Congenital Saccular Aneurysms of the Aortic Ring

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IN SPITE OF, and also because of the great progress that has been made in the last decade in both clinical and basic research in the field of cardiovascular disease and in its application to the diagnosis and care of patients with heart disease, exact autopsy findings, in the event of death, have never been more important or more meaningful. With the rapid introduction of new technics, new tools, and new drugs aimed at more accurate diagnoses, a better evaluation of the patient, and more effective treatment, it has become increasingly clear that the pathologic anatomy of the heart is no longer the sole concern of the pathologist. This is encouraging to the pathologist who is only too keenly aware that observations made at the autopsy table, like observations made in other fields of medical research, are of little value to the patient until they can be translated into usefulness by the practicing physician. Indeed, a rebirth of interest by the younger clinicians in the pathology of the heart is leading to growing awareness of the frequency, complexity, and variability of the structural manifestations of the many diseases and deformities of this organ. Furthermore, we are not only experiencing a renewed curiosity and interest in the more familiar and common patterns of disease, but also in the most uncommon. In brief, one is becoming more and more conscious of the uniqueness of the cardiovascular system of each individual and, as one would expect, no one has shown a greater interest in detailed postmortem findings than the younger well-trained cardiac diagnosticians and surgeons.

It is freely admitted that most congenital malformations of the heart have at some time been recorded in the world’s literature. In actual practice, despite all of the modern diagnostic armamentarium at one’s disposal, one does not diagnose beyond one’s realm of thought, so that the uncommon diseases and others that are not so uncommon are sometimes forgotten in one’s differential diagnosis. For this reason, it seems worthwhile to continue to report some of the less common lesions that are found from time to time at the autopsy table: particularly those that would lend themselves to specific forms of modern therapy or, if encountered unexpectedly in the course of cardiac surgery, might be better understood in the interest of the patient.

An unusual opportunity to observe three examples of congenital saccular aneurysm of varying size, in the region of the aortic ring during the past 3 years, carries with it the responsibility of bringing this type of lesion to the attention of others. Two of these were seen in adults, the third in a child 2 years of age who died suddenly shortly after admission to the hospital. This last case, which is of particular interest anatomically, is described in detail.

Report of a Case

A 2-year old white boy died suddenly 2 hours after admission for acute respiratory distress. The past history was essentially negative. There had been symptoms and signs of a low-grade respiratory infection during the month prior to admission and 10 days before admission the temperature was 104. During the week preceding admission the child received an antibiotic, and two days prior to admission, the head was noted to nod with each heart beat. Upon admission to another hospital, the child was found to be in shock and was digitalized and transferred to the Boston Floating Hospital within an hour. The temperature was 98, the pulse was 140, and respirations varied from 40 to 60. The child was pale, epyanotic, semicomatose, and moribund. The heart rhythm was irregular and the sounds were of poor quality, but no murmurs were heard. The liver was five fingerbreadths below the right costal margin and the white cell count was 49,000 per mm.3 with 74 per cent polymorphonuclear cells. A throat culture was reported as positive for *Aerobacter aerogenes*. A blood culture was sterile. Death occurred suddenly, immediately following a seizure.

Postmortem Examination. A complete autopsy
ANEURYSMS OF AORTIC RING

(A-59-149) was performed 4 hours after death. Externally there were no petechiae, splinter hemorrhages or signs of wasting. The pericardium was distended and contained twice the normal amount of clear serous fluid. The heart was large and markedly dilated. There were no pericardial adhesions, and the surfaces were everywhere smooth. The heart weighed 92 Gm. (normal 56). The structure of the tricuspid, pulmonary, and mitral valves was normal. The foramen ovale was closed, and both the interatrial and interventricular septa were intact. The wall of the right ventricle was moderately thickened, whereas that of the left ventricle was not only stretched but very hypertrophied, measuring 8 mm. in width.

Just at the raphe formed by the apposed edges of the right coronary and noncoronary cusps bordering on the position normally occupied by the membranous interventricular septum, there was a large triangular opening about 1 cm. in width, which led directly into an aneurysmal sac after the fashion of a diverticulum (fig. 1). This saccular aneurysm extended through and under the aortic valve ring and passed upward to the anterior surface of the heart in the area bounded by the pulmonary trunk anteriorly, the aorta posteriorly, and the right atrial appendage laterally. Its diameter tapered somewhat as it extended for a few millimeters to the right along the atrioventricular groove. The wall of this diverticulum-like lesion varied from 2 to 3 mm. in thickness, and its inner surface was lined by smooth intact endocardium. The right coronary artery arose in its usual position and ran below and behind the diverticulum proximally and then in front of and below the diverticulum in its course along the atrioventricular groove. This sac communicated with no other chambers or vessels of the heart. The edge of the mouth of this sac was coated by numerous tiny gray granular, friable vegetations, which extended over to the surfaces of the adjacent cusps. These vegetations seriously deformed the right cusp and could be traced to a large ragged perforation in the noncoronary cusp. The left coronary cusp was uninvolved. Smears taken directly from these vegetations revealed a mixed infection showing gram-positive cocci and occasional gram-negative rods. Blood cultures taken at the time of the autopsy remained sterile.

In summary, the autopsy revealed a congenital saccular aneurysm of the aortic ring with dilatation, deformity, and insufficiency of the aortic valve. This was complicated by an active bacterial infection that extended from the lips of the aneurysm to the adjacent cusps, leading to perforation of one. The dilatation and hypertrophy of the ventricles, particularly the left, were regarded as being symptomatic and secondary to the aortic regurgitation.

Discussion

Developmental saccular aneurysms of the aortic ring include those of the aortic sinus, those at the junction of contiguous cusps of the aortic valve, such as the case under consideration, and those that involve the membranous portion of the interventricular septum. Some are present at birth while others may develop later in life on the basis of a congenital defect as in the case of berry aneurysms of the cerebral arteries. Even those present at birth probably develop on the basis of a pre-existing structural defect. Acquired saccular mycotic aneurysms in this area, arising as a complication of bacterial endocarditis, belong in another category and are not pertinent to this paper.

According to Lev and Saphir,1 who have made a most complete review of this subject, Laennec was probably the first to describe a case of an aneurysm involving the membranous septum. Mall2,3 in a discussion of this type of aneurysm, pointed out that it could be explained by a faulty growth of the conus ridges creating a horizontal septum and a somewhat tilted aorta. Lev and Saphir found other anomalies in the parietal muscle bundle orientation, which they thought would lend support to this developmental theory. All authors consider this type of anomaly to be very infrequent, and in an autopsy series reported by Sternberg,4 only two cases involving the septum were found in 16,000 postmortem examinations.

The orifices of these aneurysms usually appear just below the right coronary cusp or just at or beneath the raphe formed by the margins of the right coronary and noncoronary cusps, or in the depth of one of the sinuses of the aorta. The diameter varies but is usually about 1 cm. In the present case and in that of Leckert and Sternberg,5 the aneurysm bulges upward to the anterior surface of the base of the heart. In other reports they tend to bulge into the right side of the heart,

Circulation, Volume XXVI, August 1962
Figure 1

The aortic valve showing a large, gaping triangular cleft, separating the right from the posterior noncoronary cusp, which lies at the level of the aortic ring and involves the adjacent membranous portion of the interventricular septum. The walls of this cleft form the mouth of the saccular aneurysm. Its edges are covered with vegetation that extends to the surfaces of the adjacent cusps.

usually at or just above or below the septal leaflet of the tricuspid valve. Aneurysms arising in the aortic sinus and at the junction of contiguous aortic leaflets can best be explained on the basis of a simple defect in the structure of these walls. In a case reported by Edwards and Burchell, a saecular aneurysm arising deep in the posterior aortic sinus protruded into the right atrium just above the septal leaflet. This ruptured, leading to sudden but prolonged and intractable cardiac failure over a period of 22 months.

Both cases of aneurysms of the membranous septum reported by Lev and Saphir, and several others they reviewed, were found in Mongolian idiots. This association has only been reported once since then. Murmurs may be heard but not commonly. Conduction abnormalities and signs of heart block were reported by Clark and White. Castoldi, in reporting an aneurysm of the septum, found an interruption in both bundle branches with borderline electrocardiographic changes, but he went on to point out that coexisting coronary disease may have played a contributing role in producing this change. In one of the cases reported by Rogers a variety of arrhythmias was reported but no histologic studies were mentioned.

The two more common complications of this lesion are bacterial endocarditis and perforation, but the actual cause of death in most cases that have come to autopsy has not been directly related to this anomaly. Associated cardiac anomalies have been reported in isolated cases by others including atrial septal defects and subaortic stenosis. In one case reported by Sternberg, an 8-year-old boy had cystic disease of the pancreas. A small septal aneurysm was simply an incidental finding.

A definitive clinical diagnosis of saecular aneurysm in this area is most unusual. Perhaps the first step toward such a diagnosis would be a recognition of its possible exist-
ence, and then, in addition to the common diagnostic aids, an angiogram might be most helpful.

Summary
Congenital sacceular aneurysms arising in the area of the aortic ring are not common. They may be located deep in the aortic sinus, at the commissure of two adjacent cusps, or in the area involving the membranous portion of the interventricular septum. They may be found at any age. This report concerns a child 2 years of age with a relatively large sacceular aneurysm of the aortic ring just at the site of the commissure between the right and noncoronary cusps of the aortic valve. Terminally, this anomaly was complicated by infection leading to vegetative and ulcerative endocarditis. Death was sudden and was apparently directly related to the complications of the deformity.

References
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Circulation. 1962;26:288-291
doi: 10.1161/01.CIR.26.2.288

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1962 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/26/2/288

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