Extensive Pulmonary Arterial Thrombi in Situ in Association With Atrial Septal Defect

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A 48-year-old woman with cyanosis was referred for investigation of atrial septal defect (ASD). Blood gas analysis on admission revealed moderate hypoxemia, and a pressure study during right heart catheterization revealed pulmonary hypertension (PH). Spiral computed tomography (CT) scan disclosed extensive thrombi in dilated large symmetrical pulmonary arteries with clear lung fields, and large strand-like thrombi on the inner surface of the pulmonary arterial wall along the vascular curvature were visualized by virtual CT angioscopic imaging. The thrombi were eventually considered to be not thromboemboli but thrombi in situ, because no segmental or larger defects were detected in the lung perfusion scan, although it showed cardiovascular imprints and an inhomogeneously decreased perfusion pattern. Pulmonary thrombi in situ are an uncommon manifestation in patients with ASD, and have not been described from the evidence of both CT and lung perfusion scans. The findings indicate that pulmonary thrombi in situ are not associated with occlusion of the large pulmonary arteries and the resultant development of PH. The patient was conservatively treated with medication, and the pulmonary thrombi did not show significant change with anticoagulant therapy. She died suddenly at the age of 51 years. (*Circ J* 2003; 67: 640–642)

Key Words: Atrial septal defect; Lung perfusion scan; Pulmonary hypertension; Pulmonary thrombi in situ; Virtual CT angiography

Extensive thrombi in situ in the large pulmonary arteries have been reported in only a small number of patients with atrial septal defect (ASD).^{1,2} We describe a case of extensive pulmonary thrombi in a patient with ASD, and discuss the usefulness of the combination of computed tomography (CT) and lung perfusion scans in the diagnosis.

Case Report

A 48-year-old woman with cyanotic lips and clubbed fingers complained of shortness of breath and exertion dyspnea. She had been diagnosed with ASD in her childhood, but had not undergone surgical correction. She had neither leg thrombophlebitis nor coagulopathy.

On admission, her chest X-ray showed an enlarged cardiac silhouette with marked prominence of the proximal pulmonary vasculature, and clear lung fields (Fig 1). Blood gas analysis disclosed a PaO₂ of 54 mmHg, a PaCO₂ of 43 mmHg, and a pH of 7.43 at room air. Echocardiography revealed a large second ostial ASD, as well as extreme enlargement of the right ventricle and the main trunk of the pulmonary artery. Right heart catheterization revealed a right atrial mean pressure of 5 mmHg, right ventricular pressure of 82/9 (systole/diastole) mmHg, and pulmonary

arterial pressure of 83/30 (systole/diastole) mmHg (mean 53 mmHg). The left-to-right and right-to-left shunt ratios were 0.17 and 0.40, respectively. The pulmonary to systemic flow ratio was 0.70, and the ratio of the peak pulmonary to peak systemic arterial pressure was 0.76. The ratio of total pulmonary arterial resistance to systemic vascular resistance was 0.94.

Pulmonary angiography demonstrated equivocal filling defects, because the pulmonary arteries were too large to be enhanced sufficiently by the usual volume of contrast media with a high concentration of iodine. Enhanced spiral CT (Siemens Somatom Plus 4) (Figs 2,3) revealed extensive filling defects in the large pulmonary arteries, which were considered to be thrombi. The left anterior chest wall protruded anteriorly because of compression by the dilated

Fig 1. Chest X-ray showing a large cardiac shadow with extreme proximal prominence of the pulmonary vasculature and clear lung fields.

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Fig2. Enhanced cross-sectional CT image demonstrating extensive thrombi in the extremely enlarged right proximal pulmonary arteries. AA, ascending aorta; DA, descending aorta; MPA, main pulmonary artery; RPA, right pulmonary artery; LPA, left pulmonary artery; SVC, superior vena cava.

Fig3. Enhanced cross-sectional CT image 5 cm above that shown in Fig2, demonstrating thrombi in the enlarged left proximal pulmonary arteries. AAr, aortic arch; LPA, left pulmonary artery; SVC, superior vena cava.

main pulmonary artery. The thrombi were located in the dilated trunk of the pulmonary artery and in the dilated right and left main pulmonary arteries. The thrombi appeared to extend to the segmental left ascending or right descending pulmonary arteries, but their full extent distally was not confirmed. The pulmonary arteries were symmetrically dilated, and there was not a mosaic attenuation pattern in the lung fields.

Magnetic resonance imaging also demonstrated the pulmonary arterial thrombi, and their signal characteristic was consistent with that of thrombi. The undulating surface of the strand-like thrombi along the curvature of the large pulmonary arteries were visualized by virtual CT angioscopic imaging (Fig4A,B). Enhanced CT scanning of the legs showed neither venous dilatation nor filling defects in the venous system.

A lung perfusion scan with ^{99m}Tc -macro aggregated albumin did not show segmental or larger defects, but there was a suspicion of a mottled perfusion pattern (Figs5,6), although perfusion defects or inhomogeneously decreased perfusion areas appeared in the fields overlapping or not overlapping the heart and dilated pulmonary arteries, respectively.

Pulmonary biopsy was not performed, because the patient refused surgery. She was conservatively treated with medications, such as digitalis, warfarin, children's bufferin and oxygen inhalation. Follow up echocardiography revealed neither regression nor progression of the pulmo-

Fig4. (A) Virtual CT angioscopic image obtained from a head to foot projection showing the smooth inner surface of the dilated pulmonary arteries and the extensive strand-like thrombi on the vessel wall following the curvature of the pulmonary arteries. (B) Schema of Fig4A showing massive thrombi (checked zone) in the pulmonary arteries. MPA, main pulmonary artery; AA, ascending aorta; DA, descending aorta; SVC, superior vena cava; RPA, right pulmonary artery; LPA, left pulmonary artery; LPV, left pulmonary vein.

nary thrombi. She died suddenly at the age of 51 years and permission for a postmortem autopsy was not granted.

Discussion

The proximal pulmonary arteries in the present case were so enormously dilated that the vascular lumen was insufficiently opacified with conventional pulmonary angiography and the filling defects were not obvious. Enhanced-CT scan was useful for detecting the central pulmonary thrombi³ but fibrosing mediastinitis, tumor invasion of major pulmonary arteries and Takayasu's disease should be excluded before the diagnosis of pulmonary thrombi is made from the CT findings⁴.

The defects in the lung perfusion scan of the present case are the imprints of the heart and dilated proximal pulmonary arteries, and the inhomogeneously decreased areas of perfusion are considered to be the mottled perfusion that frequently appears in the advanced stage of ASD with pulmonary hypertension (PH)⁵.

We have found that the thoracic vascular features associated with ASD are well shown by 3-dimensional CT angiography⁶ and the inner surface of the vascular lumen is successfully visualized by virtual CT angiography⁷ as also shown by the present case. The boundaries of the thrombi were determined by reference to the cross-sectional CT images for the appropriate presentation of the surface of the thrombi and vascular lumen.

Virtual CT angiography is superior to real angiography, because it is less invasive and has a wider angle view and

Fig5. Anterior view of the lung perfusion scan shows a mottled pattern, the imprint of the enlarged heart and the dilated pulmonary arteries. The mild uptake in the kidney and thyroid gland proved to be an intracardiac right-to-left shunt.

Fig6. Posterior view of the lung perfusion scan shows a mottled pattern, the imprint of the heart and the dilated pulmonary arteries, particularly the left upper proximal pulmonary arteries, and bilateral renal uptake, which also proved to be an intracardiac right-to-left shunt.

brighter distant lumen. In addition, the inner surface of the larger vessels can be visualized from many projections. Therefore, virtual CT angiography is an alternative to real angiography, when it is used with a rapid reconstruction time⁸

Pulmonary thrombi in situ were reported in connection with ASD in a postmortem examination¹ and Schamroth et al described the pulmonary thrombi in 8 patients with ASD and PH as being thrombi in situ; they considered that the pulmonary thrombi had been developed in progression of PH². Their conclusion was based on limited evidence from cardiac catheterization and pulmonary angiography, and not from CT or lung perfusion scans, although they considered CT scanning as a good modality for demonstrating pulmonary thrombi.

It is important to determine whether or not the pulmonary thrombi are related to the development of PH, and the CT scans usually provide useful information about whether the thrombi are in situ or embolic;⁹ for example, asymmetrical dilatation of the pulmonary arteries or a mosaic attenuation pattern of the lung fields is commonly seen in patients with chronic thromboembolic PH. The CT features of the present case differ from those seen with chronic thromboembolic PH. A lung perfusion scan without segmental or larger defects does not mean any occlusion of large pulmonary arteries, and there has not been any reports of patients with chronic thromboembolic PH in whom the lung perfusion scan does not demonstrate at least one segmental or larger defect⁴. Therefore, we consider that the pulmonary thrombi in the present case were not the major cause of the progression of PH.

We believe that PH in the present case developed not by the definite occlusion of the large pulmonary arteries, but by an increase in pulmonary resistance because of the chronic ASD. Therefore, pulmonary thrombi in situ may have no association with PH in certain patients with ASD, and such a case was reported by Murata et al¹⁰

The cause of thrombi in situ in the large pulmonary arteries is not known, although endothelial splitting in the dilated arteries is thought to be the initial process⁵; and secondary flow or flow stasis in the large pulmonary arteries

revealed by echocardiography are regarded as closely related factors in the thrombus formation!¹⁰

Anticoagulants are the therapeutic strategy for pulmonary thrombi in situ and were used in the present case². Pulmonary biopsy should be performed, if the patient consents to surgical correction!¹¹

In conclusion, we describe a rare case of extensive thrombi in situ in the large pulmonary arteries that appeared in association with ASD. CT and lung perfusion scans were helpful in the diagnosis. In addition, virtual CT angioscopic images of the large strand of the thrombi in the pulmonary arteries were demonstrated, which has not been done in such pulmonary thrombi in situ before.

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