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## 症例報告

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### RUPTURED SINUS OF VALSALVA ANEURYSM IN PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS

KAZUO NISHIMOTO, SHINICHI FUJIMOTO\*, HIDEO SHIIKI, TAKASHI FUJIMOTO,  
REIKO MIZUNO, TAKASHI UEDA\*\*, SHIGEKI TANIGUCHI\*\*,  
HIROSHI NAKANO\* and KAZUHIRO DOHI

*First Department of Internal Medicine, \*Department of Clinico-Laboratory Diagnostics,  
and \*\*Third Department of Surgery, Nara Medical University*

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*Abstract*: We report the case of a 57-year-old woman with systemic lupus erythematosus associated with a ruptured sinus of Valsalva aneurysm. The physical examination showed a holosystolic murmur at the left fourth intercostal space. Aortography revealed that the sinus of Valsalva aneurysm had ruptured into the right ventricle. Surgical closure of the ruptured aneurysm was successfully carried out. This aneurysm and its rupture may have been due to endocarditis involving the sinus of Valsalva.

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**Key words**: systemic lupus erythematosus, ruptured sinus of Valsalva aneurysm

#### INTRODUCTION

Systemic lupus erythematosus (SLE) is a disease which involves multiple organs. Cardiac involvement is especially important prognostically, and is usually manifested as pancarditis. The pathological prevalence of cardiac lesions is reported to be approximately 40~50%<sup>1)</sup>. Pericarditis is the most common cardiac involvement; its clinical diagnosis is made in only 20~30% of all cases because it is often clinically silent<sup>1)</sup>. Arterial lesions are also frequent<sup>2)</sup>, but it is frequently difficult to determine whether these are primary or secondary lesions. We report the case of a patient with SLE and a ruptured sinus of Valsalva aneurysm.

#### CASE REPORT

A 57-year-old Japanese woman with SLE had been treated with adrenal corticosteroids (prednisolone 5~10 mg/day) since the age of 47 at our hospital's out-patient clinic. At the age of 47, she was admitted to our hospital because of proteinuria, and was diagnosed as SLE because of discoid lupus, proteinuria (0.5~0.7 g/dl), positive antinuclear and anti-dsDNA antibodies, facial edema and joint pain. Early in December of 1992, she felt general malaise and anterior chest discomfort, and a holosystolic murmur at the left 4th intercostal space was noted on the 24th of December. Surgical closure of a ruptured sinus of Valsalva aneurysm was successfully carried out. Physical examination of the patient revealed a blood pressure of 164/80 mmHg and a heart rate of 102 beats/min. A holosystolic murmur (Levine 4/6) was heard

at the 4 th intercostal space at the left sternal border. The liver was palpable 1 cm below the costal margin in the midclavicular line. Peripheral edema was absent. Chest radiography showed cardiomegaly (cardiothoracic ratio of 64 %). Laboratory findings on admission were as follows : red blood cell count  $446 \times 10^4 / \mu\text{L}$ , hemoglobin 13.9 g/dL, hematocrit 40.1 %, white blood cell count  $8,700 / \mu\text{L}$ , platelet count  $16.8 \times 10^4 / \mu\text{L}$ , aspartate aminotransferase (GOT) 11 IU/L, lactate dehydrogenase (LDH) 434 IU/L, creatinine kinase (CK) 31 IU/L, Antinuclear antibodies were positive but anti-RNP antibodies and anti-SM antibodies were negative. The electrocardiogram showed high voltage in the left lateral chest leads. A color Doppler echocardiogram showed a jet across the sinus of Valsalva into the right ventricle (Fig. 1). Aortography confirmed that the sinus of Valsalva aneurysm had ruptured into the right ventricle (Fig. 2). Pathologic findings of the post-surgical specimen of the ruptured sinus of Valsalva showed thinning of the arterial wall and ruptured elastic fibres, stained by alcian blue (Fig. 3).

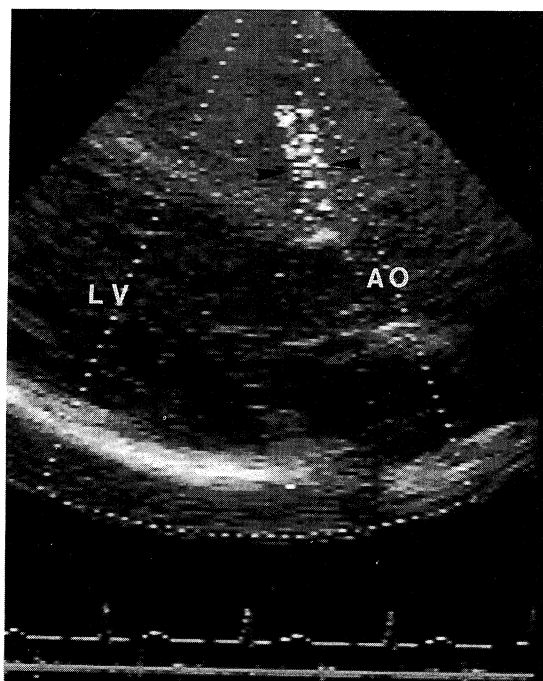


Fig. 1. Doppler echocardiography  
Color Doppler jet is seen from the sinus of Valsalva into the right ventricle (black arrow).  
(AO; aorta, LV; left ventricle)

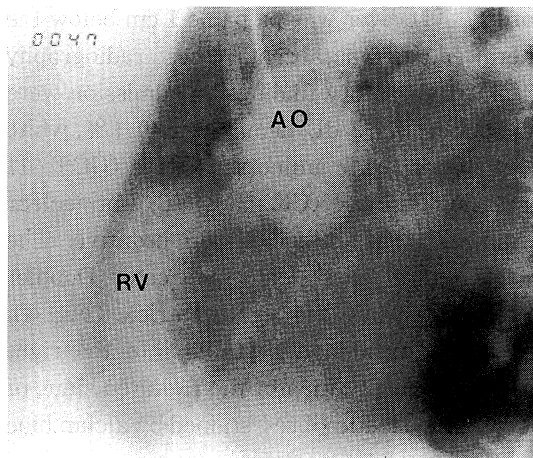


Fig. 2. Aortography  
Shunt flow is seen from aorta to the right ventricle through the ruptured sinus of Valsalva aneurysm. (AO; aorta, RV; right ventricle)

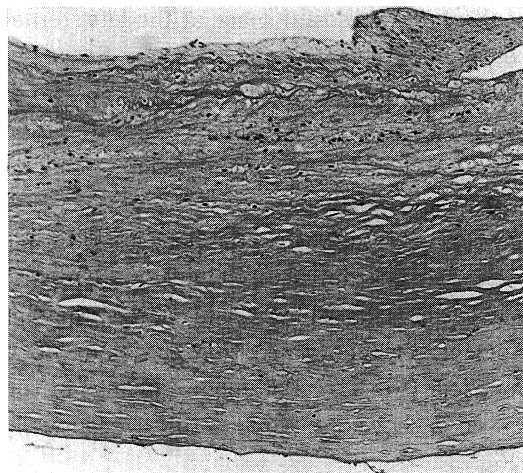


Fig. 3. Post-surgical specimen of the ruptured sinus of Valsalva. There is a little mononuclear cell infiltration. (Alcian-blue stain,  $\times 200$ )

## DISCUSSION

Although heart failure in patients with SLE is rare, mild forms of cardiac lesions are common. These cardiac abnormalities are usually silent. However, some patients develop more severe endocardial lesion. On the other hand, most of the sinus of Valsalva aneurysms are congenital except for the mycotic or bacterial infection. Because of an assumed intrinsic weakness at the union of the aorta, aortic media may separate from the aortic annulus and retract upward. Usual sites of defect are right coronary cusp in 75 % of all cases, and non-coronary cusp in 25 %<sup>3)</sup>. Vascular lesions in SLE are relatively common, both as primary lesions and also secondary to corticosteroid therapy. In patients with SLE, coronary artery stenosis is relatively more frequent than normal, approximately 30~40 %. Corticosteroid therapy can also induce cardiac abnormalities<sup>4)</sup>, such as systemic hypertension, left ventricular hypertrophy, epicardial and myocardial fat increases, and accelerated coronary atherosclerosis. It is often difficult to differentiate between primary cardiac lesions due to SLE and those from secondary complicating conditions. Sinus of Valsalva aneurysms in patients with SLE have been reported by Lokhandwala *et al.*<sup>5)</sup>. They speculated that SLE might be a cause of such aneurysms. We cannot rule out the possibility of a coincidental sinus of Valsalva aneurysm in this case. It is possible that the aortic lesions of SLE produced the sinus of Valsalva aneurysm, or that a coincidental sinus of Valsalva aneurysm ruptured due to both SLE and corticosteroid-induced atherosclerosis.

Thus, sinus of Valsalva aneurysm should be ruled out as one of the cardiovascular abnormalities in patients with SLE.

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