CONGENITAL CYST OF THE MYOCARDIUM *

L. J. SACHS, M.D., and ALFRED ANGRIST, M.D.
(From the Department of Pathology, Queens General Hospital, Jamaica, N. Y.)

An epithelium-lined cyst of the heart is a rare lesion. Such a lesion has interesting theoretical implications as to origin, and so the presentation of an additional case seems warranted.

REPORT OF CASE

The patient, G.F. (A-42-252), was a white male, 44 years old, who had worked as a counterman. He was admitted on July 16, 1942, with severe precordial pain radiating to the left arm and neck for a period of 3 hours. Mild precordial discomfort had been present for the past few days.

On admission, the patient was vomiting. He showed a gray hue to his skin and was sweating. Physical examination revealed nothing of note. The heart sounds were fair, with a regular sinus rhythm of 60. The blood pressure was 110/80 mm. of Hg. The respirations averaged 24 per minute. The urine was negative for albumin and glucose.

In spite of morphine, nembutal, caffeine and sodium benzoate, the patient expired on July 17, 1942, with the final clinical impression of an acute coronary occlusion.

The autopsy confirmed the clinical diagnosis. A fresh thrombus was found in the descending branch of the left coronary artery, approximately 1 cm. below the ostium. Both coronary arteries showed marked atherosclerosis throughout. Microscopically, areas of necrosis and also of old fibrosis were found in the myocardium. The heart weighed 280 gm.

A small cyst situated in the center of the left ventricular wall was an incidental finding (Fig. 1). The cyst measured 0.9 cm. in diameter. Its cavity was rounded and sharply delimited. The lining showed a thin, glossy smooth layer of translucent tissue. The contents of the cyst were gelatinous and translucent, with a pale greenish tint. The cyst was located well within the left ventricular musculature, failing to reach the epicardium or the endocardium. Its lower border was located about 3.5 cm. from the apex. The cyst was completely surrounded by cardiac muscle, and did not occasion any discernible bulging of the muscle toward either the epicardial or the endocardial surface. No external manifestation of its existence was present, and the structure was found only because of a fortunate routine section through its center.

On microscopic examination, an inner layer of epithelial cells was seen. The epithelium was tall columnar for the most part, with a shorter, more cuboidal appearance in some areas. The epithelial lining was distinctly ciliated (Fig. 2). The cilia were numerous, closely placed and of uniform height for each cell. In the shorter cells, the cilia

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were not so tall, but were proportionately more prominent, and, for some cells, occupied as much as one-half the full height of the cell. Most of the cilia of the taller epithelium measured less than one-third or one-fourth the height of the cell structure. The cilia were inserted on a sharp cell border, which appeared as a narrow, linear, condensed eosinophilic zone. The cytoplasm was distinctly granular and eosinophilic. The nuclei were situated in the basilar portions of the cells and were elongated and oval. The nuclei showed some variation in size, but the range of variation in both size and shape was rather narrow.

Discussion and Comparison with Other Cases

Very few similar instances of epithelial cyst formation within the myocardium could be found in the literature. Five of the published cases (Stoeckenius,[1] Kolatschow,[2] Davidsohn,[3] Yamauti [4] and Bayer [5]) represent lesions that are very similar to the one presented here. A sixth case by de Châtel,[6] and two other instances reported by Bayer, differ in many respects. The last three will be discussed separately. There were several features common to the first five cases. They were all situated in the left ventricle of the heart, near or in a papillary muscle. Yamauti and Davidsohn described unilocular formations like the one observed by us, while the cyst reported by Stoeckenius showed subdivisions into several smaller cavities "due to extension between bundles of muscle fibers." Kolatschow described two smaller confluent structures in addition to the oval main cyst. He did not state clearly whether he dealt with one large cystic structure with an irregular invaginating or undulating wall, or with three separate, closely placed cysts. The cysts described by Stoeckenius, Yamauti and Bayer, as well as the lesion presented here, were situated completely within the myocardium. The reports of Kolatschow and Davidsohn note that the cystic structure projected partially into the ventricular cavity.

The ciliated epithelial lining represents an important finding in the five cases with identical features. Its significance lies in the clue it gives to the possible origin of these cystic structures. In the cases of Kolatschow [2] and Davidsohn,[3] as well as in this instance, the contents of the cyst were gelatinous. Yamauti [4] described the contents as homogenous and "pale purple, with hematoxylin-eosin stain." Stoeckenius [1] noted a finely granular meshwork with some clumped desquamated epithelial cells in the contents. Bayer [5] described rounded, albuminoid formations in the lumen. The epithelium is described by Davidsohn as existing in two layers—an outer cuboidal and an inner higher and densely ciliated layer. The cytoplasm in his specimen was eosinophilic

and granular. The nuclei were dark, hyperchromatic and oval, some being highly irregular in shape. Yamauti found "one layer of cuboidal, distinctly ciliated epithelium, with almost oval nuclei, situated in the center of the cells." Kolatschow's case showed columnar ciliated epithelium. He failed to mention the number of layers, but his photomicrograph shows only one distinct layer of epithelial cells. He described the nuclei as "enlarged [ausgedehnten]" possibly meaning elongated, and stated that "they are smaller and more rounded toward the base." Bayer found the cyst lining to consist of one layer of high columnar epithelium with oval, deep-staining nuclei in the lower two-thirds of the cells. Stoeckenius reported columnar cuboidal and flat cells, all showing abundant cilia. Davidsohn, and also Bayer, suggested that the difference in the height of the cells in the last mentioned case may represent an artifact produced by the thickness of the section. The presence of cilia on all of the cells favors this interpretation. The cytoplasm is described by Stoeckenius as "granular, sometimes vacuolated." He described the nuclei as vesicular, elongated or rounded, sometimes hyperchromatic and rod-shaped.

Except for Stoeckenius,[1] all authors mention a thin fibrous wall situated outside of the epithelium. Such a fibrous wall was present in our specimen (Fig. 2). Davidsohn [3] found a distinct homogeneous eosinophilic basement membrane. Bayer [5] found none. A hyaline refractile basement membrane was not made out in hematoxylin and eosin stained sections in our case, but was seen after use of phosphotungstic acid hematoxylin and van Gieson's stains, and best with Verhoeff's elastica stain. No smooth muscle was present, as noted by Bayer. Included atrophic heart muscle fibers were made out, deeply imbedded in the collagenous stroma of the fibrous wall of the cyst.

In none of the reported cases were the cysts found to have clinical significance. In none of them were they diagnosed antemortem. Davidsohn [3] had only negative evidence to support an opinion of a causal relationship for the blowing systolic murmur and thrill which were heard at the base of the heart and then at the apex in his case.

Discussion of Origin

Cystic structures in the heart wall have received different explanations for their origin. All authors agree that the lesion represents a congenital anomaly. According to Stoeckenius,[1] the cyst is due to persistence of the embryologic sponge-like structure of the myocardium, corresponding to the stage of development in fish and amphibians. The cystic structures of this stage are all communicative with the main cavity of the heart, and are lined by endothelium. He assumed that

the endothelium at this stage is multipotent and can differentiate to become columnar and ciliated. The flat cells he observed reminded him of endothelial cells of the epicardium. This theory has the objection that it derives the ciliated columnar epithelium from endothelial cells. As noted above, Davidsohn [3] and Bayer [5] explained the flat cells described by Stoeckenius on the basis of a histologic artifact. This viewpoint would seem to be strengthened, as suggested by Davidsohn, by the fact that all of the cells showed cilia.

Kolatschow [2] presented another hypothesis. He placed the origin from the external part of the myocardial plate, which is the accepted origin for the epicardium, by a process of invagination into the internal plate, which is to become the myocardium proper. This proposal also derives a ciliated columnar epithelium from the mesenchyme. Bayer's [5] case of endothelium-lined cyst and diverticulum may be explained thus. The island of hyaline cartilage found in the wall of this structure may be mesenchymal, though metaplastic origin from epithelium is accepted for this tissue. We have often seen the serous epicardial covering differentiate or undergo metaplasia into rather tall, columnar cells. A cuboidal appearance of the epicardial cells is not at all uncommon. We do not recall, however, any cilia on such an altered epicardial surface layer. Yamauti [4] did not go into details concerning the origin of these cysts. Davidsohn [3] compared the structure with esophageal cysts. He noted a complete absence of embryologic data that would offer an adequate explanation for the cardiac cyst on this basis. Bayer [5] referred the origin of the cystic structure with ciliated epithelium in his case to displaced tissue of the bronchial tree. He pointed out that "as yet no relations between the heart and bronchial tree are found embryologically, but may exist." In favor of the concepts of Davidsohn and Bayer is the fact that, whereas the mesoderm ordinarily fails to produce ciliated epithelium, the entoderm, which is the source of origin for the bronchial tree and esophagus, does do so.

There exists in the early embryo an ideal stage for the heterotopic inclusion of entoderm by the mesoderm which forms the heart. This is seen in the chick embryo when the original flattened germ layers are folded ventrally to establish the body form, the gut lumen and the single heart. The bilateral cardiac Anlagen are in the region of the head at this time, and in intimate contact and partly enclosed by the endoderm to form the foregut. It is during the fusion of the bilateral cardiac primordia to form a single chamber at this juncture that the circumstances seem most favorable for such inclusions. Under this concept, the ciliated cyst of the myocardium represents a *sequestration cyst involving entoderm* in contrast to the mediastinal dermoids, which

may be derived from the ectodermal layer. De Châtel[6] suggested this mechanism of origin for his dermoid cyst.

The entoderm can give origin to squamous epithelium, as in the esophagus, and this heaping up of the epithelial cells occurs early in the embryogenesis of this organ. In some of the lower forms, such as Amphioxus, the esophagus is actually lined by ciliated columnar epithelium. This would mean that the squamous epithelial cysts and the so-called esophageal cysts of the mediastinum can also take origin from the entodermal foregut. It would simplify our problem here if we could assume that the squamous epithelial cysts represent inclusions occurring *later* in the embryonic development of this region. The ciliated cysts represent, then, inclusions occurring very early in the embryo, probably at the time of the formation of the primitive foregut. The case of de Châtel,[6] with both squamous and columnar epithelium in the wall of the same cyst, favors such theoretic considerations. By this concept, the esophageal cysts and the cardiac cysts are closely related, if not identical in origin. No direct factual evidence for any of these theoretic propositions is known to exist in the human embryo.

Conclusion

A cyst of the myocardium, lined by ciliated columnar epithelium, is described, and five similar cases in the literature are reviewed. Consideration of the theoretical implications as to origin results in the suggestion that such cysts arise through the heterotopic inclusion and sequestration of entoderm during the formation of the primitive foregut and single-chambered heart.

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[*Illustrations follow*]

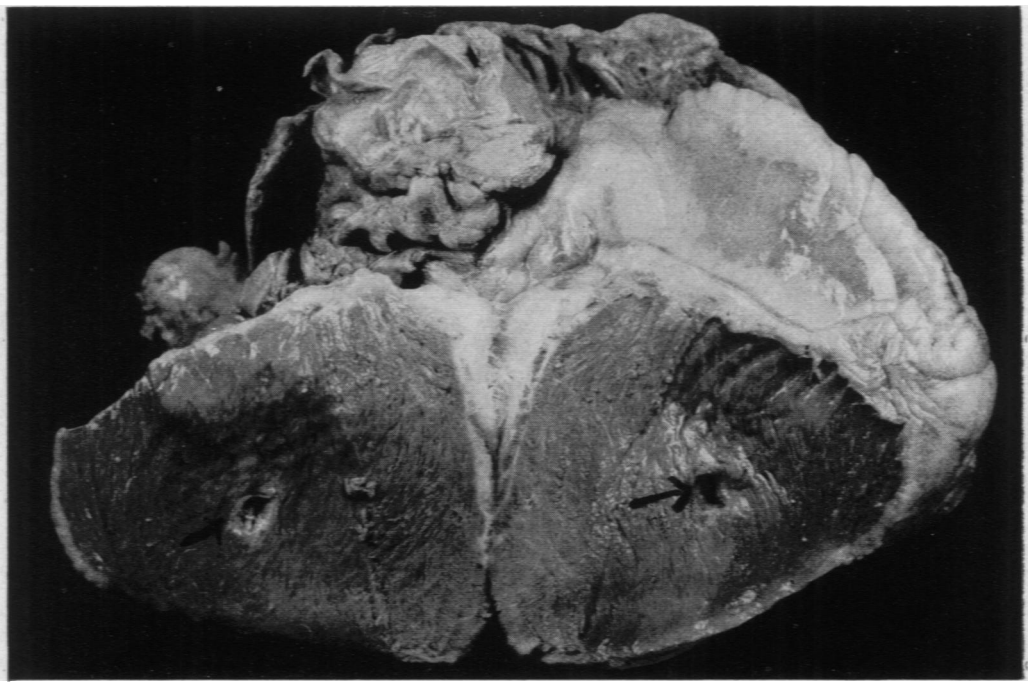
DESCRIPTION OF PLATE

PLATE 31

FIG. 1. Sectioned left ventricular wall, with transected cyst indicated by arrows.

FIG. 2. Photomicrograph of cyst wall showing the ciliated columnar epithelium which lined the cyst, and adjacent stroma. × 400.

1



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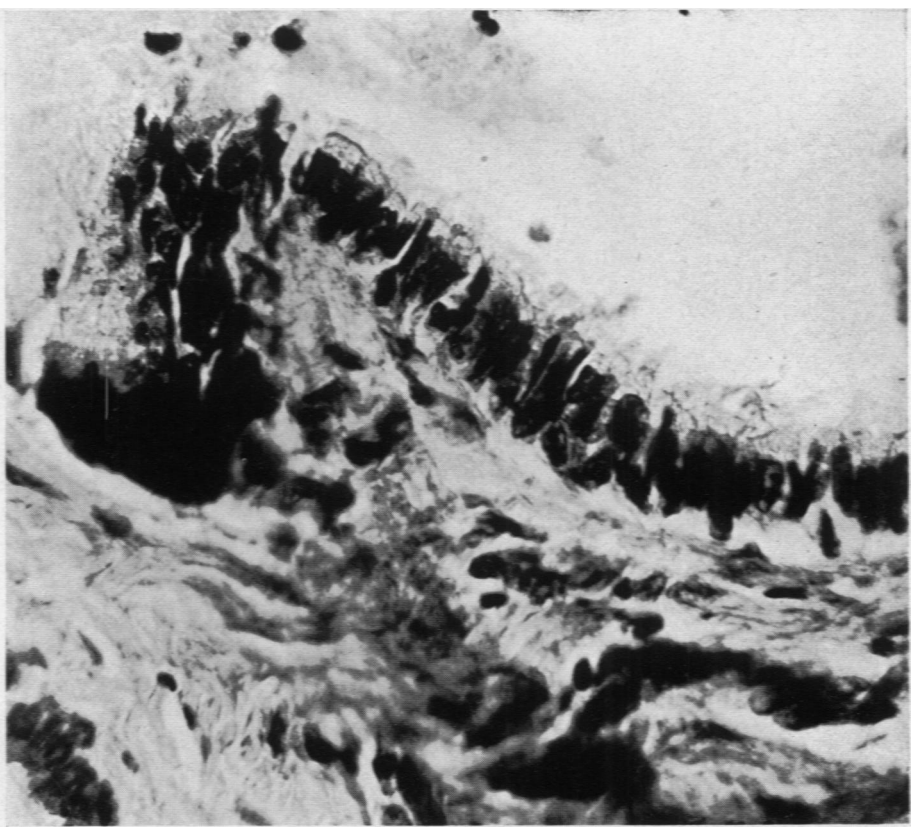

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