CYST OF THE PANCREAS ASSOCIATED WITH ECTOPIC SPLENIC ISLAND

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The first successful removal of a pancreatic cyst was reported in 1881 by Bozeman[1] before the New York Pathological Society. The next year, Gussenbauer devised the operation of marsupialization, which is still the commonest method of surgical treatment. Senn[2] reported a case successfully treated by surgery in 1885 and attempted to produce cysts experimentally by tying off the pancreatic duct. Lazarus[3] produced a cyst in the pancreas of a dog. He crushed the pancreas and caused the formation of a hæmatoma about the size of a pigeon's egg, which, after forty days, was converted into a cyst with a smooth, fibrous capsule containing 100 cubic centimetres of a watery fluid. Others attempted to produce cysts experimentally by injecting various substances into the duct of Wirsung. Opie[4] called attention to the relationship between diseases of the pancreas and obstruction to the normal outflow of bile by a stone in the common bile-duct. Eha[5] has reported a cyst the size of an orange in an infant five months old. He believed it to be a congenital cyst. Railton[6] and Shattuck[6] report similar cases in infants. Robson and Moynihan[7] refer to three cases and state that congenital cystic disease of the pancreas is exceedingly rare. The case I wish to report is especially interesting from the standpoint of etiology because of a congenital anomaly found in the pancreas post-mortem.

Case Report.—E. K., a cabinetmaker, sixty-one years of age, was admitted to the Albany Hospital December 9, 1929, into the service of Dr. A. H. Traver. His past history and family history were negative. He had always considered his health good, and his habits were temperate. Five weeks before admission he had severe pain across the upper part of his abdomen and vomited persistently for three days. He attributed this upset to some sardines that he had eaten. There was no jaundice. He had one chill but thought he had had no fever. Following this attack he had no more pain, but his appetite failed, and he felt weak. He noticed that his abdomen began slowly to increase in size. At the time of admission to the hospital his temperature and pulse were normal. He complained only of weakness and the swelling of his abdomen. There was no history of injury.

The physical examination was essentially negative except for a mass filling the upper half of the abdomen. This was smooth and was dull to percussion. No fluid wave or shifting dullness in the flanks could be made out. No definite edge could be felt. There was no apparent enlargement of the liver. The mass was not movable. There was no pulsation or bruit. A twenty-four-hour specimen of urine was negative for sugar, and each of four single specimens collected before and after operation were negative for sugar. The tests for albumen, acetone, and diacetic acid were negative. Examination of the blood showed 4,500,000 red cells and 8,600 white cells, with a normal differential. Hæmoglobin was 70 per cent. by the Tallquist scale. The blood Wassermann

was negative. Fasting blood sugar was 112 milligrams and the non-protein nitrogen was 54 milligrams per 100 cubic centimetres. X-ray examination after a barium meal showed no constrictions or filling defects. The transverse colon was displaced downward by a large rounded mass above it. The greater curvature of the stomach was indented by this same mass. The X-ray findings suggested a large mass outside the gastro-intestinal tract, probably a retroperitoneal tumor.

The pre-operative diagnosis of pancreatic cyst was based on the location of the tumor and its steady increase in size over a period of five weeks after an attack of upper abdominal pain, whose severity and radiation from right to left suggested an acute pancreatitis. In making a diagnosis, the following possibilities were considered: Retroperitoneal cyst or cyst of the mesentery, echinococcus cyst, splenomegaly, aneurism of the abdominal aorta, and ascites. Hydronephrosis or pyonephrosis seemed to be ruled out by the examination of the urine and the fact that the mass did not extend into the flank so as to fill the costovertebral angle when palpated bimanually.

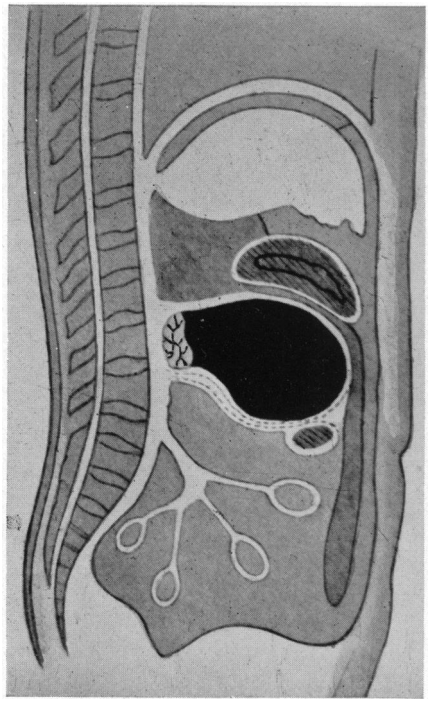

FIG. 1.—Cyst projecting forward from pancreas and presenting between stomach and colon. (From Robson and Cammidge.)

On December 11, operation was performed by Dr. A. H. Traver under ether anæsthesia. When the peritoneum was opened through an upper left rectus incision, a cystic swelling presented itself at once. It was the size of a football and protruded between the stomach and the transverse colon. There were adhesions about the gall-bladder. Palpation revealed that the cyst was separate from stomach, kidney, spleen, and liver. It was approached through the lesser peritoneal cavity by making an opening in the mesocolon. No fat necrosis was seen. The cyst was thin-walled, and fluid leaked out when an attempt was made to put in purse-string sutures, so gauze was packed about the cyst and an Ochsner's trocar was introduced. About a quart of thin, muddy fluid, which looked as if it contained old blood, was drawn into a basin; and then it was possible to enlarge the opening in the cyst and stitch it to the edge of the parietal peritoneum (*i.e.*, marsupialization). A large tube was fastened in place to drain the cyst, and fifteen ounces of fluid were collected in the first twenty-four hours. A stab wound was made at a lower point in the abdomen, and a "cigarette" wick was inserted before closing the primary incision. The fluid from the cyst was examined by Professor Arthur Knudson for amylase by the method of T. R. Brown,[8] using starch and iodine to determine the enzyme action. Only 0.8 per cent. of the amount in normal pancreatic juice was found.

The immediate post-operative recovery was good. There was profuse drainage for a week. After four or five days the tube became loose and fluid escaped around the tube in sufficient quantities to soak the dressings, the binder, and the bed linen. The skin was protected by zinc oxide ointment to prevent digestion about the wound. The patient was coöperative and took large quantities of liquids. Normal saline was given by rectum and by hypodermoclysis. When the temperature rose on the fifteenth day after operation, the wound was irrigated, and chunks of digested tissue, a cheesy material, were washed out. A week later, twenty-one days after operation, the patient died of pulmonary embolism.

Necropsy Report.—An autopsy was performed by Dr. Victor C. Jacobsen only thirty minutes after death, which was fortunate inasmuch as there are very rapid changes in the pancreas post-mortem. The important findings were: A left rectus operative incision 16 centimetres long, 4 centimetres to the left of the umbilicus. From the upper end of this wound a purulent material exuded, and the wound was partly open. To the inside of the wound the omentum was adherent, and small white areas of fat necrosis were scattered in the omentum, the anterior abdominal wall, the mesentery, and about the upper part of the abdominal cavity. There were dense adhesions about the spleen, the appendix, and the gall-bladder. Anterior to and slightly below the pancreas was a large cavity lined by an injected membrane. The cavity extended the entire length of the pancreas; it had dissected down to the pole of the left kidney, included the entire lesser peritoneal cavity, and extended into the omentum. In the lumen of the cavity was

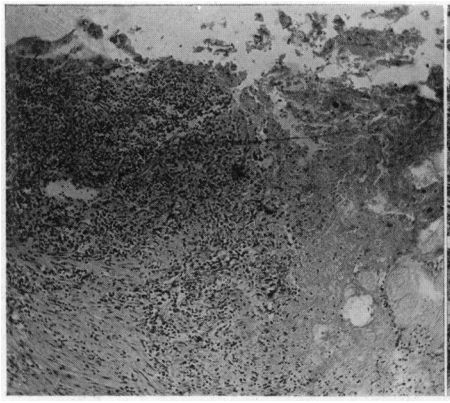

FIG. 2.—Wall of pancreatic cyst. To the left are much fibrosis and lymphocytic infiltration.

FIG. 3.—A section of the pancreas. In the central portion is an island of splenic pulp in the capsule of which are several dilated pancreatic ducts.

a grayish, caseous, soft substance, resembling necrotic fat. A sinus connected this cavity with the abdominal wound described above. The entire mass was removed and cross-sections made to show, if possible, any connection between the cavity and the pancreatic ducts, but none could be demonstrated. The larger pancreatic ducts were dilated but probing revealed no obstruction. There was considerable fat necrosis throughout the entire gland. Microscopic examination of the pancreas shows much chronic interacinar and interlobular inflammation; also dilatation of some ducts, but many are quite normal. There is hyperplasia of the lining epithelium of some ducts. In one section there is *an ectopic focus of splenic pulp entirely surrounded by pancreatic tissue.* The islands of Langerhans appear normal. In the interlobar fat and capsular fat are large areas of necrosis with moderate inflammatory reaction, but no hæmorrhage. There are cholesterin and hæmatoidin crystals and calcareous salts in some of these areas. The large tract found leading in various directions from the pancreas and down into the mesentery is lined by necrotic tissue and degenerated fat, but no definite epithelial lining is found except a few cells in one section. The conformation of the tract and the finding of ectopic splenic tissue in the pancreas suggest the possibility of some congenital anomaly of pancreatic ducts with cyst formation and subsequent rupture of the cyst, the contents dissecting in various directions and liberating fat-splitting enzymes.

The gall-bladder was markedly dilated, the bladder wall slightly thickened, and the mucosa covered with cholesterol deposits. In the lumen of the gall-bladder were about ten small cholesterol stones. The bile-ducts were markedly dilated, but patent. The right lung weighed 340 grams. There was thrombosis of nearly all branches of the pulmonary artery.

Discussion.—Pancreatic cysts are rather rare inasmuch as White[9] found only three cases in 6,078 autopsies performed at Guy's Hospital in London. They have been classified by Robson and Cammidge[10] as follows:

(1) Retention cysts, which are lined with epithelium and are caused by obstruction in the pancreatic duct, smaller ducts or acini.

(2) Proliferation cysts, due to a proliferation of glandular epithelium followed by an accumulation of fluid. These are true tumors (cystadenoma or cystic epithelioma).

(3) Congenital cysts, analogous to those found in liver, spleen, or kidneys.

(4) Hæmorrhagic cysts, due to hæmorrhagic necrosis.

(5) Hydatid cysts.

(6) Pseudocysts, produced by trauma or degenerative changes of the interstitial tissue of the pancreas. They are distinguished from true cysts in that they are not within the substance of the pancreas but are usually in the lesser omental sac.

(7) Dermoid cysts.

Contents and Location of the Cyst.—The contents of pancreatic cysts vary. The fluid is ordinarily alkaline and has a specific gravity of 1.010 to 1.020. The fluid may be perfectly clear, though usually blood is present, often having undergone marked changes to a dark chocolate or coffee-ground appearance. In the case presented above, there was evidence of old hæmorrhage. The fluid frequently contains one or more of the pancreatic ferments, and it is possible that all three may be present. However, an absence of the ferments does not in any way indicate that a cyst is not of pancreatic origin, for the ferments frequently disappear in the old cysts, often reappearing in the discharge when the cyst is drained.[11] The contents of the pseudocyst are produced by liquefaction of necrotic tissue together with a bloody and inflammatory exudate.

The cysts are frequently located between the stomach and the transverse colon or above the stomach, and less frequently between the layers of the mesocolon. However, they may occupy any part of the abdominal cavity and frequently simulate ovarian cysts.

Etiology.—The pancreas crosses the body of the first lumbar vertebra and may be injured when there is compression of the abdomen, particularly if the abdominal muscles are relaxed and the stomach is empty at the time of the accident.[12] Injury to the pancreas is frequently overlooked when there is an associated injury to other abdominal viscera. According to Heiberg,[13] in one-third of all cases of pancreatic cysts trauma was the cause of the trouble. The enlargement usually appears at once, but it may occur months later, according to Honigman.[14]

Senn classifies the causes of retention as follows:

(1) Obstruction to the outflow of the secretion from impaction of pancreatic calculi in the pancreatic duct or of biliary calculus in the ampulla of Vater.

(2) Partial or complete obliteration of a portion of the pancreatic duct from cicatricial contraction.

(3) Sudden or gradual obstruction of the duct without diminution of its lumen from displacements of the pancreas. Such a displacement may be the result of relaxation of the attachments to surrounding structures, to pressure on the gland from tumors or exudation, or to cicatricial contraction in the substance of the gland.

Hæmorrhage seems to be an important etiologic factor in many cases (Warnock,[15] Lloyd[16]), as a hæmatoma has been found in the substance of the pancreas during exploration of a pancreatic cyst. Many of the pseudocysts, according to Lloyd, are fluid effusions into the lesser peritoneal sac, the result of injury to the underlying pancreas, and not cysts of the pancreas in the proper meaning of the term. Other cases seem to follow an acute pancreatitis, which in turn may be due to a plugging of the ampulla of Vater with a gall-stone and the back flow of bile into the pancreas.[4] Experimentally, however, Mann and Giordano[17] have found it impossible to produce a pancreatitis by such means unless bile is injected directly into the duct of Wirsung under considerable pressure, and then they assume the inflammation and subsequent cyst formation are due to rupturing of some of the small ducts and an escape of pancreatic fluid rather than bile alone.

Diagnosis.—The diagnosis can be made only when the cyst has attained considerable size. Pain is not a constant symptom unless it comes from pressure or from associated conditions. In non-traumatic cases the history may suggest an acute or chronic pancreatitis or a biliary colic preceding the development of the cystic tumor. Frequently, the patient complains only of the increasing swelling of the abdomen with loss of appetite. There may be large fatty stools, and there may be sugar in the urine, but these findings are often absent if the pancreatic function is maintained.

Physical examination reveals a rounded, firm, smooth tumor of varying size in the epigastrium. It moves with respiration and is separate from liver, spleen, or kidney. The tumor may be fluctuant and may transmit the pulsations of the abdominal aorta. It is not usually tender. The stomach can usually be made out by percussion lying above the tumor and to the left, and sometimes the transverse colon can be made out passing anterior to it. When the cyst fills the abdomen, there may be a fluid wave, but there should be no shifting dullness in the flanks. A rectal or vaginal examination should help to distinguish between pancreatic cyst and ovarian cyst.

Treatment.—The treatment of these cases is always surgical and is most often simple drainage after drawing the cyst up into the wound and stitching it to the parietal peritoneum at the edge of the incision (*i.e.*, marsupialization). In some cases the operation has been done in two stages, but this is not often necessary. Aspiration by means of a trocar or needle inserted through the intact abdominal wall is no longer considered good surgery as the stomach or transverse colon may be compressed and intervene. Excision of a part or the whole of the cyst would seem advisable if this were possible

without producing an extensive hæmorrhage. Prolonged drainage is indicated.[18] The mortality of such operations is from 20 to 30 per cent. A few patients require a secondary operation because the cyst refills or because of malignant degeneration. Occasionally, the sinus formed after an operation refuses to heal; and when a total loss of pancreatic secretion occurs, this is a serious matter producing dehydration, emaciation, extreme weakness, a diarrhœa with large fatty stools, and finally death. Radium has been used with good results in a few cases to promote fibrosis and healing when there is a persistent sinus.[19]

SUMMARY

(1) A case of cyst of the pancreas is reported in a man of sixty-one years, who died twenty-one days after operation, of pulmonary embolism.

(2) The cyst had ruptured, the contents causing widespread fat necrosis. The lining of the cyst suggested an origin from pancreatic ducts, with possibly a congenital anomaly of ducts as a basis for the condition. This hypothesis is supported by the presence of another anomaly, splenic tissue within the pancreas.

(3) Other theories of etiology are discussed.

(4) Treatment consists of early operation with prolonged drainage. Complications are frequent.

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