Congenital Aneurysms of the Coronary Arteries with Report of a Case

By Ira Gore, M.D., John Smith, M.D., and Robert Clancy, M.D.

A case is presented of congenital aneurysm of a coronary artery in a patient with angina pectoris and myocardial infarction. Etiologies other than coronary atherosclerosis should be considered in young patients with clinical coronary disease; they include syphilitic ostial stenosis, coronary embolization, pulmonary origin of a coronary artery, and necrotizing arteritis in addition to aneurysm of a coronary artery. In this instance the aneurysm caused a shadow that was seen by x-ray but was not identified during life.

Ischemic heart disease, currently one of the leading causes of disability and death, is largely a manifestation or complication of coronary atherosclerosis. This is such a dominant relationship that other causes of coronary insufficiency are apt to be overlooked. Awareness of the clinical background, however, allows the clinician to suspect less common conditions such as syphilitic ostial stenosis, coronary embolization, pulmonary origin of a coronary artery, and necrotizing arteritis. Aneurysm of one or more coronary arteries, an infrequent condition as yet unrecognized clinically, is also likely to result in ischemic myocardial damage. Since technical progress may yet permit surgical remedy of some of these conditions, physicians should be alert to the possibility of their occurrence.

We wish to report the case of a young man who died of ischemic heart disease secondary to unrecognized aneurysms of the coronary arteries.

Case Report

J. S., a 26-year-old, white, male college student, was admitted to the Veterans Administration Hospital, West Roxbury, on June 21, 1957. For the preceding 10 days he had experienced increasing exertional dyspnea, orthopnea, and paroxysmal nocturnal dyspnea.

His difficulties began about 2 years earlier when, after running, he experienced a syncopal attack. In the ensuing months there were 3 or more nonepileptic syncopal episodes, each related to exertion and some associated with oppressive substernal pain that radiated to the inner aspect of the left arm. Exertional angina also occurred without syncope, and there was a progressive decline in exercise tolerance. In October 1956 he was hospitalized at the Memorial Hospital in Worcester following a syncopal episode. Electrocardiography showed evidence of a posterior myocardial infarct. Radiologic studies with a Potter-Bucky diaphragm demonstrated an irregular ovoid mass (3 by 1.5 cm.), calcified like an egg shell, adjacent to the left upper border of a normal-sized cardiac shadow (fig. 1). Fluoroscopic examination revealed that the mass pulsed synchronously with the heart. Though not typical, it was considered to be a pericardial calcification. Despite the relative youth of the patient, the other diagnoses were arteriosclerotic heart disease with angina pectoris and myocardial infarction.

At the age of 13 the patient had been hospitalized with a severe throat infection that responded to penicillin and salicylates. Streptococcus viridans was cultured from the throat. By x-ray, the heart was globular and enlarged, especially in the region of the right ventricle. Recovery was complete, however, and he was able to serve in the U. S. Navy from 1951 to 1955 without illness.

Physical examination upon admission demonstrated enlargement of the heart to the left, a protodiastolic gallop rhythm, and occasional extrasystoles. The blood pressure was 100/75 mm. Hg and the pulse rate was 88 per minute. Laboratory findings on blood, urine, and spinal fluid were unremarkable. The serologic test for syphilis (VDRL) was negative; serum cholesterol measured 153 mg. per cent of which 104 mg. per cent were esters; phospholipids were 192 mg. per cent. Serial electrocardiograms demonstrated a stable anteroapical myocardial infarct. Radiologic studies substantiated the impression of left ventricular enlargement and demonstrated pulmonary congestion initially, but not subsequently. Treatment consisted of digitalization, restriction of salt and fluids, and the administration of mercu-
Fig. 1. Lateral x-ray (Potter-Bucky) of the chest. Note the shell-like opacity delineating a circular structure at the base of the heart in the angle between the anterior cardiac border and the conus. The detail of this area, slightly magnified, is shown in the insert. This structure corresponds with an aneurysm of the circumflex branch of the left coronary artery depicted in figures 2 and 3.

On the assumption that the basic pathologic lesion was coronary atherosclerosis, despite the normal serum cholesterol values, he was placed upon a diet rich in unsaturated fatty acids. Nonetheless there was progressive deterioration, anginal attacks became more frequent, congestive failure reappeared and became increasingly difficult to control, and in November 1957, following a 4-day period of severe chest pain, he died.

At autopsy there was moderate left ventricular dilatation and myocardial hypertrophy, the heart weighed 490 Gm. A transmural scar (4 by 3.5 by 0.4 cm.) replaced a portion of the posterior left ventricular wall. The remainder of the posterior wall presented foci of muscle loss and subendocardial fibrosis which were pale gray at the base and red-gray and vascular toward the apex. The interventricular septum was thinned where its an-
CONGENITAL ANEURYSMS OF THE CORONARY ARTERIES

**FIG. 2.** Top. Dissected coronary arterial tree and aortic root viewed from above, showing diffuse aneurysmal dilatation of the right coronary and of the 2 branches of the left coronary artery. **Bottom.** Serial transections of the coronary arterial tree. The lowermost vessel is the right coronary artery; above it lie respectively the anterior descending and circumflex rami of the left coronary artery. The aneurysms are filled with thrombi of varied age and organization.

terior two thirds was largely replaced by vascularized scar tissue. Except for diffuse endocardial thickening the anterior wall of the left ventricle was unremarkable. The valves were competent with structurally intact, undeformed, pliable leaflets.

There were large fusiform aneurysms of the 3 major coronary rami (figs. 2 and 3). The circumflex and the anterior descending arteries arose from normal-sized adjoining ostia in the left aortic sinus and expanded immediately. The aneurysm of the anterior descending branch extended distally for 3.2 cm. and measured 1.5 cm. in its greatest diameter. Its wall contained egg-shell-like deposits of calcium. Serial transection after decalcification revealed a large lumen, 1.3 cm. across, completely filled with concentrically laminated, mottled gray-white and dark red, friable thrombus. Distally the aneurysm tapered gently into an 0.4-cm. artery with thin pliable walls devoid of intimal disease. The aneurysm of the circumflex artery measured 4 by 1.2 cm.; there was a small eccentric residual lumen, but in all other respects it was identical with the anterior descending ramus. The right coronary artery arose from a normal-sized
Fig. 3. Low-power photomicrograph of the left circumflex coronary aneurysm in situ in the left-atrioventricular sulcus. A transsection of the mitral valve at the left distinguishes the endocardial aspect. Except for a relatively small lumen containing fragments of fresh thrombus, the aneurysm is filled with degenerating blood clot. The space at the upper left of the aneurysm is an artefact. At the lower right, partially encircling the persistent channel, the thrombus is organized.

Aortic ostium. There was a small accessory right coronary ostium which fed a short vessel to the right ventricle. After a course of 0.8 cm. the main ramus expanded into a fusiform aneurysm 5.1 cm. long and 1.2 cm. in greatest diameter. Distally it tapered into a channel 0.4 cm. wide which exhibited focal thickening by firm gray-white tissue and severe luminal narrowing 3.2 cm. beyond the aneurysm. This aneurysm, like that of the descending ramus of the left coronary artery, was totally occluded by concentrically laminated, gray-tan to red thrombus.

The aorta was smooth with only a moderate quantity of linear flat yellow intimal deposits in its abdominal portion.

The lungs and viscera were moderately congested.

Microscopically, the left ventricular myocardium presented extensive areas of subendocardial and transmural scarring; additionally, there were small foci of recent ischemic necrosis involving the right ventricular myocardium. The walls of the coronary aneurysms were essentially fibrous with focal calcific deposits and only tiny vestiges of elastica or medial musculature. Except for a small lumen in the left circumflex the 3 aneurysms were completely filled with thrombus of varying age and organization. Related to the organization of the thrombi were small accumulations of lymphoid cells about thin-walled vascular channels in the aneurysmal wall. Uninvolved segments of the coronary arterial tree presented small focal gaps and defects in the medial musculature without associated inflammation.

Discussion

In the present case the structure of the aneurysms and the absence of septicemia or of endocarditis clearly rule out bacterial infection as an etiologic factor. Similarly, the clinical history and course as well as the anatomic findings negate the possibility of syphilis, rheumatic fever, or necrotizing arteritis. The distinction, accordingly, lies between congenital and arteriosclerotic aneurysm. In this instance, with organizing thrombi filling the dilated channels, this differential cannot be made on structure alone. The secondary intimal changes, which result from the organization of mural thrombi, duplicate the usual features of atherosclerosis. The replacement of specific vascular structure by fibrous tissue with calcific deposits too has no differential diagnostic value. Atherosclerosis, however, is a generalized pathologic process though its clinical manifestations may be local, and it is extremely unlikely that it may be so advanced that it causes aneurysms in the coronary arteries while there is only insignificant or early disease in the aorta and elsewhere. Indeed it has been demonstrated that atherosclerosis is generally more severe and advanced in the aorta than in other areas. Accordingly, we think that the aneurysms were congenital rather than arteriosclerotic. Obviously, this distinction could not have been made clinically, but an awareness of coronary aneurysm as an entity might have allowed us to interpret the radiologic findings differently and to place more significance upon the normal serum cholesterol values. There are undoubtedly many exceptions to the gen-
eralization that there is a positive correlation between elevated serum cholesterol levels and coronary atherosclerosis. In the younger age groups, however, exceptions of this nature are fewer and one must urge that other possibilities be considered before concluding that a given case is such an instance.

The basis for the formation of any aneurysm is a structural weakening of the vessel wall; the actual causative mechanism, of course, is variable. Only too often, as in this case, the aneurysmal wall is essentially scar tissue, and except for lesions associated with violent or even necrotizing inflammation, little or no evidence remains to indicate the nature of the original lesion. Clues are sometimes to be found in relatively uninvolved segments of the diseased vessels. We consider that the small focal medial defects of "uninvolved" segments are representative of the lesions which, in greatly augmented form, weakened the affected portions of the coronary arteries sufficiently to lead to aneurysmal dilatation. It will be recalled that Forbus demonstrated somewhat similar lesions as the basis for congenital berry aneurysms at the bifurcations of cerebral arteries.

The designation of an aneurysm as congenital implies only that the basic cause is developmental. The actual exploitation of the structural weakness by aneurysmal dilatation may occur during the neonatal period but may not appear for decades. Dilatation of the affected channel creates an additional burden that contributes to further growth of the aneurysm, since, hydrodynamically, the tension to which the wall of a channel is subject increases with its diameter. In the process there is progressive attenuation and atrophy of residual elements of the vessel wall. Fibrous repair and reinforcement tend to limit expansion, but usually without success. Even the occurrence of calcification within the wall of the aneurysmal sac, since it is patchy, does not prevent further growth. Thrombosis, a complication to which aneurysms are vulnerable because of slowing and turbulence of the blood stream, may through organization contribute to fibrous reinforcement of the sac wall. In the case of an aneurysm of the coronary artery, however, this sequence may be lethal, as in our case. Even nonocclusive mural thrombosis within a coronary aneurysm is not without hazard. First, the propensity of a clot to propagate may lead to complete occlusion. This situation is exemplified again by our case, in which the occluded aneurysms showed evidence of episodic clotting with recent thrombi superimposed upon older organized and organizing thrombi. Second, dislodgment of a mural thrombus, favored by the expansile pulsation of the aneurysmal sac, may lead to distal embolization. One wonders if this could not have been true in some of the cases reported with thrombosis distal to a coronary arterial aneurysm.

According to Packard and Wechsler, the first report of an aneurysm of the coronary artery was made by Bougon in 1812. By 1929, 2 probable cases of periarteritis nodosa being excluded, 28 cases had been described; by 1948 the number had risen to 45, and by 1957 there were 68. Etiologically, as shown in the accompanying table, more than half the reported cases are of congenital or arteriosclerotic origin. Current experience, moreover, suggests an even greater predominance, since the septic states that cause mycotic embolic aneurysms are now much more readily managed and there is a reduced incidence of syphilis and rheumatic fever. Similarly, other differences between older and more recent experience may be related to the diminished frequency of infectious forms of aneurysm. Scott had noted that coronary occlusion predominated as a cause of death, whereas aneurysmal rupture was more frequent in the earlier series of Packard and Wechsler. As is evident in table 1, the left coronary artery has been involved much more frequently than

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the right, but Crocker et al. did not find this to be true in the most recent series of 20 cases.

There were aneurysms of both coronary arteries in 11 cases; in 6 of that group the lesions were congenital. As shown in table 2, aneurysms of the coronary arteries are several times more frequent among males than females. The sex disparity is even greater if consideration is limited to the most frequent forms, congenital and arteriosclerotic. Age-wise, arteriosclerotic aneurysms are essentially lesions of the older age groups, whereas those of congenital origin occur at any age. One of the lesions tabulated herein as congenital was considered by the author to be arteriosclerotic as well. For those congenital lesions that do not appear until late in life at least, it seems reasonable to suspect that factors associated with aging are an important complement to the basic developmental lesion.

**Summary**

We have reported a case, the sixty-ninth of coronary artery aneurysm, the twenty-first of congenital aneurysm of the coronary artery, the sixth in which both coronary arteries were involved, and the first in which a coronary aneurysm was visualized, though not recognized, during life.

The patient had symptoms of myocardial anoxia for 2 years prior to his death from myocardial infarction. One year before his demise an x-ray film of the chest, taken with a Potter-Bucky diaphragm, had demonstrated a calcified ovoid mass, which subsequent autopsy proved to be an aneurysm involving the circumflex branch of the left coronary artery.

We have discussed the differential pathologic features of congenital and arteriosclerotic aneurysms and have tabulated some of the statistical information of the reported cases, including for completeness, other etiologic causes of the lesion.

**Summario in Interlingua**

Le caso que nos reporta es le sexanta-nine de aneurysma de arteria coronari, le vinti-prime de aneurysma congenite de arteria coronari, le sexe in que ambe arterias coronari
es afficite, e le prime in que un aneurysma coronari esseva visualisate—ben che non recognoscite—durante le vita del patiente.

Le patiente habeva symptomas de anoxia myocardial durante 2 annos ante su morte ab infarceimento myocardial. Un anno ante su morte, un roentgenogramma thoracie, obtenite con un diaphragma Potter-Bucky, demonstrava un calcificata massa ovoide. Le necropsia revelava subentemente que ille "massa" esseva un aneurysma que afficeva le branca circumflexe del arteria sinistro-coronari.

Nos ha discutite le aspectos pathologic de interesse pro le differentiation de aneurysmas congenite ab aneurysmas arteriosclerotic e ha summarisate in forma tabular le informatione; statitic del casos reportate in le litteratura. Pro render le presentation complete, altere causas etiologic del lesion es includite.

REFERENCES


One hundred eleven proved cases of polyarteritis nodosa were classified according to the presence or absence of lung involvement. In the cases with lung involvement, a respiratory infection preceded and dominated the illness. Blood and tissue eosinophilia was present and, microscopically, a granulomatous polyarteritis or necrotizing lesions not necessarily related to arteries were demonstrated. The theories of the pathogenesis are reviewed. Evidence points to an association with preceding respiratory infections, especially those due to streptococcus and with rheumatic fever. Of the many drugs inerminated, sensitivity to thiouracil drugs seemed to be the only valid relationship. Necropsy findings were reviewed in 86 patients in whom blood pressure records were adequate. Among 48 patients whose blood pressure remained normal, recent renal polyarteritis and glomerulitis were common; but healed lesions were rare. In 14 of 17 patients who developed hypertension during the disease and 21 patients whose blood pressure was high when first measured, healed renal polyarteritis was found.

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