Production of Aortic Regurgitation by Unperforated Aneurysm of the Sinus of Valsalva

By Seymour B. London, M.D., and Rose E. London, M.D.

Recently we observed an elderly man with the classical features of aortic regurgitation and progressive left ventricular failure. Postmortem studies revealed the cause of the regurgitation to be an unperforated aneurysm of a sinus of Valsalva. Since aneurysm of the sinus of Valsalva is generally unrecognized until perforation occurs, the presence of aortic regurgitation, unexplained by other causes, may be a diagnostic clue to this potentially lethal lesion. Surgical repair of the unperforated aneurysm may prevent the complications of aortico-cardiac fistula, subacute bacterial endocarditis, and cardiac failure.

Report of Case

A 76-year-old retired pharmacist 5 years prior to his terminal illness developed progressive cardiac disability, marked by dyspnea, orthopnea, and peripheral edema. The onset was associated with a partial paralysis of the left hand and leg. For 2 years prior to his death, he suffered bouts of severe chest pain over the right anterior chest, with radiation through both shoulders into the hands. The only significant past history was of a "chest condition," discovered over 40 years ago, for which restricted activity was advised.

On physical examination a nodding motion of the head was observed with each heart beat. The heart was enlarged with the apical impulse in the sixth left interscostal space at the anterior axillary line. The sounds were of good quality and the aortic second sound was louder than the pulmonic second sound. A loud diastolic murmur was heard over the precordium and was loudest at the aortic area, where a short systolic murmur was also heard. The blood pressure was 210/60. The lungs were resonant but many rales were present at both bases. No abdominal organs were palpable. The left arm and leg were weak, and there was moderate pitting edema of the legs. Peripheral pulsations were symmetrical and of a "water hammer" collapsing type, with capillary pulsations of the lips and nail beds.

Electrocardiograms showed marked left ventricular hypertrophy (fig. 1). X-ray of the chest indicated left ventricular enlargement; the aorta was tortuous but not dilated (fig. 2). The hemogram, Kahn test, urine examination, and serum electrolytes were normal.

For the 4 months prior to his death the patient experienced a relentless worsening course, marked by severe left ventricular failure responding poorly to salt restriction, digitalization, and diuretics. There were frequent attacks of severe prolonged chest pain attributable to coronary insufficiency and one episode associated with electrocardiographic evidence of anteroventricular myocardial infarction. He died during an acute bout of severe chest pain, orthopnea, and peripheral vascular collapse.

Autopsy showed an unperforated aneurysmal dilatation of the right sinus of Valsalva, measuring 2 by 1½ cm., which was eccentrically oriented, extending posteriorly behind the commissure of the right coronary and noncoronary leaflets (fig. 3). The aneurysm protruded down to the right atrium, indenting the tricuspid valve ring but not perforating through the thin glistening endocardium. There was no evidence of syphilis of the aorta. The aortic and mitral valves showed slightly nodular thickening commensurate with age but the chordae tendineae were thin. The endocardium of the left ventricle just beneath the valve of the right coronary sinus demonstrated thickening with formation of "diastolic pockets" (fig. 4) at several levels, pathognomonic of aortic valvular regurgitation. Left ventricular hypertrophy was present. There was arteriosclerotic narrowing in all coronary vessels with occlusion of the left anterior descending artery and old and recent anterior myocardial infarction. There were chronic hyperemic changes in the lungs and abdominal organs compatible with congestive heart failure.

Discussion

Congenital aneurysm of sinus of Valsalva is a developmental defect due to the failure of the aorta to fuse firmly with the membranous ventricular septum at the annulus.2,3 The high intraluminal aortic pressure causes thinning and protrusion of the weakened structure. Since the location of the aortic annulus is intracardiac, aneurysms of the different sinuses bear unique relationships,4,5 the
right coronary sinus being related to the right ventricle, and the noncoronary sinus to the right atrium. The left coronary sinus, which generally is not involved in congenital aneurysm, is encased in the left ventricle.

Perforated aneurysm of the sinus of Valsalva has received considerable attention recently because of the dramatic nature of the
Endothelial pockets formed by the regurgitant jet are indicated by the arrow. The normal commissure between the right coronary sinus and posterior (noncoronary sinus) and the mouth of the aneurysm is seen above the arrow.

illness, the outstanding physical signs, the confirmation by contrast aortography, and the recent feasibility of surgical repair. On the other hand, unperforated aneurysm of the sinus of Valsalva has received scanty mention because of the lack of characteristic features. Outstanding in our case was the classical aortic regurgitation, manifested by clinical, electrocardiographic, and x-ray findings and confirmed by postmortem studies. While there has been occasional association of aortic regurgitation with unruptured aneurysm of the sinus of Valsalva of the congenital type, the significance and the mechanism of production of aortic regurgitation in this situation have not been elucidated. Our concept based on the anatomic findings in this case, is that the failure of the aorta to fuse to the annulus fibrosis not only provides a weakened area, so that aneurysms can be produced, but that the annulus itself is not supported by the suspending action that the aorta normally provides. As the high intraluminal pulse pressure is exerted against the weakened wall, aneurysmal formation occurs with protrusion into the adjacent structure. Since the annulus is unsupported, it eventually is involved in this protrusion with the result that buckling of the rim occurs. This interferes with appo-

Figure 4

Endothelial pockets formed by the regurgitant jet are indicated by the arrow. The normal commissure between the right coronary sinus and posterior (noncoronary sinus) and the mouth of the aneurysm is seen above the arrow.

Figure 5

Relationship of the unperforated aneurysm of the sinus of Valsalva to the annulus of the aortic ring. A. Normal relationships of sinus of Valsalva to adjacent structures at the annulus of the aorta. B. Congenital weakness of sinus of Valsalva area is due to failure of fusion of aorta to annulus. Arrow indicates direction of pressure against the weakened wall. C. Protrusion of aneurysm (S.V.A.) into right atrium and base of tricuspid valve; displacement of annulus laterally and failure of apposition of valves causes regurgitant jet; production of endothelial diastolic pockets (D.P.) on the ventricular septum results. A, aorta; Ann, annulus; A.S.V., aortic semilunar valves; l.v., left ventricle; r.a., right atrial wall; r.v., right ventricular wall of septum; S., septum; S.V., sinus of Valsalva; T.V., tricuspid valve.
sition of the valves and leads to aortic regurgitation (fig. 5).

The clearly audible aortic second sound despite clinical aortic regurgitation is consistent with the normal, pliable valves found at autopsy and serves as a distinguishing feature from rheumatic valvulitis and calcific aortic disease, in which the valve structure is fixed and deformed, with loss of the aortic second sound. Syphilis of the aorta results in separation of the commissure with resultant aortic regurgitation. The second aortic sound eventually disappears when the valves, which are otherwise normal, are unable to achieve any approximation.

Since aortography clearly identifies the presence and magnitude of the aneurysm of the sinus of Valsalva, it serves ante mortem as the single diagnostic criterion in distinguishing the etiology of aortic regurgitation. Clinical suspicion, however, should be aroused in all cases of aortic regurgitation in which the etiology is not obvious.

Conclusions

A case report is presented of an elderly man with severe aortic regurgitation and cardiac failure in which the etiology, revealed by postmortem studies, was due to the effects of unperforated aneurysm of the sinus of Valsalva.

The mechanism of production of aortic regurgitation due to aneurysm of the sinus of Valsalva is discussed.

Unperforated aneurysm of the sinus of Valsalva should be considered as a possible cause of aortic regurgitation and cardiac failure when the etiology has not been established. The usefulness of contrast aortography is suggested in those cases in which surgical repair might be desirable.

References

Production of Aortic Regurgitation by Unperforated Aneurysm of the Sinus of Valsalva
SEYMOUR B. LONDON and ROSE E. LONDON

Circulation. 1961;24:1403-1406
doi: 10.1161/01.CIR.24.6.1403

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1961 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/24/6/1403.citation

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org/subscriptions/