

Immobile right aortic leaflet in a case of the aortitis syndrome complicated by a left coronary sinus aneurysm

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Summary

A 37-year-old housewife had the aortitis syndrome complicated by acute aortic regurgitation and severe coronary insufficiency. Her echocardiogram at the level of the aortic valve ring revealed an enlarged left aortic sinus and an oscillating membranous structure centrally in the lumen of the aorta. At emergency surgery, there was no intimal flap in the ascending aorta, but there was a 4 cm diameter aneurysm of the left aortic sinus. The leaflets of the aortic valve *per se* were normal. Following its surgical repair and an aortic valve replacement, prednisolone was administered, and all abnormal laboratory data were corrected. The oscillating structure in the ascending aorta was considered to be the right aortic leaflet whose reduced mobility was caused by a distorted aortic valve ring due to the aneurysm. An immobile aortic leaflet is an extremely rare echocardiographic finding worthy to be reported.

Key words

Immobility of the aortic leaflet

Intimal flap

Left coronary sinus aneurysm

Aortitis syndrome

Dilatation of the ascending aorta is not infrequent in the aortitis syndrome¹⁾. Usually, such dilatation is diffuse rather than localized¹⁾. Should localized dilatation occur near the aortic valve, the surrounding structures may become distorted, causing valvular dysfunction such as aortic regurgitation. Described here is a case of the aortitis syndrome with immobility of the right aortic leaflet secondary to distortion of the aortic valve ring by an aneurysm of the left aortic sinus.

Case report

This 37-year-old housewife was in good health until she first noticed high fever of several weeks' duration ten months previously. The cause of the fever was not determined. Since the middle of April, 1982, she experienced the onset of shortness of breath at rest and exertional dyspnea with increasing severity. A local practitioner diagnosed aortic regurgitation with severe inflammation. After her recovery from acute left heart failure, she was trans-

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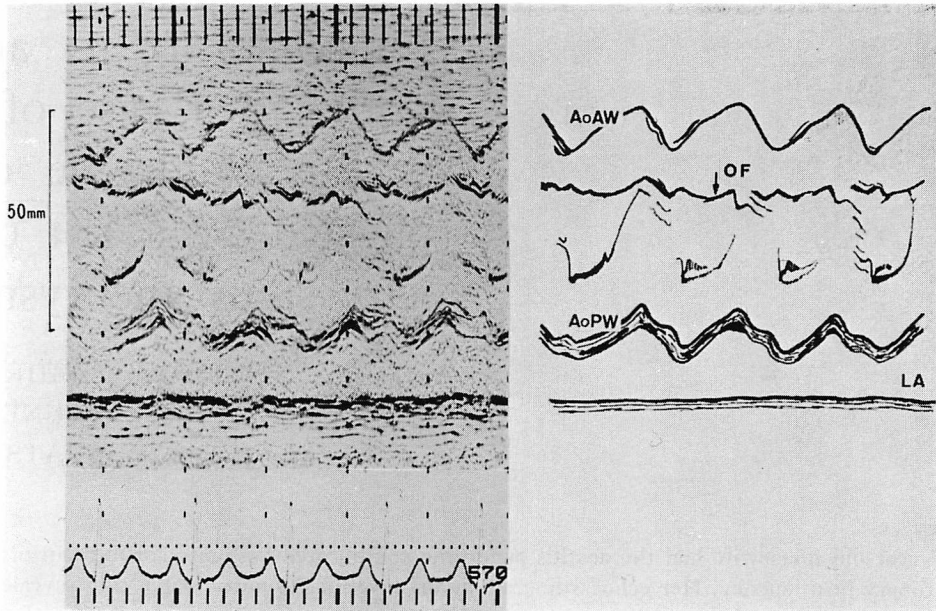


Fig. 1a. M-mode echocardiogram of the aortic root shows an oscillating flap and a markedly dilated aorta compressing the left atrium.

AoAW=anterior wall of the aorta; AoPW=posterior wall of the aorta; OF=oscillating flap; LA=left atrium.

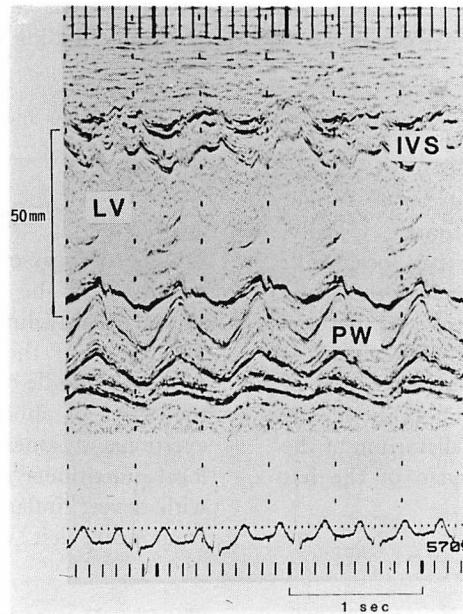
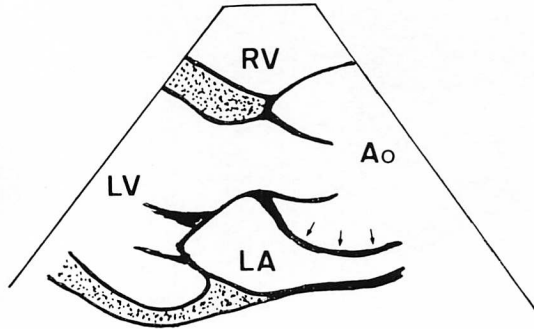
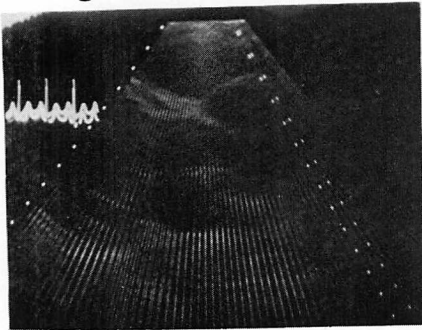


Fig. 1b. M-mode echocardiogram of the left ventricle.

Fine fluttering of the interventricular septum and the anterior mitral leaflet are noted. Contractility of the left ventricle is not impaired.

LV=left ventricle; IVS=interventricular septum; PW=posterior wall of the left ventricle.

Long Axis



Short Axis

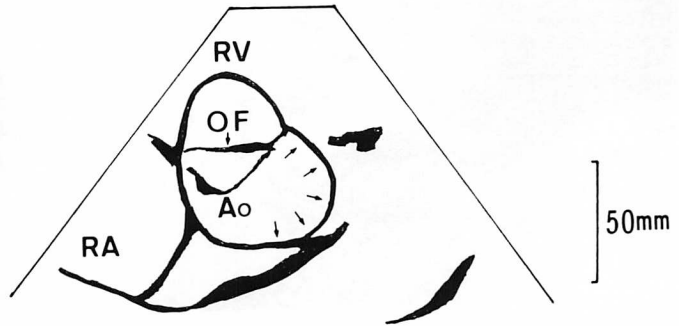
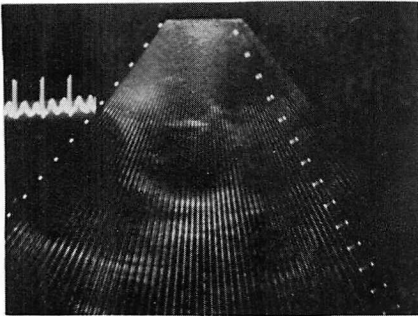


Fig. 2. Two-dimensional echocardiograms of the aortic root showing distorted, dilated aortic valve ring (arrows in the schema).

The left atrium is compressed by the enlarged aorta.

RV=right ventricle; LV=left ventricle; Ao=aorta; LA=left atrium; RA=right atrium; OF=oscillating flap.

ferred to Tokyo University Hospital on May 10, 1982 for further evaluation.

Physical examination on admission revealed a slightly thin, pale woman in mild respiratory distress. Blood pressure was 132/10 mmHg, with no difference between extremities. Her pulse was 100 beats per min. and regular; her temperature, 36.6°C. An aneurysmal pulsation of the left common carotid artery was observed. A grade 3/6 blowing aortic regurgitant murmur was audible over the third interspace at the left sternal border. No rales were audible in the lungs, and no vascular bruit was audible over the abdomen. The optic fundi were normal.

The laboratory data were normal except for

an erythrocyte sedimentation rate of 82 mm per hour and a C reactive protein of 4+. The serologic tests for syphilis were negative and no bacteria were cultured from the blood, urine, sputum and gastric secretions. A chest radiograph revealed marked cardiomegaly and moderate accentuation of the hilar markings. An electrocardiogram was normal.

An M-mode echocardiogram revealed marked enlargement of the aortic dimension (53 mm) and marked reduction of the left atrial dimension (17 mm). In the dilated aorta, an oscillating poorly mobile flap was observed. The right coronary cusp was not visualized, and the left and non-coronary cusps opened from, and closed

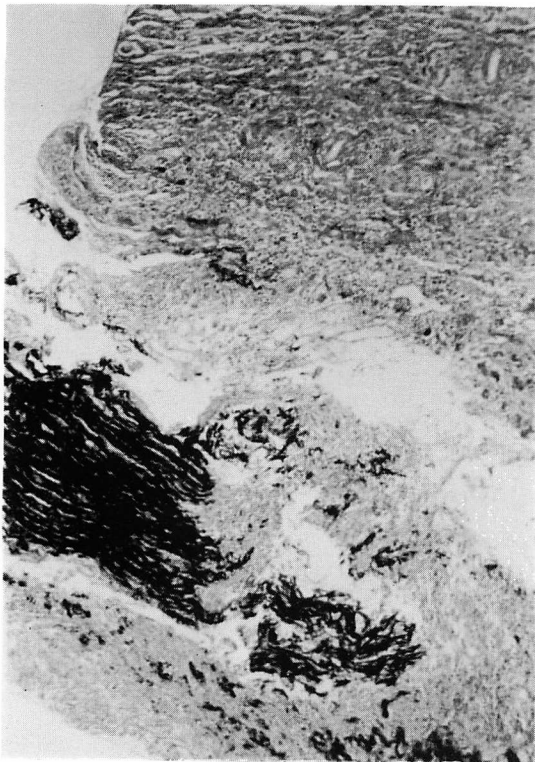


Fig. 3a. Photomicrograph of the wall of the ascending aorta which shows marked thickening of the adventitia (upper portion) and disruption of elastic lamellae with inflammatory cell infiltration in the media (Elastica, Van Gieson stain 40 \times , reduced to 57%).

toward the flap (**Fig. 1a**). The left ventricular dimension in end-diastole and in end-systole, and ejection fractions were 53 mm, 36 mm and 0.71, respectively. Fine fluttering of the inter-ventricular septum and the anterior leaflet of the mitral valve were clearly observed in diastole (**Fig. 1b**). A two-dimensional echocardiogram at the level of the aortic valve ring revealed a distorted enlarged left coronary sinus, which compressed the left atrium. From the anterior portion of the aortic valve ring to the center of the ascending aorta, a membranous structure with reduced mobility was observed, which was thought to be an oscillating flap (**Fig. 2**).

The oscillating membranous structure ob-

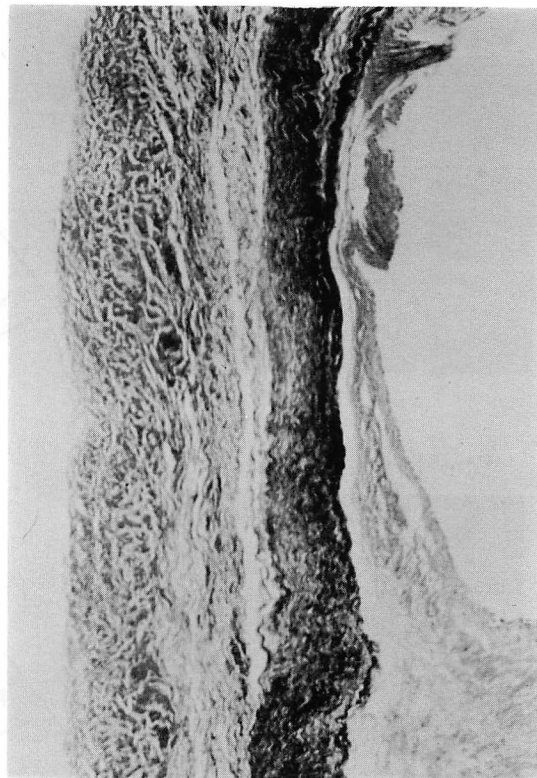


Fig. 3b. Photomicrograph of the right aortic leaflet with no remarkable change (Elastica, Van Gieson stain 40 \times , reduced to 57%).

served echocardiographically was suspected to be an intimal flap due to aortic dissection; therefore, aortitis syndrome complicated by a dissecting aneurysm of the ascending aorta and acute aortic regurgitation were diagnosed. Treatment with 30 mg prednisolone was begun, but three days after its initiation, she experienced severe recurrent substernal oppression with marked ST depressions in leads I, aVL, V₄, V₅ and V₆. Her rapidly worsening coronary insufficiency was considered closely related to the aneurysm of the ascending aorta; therefore, emergency surgery was performed. Angiography was not performed because of her allergy to iodine.

At surgery, there was no dissection in the ascending aorta, but there was a 4 cm diameter

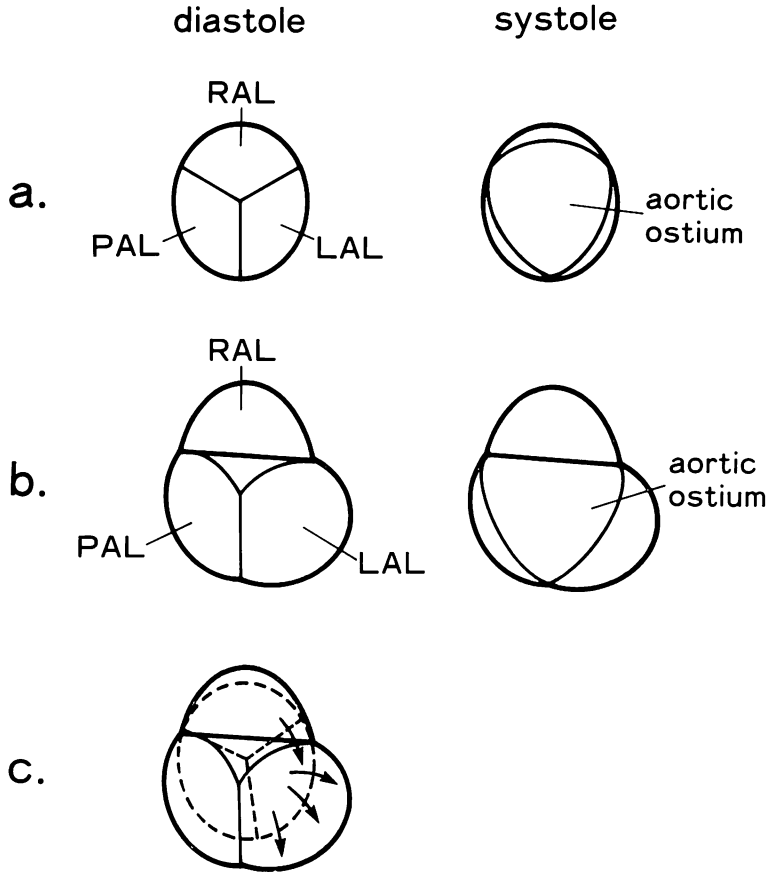


Fig. 4. Schematic illustrations of the aortic root in diastole and in systole.

Opening and closing movements of the right aortic leaflet are greatly restrained in the present case (b), while those of a normal case (a) are very mobile without limitation. Right aortic leaflet is stretched by distortion of aortic valve ring caused by distension of the left aortic sinus. Arrows in the overlapping view (c) show the direction of stretching.

RAL=right aortic leaflet; LAL=left aortic leaflet; PAL=posterior aortic leaflet.

aneurysm of the left aortic sinus. The wall of the ascending aorta appeared hyperemic and edematous, indicative of active inflammation. The orifices of the left and right coronary arteries were normal, but the main trunk of the left coronary artery was compressed by the left aortic sinus aneurysm. The leaflets of the aortic valve were neither inflamed nor sclerotic. The aortic valve was replaced with a Hall-Kaster's prosthetic valve, and the aneurysm was repaired by suturing.

Histologically, the ascending aorta had mark-

edly thickened adventitia with fibrous changes. The media consisted of fragmented elastic fibers and infiltrating inflammatory cells (**Fig. 3a**). The leaflets of the aortic valve were normal (**Fig. 3b**).

Her postoperative course was uneventful. Prednisolone therapy was reinstated and all abnormal laboratory data improved.

Discussion

There are numerous reports of the echocardiographic diagnosis of aortic root dissec-

tion²⁻⁴⁾, and a number of false positives were also reported^{5,6)} in the past. Recently, however, the capability of visualizing the intimal flap has greatly improved, and false positives have been markedly diminished by detailed observations using two-dimensional echocardiography^{7,8)}. Thus, an oscillating aortic flap is regarded characteristic of a dissecting aneurysm of the ascending aorta; once an oscillating membranous structure is observed in the ascending aorta, there is no alternative means of diagnosing an aortic dissection.

However, this case had an oscillating membranous structure without aortic dissection. Since no abnormal structure was found in the ascending aorta at surgery, and since the right aortic leaflet was normal, the echo source is considered to have been the right aortic leaflet itself. A possible explanation for the immobility of the right aortic leaflet is the stretching of the cusp secondary to the dilatation of the left aortic sinus (Fig. 4). Such a distorted aortic valve ring is theoretically possible with dilatation of the specific coronary sinus. Chisholm⁹⁾ advanced a theory of trigonoidation of the semilunar valves to explain the causes of functional systolic murmurs. Although his theory is not generally accepted, his concept can be applied to explain the observations in our case. Since aneurysm of the sinus of Valsalva, especially that of the left coronary sinus, is infrequently encountered¹⁰⁾, immobility of a specific aortic valve leaflet must be very rare.

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心エコー図上右冠尖の開閉運動の消失を認めた
左冠洞動脈瘤の1例

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大動脈炎症候群に合併した左冠洞動脈瘤の症例
において, 心エコー図上, 右冠尖の開閉運動の消
失が観察されたので報告する.

症例は37歳, 主婦. 大動脈炎症候群に急性大動脈弁閉鎖不全症と重篤な冠不全を併発したため入院した. 心エコー図では大動脈弁輪部において左冠洞の著明な拡張を示し, 大動脈内には可動性に乏しい, 細かい動きを示す膜様の構造物 (oscillating flap) を認めた. 容態が急速に悪化したため行なった緊急手術において, 大動脈内にはピンポン玉大の左冠洞動脈瘤を認めたが, 異常な構造物はなく, 大動脈弁も正常であった. 弁置換と瘤縫縮を施行後, prednisolone 30 mg/day の投与にて状態は著明に改善した. この oscillating flap とみたものは右冠尖そのものと考えられ, 右冠尖の開閉運動の消失は, 左冠洞動脈瘤による弁輪の変形により, 弁帆がテント状に張られたためと考えられた.

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