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CASE REPORT

An Unusual Presentation of Intestinal Duplication with a Literature Review

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KEY WORDS: intestinal duplication; presentations; complications; review.

There are several types of cysts associated with the alimentary tract, including lymphangioma, nonpancreatic pseudocyst, enteric duplication cyst, and mesothelial cyst. Classification is based on the histological appearance of the cyst lining and wall (1).

Duplications of the alimentary tract are uncommon congenital abnormalities. The rate of occurrence is not accurately known, but one survey reported its occurrence in 67 patients during a 22-year period at one pediatric institution (2). A second report cited 63 affected adults over a 10-year period (3). Duplications are usually observed early in life, but a minority remain unsuspected until adulthood (4). The clinical presentation may depend on the location of the duplication and adjacent structures. The symptoms and signs produced by duplications are often vague, so that a correct interpretation of clinical findings is rare. Patients may previously have been misdiagnosed, erroneously labeled as having functional bowel problems (5, 6). The correct diagnosis is more frequently suggested by ultrasound scan, computerized tomography, or established at operation and postmortem.

CASE REPORT

A 52-year-old white male, previously diagnosed as having irritable bowel syndrome, presented to his general practitioner because of the urinary symptoms of frequency and hesitancy. Prostate size was estimated to be minimally enlarged on digital examination; ultrasound and radiographic investigations were arranged. These indicated the presence of a cyst, which computerized tomography confirmed was contiguous with small bowel loops, arising in the mesentery. The cecum appeared displaced superiorly, while in the pelvis, the bladder was impressed from above by the mass. Using ultrasound guidance, the cyst was aspirated. Cytological examination of the aspirate showed only amorphous debris; with no cellular elements present.

At laparotomy, approximately 2 ft proximal to the ileocecal valve, the ileum appeared narrowed and stretched over a smooth-surfaced thin-walled cyst (Figure 1). This portion of ileum was resected with end-to-end anastomosis. The patient's recovery was uneventful, and the urinary symptoms abated.

Histology. Macroscopic examination showed 450 mm of ileum stretched over an intimately associated grey mass that measured 180 × 120 × 80 mm. Slicing revealed a thin-walled cyst with a smooth inner surface and containing viscous putty-like material.

Light microscopy showed that the cyst was composed of the normal constituents of alimentary tract wall. It was lined by stratified squamous, ciliated, and gastric type epithelium (Figure 2) with a muscularis mucosa and a muscularis propria of which the outer longitudinal layer was "shared" with the adjacent ileum. No dysplasia, malignancy, or intrinsic ileal abnormality was present.

DISCUSSION

Duplications are cystic, spherical, or tubular structures and lie either within the wall of the alimentary tract (intramural) or in the mesentery (7, 8). Occasionally the duplication may possess its own mesentery. More than two thirds of duplications are spherical, while the rarer tubular variety often communicate with the bowel lumen (5). Closed duplications tend to enlarge progressively as secretions from the mucosal cyst lining accumulate, resulting in thick clay-like contents (5, 7). There is a cyst epithelial lining with surrounding smooth muscle, inner circular, and outer longitudinal layers. The latter may be continuous with the muscularis propria of the gut. The epithelial lining is variable and resembles some part of the alimentary or respiratory tract, but it does not necessarily correspond to the mucosa at the level at which the duplication is found. Ciliated columnar cells are found in the fetal gut, and the respiratory tract has a foregut derivation (5).

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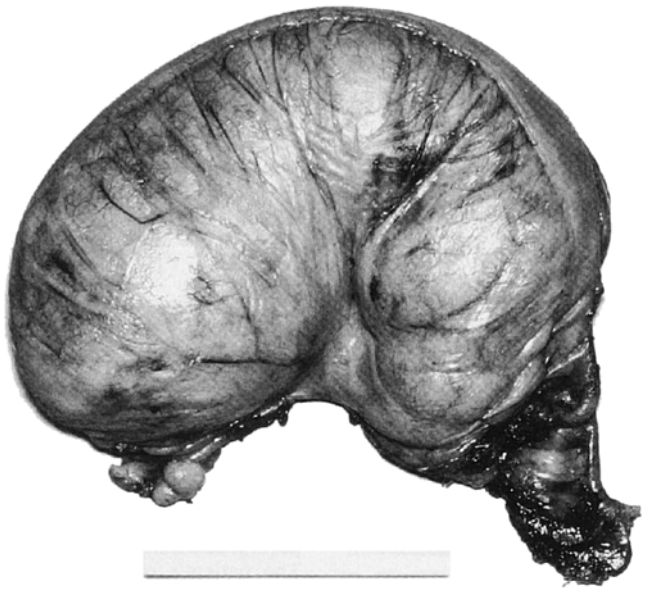

Fig 1. Ileum stretched over the enteric duplication cyst. (10-cm scale included).

The cause of duplication is not accurately known; explanations include persistence of the vitelline duct, out-pouching and incarceration of embryonic bowel, and most likely a neurenteric canal communication (4, 5, 10). There may be nonenteric associations; spinal defects include complete spinal column duplication, spina bifida, but more commonly hemivertebrae (5). Other associations include mental retarda-

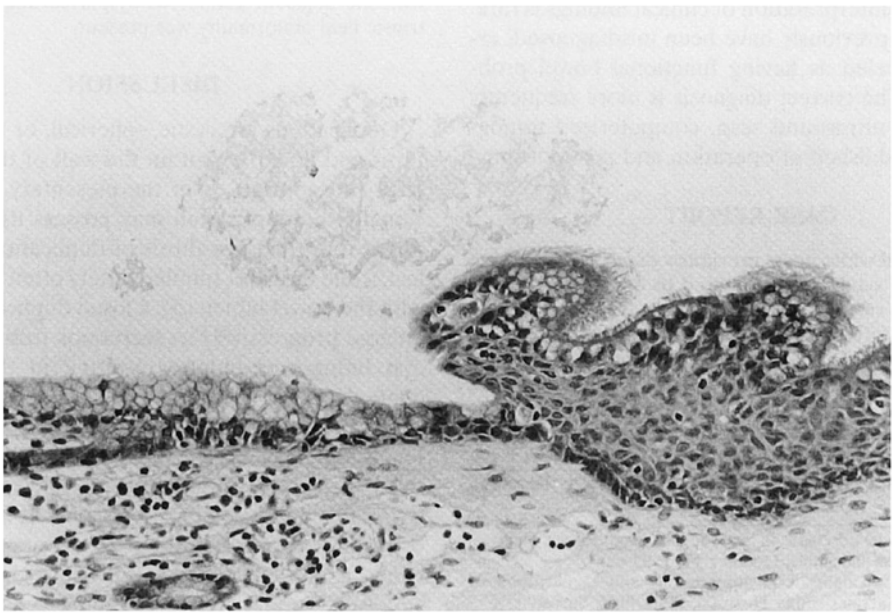

Fig 2. Epithelial lining of the duplication cyst showing ciliated and glandular epithelium (original magnification ×160).

tion, hernias, club foot, meningocele, bilobar right lung, hydrocephalus, Meckel's diverticulum, and doubling of the genital and urinary organs (3–5).

The manifestations of duplications include abdominal mass, distension, pain, and now in our case urinary symptoms. Complications may be perforation, intussusception, volvulus, malignant change, and intestinal, ureteric, biliary, and inferior vena caval obstruction (3–5, 7, 9). The symptoms and signs produced are often difficult to assess correctly. Plain x-ray, barium studies, and intravenous urography have been used with limited success (11), but the findings of an echogenic inner mucosal layer and a hypoechoic outer layer on ultrasound confidently identify duplication cysts (12). Computerized tomography is useful in demonstrating more precisely the relationship between the cyst and surrounding structures (11, 13).

The treatment of choice is surgical with removal of the cyst and adjacent bowel determined by the shared blood supply. Marsupialisation with cauterization, has also been used with less success (2, 4).

CONCLUSION

Duplications of the alimentary tract are uncommon congenital abnormalities, and consist of cystic spherical or tubular structures lying within the wall of the alimentary tract or in the mesentery. They may arise at any level from the mouth to the anus and may be multiple. Although usually observed early in life, a minority may remain unsuspected until adulthood. The clinical presentations may be vague and diverse depending on the location of the duplication. Presentations include pain, distension, dysphagia, and dyspepsia, and complications involve bleeding, perforation, obstruction of viscera, and malignancy.

Plain x-ray investigations are of limited use in the diagnosis of duplications, but ultrasound findings may be diagnostic, with computerized tomography useful in delineating surrounding structures. Once the diagnosis is established, surgical correction is the treatment of choice, preferably with complete removal.

SUMMARY

A 52-year-old male presented with urinary symptoms of frequency and hesitation. X-ray, ultrasound, and computerized tomography investigations were performed that indicated the diagnosis and position of an enteric duplication cyst. Elective surgery was performed to completely remove the duplication cyst. Histological examination showed that the cyst was lined by stratified squamous, ciliated, and gastric-type epithelium, with a muscularis mucosae and a muscularis propria. No malignancy or dysplasia was seen. Duplications of the alimentary tract are uncommon congenital abnormalities. They may be multiple and arise at any level from the mouth to anus. Usually observed early in life, a minority may remain unsuspected until adulthood. The clinical presentations may be vague and diverse depending on their location. These include pain, distension, dysphagia, dyspepsia, and complications involve bleeding, perforation, malignancy, and obstruction of the alimentary tract and vessels. Plain x-rays are of limited use in the diagnosis of duplications but ultrasound findings may be diagnostic, with computerized tomography useful in delineating surrounding structures. Once the diagnosis is established, surgical correction is the treatment of choice, preferably with complete removal.

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