ANNULAR PANCREAS: REVIEW AND CASE REPORT

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Annular pancreas is a rare congenital anomaly of some surgical importance. Although well over 100 cases have been reported since 1818 when it was first described by Tiederman,[1] more interest has been shown in the condition in recent years and most of the cases treated surgically have been described in the last decade. At first regarded as an anatomical curiosity sometimes with pathological consequences, it has now become a condition which can be diagnosed preoperatively and satisfactorily treated. The condition has been described at all ages, ranging from a 7-week embryo[2] to an elderly man of 74.[3] A large proportion of cases have no symptoms at any time: on the other hand it may present in the newborn as a cause of high intestinal obstruction which requires immediate surgery. Most of the reported cases have been in adults. It is interesting to observe that this anomaly, having existed since birth, in many cases causes symptoms only after many years.

Associated pathological conditions have been described in a minority of cases requiring surgery. Of these co-existing conditions the more important have been chronic pancreatitis, biliary diseases and peptic ulceration. Theorectically, both pancreatic and biliary disease could be explained by stasis in the respective duct systems caused by obstruction. However, in the case here presented the gall-bladder condition appears on X-ray findings to be unrelated to obstruction of the common bile-duct by the annulus. It is possible that chronic duodenal obstruction at the level of the annulus may disturb the acid-base relationship at the pylorus and so predispose to peptic ulceration.

A high incidence (25%) of other congenital abnormalities such as cardiac anomalies, aplasia of kidney, duodenal atresia and accessory pancreas is found together with annular pancreas.

Anatomy and Pathology

The annulus surrounds the second part of the duodenum, extending from the head of the pancreas (Fig. 1). It is composed of normal pancreatic tissue. The pancreatic ring may be deficient anteriorly, and in some cases the ring may be completed by fibrous tissue. It has its own duct, which commences anteriorly next to the head of the pancreas and then runs to the right and around the duodenum to end up posteriorly, where it usually terminates in the main pancreatic duct. There may also be actual stenosis and thickening of the portion of the duodenum that is surrounded by pancreatic tissue. If obstruction is present it may lead to dilatation

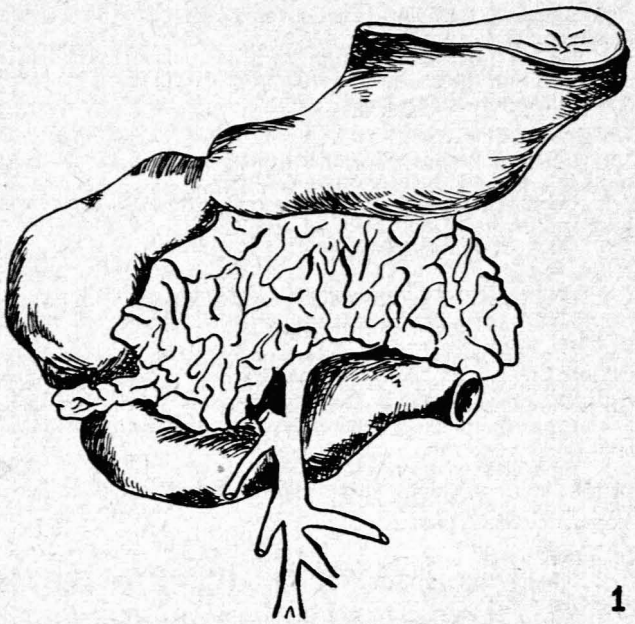

Fig. 1. Sketch illustrating pylorus and duodenum with the annulus surrounding the second part. An attempt has been made to illustrate the dilatation of the first part of the duodenum.

of the first part of the duodenum and occasionally of the stomach also.

Embryology

It is of interest to note that the annular pancreas is a normal finding in birds. In man the pancreas develops from a dorsal and ventral outgrowth from the primitive duodenal wall. The dorsal pancreas forms the body and tail of the adult gland. Rotation and unequal growth of the duodenal wall results in a rotation of the ventral pancreas with its duct to the right and then posteriorly, to fuse with the dorsal portion and form the head of the adult pancreas. With the fusion of the two parts of the foetal pancreas an anastomosis between the ducts occurs and, although the ventral pancreas forms only a small part of the adult gland, its duct becomes the main pancreatic duct. A most likely explanation for the development of this anomaly is a failure of the ventral anlage of the pancreas to rotate with the duodenum. As the proximal end of the duct rotates to the right and posteriorly, a band of pancreatic tissue is drawn circumferentially around the duodenum. This theory is in keeping with the described course of the annular duct and the fact that the duct normally empties into the main pancreatic duct.

Clinical Findings

The clinical features are often coloured by the symptoms and signs of related pathological conditions. Otherwise most of the patients present with obstructive symptoms of insidious onset and lasting over a period of years. Bouts of nausea and vomiting after meals and epigastric pain, which may be severe, are the presenting symptoms. The pain is not relieved by alkalis. Jaundice may, rarely, be a feature. On examination there may be epigastric tenderness only and the annulus is not palpable. Efforts to establish a diagnostic syndrome have been disappointing.[4] Clinical examination is of little help except to exclude other conditions.

Diagnosis

The condition is frequently missed, because the symptoms are not typical of any particular disease and because, owing to its rarity, the condition is not thought of. The most valuable pre-operative evidence is given by the radiologist, who also must be aware of the condition in order to diagnose it. Further, it may be missed at operation as well, if the surgeon does not bear it in mind.

Radiological Features

The lesion can be radiologically detected only by barium meal examination, and the most important and constant feature is the narrowing which is seen in the second part of the duodenum. As the narrowing is due to an external pressure, there is no mucosal irregularity and the contours are smooth. It is usually more marked on the lateral aspect of the duodenum than on the medial aspect, but may occasionally be equally prominent on the two sides. The point of obstruction may, on rare occasions, be obscured by the overhanging distended duodenal bulb.[5] The narrowing usually affects only a very small segment of duodenum, most of the described cases being 1-3 cm. in longitudinal extent.

Proximal to the constricted portion of the duodenum varying degrees of obstructive dilatation occur. The obstruction may be almost complete, with a large food residue remaining in the stomach after many hours,[6,7] or it may cause only slight hold-up, or even none.

The duodenum distal to the constriction is usually described as being of reduced lumen, but it may even be markedly distended.[4,8]

The second part of the duodenum is retracted medially as though hooked slightly to the right by the encircling finger of the pancreas.

Associated peptic ulcer in the duodenum, and less frequently in the stomach, may also be detected during the examination (18 out of 56 cases reported—Whelan and Hamilton[5]).

In the differential diagnosis the main diseases to be considered are duodenal polyps, post-bulbar ulcers, congenital bands, and malignant tumours. Lehman[8] and Whelan and Hamilton[5] describe cases which on barium examination were first thought to be simple duodenal tumours, and it may be impossible to distinguish the two conditions pre-operatively if there is no sign of obstruction. As a small polyp up to about 1 cm. in diameter is unlikely to cause any hold-up of duodenal contents, any obstructive signs are thought to favour the diagnosis of annular pancreas.

Of the congenital obstructing agents the cholecystoduodeno-colic band is the one which has caused confusion, and Whelan and Hamilton describe a case in which this condition was erroneously diagnosed as annular pancreas. These bands, however, usually obstruct the first part of the duodenum, whereas annular pancreas always affects the second part. They may of course be indistinguishable without direct visualization.

Malignant tumours of the duodenum or invading the duodenum from periduodenal tissue often cause narrowing of the lumen, but in these diseases the destruction of the mucosa and irregularity of contour are early radiological features and will readily distinguish the malignant from the benign.

Post-bulbar ulcer may cause an appearance almost indistinguishable from that of annular pancreas owing to the asymmetrical narrowing of the duodenum caused by spasm

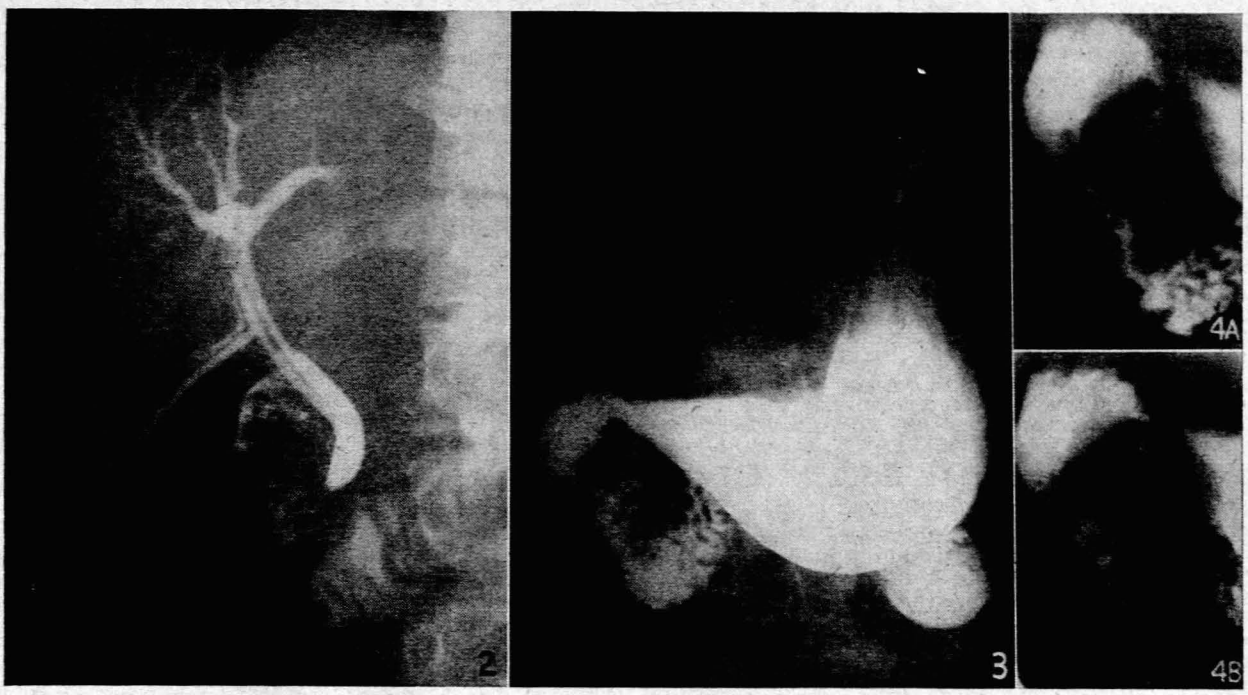

Fig. 2. The T-tube cholangiogram done after the duodenal short-circuit operation. The common bile-duct is unobstructed. The contrast medium outlines the mucosa of the second and third parts of the duodenum. At the proximal end of that part of the duodenum which is outlined, the impression of the pancreatic annulus is seen. It is obviously not affecting the common bile-duct.
Fig. 3. The appearances on barium meal after operation. The constriction of the second part of the duodenum by the annulus is clearly demonstrated; some hold up of barium proximal to the constriction is present. The short-circuit loop of the duodenum which by-passes the constriction is also shown. The medial displacement of the second part of the duodenum is due partly to the anomaly and partly to the operation.
Fig. 4. Serial spot films of the second part of the duodenum showing the constriction, with hold-up of barium proximally, and intact mucosa.

in the wall opposite the ulcer. If a crater is seen the diagnosis is obvious; co-existent post-bulbar ulcer and annular pancreas have not yet been described.[9] But a crater may not be visible, although spasm is present in almost every case of post-bulbar ulcer. Kaufman and Levene[10] publish illustrations of post-bulbar ulcers which on barium meal resemble the appearance of annular pancreas very closely.

The diagnosis in these cases may have to depend on secondary manifestation of ulceration, such as irritability of the bulb and absence of signs of obstruction. Ball, Segal and Golden (quoted by Kaufman and Levene[10]) state that all their cases of post-bulbar ulcer showed irritability of the bulb.

Treatment

Surgery affords the only relief and the procedure undertaken will depend on associated conditions if present. The most attractive procedure at first thought is a *direct attack* on the annulus, namely, division and resection of a portion of the ring, but in the earlier cases described this led to fistula formation in some, and a failure to relieve the obstruction in others. The former complication may be overcome by remembering the anatomy and commencing the resection anteriorly and as close to the head of the pancreas as possible. This ensures that the duct will be resected near its origin so that little or no secreting tissue will be distal to the point of division. Furthermore, the use of non-absorbable sutures will also diminish the chance of fistula formation. Before considering the operation as completed it is necessary to inspect the affected duodenal segment in order to eliminate the possibility of stenosis, and it may be advisable to open the duodenum proximally and palpate the size of the lumen. If stenosis exists it can be overcome by incising the duodenal wall longitudinally and closing the lumen in the transverse diameter in the Heineke-Mukulicz pyloroplasty. When considering all that this entails and the obvious hazards, we feel that a simpler procedure is to be preferred.

Posterior gastro-enterostomy has been employed. An objection to this procedure is that the operation does not necessarily relieve the distention of the duodenum, nor consequently the tendency to pain and accasional vomiting. Furthermore, the procedure exposes the patient to the danger of stomal ulceration.

Polya gastrectomy is effective in relieving the obstruction but obviously should only be performed when there is an associated peptic ulcer. The duodenum is resected just proximal to the annulus which need not be disturbed.

Retrocolic duodeno-jejunostomy is a relatively simple and safe procedure and appears to be free from complications. It has so far been the most satisfactory procedure in the cases reported.

If jaundice has been present or suspected, the common bile-duct should be explored and any stenosis of the duct dilated. An indwelling T-tube should be left in for drainage.

Finally in the most recent cases reported,[4] including the case we are now reporting, *duodeno-duodenostomy* has been performed with good results. After mobilizing the duodenal loop the anterior wall of the distended duodenum proximal to the annulus is anastomosed to the anterior wall of the duodenum distal to the obstruction.

CASE REPORT

An adult European male aged 38, occupation physiotherapist, was admitted to the Johannesburg General Hospital in August

1957 for investigation and treatment of epigastric pain. The patient had had bouts of pain since the age of 12 years. These attacks were usually severe, and used to occur about once a day, with occasional periods of relief. The attacks were not related to meals, nor were they relieved by alkalis. He generally felt nauseous, but vomiting was only an occasional feature. Over the years he was treated by many doctors, and was eventually labelled a neurotic. At the age of 18 he underwent appendicectomy for 'chronic appendicitis'.

In 1943, while serving in the South African Army in East Africa he developed severe abdominal pain associated this time with profuse vomiting. At this time also, he was slightly jaundiced. He was diagnosed as a case of malaria and was treated for this.

In 1947 a cholecystogram showed stones in the gall-bladder. His attacks of pain, vomiting and slight jaundice were attributed to cholelithiasis, and for this he underwent a cholecystectomy.

Between 1955 and 1957 he again underwent numerous investigations for recurrent upper abdominal pain and vomiting. He was losing weight and was contemplating suicide. These attacks of pain were associated with slight jaundice and were similar to those occurring before the cholecystectomy. The pain was not related to meals, was again epigastric in situation, radiating through to the back.

When examined on admission the patient presented as a well-built adult male in no obvious distress. He was not clinically jaundiced or anaemic. He bore the scar of his previous Kocher incision and the lower abdominal scar of the previous appendicectomy incision. The abdomen was soft, was not distended, and was tender to deep palpation in the epigastrium. No abnormal masses were palpable. His other systems were normal. His blood pressure was 130/84 mm. Hg.

The following investigations were made:

Liver Function Tests showed some abnormality indicating slight parenchymatous dysfunction without an obstructive element. The results were as follows (the normal values are shown in brackets):

Thymol turbidity, 2·0 (0-2). Thymol flocculation, negative (negative). Colloidal red, + (negative). Cephalin cholesterol flocculation (24-hour reading), ++ (negative). Taka Ara (Ucko's modification) (negative) (negative). Zinc sulphate turbidity, 15·8 (<12·5). Total lipid, 491 (500-700 mg. %). Alkaline phosphates (King Armstrong), 6·8 (4-13). van den Bergh, delayed direct (negative). Bilirubin, direct, 0·3 (0·2 mg. %). Bilirubin, total, 1·5 (1·2 mg. %). Total protein, 7·1 (5·6-8·5 g. %). Albumin, 3·5 (4·3-5·7 g. %). Globulin, 3·6 (1·3-3·0 g. %). Gamma globulin, 1·27 (0·6-1·25 g. %). Cholinesterase, 86% of the average normal activity.

Urine contained urobilin +++ and no bile.

Full blood count normal. Prothrombin index normal.

Intravenous Cholangiogram: Normal calibre of duct, no stones or stenosis evident.

Blood urea, 21 mg. %.

Intravenous pyelogram normal.

Serum amylase 28 Street-Close units (normal 8-38).

X-ray of chest normal.

Barium Meal. Pre-operative barium meal was said to be normal, but details were not available (report or films not traced).

Operation

On 30 July 1957 under general anaesthetic the abdomen was opened *via* a right subcostal (Kocher's) incision. Numerous upper abdominal adhesions from the previous cholecystectomy were encountered, making the initial dissection difficult. The common bile-duct was perhaps slightly dilated. The liver, stomach and first part of the duodenum appeared normal. The annulus was not seen at this stage. The common bile-duct was opened and explored with bougies. No stones or stenosis were encountered, the bougies passing freely into the duodenum.

It was then decided to expose and palpate the head of the pancreas and ampulla. The hepatic flexure was mobilized and it was then that an annular pancreas was found encircling the second part of the duodenum. It was a complete ring of pancreatic tissue. The duodenum proximal to the annulus appeared more dilated than that distal to it.

The duodenum was mobilized by the Kocher manoeuvre, i.e. an incision around its convexity and brought forwards. The annulus was not disturbed and a duodeno-duodenostomy was performed anterior to the annulus. A T-tube was inserted into the common bile-duct and the abdomen was then closed.

Recovery from the operation was uneventful.

A cholangiogram through the T-tube was performed on 12 August 1957 (Fig. 2). This shows completely normal hepatic duct and common bile-duct with no obstruction to the flow of contrast medium from common duct to duodenum. The proximal limit of the contrast medium outlines the duodenal constriction resulting from the pancreatic annulus and this is seen to be about 2 inches proximal to the ampulla of Vater. It is evident from this that the biliary disease cannot be explained solely on the basis of ampullary obstruction.

The T-tube was removed 4 days later and the patient left hospital completely free of pain. He has since remained in good health.

A post-operative barium-meal examination was done on 4 November 1957 (Figs. 3 and 4). This showed that the stomach was of normal size and appearance. The duodenal cap was not distended but its normal outline had been altered by the operation. From the duodenal cap the barium passed *via* two routes, firstly by the short-circuit loop, and secondly by the normal route to the second part of the duodenum, where it was held up by a very narrow constriction through which it passed with some difficulty. The duodenal indentation was present both medially and laterally but was more marked on the lateral side. There was no mucosal destruction. The medial displacement of the duodenum, said to occur at the level of the annulus, was seen but must be partly accounted for by the operation.

SUMMARY

1. A case of annular pancreas is described in which the diagnosis was made at operation. It had been missed pre-operatively in spite of numerous investigations extending over many years.

2. The embryology, anatomy and pathology are described.

3. Pre-operative diagnosis depends mainly on radiology. The differential X-ray diagnosis from such conditions as duodenal polyps, post-bulbar ulcers of duodenum, congenital bands and malignant tumours is discussed.

4. The treatment carried out in this case was duodeno-duodenostomy; other operations are discussed.

5. Associated biliary disease was present. Post-operative T-tube cholangiogram showed that there had been no obstruction to the common bile-duct to account for this.

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