Rupture of Aneurysm of Aortic Sinus of Valsalva Associated with Acute Bacterial Endocarditis

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While aneurysms of the aortic sinus of Valsalva are uncommon, the importance of the correct diagnosis has been emphasized by recent reports of successful surgical correction in patients with rupture of such an aneurysm.1, 2

A patient with a ruptured sinus of Valsalva associated with acute bacterial endocarditis recently seen at the Beth Israel Hospital, Boston, illustrates the problem of differential diagnosis in such instances.

J. H. (Beth Israel Hospital No. M84990), a 68-year-old white man entered the hospital on March 20, 1957, because of fever, chills, and dysuria for 3 weeks. Urine sediment was reported to have contained numerous white and red blood cells at the onset of the illness and sulfasoxazole (Gantrisin) and tetracycline therapy was instituted. During the next 2 weeks, the fever and dysuria subsided and therapy was discontinued. During the week prior to admission, however, the patient noted increased weakness, occasional chills, fever up to 102°F, and extreme dyspnea on slight exertion. A platelet count on the day of admission was reported as 18,000 per mm.³ and the hemoglobin was 9 Gm. per cent.

From 1941 to 1951 the patient received periodic phlebotomies with relief of mild symptoms of polycythemia vera. Between 1952 and 1954 increasing hepatosplenomegaly was noted, the hemoglobin and white blood cell count fell, and occasional metamyelocytes and myelocytes appeared in the peripheral blood. The diagnosis of myeloid metaplasia was tentatively made.

By August 1955, the platelet count was low, there were transient bleeding episodes and the hemoglobin was 7.6 Gm. per cent. Prednisone therapy was instituted. A chest film in December 1956 was unremarkable. The last known examination of the heart 4 weeks prior to admission indicated no change. A grade-II harsh blowing systolic murmur over the precordium, which had been present since 1941, was heard.

The patient on admission was acutely ill and in moderate respiratory distress. The blood pressure was 110/40 in both arms, the pulse was 90 and regular. Rectal temperature was 101.6°F.

There was cyanosis of the lips, and there were numerous ecchymoses over the abdomen and petechiae in the mouth. Crackling rales were heard at the right base. A strong thrill was felt maximally at the lower left sternal border in both systole and diastole. Grade-IV systolic and diastolic murmurs were heard over the entire precordium, loudest at the left sternal border in the fourth intercostal space. These murmurs radiated mainly to the right and downward. At times, the murmurs were continuous with a machine-like quality; at other times, there was a short silent period between the systolic and diastolic murmurs. First and second heart sounds were inaudible at the point of maximal intensity of the murmur. There was hepatomegaly, and the spleen was palpable 8 fingerbreadths below the left costal margin. Peripheral pulses were of the Corrigan type.

Repeated urinalyses showed intermittent hematuria and albuminuria. Hemoglobin on admission was 11 Gm. per cent but 4 units of packed cells were required over a 20-day period to maintain it there. White blood cell count was 5,200 with 80 per cent polymorphonuclear leukocytes, 5 per cent band forms, 11 per cent lymphocytes, 2 per cent monocytes, 1 per cent myelocytes, and 1 per cent metamyelocytes. Platelet count was 60,000 and reticulocyte count 3 per cent. Stool guaiac

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on admission was negative; subsequently it was consistently 2+. The nonprotein nitrogen was 57 mg. per cent. Liver function tests showed parenchymal damage. Three blood cultures taken on admission were positive for *Staphylococcus aureus*, coagulase positive; subsequent blood cultures were negative. The venous pressure was 75 mm. of water.

Chest x-ray was consistent with pulmonary edema and slight enlargement of the heart. Electrocardiograms showed right bundle-branch block, first degree atrioventricular block, and left ventricular hypertrophy.

A regimen was started of bed rest, aqueous penicillin, 1,200,000 units intramuscularly every 4 hours, and streptomycin 0.5 Gm. intramuscularly twice per day, prednisone 12.5 mg. twice per day, and an ulcer diet. Lanatoside-C, 1.2 mg., was given over a 4-hour period followed by maintenance doses of digitoxin. One unit of packed red cells was also given.

By noon of the next day, the patient was afebrile and complained only of anorexia. Chest x-ray showed marked pulmonary congestion, which responded to a low-sodium diet and frequent injections of mercurhydalin. Positive blood culture reports of *Staph. aureus*, coagulase positive, supported the initial impression of bacterial endocarditis; penicillin and streptomycin were continued, since the organism was sensitive to these drugs. The most likely site of the rupture of the sinus of Valsalva if present was considered to be into the right ventricle or pulmonary artery rather than the right atrium or superior vena cava, since there was no pulsation of the neck veins.

During the first week, the patient continued to improve symptomatically. The blood pressures varied between 130/60 and 120/0. The murmurs changed only in minor degrees from day to day. On the ninth hospital day, a splinter hemorrhage was seen under the right thumbnail, and the aortic murmurs were louder. On the tenth hospital day, pulsation of the pulmonary artery was not seen during chest fluoroscopy. This was considered to be evidence against an aorta-to-pulmonary artery fistula.

On the seventeenth hospital day, the patient suddenly developed dyspnea at rest and marked orthopnea, and physical signs consistent with pleural effusion were noted at both lung bases. Digitoxin dosage was increased and ammonium chloride and actazolamide (Diamox) were added to the diuretic regimen. On the twenty-first hospital day, 1,200 ml. of straw-colored fluid were removed from the right pleural space and on the following day 500 ml. were removed from the left pleural space. Following these procedures, there was some relief of dyspnea; however, the fluid reaccumulated in each pleural cavity and required removal on the twenty-sixth hos-
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hospital day. At 3:30 a.m., on the twenty-seventh hospital day, the patient died suddenly, having remained afebrile since the first hospital day.

Pathologic Examination

At autopsy, the heart was slightly enlarged weighing 410 Gm. There was an aneurysm of the right coronary sinus of Valsalva, 3.5 cm. deep and 2.5 cm. in greatest diameter (figs. 1 and 2). Most of the lining of the aneurysm, including the right coronary cusp, was yellowish and ragged, but a portion was smooth.

There were 2 communications between the aneurysmal sac and the right ventricle, each about 0.4 cm. in diameter. One of these, within the ragged zone, had an irregular edge (fig. 3c); the second, within the smooth area, had a regular smooth edge (fig. 2). There was also a communication with the left ventricle through the right coronary cusp, measuring 0.5 cm. in diameter (fig. 3b). The commissure between the right and noncoronary cusps was excavated, yellowish, and ragged. On section, this alteration was seen to extend upward in the subepicardial tissue and into the right atrium, where it presented as a nodule immediately above the posterior commissure of the tricuspid valve.

Microscopic examination revealed necrotizing inflammation in the aneurysm, the right coronary cusp, and the abnormal commissure, with extension into the right atrium and the adventitia at the root of the aorta. These areas contained numerous inflammatory cells and many colonies of cocci, embedded in a fibrin mesh. The grossly smooth part of the aneurysm showed a fibrous wall with little active inflammation. The entire picture was consistent with an acute bacterial endocarditis in an aneurysm of the sinus of Valsalva with several perforations. It could not be determined whether the aneurysm was congenital or was an erosive aneurysm caused by the acute bacterial endocarditis.

Other pertinent findings included an enlarged liver and spleen (2,400 and 1,120 Gm., respectively) which showed myeloid metaplasia. The bone marrow was hyperplastic. An occasional focus of hematopoiesis was noted in the right kidney. The lungs were acutely congested and edematous. There was obstructive benign prostatic hypertrophy, with bilateral nephrolithiasis and cystolithiasis. There was stenosing arteriosclerosis of the ostium of the right renal artery associated with an atrophic right kidney (70 Gm.) and compensatory hypertrophy of the left kidney (270 Gm.). Petechiae were present in the left kidney, bladder, bowel, and skin of the back, and there were large ecchymoses on the extremities.

Cultures of the aortic valve lesion grew out coliform bacilli, Bacillus subtilis, and coagulase negative Staph. albus; these were considered contaminants. Culture of heart blood taken at autopsy was sterile.

Discussion

The sinuses of Valsalva are 3 small dilatations in the wall of the aorta immediately above the aortic valves. They are in close relation to all of the cardiac chambers, particularly the right atrium and right ventricle.

Aneurysms of a sinus of Valsalva are either congenital or acquired. Congenital aneurysms are thought to be due to developmental defects in either the aortopulmonary segment or the elastic tissue of the aortic sinuses. Jones and Langley list the following criteria for differentiating the 2 types. In the congenital type, the aneurysm (1) is confined to the
right coronary sinus and adjacent two thirds of the noncoronary sinus; (2) is usually small; (3) commonly ruptures into the right ventricle or right atrium; (4) remains intra-cardiac; (5) is usually associated with other developmental defects. In the acquired type, the aneurysm (1) may arise from any sinus; (2) tends to extend upward; (3) is often extracardiac; (4) cardio-aortic fistulae are rare; (5) congenital cardiac defects are rare; (6) it is always associated with acquired heart disease, usually syphilis or bacterial endocarditis.

Diagnosis of nonperforated aortic sinus aneurysm is difficult. Falhot and Thomsen in 1953 described a case of unruptured congenital right aortic sinus aneurysm diagnosed by retrograde aortography. Steinberg and Finby made the diagnosis of unruptured aortic sinus aneurysm in 16 cases by angiocardiography. Six were thought to be congenital: 3 were patients with arachnodactyly and 3 were associated with coarctation of the aorta. In 9 of the remaining 10 cases, the aneurysm was due to syphilis. Steinberg suggested that the only clue to diagnosis on physical examination is a grade-III to IV systolic murmur over the heart, especially to the left of the sternum.

Perforation of an aneurysm is ordinarily accompanied by sudden intense dyspnea, sometimes associated with chest pain, especially on minimal exertion; increase in pulse pressure; sudden appearance of continuous or “to-and-fro” murmurs usually at the fourth intercostal space to the left of the sternum.

Diagnostic methods which have been used to establish the diagnosis include cardiac catheterization and surgical exploration.

In our case, the diagnosis of ruptured aortic sinus of Valsalva into the right ventricle was strongly suggested by the history of sudden onset of extreme dyspnea on minimal exertion, the wide pulse pressure suggesting a lesion at or above the aortic valve, the finding at the lower left sternal border of a machinery murmur which radiated mainly to the right and downward, and the known association of acute bacterial endocarditis with this condition. Absence of facial edema and other features of a superior mediastinal syndrome was evidence against a fistula between the aorta and superior vena cava. Absence of pulsation in the pulmonary artery was evidence against rupture into that structure. Absent cervical or peripheral venous pulsations were evidence against rupture into the right atrium.

Other conditions considered in the differential diagnosis were patent ductus arteriosus and aortic stenosis and regurgitation secondary to bacterial endocarditis. These diagnoses were considered unlikely on the basis of the character, location, and radiation of the murmurs.

It cannot be ascertained whether the aneurysm was congenital or acquired. In favor of a congenital origin are presence of a systolic murmur over the precordium for at least 15 years, presence of the aneurysm in the right aortic sinus, and rupture of the aneurysm into the right ventricle.

As is illustrated by our patient, the course of the disease is almost always rapidly deteriorating with most patients dying of intractable congestive heart failure within 6 months.

In view of a recent case report of successful diagnosis and surgical treatment of rupture of an aortic sinus aneurysm, surgical correction was briefly considered in the case presented here, but the idea was abandoned because of the patient’s poor general condition.

Summary

A case of ruptured aortic sinus of Valsalva aneurysm secondary to acute bacterial endocarditis is presented. In diagnosis of this entity emphasis is placed on the character, location, and radiation of the murmur. Importance of correct diagnosis is mentioned, since in some cases surgical correction may be carried out.

Summario in Interlingua

Es presentate un caso de ruptura de un aneurysma del aortic sinus de Valsalva,
secundari a acute endocarditis bacterial. In
le diagnose de iste entitate, le accento debe
esser ponite super le studio del character,
del location, e del radiation del murmure.
Le importantia del obtention de un diagnose
correcte es augmentate per le facto que in
certe casos un correction chirurgic pote esser
effectuate.

REFERENCES

Quotation from “ART AND LIFE”
LEONARDO DA VINCI
Italian painter, sculptor, and scientist; 1452-1519
The supreme misfortune is when theory outstrips performance.
Poor is the pupil who does not surpass his master.
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