HAEMORRHAGE IN AN ENTEROGENOUS CYST OF THE DUODENUM

H. J. DU TOIT, Dip. Surg., F.R.C.S. (EDIN.) and J. W. PONT, F.R.C.S. (EDIN.)

Department of Surgery, Baragwanath Hospital, Johannesburg

The anomaly of an enterogenous cyst of the intestine is a well-recognized entity. In the duodenum, however, it is of rare occurrence. Since the first description of a case of enterogenous cyst of the duodenum by Sanger and Klopp[1] in 1880, only 25 cases have been described in the world medical literature. Of these, only 2 have presented the rare complication of intracystic haemorrhage.

These cysts of the duodenum usually manifest themselves during the first decade of life by symptoms resulting from duodenal obstruction. The clinical picture therefore closely resembles that of congenital hypertrophic pyloric stenosis. The present case is reported on account of the rarity of the condition and the unusual features of the case.

CASE REPORT

M.T., an 8-year-old Bapedi girl, was admitted to hospital on 24 February 1958, complaining of recurrent cramp-like upper abdominal pain associated with bouts of vomiting for 3 days. Her mother reported that at the age of 11 months the child had been taken to hospital because of repeated vomiting. At that time an upper abdominal mass had been noted. No operation was performed. At the age of 2 years the child had been readmitted to hospital, again because of vomiting, and an operation was performed. A cyst the size of an orange had been noted to the right of the second part of the duodenum, and the surgeon had performed an antecolic gastro-enterostomy.

Since the time of operation the child had suffered from monthly attacks of pain and vomiting, but the mother had never noticed any evidence of melaena stools or the passage of blood per rectum.

On examination the patient was found to be well-nourished and normally hydrated. The abdomen presented with a right-sided paramedian scar overlying a rounded mass, the size of a large orange, which on clinical examination could not be dissociated from the liver. Increased peristaltic sounds were heard, coinciding with cramp-like abdominal pain. No melaena stools were observed. On roentgenographic investigation no calcification or fluid levels were noted.

Observation for 48 hours revealed no essential change in the patient's good general condition. Early on the 3rd day the local signs changed somewhat; the mass increased in size, it became tender to palpation, and pyrexia developed. Laparotomy was decided on.

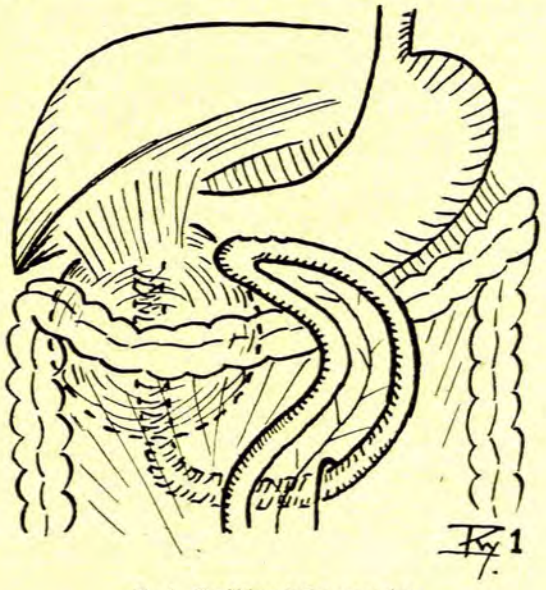

Fig. 1. Conditions before operation.

At operation the stoma of the previous antecolic gastro-enterostomy was found to be normal and no dilated loops of bowel were observed. The hepatic flexure of the colon was adherent to, and stretched over, the anterior surface of a swelling, the size of an orange, and overlying the second part of the duodenum. Multiple dense adhesions anchored the superior surface of the mass to the inferior surface of the liver (Fig. 1).

The mass was then mobilized from the right-hand side, and during dissection a small perforation occurred in the lateral wall, from which black blood and clot escaped. Further dissection showed that the cyst was intimately associated with the antero-lateral wall of the second part of the duodenum. This part of the duodenal wall was oedematous, and there was blood clot adherent to it, the region having an appearance and consistency very suggestive of a bleeding peptic ulcer with clot in the crater. No communication between the cyst and the lumen of the duodenum could be demonstrated. Further exploration and attempts at a curative procedure were precluded by deterioration in the patient's condition, and we were advised by the anaesthetist to abandon further surgery. A section of the wall of the cyst, which had by that time contracted down to one-quarter of its original size, was excised for histological examination. The ulcer-crater was covered and oversewn with the remaining walls of the cyst. The adjacent pyloric antrum was sutured to the anterior wall of the cyst to cover this suture-line. The conditions after the operation are shown in Fig. 2. Post-operative recovery was uneventful.

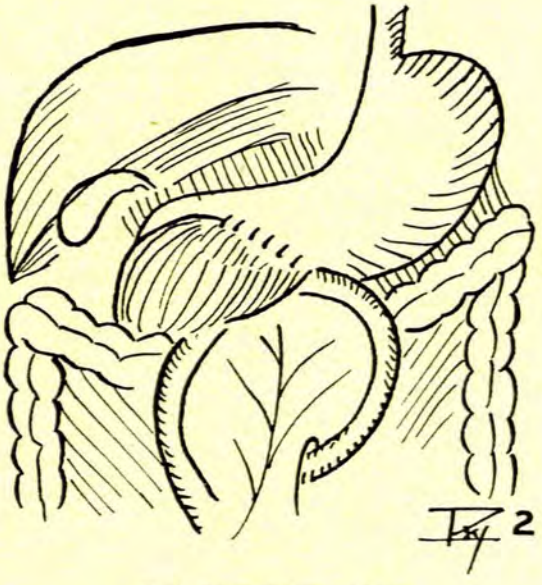

Fig. 2. Post-operative conditions.

Histological report. 'Section of this tissue from the cyst wall submitted shows that it is composed of bundles and strands of smooth muscle running in various directions. The stroma is loose and contains ganglionic nervous tissue elements and a few iron-laden macrophages. The whole area is infiltrated by lymphocytes. Some of the vessels contain thrombi. A small surface covered by histiocytes is observed. No epithelium is observed. The features are those of recent haemorrhage, and are consistent with those of an enterogenous cyst.'

DISCUSSION

During the original admission this case presented classical features, and received recognized treatment. The subsequent course, however, was unusual in that some element of obstruction remained, and an acute episode occurred 6 years after operation. These acute symptoms were undoubtedly due to haemorrhage into the cyst, causing local pressure, because no communication existed between the cyst and the duodenal lumen.

An analysis of the literature brought the following interesting information to light:

Sanger and Klopp,[1] in 1880, reported the first case of an enterogenous cyst of the duodenum, in a newborn infant who had died during a difficult delivery. At autopsy it was found to have transposition of the viscera and 5 cysts were present, viz. one of an accessory liver, an accessory bile duct, and 3 intestinal cysts, one of which arose from the duodenum and was the size of a walnut.

Roth,[2] in 1881, reported the case of a newborn male infant who had died a few hours after birth and was found to have suffered from a large thin-walled enterogenous cyst of the duodenum. A similar cyst was observed in the posterior mediastinum close to the oesophagus. The cyst did not communicate with the duodenal lumen.

In 1919 Meyer[3] reported on a 3-weeks-old female infant, who presented with the signs and symptoms of congenital pyloric stenosis and died 4 days after this diagnosis had been made. No operation was performed. At autopsy a cyst the size of a hen's egg was found attached to the medial wall of the duodenum, from the pylorus to the ampulla of Vater. No communication between the cyst and the lumen of the duodenum could be demonstrated.

It was in 1923 that Waugh[4] reported a case of a 19-day-old female infant who presented the signs and symptoms of pyloric stenosis associated with a palpable mass in the right hypochondrium and was found at operation to have an enterogenous cyst lying lateral to the second part of the duodenum. It had pushed the hepatic flexure of the colon downwards and forwards. Again there was no evidence of communication between the duodenal lumen and this cyst. An attempt was made to cure the cyst by packing it with gauze but it filled up again after 6 weeks. Suture of the cyst wall to the aponeurosis of the abdominal wall was subsequently performed in an attempt to obliterate the cyst, but the infant died 6 days after operation, from a pulmonary infection.

Similarly, in 1927, Maddox[5] reported on a case with the same clinical picture as that recorded by Waugh. In this case the cyst was associated with the second and third part of the duodenum, and was the size of a goose's egg. Poor condition of the child did not allow any curative form of surgery to be performed, and death occurred 16 hours after operation.

The sixth report of this type of intestinal cyst came from the hand of Smith[6] in 1930, when he found a cyst attached to the anterior wall of the duodenum, and again he was unable to find any evidence of communication with the intestinal lumen. Smith established external drainage, which proved of no avail, as the infant died 1 week after operation.

By this time about 50 years had elapsed since the report of the first case, and no successful surgical procedure was available for the cure of this rare condition.

Gardner and Hart[7] were the first surgeons to come forward with positive and systematic teaching for the cure of enterogenous cysts of the duodenum. In their paper, published in 1935, they reported a duodenal cyst in the medial wall of the second part of the duodenum in a female child aged 15 years. There was no communication with the intestinal lumen. An anastomosis between the cyst and duodenal lumen was established, and this proved to be a satisfactory method of treatment as shown by their follow-up report 3 years after operation. Gardner and Hart suggested the following methods of treatment:

1. Excision.
2. Permanent internal drainage into the intestinal lumen by (*a*) anastomosis between the cyst and duodenum or (*b*) anastomosis between the cyst and jejunum, combined with jejuno-jejunostomy.

Ripstein,[8] in 1949, stressed the fact that as intestinal duplications are a frequent cause of massive intestinal haemorrhage in infancy, and because they usually cause acute symptoms, it is essential for the surgeon to be familiar with their mode of presentation and the treatment of choice.

Altogether 21 cases of enterogenous cyst of the duodenum were found described in the literature up to 1953; 4 more cases have since been reported.

In 1955 Pinkerton and Annamunthodo[9] described a case of post-eclamptic anuria which was complicated by haemorrhage into an enterogenous cyst of the duodenum.

The presentation and diagnosis of enterogenous cyst of the duodenum was adequately reviewed by Mendl and Tanner[10] in 1954, and they mention the fact that marked distension of the cyst occasionally produces atrophy of its muscular coats, so that a fibrous-walled cyst may remain. We believe that the distension occasioned by intra-cystic haemorrhage had caused the disappearance of a mucosal lining in our own case.

Our own case will be the 26th in the series of publications. Of these 26 cases only 2 have shown frank intracystic haemorrhage.

According to Shallow *et al.*[11] the mortality in the first 14 cases was 50%.

Our impression from the literature is that gastro-enterostomy is a life-saving procedure in the infant with obstruction. It seems essential, however, to make a subsequent direct attack on the cyst itself in order to prevent later complications. In our case the gastro-enterostomy that was performed when she was 2 years old was insufficient for a lasting cure. Eradication of the cyst or the establishment of permanent internal drainage is an essential part of the treatment.

SUMMARY

1. A case of enterogenous cyst of the duodenum complicated by intracystic haemorrhage is described.
2. The relevant literature is reviewed.
3. It is emphasized that enterogenous cyst of the duodenum should be considered in the differential diagnosis of congenital hypertrophic pyloric stenosis.
4. The treatment of choice is briefly discussed.

OPSOMMING

1. 'n Geval van 'n derm-sis van die duodenum met bloeding in die sis is beskrywe.

2. 'n Kort oorsig van die betrokke literatuur word aangehaal.

3. Die feit dat derm-sis van die duodenum oorweging moet geniet by diagnose van aangebore hipertrofiese stenose van die piloris word benadruk.

4. Die beste behandeling word kortliks bespreek.

We wish to express our gratitude to Dr. I. Frack, Medical Superintendent of the Baragwanath Hospital, for his permission to publish this case, and to Dr. I. J. P. Burger for his histological report.

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