RARE FORM OF SACCULAR CARDIAC ANEURYSM WITH SPONTANEOUS RUPTURE *

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A not uncommon late complication of coronary arterial disease and cardiac infarction is ventricular aneurysm, which rarely ruptures except in the presence of reinfarction of the old scar or adjacent myocardium. Commonly such aneurysms develop only at the site of large and healed infarcts, are of considerable size and have a wide communication with the cavity of the chamber involved. Actually many of them represent merely a dilatation of the heart wall rather than aneurysm in the more restricted sense of the term.

Among a series of 40 cases of spontaneous rupture of the heart reported by us in a previous paper [1] there was encountered one example of rhexis of a true saccular aneurysm communicating with the left ventricular cavity by only a small opening, so that the aneurysm lay over the external surface of the heart. In view of the unique character of the lesion, our failure to encounter an identical example among the exhaustive literature dealing with cardiac rupture, and because the length of the first communication necessitated only a brief consideration of individual instances, we have felt it advisable to make a more detailed account of this particular case

A similar aneurysm which, however, failed to rupture was described by Corvisart [2] in 1797. It occurred in a negro 27 years of age who entered the hospital "in inexpressible anguish and anxiety; breathing was laborious and interrupted; he suffered little pain in the thorax, which besides, sounded well in its whole extent; he complained of feeling a violent pain both toward the region of the stomach and of the liver." Death occurred the following day. The heart was reported to be of natural size, "but the superior and lateral portion of the left ventricle was surmounted by a tumor almost as large as the heart itself." This tumor was of cartilaginous consistence but of the appearance of muscle, communicated with the ventricle by a small opening, and contained coagula.

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Report of Case

Clinical History: W. E. J., Male, aged 45, a lieutenant in the Portland fire department, died suddenly while driving his car. An autopsy was made at the direction of the coroner. Three months prior to death the man had consulted a physician because of severe pain in the stomach, radiating to the left arm, pain in the back and headache. On this occasion he appeared very ill and suffered intensely, requiring a half grain of morphine hypodermically for relief. The heart was irregular, the valve tones indistinct, the pulse feeble and the arteries were inelastic to palpation. The systolic blood pressure was recorded as 160 but there is no notation of the diastolic reading. A hasty urinalysis revealed a trace of albumin but nothing else of consequence. The past history was irrelevant except for loss of appetite and headache extending over a period of five years. In early life he drank to a moderate extent but for the past few years had indulged but very little. On the day of death he was on duty and feeling unusually well.

Postmortem Examination *

For the sake of brevity only the more pertinent findings will be described.

Even before opening the pericardial sac the presence of hemorrhage was anticipated by the dark color and tenseness of the membrane. Upon incising the pericardium the sac was found to be filled with blackish clotted blood, obscuring from view the empty and contracted heart and forming a cast about it. When the clot was removed there was seen over the anterolateral aspect of the left ventricle an area of fibrous adhesions 7.8 by 4.5 cm. in extent binding the epicardium and pericardium. Similar bands obliterated the pericardial space over the surface of a mushroom-like mass situated well up toward the atrioventricular sulcus and between the anterior interventricular and circumflex branches of the left coronary artery (Fig. 1–A). Along the inferior border of this pouch and 1 cm. lateral to a line drawn through its center was a slit-like, ragged-edged opening 4 mm. in length. A probe passed into this defect led through the 1 mm. wall at the base of the aneurysm into its cavity which lay over the external surface of the left ventricle. On separating the patch of pericardial adhesions lying inferior to the aneurysm a layer of clotted blood was exposed in the fibrous tissue holding the pericardia together. Tracing the hemorrhage cephalad one found it leading to the rent in the inferior wall of the aneurysm mentioned previously. It would thus appear that rupture took place at this

* Path. No. 10,725, Group F, Case No. 35 (see Ref. 1).

point and that the escaping blood then dissected through the adhesions for some distance before finally breaking out into the unobliterated portion of the pericardial sac (Fig. 1–A). Further fibrous bands obliterated the pericardial space over the anterior and left aspects of the left atrium and the greater part of the posterior surface of the left ventricle, stopping abruptly 2 cm. from the interventricular septum posteriorly and 2.5 cm. from the apex inferiorly. In addition to dissection along the course already described the extravasated blood had also made its way along the base of the aneurysm over the surface of the left atrium as far as the point of entrance of the left pulmonary veins and into the space separating the root of the aorta and pulmonary artery, tearing adhesions wherever these existed along its path.

The aneurysm measured 4 by 4 by 3 cm., the last mentioned figure being the height above the surface of the heart. The wall varied between 0.1 and 0.7 cm. in thickness and appeared to consist solely of pericardium, fibrous connective tissue, epicardium and possibly endocardium, bound intimately together. The lining of the sac was generally smooth and whitish, except for linear grooves and ridges radiating outward in a fan-like fashion from an opening measuring 0.5 by 0.3 cm., located at the base of the cavity and leading directly through the myocardium into the left ventricular chamber. In several places the smooth and glistening white lining of the pouch was coated with a thin, yellowish brown film of fibrin; otherwise the aneurysm was empty.

An incision carried through the opening leading through the left ventricular wall disclosed patchy, whitish scar tissue in the myocardium immediately around the defect and for approximately 2.5 cm. outward into the muscle on all sides of it (Fig. 1–B). This was the only macroscopic lesion in any part of the heart muscle. Moreover, after sectioning it became evident that the fluted ridges around the margins of the opening into the aneurysmal sac were directly continuous with several greatly attenuated trabeculae carneae. Elsewhere these muscle bands were of average normal size except along the septal surface of the ventricle where there was some flattening and atrophy. The internal end of the communication between the ventricle and sac on its surface was situated 1.5 cm. from the attachment of the posterior mitral valve cusp to the mitral ring. The anterior and posterior walls of the left ventricle averaged

2 cm. in thickness, decreasing to 1.5 cm. at the point of the opening leading to the aneurysm and to 1 cm. at the apex of the chamber. The several valves of the heart and the endocardium of the different chambers displayed no evidences of disease.

The ostium of the right coronary artery appeared almost closed by sclerosis, while that of the left was greatly narrowed by the same process. Extending cephalad from the commissure separating the anterior and left posterior aortic leaflets were several shallow linear wrinkles in the intima but nothing further suggestive of syphilis was observed at any other point along the course of the aorta. Aside from the plaques obstructing the coronary orifices there were a number of small and beaded yellowish streaks in the intima of the sinuses of Valsalva. Similar plaques were scattered along the intima of the thoracic aorta.

The main left coronary artery was a large and freely patent vessel exhibiting only small atheromatous plaques. Its anterior interventricular branch was likewise of ample size, but in places the lumen was moderately obstructed by whitish sclerotic patches. The same was true for the circumflex division and the right coronary artery. The latter vessel did not extend for any appreciable distance past the margo acutis, and to compensate for this the left circumflex gave off a posterior interventricular branch which coursed toward the apex and formed an adequate anastomosis with the anterior descending artery. At the point of bifurcation of the left coronary there originated two good sized twigs which supplied the portion of the left ventricle where the aneurysm and myofibrosis were located. The smaller of these passed directly to the area of scarring about the opening leading through the ventricular wall, and macroscopically its lumen at one point seemed to be occluded by an old and organized thrombus. No fresh thrombi were discovered in any of the coronary vessels.

The heart, together with the adherent portion of the pericardium and ascending aorta, weighed 640 gm. The kidneys had a combined weight of 440 gm. The capsule of each stripped readily, revealing a faintly and finely granular surface. On section the cortex appeared to be of normal thickness and the rays and labyrinths were everywhere plainly outlined. The lungs, spleen and liver displayed nothing other than acute passive hyperemia.

Microscopic Examination

Heart: Sections from blocks cut to include the opening in the left ventricular wall and the adjacent myocardium for 2.5 cm. on either side of it reveal irregular patches of connective tissue interrupting the muscle substance (Fig. 1–B). For 5 to 7 mm. around the margins of the hole the tissue is almost wholly fibrous but becomes progressively less so farther away from the defect. Several of the trabeculae carneae in this region are reduced to dense scar tissue or show only occasional persistent muscle cells. Between two of these muscles small thrombi consisting of fibrin and leukocytes are discovered. Near the opening the fibrous connective tissue nuclei are larger and more numerous than elsewhere and yet are widely separated by an abundance of coarse and compact intercellular collagen, among which is a fair number of thin-walled and engorged blood vessels. More peripherally the vessels are larger and possess a definite muscularis. None are occluded by thrombi nor do they exhibit evidences of arteriolosclerosis. Approaching the margin of the aneurysmal opening one finds the scar replacing the myocardium passing uninterruptedly from the sac on the external aspect of the heart along the defect to become continuous with the endocardium. At the junction of the scar and endocardium there is edema, engorgement of capillaries and hemorrhage by diapedesis. The connective tissue cells forming the immediate lining of the communication uniting the ventricular cavity and the aneurysm have very large and hyperchromatic nuclei whose long diameter lies parallel to the opening and at a right angle to the myocardial cells. In this location the tissue is compact rather than loose, possibly indicating a compression by the pulsation of the blood passing through the opening. A true endothelial lining is lacking but the inner surface is nevertheless smooth and not covered by thrombi. As in other parts of the scar there are moderate numbers of capillaries about which are lymphocytes, plasma cells and histiocytes.

In the wedge-shaped area between the epicardium and base of the aneurysm (see Fig. 1–B) there is a rich fibroblastic proliferation and the whole zone consists of these cells and histiocytes. The muscle cells adjacent to the epicardium are hypertrophic, as evidenced by their great size and large hyperchromatic nuclei. Quite generally the muscle fibers are separated by very loose and apparently edematous

connective tissue. A deposit of fibrin covers the epicardium along this angle.

Sections from a number of different parts of the aneurysm wall reveal the lining to be undulating, interrupted by occasional sharp clefts, and consisting of compressed fibrous connective tissue, at times coated with a thin layer of fibrin or blood platelets (Fig. 2). Nothing resembling normal endocardium is identified. Passing from within outward there is encountered first a zone of solid connective tissue with abundant intercellular substance, followed by loose edematous tissue of the same nature containing numerous but irregularly distributed capillaries distended with blood and sometimes surrounded by fresh hemorrhage. At least one of these vessels exhibits a definite rupture of its wall, indicating that in this instance at least the surrounding hemorrhage is due to rhexis. Next there is a very active fibroblastic zone displacing almost entirely the fat cells of the subepicardial region. The blood vessels here are distended and collared by numerous lymphocytes, plasma cells, histiocytes, a sparse sprinkling of neutrophilic polymorphonuclear leukocytes and multinucleated giant cells engorged with yellowish blood pigment. The pericardium and epicardium are so intimately adherent that it is practically impossible to distinguish one from the other. Two coronary arterioles included in the subepicardium display proliferation of the intimal cells with resultant narrowing of their lumens.

Still another section, coming from the vicinity of the rupture through the aneurysm wall, discloses extensive recent hemorrhage into the fibrous adhesions between epicardium and pericardium and probably represents another ramification of blood from the point of rupture. More inferiorly and toward the apex of the ventricle the hemorrhage spreads out extensively and diffusely, breaking up and widely separating the adhesions. By these routes the dissecting hemorrhage must eventually have reached the nonobliterated portion of the pericardial sac and led to fatal hemopericardium.

The main left anterior interventricular coronary artery is the seat of intimal atherosclerosis, which has produced eccentric narrowing of the lumen. The deeper and more external parts of the plaques are often the seat of fatty degeneration and contain dust-like particles of lime salt. There are no apparent breaks in the endothelium. Lying free within the lumen and only partially occluding it is a small

ante mortem clot consisting of fibrin and leukocytes. The media is thin and atrophic while the adventitia is unaltered.

Microsections from three different points along the course of the small artery originating at the bifurcation of the main left coronary artery and coursing directly toward the scar about the aneurysm opening disclose advanced atherosclerosis of the intima with pronounced narrowing and eccentric displacement of the lumen. Thrombosis is lacking, although a few scattered erythrocytes and leukocytes are present. The deeper parts of the various patches of sclerosis are fatty, containing fat-laden phagocytes and large plates of calcium, not infrequently encased by multinucleated foreign body giant cells. Also there are several endothelial-lined, blood-containing channels in the degenerated parts of the plaques, but nothing whatsoever to indicate the existence of an organized and recanalized thrombus. The media of this vessel is thin and in places infiltrated with lymphocytes and plasma cells, yet without evidence of necrosis or scarring such as might be anticipated in chronic syphilis. Similar but larger cell collections collar the adventitial vessels in which evidences of endarteritis are lacking.

Microscopically the right coronary artery displays the same atherosclerotic process present in the other arteries with the additional finding of atheromatous ulceration. Although there is stenosis the lumen is not thrombosed. Perivascular accumulations are lacking.

The histopathological changes in the aorta are quite the same as those described for the coronary vessels, namely, varying degrees of intimal atheroma with softening, collars of lymphocytes and plasma cells about the vasa vasorum of the media and adventitia, sometimes with minute flame-like scars but never obliterative endarteritis.

Kidneys: A microscopic study of these organs reveals occasional completely hyalinized glomeruli, hyperemia of all other glomerular tufts, and subintimal hyaline thickening of certain intertubular and afferent glomerular arterioles, leading in some instances to moderate stenosis.

Pathological Diagnoses: Arteriosclerotic narrowing of coronary arteries, especially pronounced in one branch of the left; patchy myofibrosis of left ventricle with formation of saccular aneurysm, spontaneous rupture of this aneurysm and fatal hemopericardium; focal

fibrous pericarditis; atherosclerosis of aorta; possible rheumatic arteritis and aortitis; hypertrophy of heart; slight arteriolosclerosis of kidneys; probable hypertensive cardiovascular renal disease; generalized acute passive hyperemia.

Discussion

From the foregoing description it is clear that death was the result of spontaneous rupture of the thin and fibrous wall of a very unusual type of cardiac aneurysm and the pericardial adhesions about it. The mechanism by which the aneurysm was produced is less easily explained. Although meager, the clinical data suggest that on the one occasion, three months prior to death, when the man was seen by a physician he was suffering from cardiac symptoms referable to the heart and compatible with coronary thrombosis. The one other significant observation at this time was the systolic blood pressure of 160 recorded during the seizure. It is common knowledge that the blood pressure decreases after a coronary accident and it is probably not erroneous to postulate that prior to the attack of cardiac distress the tension was greater than 160 mm. Hg. In any event the man had hypertension during the seizure and it is unnecessary to prove that the blood pressure was ever greater. If we accept Bell and Clawson's [3] criteria for the clinical and pathological diagnosis of essential hypertension this case becomes a clear-cut instance of hypertension, for not only was the blood pressure elevated beyond the normal limit of 150 mm. of mercury during life, but at autopsy there was found cardiac hypertrophy (640 gm.) without demonstrable cause other than possibly from pericardial adhesions and the characteristic renal arteriolar lesions of essential hypertension.

Granting that primary hypertension formed the basis of the vascular disease there yet remains the problem of genesis of the aneurysm. In the absence of demonstrable coronary thrombosis one must seek to explain the myofibrosis on other grounds. Slow and progressive ischemia of a small area of myocardium brought about by high grade atherosclerosis of the vessel supplying the myocardial scar may have led to disappearance of patches of heart muscle and finally replacement fibrosis. Less probably the scar may represent the end-result of focal rheumatic or syphilitic myocarditis. Definite proof of

the existence of either of these diseases is lacking but certain features about the perivascular aortic and coronary arterial lesions are more suggestive of the former.

When and how the aneurysm formed is a question to which we can give no definite answer. In view of the clinical history there is reason to believe that it may have had its inception three months prior to death either during or shortly after the attack of severe thoracic pain and cardiac embarrassment. It seems possible in the presence of hypertension there may have been sufficient increase of intracardiac pressure exerted over the small area of already existent fibrosis to cause stretching or dissection until the external surface of the ventricle was reached, after which instead of immediate rupture the scar became adherent to the pericardium and these structures continued to hold for a time before finally giving way. Still another possibility is that of cardiac infarction, partial rupture during the acute stage of necrosis, followed by the formation of pericardial adhesions and finally rupture.

Although ordinarily cardiac aneurysm develops along the whole extent of the scar following healing of an infarct we have seen at least one striking exception to the rule. This case (No. 8370) (Fig. 3) may possibly represent an earlier stage of saccular aneurysm than the one forming the basis for our communication, and if so serves to show that not all cardiac aneurysms demand a large area of scarring for their development. In the example just mentioned a portion of a massive healed infarct involving the anterior wall of the left ventricle and interventricular septum was separated from the main cicatrix by a zone of intact myocardium and in the small part there formed a little pouch near the apex and adjacent to the interventricular septum. The peculiar and unusual feature was the fact that on either border of the wholly fibrous wall of the sac the ventricular muscle was of normal structure and thickness so that it appeared as if the aneurysm had been punched out over only a small area. As in the first case (No. 10,725) the neck of the pouch was somewhat narrower than the sac itself and the aneurysm wall externally consisted solely of fibrous tissue and epicardium. What would have been the later history of this aneurysm had the individual survived for a longer time is conjectural, but it seems possible that it might later have ballooned out over the external aspect of the ventricle as in the

first case, although the presence of the tampon thrombus that filled the sac may conceivably have organized and precluded such a complication.

SUMMARY

A rare and almost unparalleled type of cardiac aneurysm situated over the external aspect of the left ventricle is described and the possible causes for its development are discussed.

REFERENCES

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DESCRIPTION OF PLATES

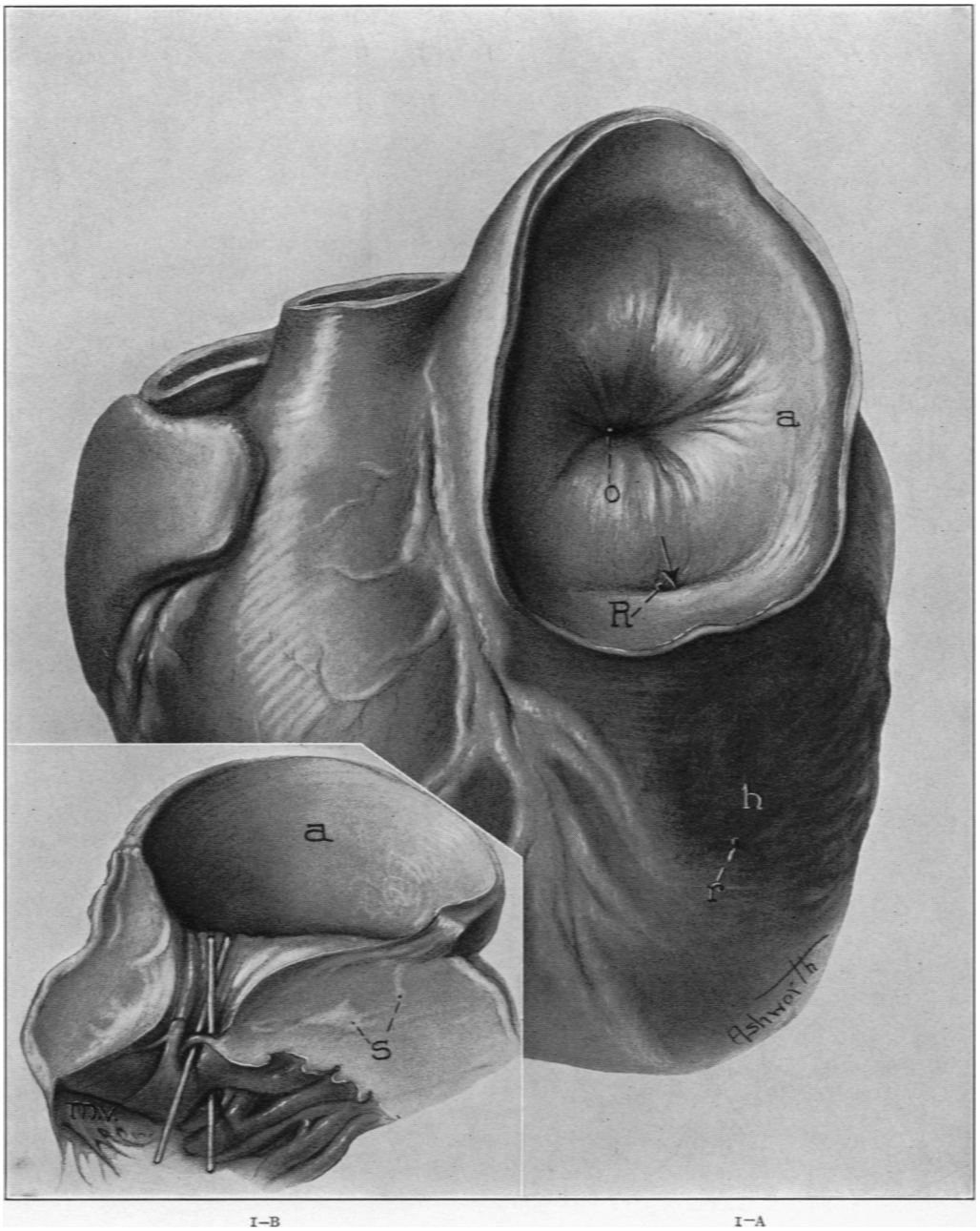
PLATE 99

FIG. 1–A. Case No. 10,725. Drawing, actual size. Anterior aspect of heart showing position and appearance of saccular aneurysm after removing a portion of its wall.

a = aneurysm.
o = opening with fluted margins, leading to left ventricular cavity.
R = point where rupture began at base of aneurysm wall.
r = fresh dissecting hemorrhage in pericardial adhesions.
h = torn fibrous adhesions over epicardium.

FIG. 1–B. Actual size drawing of aneurysm and left ventricular wall after hemisection of the sac to display the size and character of the defect and structure of adjacent heart muscle.

a = aneurysm.
s = patchy myofibrosis in vicinity of opening leading to aneurysm.
m.v. = posterior cusp of mitral valve.
Marker rods indicate ramifications of the channel between the atrophic trabeculae carneae and the main communication.



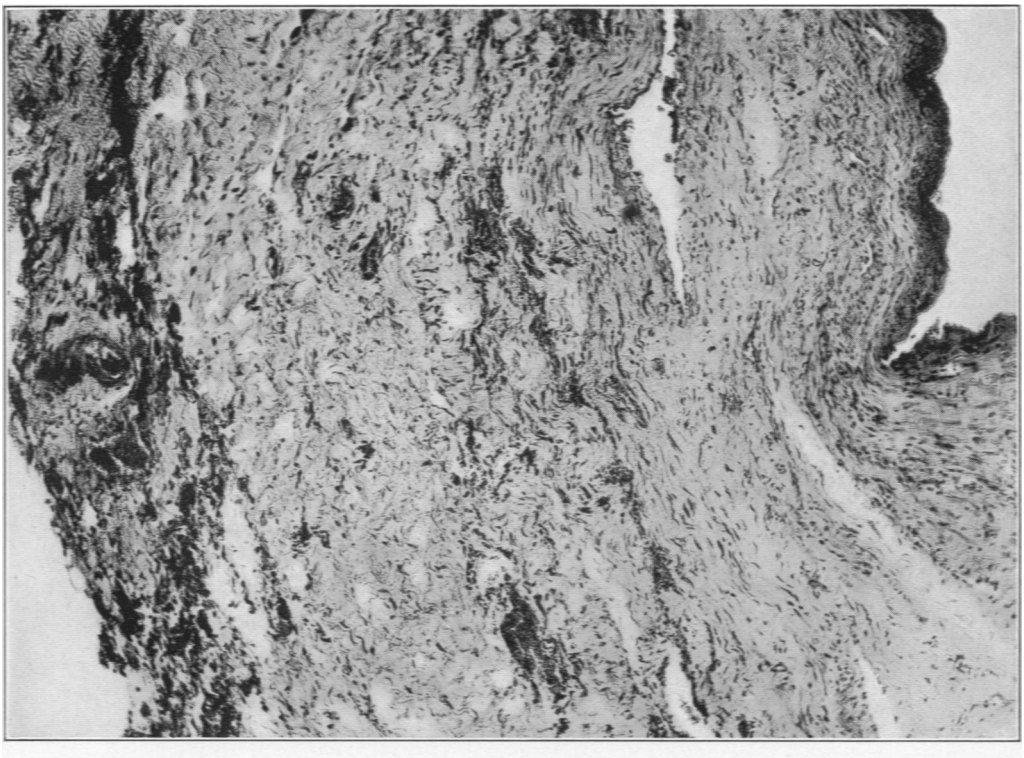
I–B I–A

Hunter and Benson Rare Form of Saccular Cardiac Aneurysm

PLATE 100

FIG. 2. Low power photomicrograph showing entire thickness of cardiac aneurysm in case No. 10.725. To the right is seen a small portion of the undulating lining of the sac, at this point coated with fibrin. On the left border are torn vascular fibrous adhesions replacing the epicardium. All of the remainder of the wall consists of relatively young fibrous connective tissue in which a number of distended capillaries are visible. The complete absence of myocardium is evident.

FIG. 3. Case No. 8370. Photograph displaying septal surface of left ventricle. The interventricular septum (sectioned) and anterior wall of the ventricle are the seat of a healed infarct extending to the endocardium, which is greatly thickened and whitish. Near the apex is a small aneurysm surrounded by intact myocardium of normal thickness. Only the tamponing thrombus, thin fibrous wall of the sac and epicardium separate the cavity of the ventricle from the external surface of the heart.



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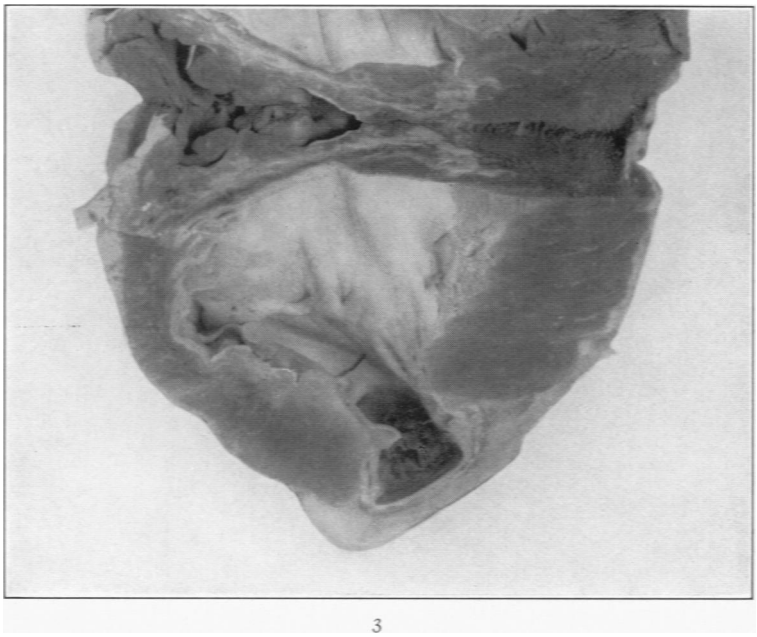

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Hunter and Benson Rare Form of Saccular Cardiac Aneurysm