globulin was then administered in doses of 0.1 g. per kg. body weight every six weeks. On May 14, 1955, four weeks after an injection of gamma globulin, the first patient was readmitted to hospital with a septic arthritis of the left knee joint. Fluid aspirated from this joint grew a pure culture of *Staphylococcus aureus hæmolyticus*. He again recovered promptly. Apart from this episode, both boys have been entirely free from clinical infections for a period of six months.

The family history is significant in that an only uncle on the maternal side died at the age of five years (1935). Autopsy records reveal the cause of death as being incompletely resolved lobar pneumonia, fibrino-purulent pericarditis and plastic peritonitis. With the exception of the above-noted cases, review of the family history for three generations revealed only good health and longevity, in all siblings, both male and female, on both maternal and paternal sides.

DISCUSSION

The findings in our cases conformed with those already reported in the literature. The history and autopsy findings in the maternal uncle strongly suggest that he too was afflicted, giving further support to the theory that the defect is a recessive sex-linked characteristic similar to hæmophilia. The modus operandi of this defect is receiving considerable study. Many workers believe that immature plasma cells and lymphoblasts participate in the formation of antibodies, i.e. gamma globulins, and that the absence of the latter substances may be accounted for by the observed failure of those cells to react normally to antigenic stimulation.

Absence of isohæmagglutinins for appropriate blood groups or a positive Schick test after adequate immunization are good screening tests in suspected cases. Early recognition of this condition is imperative since effective replacement therapy with gamma globulin is now available. Further experience will show whether the minimum recommended dosage of 0.1 g. per kg. of body weight every six weeks might be inadequate in some cases, as in Case 1.

SUMMARY

1. Two cases of primary or congenital agammaglobulinæmia have been described. They are characterized by recurrent infectious processes, absence of plasma gamma globulin, absence of isohæmagglutinins, and the inability to produce normal immunological response to immunization.

2. The condition is a serious and potentially fatal defect which, if diagnosed and treated, appears to be compatible with normal life.

3. The treatment consists of administration of purified gamma globulin in doses of 0.1 g. per kg. of body weight every six weeks, although this may prove inadequate.

We are indebted to Dr. S. Hanson, Director of Laboratories, Edmonton General Hospital, for his academic and technical assistance in establishing the diagnosis and to Dr. Robert A. Good, University of Minnesota Medical School, for confirming the diagnosis.

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PAPILLARY CYSTADENOCARCINOMA OF THE PANCREAS*

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THE PURPOSE of this paper is to report a case of papillary cystadenocarcinoma of the pancreas treated successfully by surgical excision, and to review briefly the literature on this subject.

Mrs. N., aged 47 years, was admitted to the Montreal General Hospital on July 21, 1949. One month before admission she had discovered a firm rounded mass the size of a grapefruit in the left upper quadrant of the abdomen. There had been no abdominal discomfort of any kind and no symptoms referable to the gastrointestinal or urinary tracts.

On examination, she appeared healthy. The cardiovascular and respiratory systems were normal. Examination of the abdomen showed the spleen to be enlarged, the lower border being one inch below the left costal margin.

A firm, rounded mass about 5 inches (12.5 cm.) in diameter was palpable in the left upper quadrant of the abdomen. This mass was quite mobile in a lateral direction, but only slightly so in a vertical direction. There was no tenderness, and the temperature was normal.

Laboratory data.—Urea nitrogen 17 mg. per 100 c.c.; blood sugar (fasting) 0.108 g. per 100 c.c.; bilirubin 0.2 mg. per 100 c.c.

Hæmogram.—Erythrocytes 4,370,000; white cells 5,750; Hb 74% (11.5 g.); red cell diameter 7.3 μ; red cell volume 41%; erythrocyte sedimentation rate 40 in one hour; variation in size, shape, and staining of erythrocytes.

X-ray studies.—A flat plate of the abdomen demonstrated a large ovoid area of increased density in the region of the lower pole of the left kidney.

*From the Surgical Service of the Montreal General Hospital.

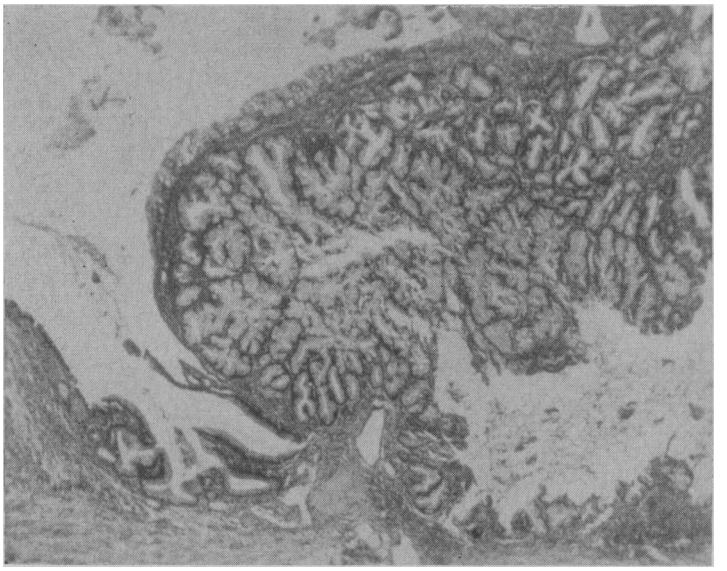

Fig. 1.—A portion of cyst wall with papillary in-growth from more simple lining. × 35.

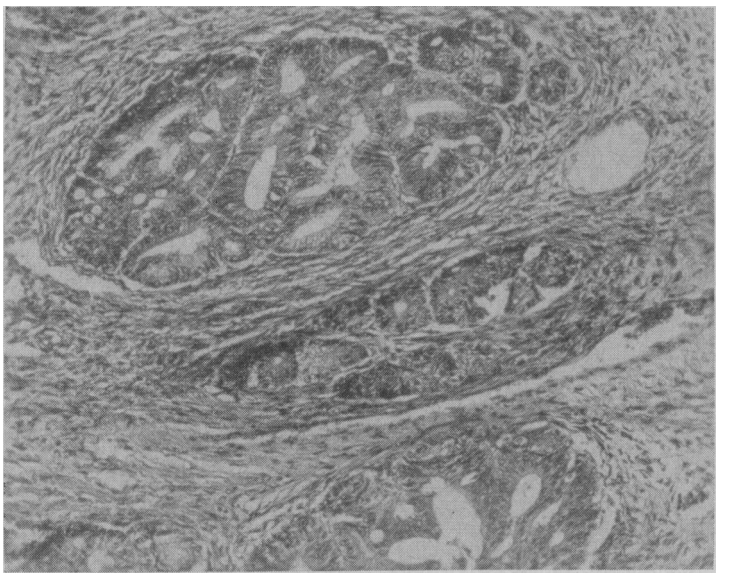

Fig. 2.—Atypical gland structures infiltrating deeply within the fibrous wall of the cyst. × 100.

An intravenous pyelogram showed a marked increase in the size of the calyceal pattern on the left side as compared with the right. However, the ovoid area of increased density seen in the previous films was now found to overlie the lower pole of the left kidney, from which it was distinctly separate.

Diagnosis.—Because of the mobility of the mass and the absence of any gastrointestinal symptoms, a clinical diagnosis of mesenteric cyst was made.

Operation.—On July 25, 1949, under general anæsthesia, the abdomen was explored through a transverse incision on the left side at a level one inch (2.5 cm.) above the umbilicus. A large mass was found between the stomach and the transverse colon. The gastro-colic omentum was incised and the mass was then seen to be a cyst springing from the tail of the pancreas. This measured 5 inches in diameter and 6 inches in an antero-posterior direction. The cyst had a thick fibrous wall which was densely adherent to the hilum of the spleen. The splenic vein was enormously dilated as a result of compression by the cyst, and the spleen was grossly enlarged due to passive congestion. The liver was not enlarged and its surface smooth. There was no enlargement of the regional lymph nodes.

The cyst was enucleated without difficulty from the tail of the pancreas, the splenic artery and vein were ligated, and the spleen was then removed together with the cyst. The rent in the gastro-colic omentum was repaired, a Penrose drain inserted and the abdomen closed.

Convalescence was uneventful and the patient was discharged from hospital on August 10, 1949. Though she has since required treatment from time to time for the anæmia, her general health has remained good and there has been no evidence of any recurrence of malignancy.

Pathology.—Gross examination showed a large cystic mass having a slightly lobulated appearance. The wall of the cyst was thick and fibrous. When opened, the cyst was found to be made up of a number of communicating cysts of varying size containing a brownish mucoid material. The lining of the cyst varied greatly. In some areas it was of a dull white colour and devoid of epithelium. Other areas were lined with an epithelium granular in appearance and definitely hæmorrhagic. In still other areas the lining epithelium exhibited a marked papillary appearance.

Microscopic study of sections taken from different portions of the specimen showed this to be a multicystic, mucus-secreting columnar cell papillary cystadenocarcinoma of the pancreas of low-grade malignancy.

It is customary to classify neoplastic or proliferative cysts of the pancreas into three general types: (a) cystadenoma, (b) cystadenocarcinoma, (c) teratomatous cysts.

The cystadenoma is a benign lesion but there is considerable clinical and pathological evidence that it may undergo a malignant transformation.[1, 4, 5]

Cystadenocarcinoma is the malignant variant of the cystadenoma, and whereas the cystadenoma may occasionally show papillary infoldings and projections of the lining epithelium, this papillary tendency is almost invariably present in the cystadenocarcinoma.

INCIDENCE

As compared with pseudo-cysts and retention cysts of the pancreas, the cystadenoma is extremely rare and the cystadenocarcinoma even more so.

It is almost impossible to obtain definite information as to the incidence of these lesions or the actual number of cases on record because of the confusion which exists in the literature.

For example, Kennard[3] reported 25 cases of cystadenocarcinoma which he had collected from the literature, but this included 12 cases which had previously been excluded by Lichtenstein[4] because of inconclusive data. In 1952, Sawyer, Spencer and Lubchenco[6] collected 47 cases of cystadenoma and 29 cases of cystadenocarcinoma from the literature and reported an additional case of each variety. Later in the same year Burk and Hill[1] reported a case of cystadenocarcinoma.

The case reported here is the first proven cystadenocarcinoma of the pancreas in more than 300,000 admissions to the Montreal General Hospital. It brings the total of reported cases to 32.

ETIOLOGY

The etiology of neoplastic cysts has been presented in some detail by Jemerin and Samuels[2] and it would seem unnecessary to review this again.

CHARACTER

These cysts do not become as large as pseudocysts, but several have been reported as being 15 cm. in diameter. Characteristically they present a lobulated appearance and when opened are seen to be multilocular. The contents vary from a thin clear or hæmorrhagic fluid to a mucoid or gelatinous material.

Both cystadenoma and cystadenocarcinoma occur most commonly in the tail and only rarely involve the head of the pancreas. With increase in size they usually present between the stomach and transverse colon.

SYMPTOMS

Symptoms when present occur as a result of pressure on surrounding structures. Pain is the chief symptom and is usually located in the left upper quadrant of the abdomen. Occasionally it may be referred through to the back. Pressure on the stomach may cause discomfort after meals and a feeling of nausea. Not uncommonly there are no symptoms, the lesion being discovered during routine physical examination by the finding of a rounded, somewhat movable mass in the upper abdomen.

DIAGNOSIS

Intravenous or retrograde pyelography will be of assistance in ruling out lesions of the kidney. A barium meal may be of value by revealing extrinsic pressure on the stomach. However, it is usually impossible to differentiate proliferative cysts of the pancreas from mesenteric or omental cysts, and a correct diagnosis can be established only by surgical exploration of the abdomen.

TREATMENT

Total excision is obviously the treatment of choice not only for the cystadenocarcinoma but also for the cystadenoma because of its reputation for malignant degeneration.

It is fortunate that these cysts almost invariably arise from the tail of the pancreas since this makes them more amenable to surgery. It is also fortunate that, unlike pseudo-cysts, the proliferative cysts are non-inflammatory lesions and therefore are not densely adherent to surrounding structures. In some cases splenectomy may be necessary because of involvement of the splenic vein.

SUMMARY

1. The literature on proliferative cysts of the pancreas is reviewed briefly.

2. A case of papillary cystadenocarcinoma of the pancreas with successful surgical excision and a six-year survival is presented.

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CONSULTANT BY DECREE

"The National Health Service Act abolished the conception of the apprentice or the beginner. In 1948 the Minister told the country that henceforth everybody could get treatment by a specialist. This was another way of saying that everybody would be called a specialist, and not merely a specialist but a consultant, one fit to be consulted. In this brave new world the surgical trainee, if he is fortunate, is summoned one day to a board meeting as a registrar, and leaves it ten minutes later as a consultant. Having treated his wife to the best dinner he can buy, and treated himself to a better car than he can afford, he proceeds without delay to invent some new operations. He may not be very clever, and he may not have studied very deeply, but he can hardly fail to think out a new operation or a modification of an old one."—Sir Heneage Ogilvie, *Lancet*, 1: 115, 1956.