A Case of Intestinal Obstruction from Spontaneous Subserosal Hemorrhage with Angiomatous Malformation Associated with Reduplication of the Jejunum*

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INTESTINAL DUPLICATIONS are still uncommon enough to be interesting. Hemangiomata of the bowel have been reported from time to time, but the association of the two in juxtaposition, and the occurrence of a spontaneous subserosal hemorrhage in the hemangioma which produced intestinal obstruction by external pressure on the submucosa without bowel hemorrhage, is believed to be unique. Duplications of the intestinal tract have been reported on rare occasions in the earlier literature under a great variety of names, such as enterogenous cysts, enteric cysts, ileum duplex, giant diverticula and inclusion cysts. Ladd and Gross have been most influential in establishing the term duplications to designate these strange structures. Until recent years they have been considered extremely rare, but as the surgical profession becomes more familiar with them they will doubtless be recognized more frequently. This has already happened at the Children's Hospital in Boston, where Gross, Holcomb and Farber[4] reported 67 instances from 1928 to 1950. I have found no other series of comparable size. The average surgeon will probably see only one or two or none in a lifetime, but certainly a man doing a good deal of pediatric surgery should know what they are and how to deal with them. They usually manifest themselves in the first few days or months of life and almost always in childhood, although rare instances in older people are reported.

Duplications appear in a great variety of forms. The most frequent and classical type is in the lower ileum[2] and may be inches, or feet long, lying in the mesentery alongside the normal gut and having a mucosal lining and a smooth muscle wall, intimately fused with the adjacent gut muscle and having a common blood supply with the neighboring bowel.[1] They may be blind closed loops or communicate at either, or both ends with the bowel, most commonly at the lower end. This type may be found in the colon and jejunum also. The next most common type is more or less cystic and globular in the leaves of the mesentery, usually in apposition to the adjoining intestine and distinguished from a mesenteric cyst by its lack of translucency and its thick muscular wall. Such structures have occurred from the base of the tongue to the retrorectal area.[4] A third rarer type may communicate with the small bowel almost anywhere in its course and extend up through the right diaphragm to a pouch in the chest, often resembling a stomach. A number of duplications of the esophagus have been reported.[4] It is remarkable that the type of mucosa lining them is of such variable and unpredictable structure. One at the base of the tongue contained colonic mucosa and one in the retrorectal space, gastric mucosa.[4] Many in almost any location have shown gastric mucosa and some have had, as would normally be expected, mucosa almost exactly resembling that of the neighboring bowel. Some have had two or three types of mucosa in their lining and in others containing gastric like fluid, there was occasional peptic ulceraton or diffuse inflammation and some have been so distended from a secretory pressure that the mucosa has sloughed.

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Truly nature moves in mysterious ways, her wonders to perform.

SYMPTOMS

Symptoms usually appear early in life and are predominantly of three types: (1) Obstructive. This is usually due to swelling from retained secretion in a closed duplication causing mechanical pressure on the bowel or from damage to the intestine from interference with its circulation from pressure or less commonly from a small duplication becoming the leading point of an intussusception. (2) pain either from distention in a closed loop or from chemical irritation or ulceration due to the presence of gastric-like juices. (3) bowel hemorrhage. This has been prominent in many case reports and has usually been shown to come from peptic ulcer in, or adjacent to, ectopic gastric mucosa in the cases operated upon.

TREATMENT

First it should be realized, that, because of the inextricable mingling of the muscular walls of these structures with that of the gut and the common blood supply, they can practically never be dissected out without hopeless damage to the companion bowel. Wherever possible the duplication along with the adjacent intestine should be resected and an appropriate anastomosis done. Some of the old procedures such as marsupialization are now rarely considered. Some which cannot be safely resected have been anastomosed at one or more points to the adjacent gut, and Gardner and Hart[3] reported a celebrated case some years ago with a globular duplication just below the pylorus in the concave curve of the duodenum, where they made a rather large window between it and the duodenum, with relief of symptoms. The ones in the chest communicating with the gut through the diaphragm have usually required staged operations, removing the abdominal portion and marsupializing its upper end temporarily and later removing the chest component transthoracically.[5] The retrorectal ones can usually be got at by removing the coccyx and lower sacrum.

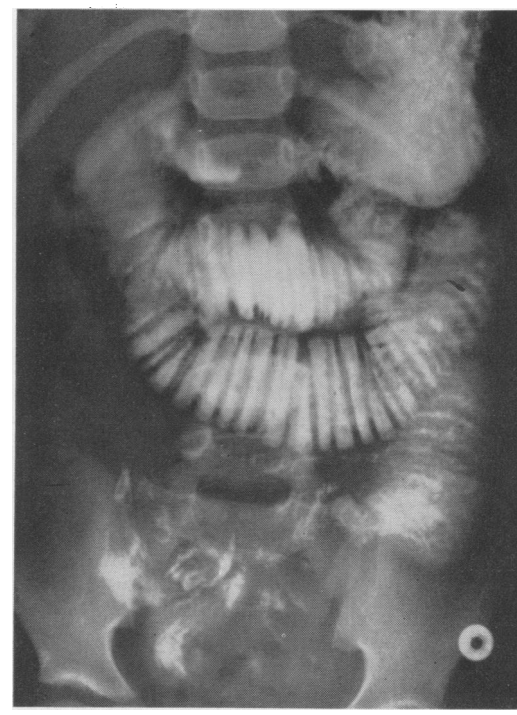

FIG. 1. Barium study showing chronic obstruction of upper jejunum.

Hemangiomata in the digestive tract have usually manifested themselves by massive bowel hemorrhage or a slow occult loss of blood with profound anemia. I recall a child upon whom I operated some years ago because of several large rectal hemorrhages, in full expectation of finding a peptic ulcer in a Meckel's diverticulum, who had an extensive hemangioma of the terminal ileum. They are by no means always easy to recognize, even at laparotomy. The hemangiomatous intestine should of course be resected.

CASE REPORT

ST. JOSEPH'S HOSPITAL—NO. 62988—R. C.

A six-year-old boy was admitted by Dr. H. C. Thomas on March 22, 1955, with a history of several episodes of nausea and vomiting during the past 3 months. These attacks were associated with moderate abdominal pain without fever or

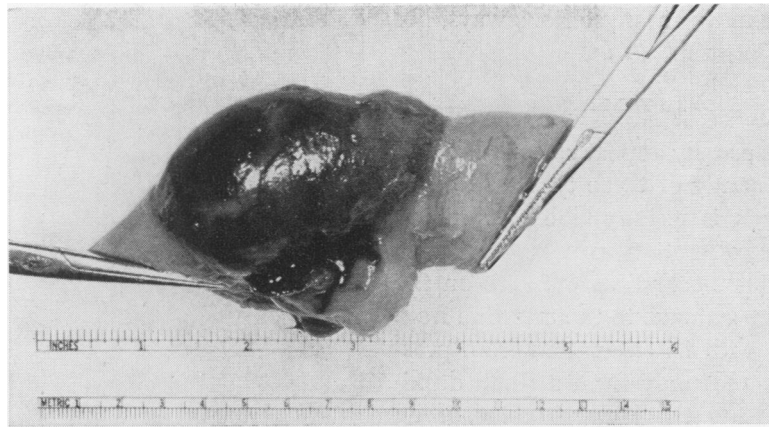

FIG. 2. Gross specimen—spontaneous hemorrhage in hemangioma of jejunum causing intestinal obstruction.

local signs. He had not had diarrhea or melena. On March 1st, he had had a more severe attack of nausea and vomiting than he had previously experienced, with a low grade fever for a few days. He was admitted to the hospital and appendicitis was suspected, but this seemed doubtful and he was not operated upon. He was readmitted a few days later, after vomiting food and later greenish material, and complaining of colicky abdominal pain. His urine showed a trace of albumin and an occasional coarse granular cast. R.B.C., 4,400,000; Hg., 12.3 gm.; W.B.C., 12,300; stab forms, 6; segmented cells, 67; lymphocytes, 21; monocytes, 2. He was a rather thin, poorly nourished child who did not appear to be in acute distress. The abdomen was flat with slight muscle guarding. The liver, spleen and kidneys were not felt. No definite mass was made out and rectal examination was negative. A gastro-intestinal series suggested a possible partial obstruction of the upper jejunum and a special small bowel study the next day demonstrated a definite small bowel obstruction involving the jejunum approximately 30 to 45 cm. from the ligament of Treitz (Fig. 1). The obstruction, the radiologist reported, did not appear to be on the basis of an adhesive band since various films demonstrated involvement of a segment of bowel distal to the point of obstruction for a length of approximately 7.5 cm. and this involved segment had the appearance of distended small bowel due to something within its lumen. The most likely possibilities were believed to be a bezoar, intussusception or some type of neoplasm growing for the most part intraluminally. There was moderate distention of the jejunum proximal to the point of obstruction. At this point I was asked to see the patient. Even after seeing the roentgen films I could not be sure that I felt a mass. It was agreed that he should be explored. The lesion did not suggest intussusception to me and I thought a lymphoma was most likely.

The next morning under general anesthesia, a mass was embarrassingly obvious. It felt at least 7.5 cm. long and half as wide and lay in midabdomen, slightly to the left side. A midrectus incision was made and a rather startling picture presented. An intensely hemorrhagic segment of gut, some 10 cm. long, greatly swollen and lightly adherent to the anterior abdominal wall and adjacent mesentery was easily freed by blunt dissection. The hemorrhage was subserosal and fairly well demarcated above and below and surrounded at least 2/3 of the circumference of the bowel. There was an anomalous structure in the mesentery next to the bowel, measuring roughly 4 x 3 cm. and the mesentery was thickened. My first thought was of a duplication, though a jejunal diverticulum was considered. The subserosal hemorrhage had obviously compressed the mucosa and brought about a partial obstruction. The proximal jejunum was dilated and thickened. I immediately thought of a picture—in a case report in a recent number of the ANNALS OF SURGERY by Lampert *et al.*[6] entitled "Traumatic Subserosal Hemorrhage Causing Small Bowel Obstruction"—a beautiful illustration of high grade obstruction of the jejunum from an encircling subserosal hematoma following a basketball injury. The patient's physician knew of no injury nor, on further inquiry, did the child's parents. It was obvious that a resection had to be done. The lumen of the bowel above the lesion was nearly twice that of the gut below it. The diseased area with the mesentery containing the abnormal structure was excised and the vessels secured. The distal bowel was divided quite obliquely which enlarged its lumen greatly and a 2 layer end to end anastomosis was accomplished with catgut for the inner layer

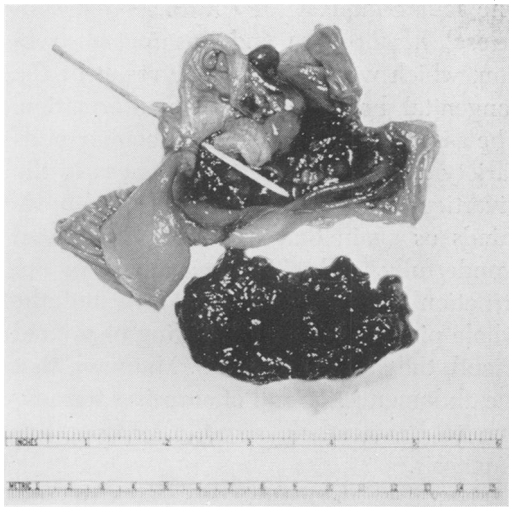

FIG. 3. Dissecting hematoma evacuated communicating reduplication of jejunum shown.

and interrupted silk for the outer. 150 ml. of blood were given and the abdomen closed and gastric suction instituted. He made a rapid convalescence but had a minor wound infection. He soon began to eat eagerly, rapidly regained his lost weight and is reported to be in excellent condition at present.

PATHOLOGICAL REPORT

GROSS: The specimen consisted of a segment of jejunum measuring 13 cm. in length and 5.5 cm. in circumference at both the proximal and distal ends (Fig. 2). The circumference of the central portion measured 3.5 cm. Along the mesenteric attachment and communicating with the lumen of the jejunum was a duplication of the intestine 3.5 cm. in length and 2.2 cm. in circumference. The communicating ostium measures 0.7 cm. and was occluded by a small mass of barium. The mucosa of the duplicated bowel was intact, slightly granular and hyperemic. A distinct mucosa, submucosa, muscularis and serosal layer was identified. A subserosal hematoma arose along the mesenteric attachment of the jejunum, extended around the jejunum for approximately three-fourths of the circumference and involved 8 cm. of the long axis of the jejunum. The subserosal hematoma contained both fresh and old blood (Fig. 3). The duplication was not involved in the dissecting hematoma. Just adjoining the neck of the duplication was a lymph node 1.2 cm. in diameter and of uniform gray color and moderately firm consistence. Just proximal to this lymph node was a smaller nodule 0.8 cm. in diameter, which on section contained a central area of calcification. The attached segment of mesentery appeared to contain normal arteries and veins in which no thrombi could be demonstrated.

HISTOPATHOLOGY: The duplicated segment contained a well formed mucosa, submucosa, muscularis and serosal layer. The mucosa was moderately thin, the superficial portion being a tall columnar cell and in the crypts of Lieberkuhn mucus secreting cells of the goblet type were seen interspersed with large eosinophilic granular cells of Paneth. There were no villi. The mucosa con-

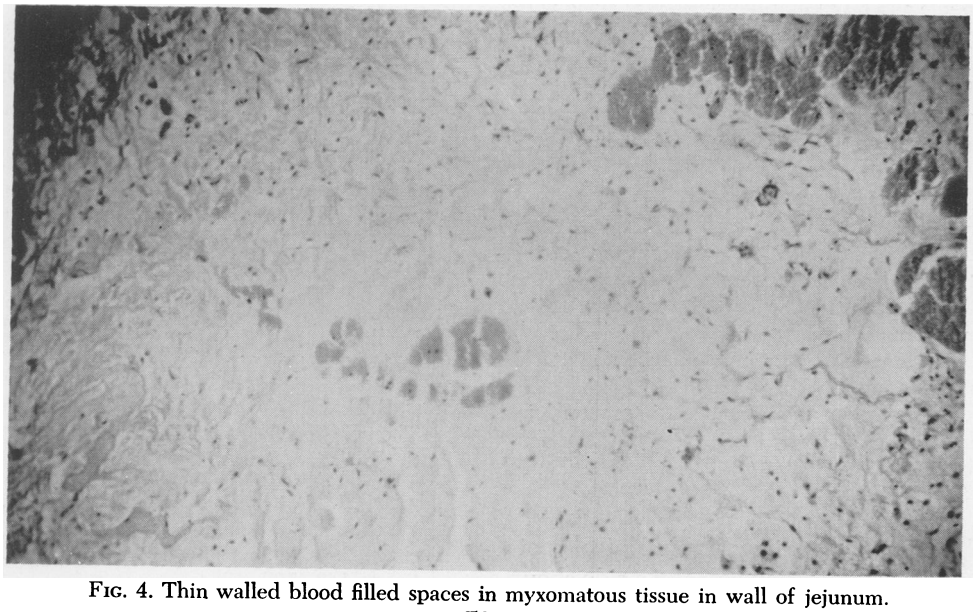

FIG. 4. Thin walled blood filled spaces in myxomatous tissue in wall of jejunum.

tained a mild inflammatory reaction in which a few eosinophils were prominent. The lamina muscularis mucosa was hypertrophied while the submucosa contained an increased amount of collagen and increased vascularity. No significant inflammatory reaction was present in the submucosa. The tunica muscularis also hypertrophied and contained a recognizable inner longitudinal layer, an outer spiral layer, and in some areas, the outer layer resembled the taenia coli. At the junction of the duplication with the jejunum the muscular layer underwent transition from the normal small intestinal muscular coat to a thickened muscular coat of the duplicated large intestine. Occasionally only a few irregular muscle bundles formed the muscular connection. There was a rich vascular supply at this junction, occasionally having an angiomatous pattern existing in a rather loose, or myxomatous appearing connective tissue of the serosa. This angiomatous and myxomatous appearance became increasingly prominent at the margin of the subserosal hematoma (Fig. 4). The subserosal hematoma involving only the jejunum had dissected into the muscularis and in some areas had extended through the submucosa into the lamina muscularis mucosae. It had not penetrated the mucosa at any point. Various stages of fibroblastic repair were evident. No arteries, or phlebitis were identified.

The identified lymph node adjacent to the neck of the duplication contained erythrocytes in the dilated sinusoids but otherwise was unaltered. The smaller nodule contained a central area of calcification about which were uniform stellate cells scattered through an amphophilic, granular and fibrillary mucinous substance. At the periphery of the nodule a rich vascularity was present along with fibroblasts which appeared to be forming a capsule. This myxomatous and calcifying nodule had a histological appearance similar to the subserosal tissue at the neck of the duplication and the angiomatous tissue bordering the dissecting hematoma.

PATHOLOGICAL FINDINGS

1. Luminal communicating reduplication of the intestine, mid jejunum, colonic type mucosa.
2. Jejunal obstruction partial, secondary to intramural hemorrhage.
3. Myxoma circumscribed, mesenteric attachment of jejunum.
4. Angiomatous malformation, intramural, jejunum.

COMMENT

This case presents a pathological potpourri. The duplication is really a side show. The major interest, of course, is the subserosal hemorrhage and ensuing obstruction, which was evidently the result of the congenital hemangiomatous malformation. The associated myxoma, while benign, adds variety and the colonic type mucosa in the diverticulum adjacent to the jejunum reminds us again that we are fearfully and wonderfully made. This mechanism of obstruction must be extremely rare and the whole picture bears out a saying of my old friend, the late Dr. Frank S. Mathews, that the abdomen is too full of surprises for anyone to be arrogant about a preoperative diagnosis.

SUMMARY

A case is presented of congenital hemangiomatous malformation of the jejunum with a communicating reduplication of the jejunum containing colonic type mucosa in juxtaposition to the hemangioma, with the occurrence of chronic intestinal obstruction from spontaneous subserosal hemorrhage in the hemangioma with external compression of the submucosa. This is believed to be very rare.

BIBLIOGRAPHY

[1] Fecher, M. P., M. Day and F. G. King: Ileal Duplications. Ann. Surg., **135**: 554, 1952.

[2] Fisher, H. C.: Duplications of the Intestinal Tract in Infants. Arch. Surg., **61**: 957, 1950.

[3] Gardner, C. E., Jr., and D. Hart: Enterogeneous Cysts of the Duodenum. J.A.M.A., **104**: 1809, 1935.

[4] Gross, R. E., G. W. Holcomb, Jr. and S. Farber: Duplications of the Alimentary Tract. Pediatrics, **9**: 449, 1952.

[5] Gross, R. E., E. B. Newhouser and L. A. Longino: Thoracic Diverticula Which Originate from the Intestine. Ann. Surg., **131**: 363, 1950.

[6] Lampert, E. G., J. G. Goodfellow and T. J. Wachowski: Traumatic Subserosal Hemorrhage Causing Small Bowel Obstruction. Ann. Surg., **140**: 768, 1954.