POLYCYSTIC DISEASE OF THE PANCREAS (DYSONTOGENETIC CYSTS)

REPORT OF A CASE WITH PARTIAL PANCREATECTOMY

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CYSTS of the pancreas may be considered comparatively rare. This condition most frequently presents interesting points as regards the clinical diagnosis or the pathologic picture found at exploration or at necropsy as well as ample reason for wondering as to the possible etiology and pathogenesis.

Schmieden and Sebening,[3] in 1927, reported 128 cases in which operations were performed for cysts of the pancreas. In 73 cases the origin of the cysts was not known, in 16 the cysts originated from previous trauma, in 28 they occurred following acute disease of the pancreas, and in 11 they were found in a tumor of the pancreas. According to Robson and Moynihan,[2] pancreatic cysts may be classified as follows: (1) Retention cysts; (2) proliferative cysts, cystic adenoma, and cystic epitheliomata; (3) hydatid cysts; (4) congenital cysts; (5) hemorrhagic cysts; and (6) pseudocysts.

Rarest among these cysts are the congenital cysts of the pancreas, which for the sake of description have been termed "polycystic disease of the pancreas." Pathologists have used the term "dysontogenetic cysts of the pancreas," which is a better term than "polycystic disease of the pancreas." It is a known fact that this type of cyst frequently occurs with other cysts, especially cysts of the kidneys, liver, and central nervous system. This association suggests a definite etiologic relationship between these different cysts. Lindau,[1] in his classic monograph on this subject, has furnished a new understanding of the pathologic picture of hemangiomata of the central nervous system and their relation to dysontogenetic cysts of the pancreas and kidney. In addition to five cases which he observed personally, Lindau collected ten cases from the literature. Dysontogenetic cysts of the pancreas were present in eight, cysts of the kidneys were present in ten, hypernephromata were present in six, and tumors of the suprarenal glands were present in two of these 15 cases. At the present time in the files of The Mayo Clinic there are histories of four cases presenting this syndrome, which in the recent literature has been described as "Lindau's disease." These cases will be reported later.

In the present communication we wish to present a case of dysontogenetic

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cysts of the pancreas in which there was no clinical indication of any lesion of the central nervous system. From the scattered reports in the literature it is very difficult to get a definite idea as to the frequency with which a polycystic disease of the pancreas is found without simultaneous lesions of the central nervous system. This case, however, will illustrate the necessity of ruling out such a possible combination. When the patient entered the clinic the symptoms predominantly were referable to the abdomen.

Case Report.—A woman, age 39, registered at the clinic February 12, 1936, because of abdominal pain. Her family history was essentially negative. She had been married 21 years and had had two children. The patient had never been very strong, but had always worked hard. Since 1934 she had felt much weaker than previously. About September, 1933, she first had noted a slight soreness in the left side of the abdomen when carrying anything heavy. This soreness had disappeared when she had rested or had assumed the recumbent position. Six months later she accidentally had felt a hard lump in the upper left side of her abdomen. She had noted that this palpable tumor had been gradually getting larger, and that the abdominal soreness had increased moderately. About February, 1935, she had detected a similar tumor in the upper right quadrant of her abdomen. This tumor had been growing more rapidly than the one on the left side, especially during the last two months before the patient came to the clinic. There had been an associated soreness on the right side, which had been of a pinching character. During 1935 she had had a constant sensation of abdominal fullness and an occasional attack of nausea, but she had not vomited. Her appetite had remained good except for the last three weeks before she came to the clinic. There had been some shortness of breath and she had been cold and restless most of the time. During 1934 and 1935 she had lost about 14 pounds (6.4 Kg.). Since 1921 the patient had had "rheumatism," especially in both arms, in the back of her neck, and in the small of her back. During 1935 the "rheumatic" pains in her arms had been associated with a rather marked numbness of both hands and arms, which had been aggravated by the occasional pains in her back. There had not been any marked headache, any dizziness, or any visual disturbances.

Physical Examination.—The patient was in general good physical condition. Blood pressure 112/80. The abdomen appeared to be moderately distended. Palpation revealed a large, irregular, somewhat hard mass, the size of a grapefruit, in the left upper quadrant. Palpation of the right upper quadrant revealed a similar mass which was slightly larger. The tumors moved with respiration and when the patient changed her position. Photo-electric estimation of the hemoglobin was 13.6 Gm. R.B.C. 4,150,000, W.B.C. 9,100. The urine was normal.

Roentgenologic Examination.—The thorax and colon were normal. There were slight hypertrophic changes in the lumbar vertebrae. An intravenous urogram, which was made in order to exclude the possibility of a polycystic kidney, revealed a ptosis of the right kidney and an extrarenal shadow below and adjacent to the lower pole of the left kidney. The origin and the nature of the palpable masses were still not entirely clear. The impression remained that this might be a case of either a mesenteric or a pancreatic cyst. A retroperitoneal tumor was not thought likely because of the rather free mobility of the masses.

Operation.—An exploratory laparotomy was performed (W. W.) February 2, 1936. The kidneys and liver were normal and did not contain any cysts. The entire pancreas was filled with cysts which varied from one to three centimeters in diameter. The two masses felt in the upper quadrants of the abdomen were groups of cysts which contained colloidal material. A resection of the lateral fourth of the pancreas was carried out, and a mass of cystic and colloid material 10 to 12 cm. in diameter was removed. The remaining fourth of the pancreas, the tail, was sutured to the proximal two-fourths. The

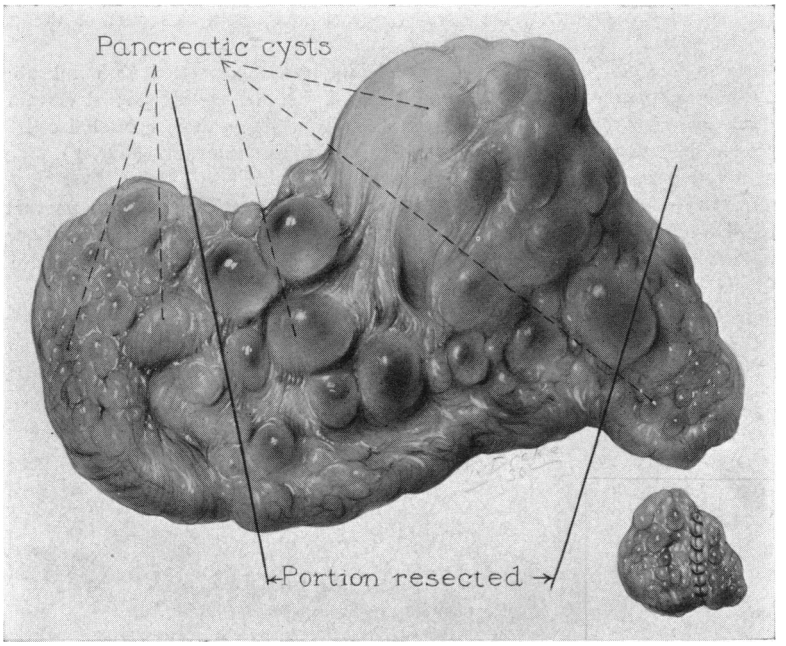

FIG. 1.—Drawing of the pancreas as seen at operation, showing the general involvement by multiple small and larger cysts. The part of the pancreas lying between oblique lines was resected. Insert shows continuity of pancreas which was effected after resection of its middle part.

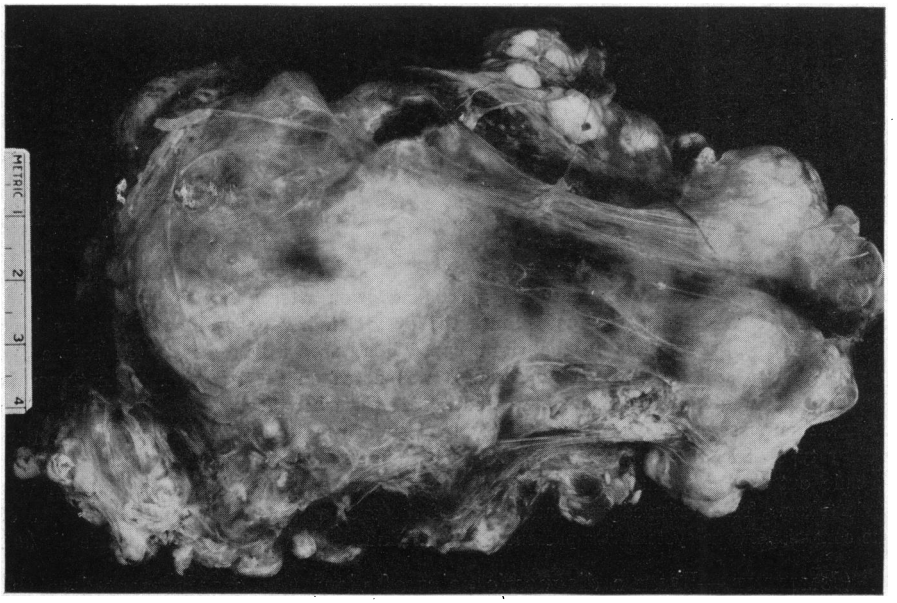

FIG. 2.—Photograph of the part of the pancreas resected.

head of the pancreas was also polycystic but, because of the anatomic relationship to important neighboring organs, the cysts (Figs. 1 and 2) in this part of the pancreas were not removed.

Microscopic Examination revealed small and large cysts filled with a hyaline substance. The walls were covered by one layer of cuboidal epithelium; at certain places the epithelium was cylindrical in character. At other places these epithelial cells formed papillary proliferations which extended into the lumen of the cysts (Fig. 3). Very few islands of Langerhans were observed.

Postoperative Course.—Following the operation the patient made an uneventful recovery. The diastase in the urine was determined daily following the operation, but at no time was there any definite increase of it. On the fourteenth postoperative day a

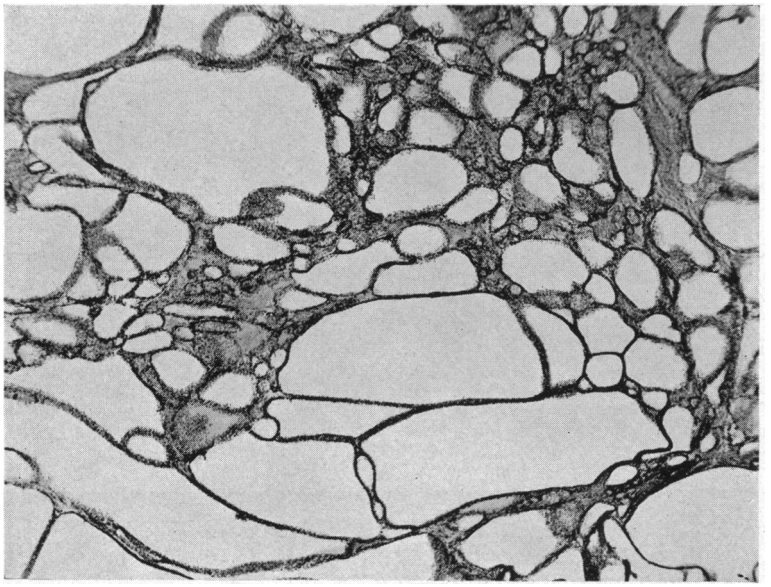

FIG. 3.—Photomicrograph showing small and large cysts, the walls of which are lined by a layer of cuboidal epithelium which forms papillary proliferations at certain points (×30).

glucose tolerance test was moderately positive and a restriction of her carbohydrate intake was advised. At the time of her dismissal, March 7, 1936, her general condition was excellent. The palpable tumor in the right upper abdominal quadrant was found to be definitely reduced in size; it was as large as a good-sized plum. The patient was advised to return to the clinic for a reëxamination after a few months.

Follow Up.—August 13, 1936: Since the operation her general condition has been satisfactory. The soreness in the right upper quadrant, which was noted before the operation, was still present, but no soreness was noted on the left side. The tumor in the right upper quadrant had not increased in size since her discharge. The examination otherwise was essentially negative. The blood sugar was normal. Because of the particular type of cysts of pancreas which had been present, it was deemed very important to rule out the possibility of Lindau's disease. A careful neurologic examination was undertaken. This included an examination of the eyes and ocular fundi. No disease of the central nervous system was present. This excluded the possibility of Lindau's disease.

COMMENT.—From a diagnostic point of view, this case presented to the clinician most of the difficulties associated with obscure tumors of the ab-

dominal cavity. It demonstrates very well that an approach to a tentative diagnosis can only be made by the application of all diagnostic skill. The value, for instance, of the excretory urogram in this case is obvious as it eliminated the possibility of polycystic kidneys. The resection of the pancreas was rather extensive. It is interesting to note that in spite of this the amount of urinary diastase did not increase postoperatively beyond normal limits. Another interesting fact was the moderately positive glucose tolerance test two weeks after the operation. In regard to the comparatively few islands of Langerhans found in the removed specimen, it may be assumed that the resected portion did not include a very active part of the pancreas, and that sufficient islands of Langerhans remained to enable the body to carry on a comparatively normal carbohydrate metabolism. Lindau mentioned that cysts of the pancreas were found in eight of 15 cases of Lindau's disease. In only two cases was there evidence of glycosuria.

The ultimate fate of the patient in the present case is doubtful. The pathologic condition must be considered congenital and the lesion no doubt required years to reach its preoperative size. It is a question whether or not this patient has reached a period of very active growth of the cysts, as seems indicated by the clinical history. Another possibility which warrants a guarded prognosis in this case is the possibility of future involvement of the central nervous system. Although at the present we have no experience as regards this possibility, it seems at least theoretically possible that the abdominal symptoms may precede the neurologic symptoms, which would make the ultimate prognosis unfavorable.

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