differential diagnosis. However, temporal arteritis is unlikely because she did not suffer from headache and her temporal artery pulsation was normal. A diagnosis of APS seemed unlikely. The patient did not have any problems during pregnancy. Her APTT was not prolonged. Antinuclear antibody and anti-cardiolipin antibody were negative.
Takayasu’s arteritis, a chronic nonspecific inflammatory disease of the large vessels, affects arteries with abundant elastic tissue in the walls, such as the aorta and its major branches. Pulmonary artery involvement is reported to occur in 50-80% of unselected patients with Takayasu’s arteritis, which was identified using angiography and/or MRI (1-7). Pulmonary arterial abnormalities are distributed widely but unequally from the trunk to peripheral branches. Upper lobe pulmonary artery branches are most frequently involved and most often on the right side. Segmental branches are the most frequent site, followed by subsegmental branches. Stenosis and occlusion in the pulmonary artery are the most common lesions in Takayasu’s arteritis, occurring late in inflammation (10, 11). CT and MRI play an important role in detecting inflammatory processes early (5, 12), showing the wide spectrum of Takayasu’s arteritis lesions including stenosis, dilatation, aneurysm, and mural thrombi. MRI is especially well suited for demonstrating thickening of the pulmonary artery, which is difficult to appreciate on angiography. MRI is also useful in diagnosing pulmonary infarction (5). It demonstrates the presence of subacute alveolar hemorrhaging as T1-weighted hyperintense infiltrates, suggesting pulmonary infarction. In our case, CT and MRI were decisive in diagnosing pulmonary infarction in Takayasu’s arteritis.

Clinical manifestations of Takayasu’s arteritis vary widely, depending on the site and grade of arterial lesion. In addition to the general symptoms of asthenia, weight loss, and fever, predominant clinical features are reduced peripheral arterial pulse amplitude, vascular bruit, elevated blood pressure, and dyspnea on exertion (2, 13). These findings are mostly due to lesions of the aorta and its main branches. Although Takayasu’s arteritis frequently involves pulmonary arteries, symptoms rising from pulmonary artery lesions are less frequent (1-3, 6, 7, 13, 14). Since pulmonary artery stenosis develops chronically, most patients only show signs of mild to moderate pulmonary hypertension. Lupi et al retrospectively analyzed 22 patients with Takayasu’s disease and found that 50% of cases had pulmonary involvement (1). No patients had pulmonary symptoms, but 63% had clinical, radiological, and electrocardiographic findings suggesting pulmonary hypertension or right heart strain. Pulmonary artery obstruction is sometimes accompanied by severe symptoms. Some case reports have discussed patients with initial symptoms mimicking pulmonary thromboembolism (5, 6, 8, 9), mainly chest pain, shortness of breath, and hemoptysis. Some misdiagnosed with pulmonary thromboembolism were not treated with corticosteroids for several months to years. This delay in diagnosis led to severe dyspnea due to pulmonary hypertension and heart failure. Communication between the systemic and pulmonary arteries is another well-documented incident in Takayasu’s disease (7, 11). In a few cases, infiltrative shadows occurred due to pulmonary hemorrhage from collateral circulation between pulmonary arteries and bronchial arteries (6).

In the present case, chest CT and MRI findings were consistent with typical pulmonary artery involvement (5, 8, 10, 12). She initially had mild chest pain and infiltrative shadow derived from pulmonary infarction. She had no symptoms or signs suggesting pulmonary hypertension or right heart distress. Although many patients with Takayasu’s arteritis have pulmonary arterial stenosis, only a few complain of symptoms associated with pulmonary infarction. This is related to the fact that artery obstruction occurred gradually and with parallel development of collateral circulation (2). As the mechanism of pulmonary infarction in this case, the following possibility is considered: 1) thrombus of the main pulmonary artery wall was exfoliated, and occluded to peripheral pulmonary artery, 2) inflammation of peripheral pulmonary artery induced rapid occlusion, 3) hypercoagulopathy associated with Takayasu’s arteritis induced thrombosis in the peripheral pulmonary artery (15). Despite an intensive search of the English literature, we found only 2 patients reported to have pulmonary infarction associated with Takayasu’s arteritis (3, 5). Patients mainly reported sudden-onset chest pain and shortness of breath, but no pulmonary hypertension was noted in either case. As in the present patient, pulmonary infarction was the initial manifestation of Takayasu’s arteritis in one patient (5), whose clinical course was favorable owing to a good response to corticosteroids and heparin. Together with our experience, this suggests that it is important to consider Takayasu’s arteritis as an underlying disease in pulmonary infarction, where early treatment leads to a favorable clinical course. Examinations using enhanced CT and MRI are useful in differentiating among conditions.

References

8. Shlomai A, Hershko AY, Gabbay E, Ben-Chetrit E. Clinical and radiographic features mimicking pulmonary embolism as the first symptom.

© 2006 The Japanese Society of Internal Medicine
http://www.naika.or.jp/imindex.html