PRESACRAL ENTEROGENOUS CYST*

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THIS PAPER presents a case of enterogenous cyst located in the presacral area. It is also our desire to present each case of histologically proved enterogenous cyst that has been reported in the literature as occurring below the peritoneal reflexion.

Enterogenous cyst is a term applied to congenital cysts which have their origin from some portion of the digestive tube. The literature is filled with reports of these cysts arising from the stomach and from the small and large bowel, but reports of cases involving the rectum are scant. It is our desire to report only the cases that have arisen ventral to the sacrum and below the peritoneal reflexion.

Ten reported cases that carry histologic proof of enteric origin have been found in the literature. To this number we wish to add the following case report.

CASE REPORT

Mrs. L. L., a white woman age 33, consulted us on May 12, 1948, complaining of indigestion of two years' duration. Her history was suggestive of duodenal ulcer. There was no complaint of any rectal difficulty, but in the course of physical examination a smooth, fluctuant tumor was felt posterior to the rectum and in close approximation to the coccyx. The tumor was slightly movable in all directions and was not excessively tender to pressure. The finger, on rectal examination, could reach the dome of the tumor and its size was estimated to be that of an English walnut. Proctoscopic examination revealed only a smooth rectal wall over the tumor. Air contrast roentgen-ray studies of the rectal ampulla showed no tendency for the tumor to protrude into the posterior rectal wall. Roentgen-ray studies of the sacrum revealed an indefinite soft tissue mass just ventral to the coccyx. The tip of the coccyx was bent forward at a right angle from the lower sacral segment. All laboratory examinations fell within normal limits except that stool examinations were positive for occult blood. Roentgenographic visualization of a duodenal ulcer confirmed a typical ulcer history.

She was placed on medical management for the ulcer, and the systemic response was rapid. During the course of her early ulcer management we had time to become familiar with lesions occurring in the presacral area by study of the literature. A case report by McLanahan and Stone[3] made the tentative diagnosis of enterogenous cyst tenable in our case. The patient readily agreed to the removal of the tumor. Because of the close approximation of the tumor to the posterior rectal wall, it was thought wise to administer sulfasuxadine for ten days prior to surgery.

OPERATION

On June 8, 1948, the tumor was removed through a posterior three inch midline incision from the anus to the coccyx. It was necessary to remove the distal segments of the coccyx to uncover the tumor in its entirety. With a finger against the tumor from the inside of the rectum, the mass was dissected free from its attachment to the posterior rectal wall, and after enucleation, the gloved finger could be seen clearly through the bowel mucosa. No tissue being available for obliteration of the space occupied by the

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tumor, the wound was dusted with sulfathiazole and a soft rubber drain inserted beneath the skin. Drainage persisted for ten days, when healing became complete.

PATHOLOGIC DIAGNOSIS

Gross Examination. The specimen consisted of a well-encapsulated, thin-walled cyst measuring 2½ x 1½ centimeters. Cut section revealed the cyst to be filled with thick, mucoid material. The wall was lined with a pink, soft, mucosalike tissue and measured 0.2 centimeters in thickness.

Microscopic Examination. Sections were made from many different areas of the cyst, and were stained with hematoxylin-eosin, azocarmin and mucicarmin, respectively.

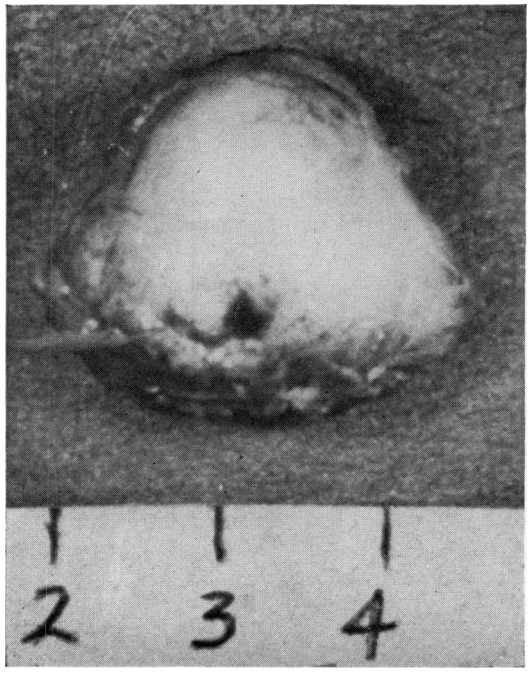

FIG. 1.—Gross appearance of the enterogenous cyst.

Sections stained with hematoxylin-eosin showed the cyst wall as built of the following layers: an epithelial layer and a sub-epithelial layer, loosely arranged, containing small vessels, cell formations like plasma cells, large mononuclear lymphocytes and wandering cells. This layer was followed by two coats of tissue consisting mainly of smooth muscle layers. In addition to these muscle layers, collagenous and reticular connective tissue were present, as indicated by azocarmin stain.

The outermost layers of the cyst were indistinct, and peritoneum could not be demonstrated. Between the outer and inner layers, we found bundles of nerves and in one instance a collection of ganglion cells. By this description the tissue is that of intestinal wall if the epithelium presents the characteristic features of bowel mucosa. Under normal conditions, the mucous membrane of the intestine consists of high columnar, mucus-secreting cells arranged to a certain pattern. In our case, the covering epithelium also consisted of several layers of squamous or transitional epithelium. The cells in the squamous area were in several layers, the lower one more columnar in type, while the uppermost layer showed a more balloonlike cytoplasm. There were areas where this cell layer did not

consist of squamous epithelium but of mucus-producing columnar epithelium, proved by the use of mucicarmin stain.

Stains with iron hematoxylin and with mucicarmin prove furthermore the presence of a cuticulum. Cells of this type are met with in the whole intestinal tract from the cardia to the anus.

Practically all layers characteristic for intestine were demonstrable, namely, mucus-producing columnar epithelium, smooth muscle layers with vessels and nerves. The presence of squamous epithelium is attributed to the effect of metaplasia.

Diagnosis. Enterogenous cyst.

TABLE I.

Author	Year	Author's Diagnosis	Pathologic Diagnosis	Age and Sex	Symptoms	Treatment
Middledorpf	1885	Cyst of postanal gut origin	Small loop of vestigial intestine	1 yr., F		Posterior excision by Kraske
Ballantyne	1932	Cyst of postanal gut origin small area of adenocarcinoma	Cyst of intestinal origin with early adenocarcinoma	38, F	Pain in left buttock	Posterior excision
McLanahan and Stone	1934	Presacral enterogenous cyst	Enteric cyst	48, F	Lump in rectum	Posterior excision
McLanahan and Stone	1934	Presacral enterogenous cyst	Enteric cyst	1 mo., M	Vomiting and constipation	Aspiration and subsequent posterior excision
Thomason	1934	Cyst of postanal gut	Cyst of intestinal origin	34, F	Pain, draining sinus	Repeated posterior excision
Raven	1935	Cyst of postanal gut	Cyst lined with stratified columnar and ciliated epithelium	2½ mo., F	Acute obstruction	Demonstrated at autopsy
Guis and Stout	1938	Cyst of vestigial gut	Cyst of intestinal origin	26, F	Pain and pressure in rectum	Intrarectal marsupialization
Guis and Stout	1938	Cyst of vestigial gut	Cyst of intestinal origin	36, F	None found on routine exam.	Posterior excision
Ladd and Gross	1940	Duplication of rectum	Cyst of intestinal origin with mixed mucosa	6 mo., F	Recurrent obstruction	Repeated aspiration and subsequent posterior excision
Custer, et al.	1946	Enterogenous cyst involving rectum	Enterogenous cyst	29, M	Symptoms of bladder pressure	Excision by laparotomy

REVIEW OF LITERATURE

We have been able to find ten cases of histologically proven presacral enterogenous cysts reported in the literature. (Table I).

Middledorpf,[1] in 1885, was the first to describe a presacral cyst of intestinal origin. The patient was a one-year-old female, and when the cyst was removed by Kraske, it was found microscopically to have all the structure of intestinal wall. Diagnosis: cyst of postanal gut origin.

In 1932, Ballantyne[2] reported a presacral cyst in a female of 38 years. The cyst contained 475 cc. of fluid and was removed by posterior excision. Microscopically, the cyst was lined with columnar epithelium. Goblet cells secreting mucin were present. There was a small area showing adenocarcinoma. Diagnosis: cyst of postanal gut with area of adenocarcinoma.

In 1934, McLanahan and Stone[3] described two cases of presacral enterogenous cyst. The first case was a woman of 48 years who complained of a lump in the rectum which at times would protrude from the anus but was easily reduced. Digital examination revealed a small cystic mass lying about two inches within the anal orifice and situated between the posterior wall of the

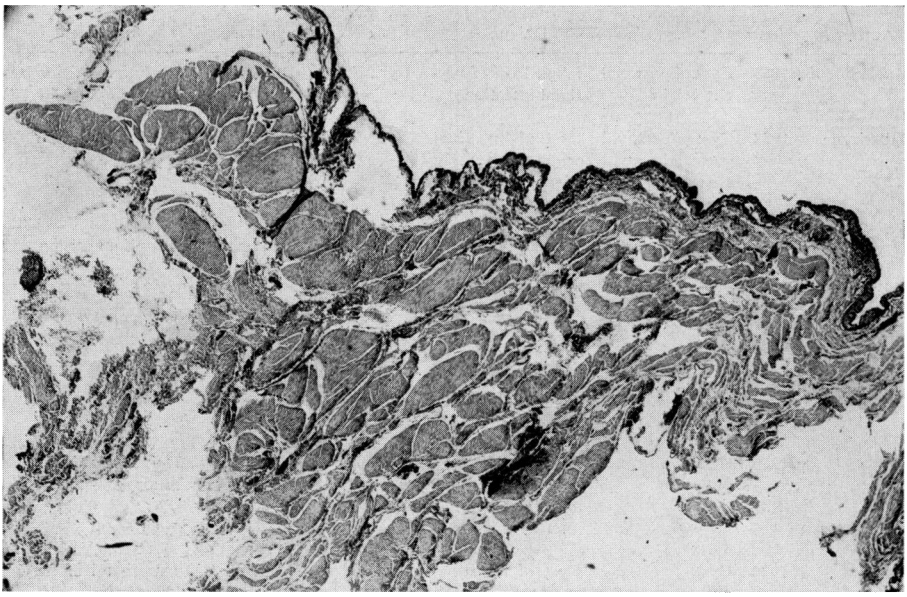

FIG. 2.—Low power microphotograph of cyst wall demonstrating the different layers of epithelium, subepithelium, and muscularis.

rectum and the coccyx. The mass was about the size of a walnut, was fluctuant, freely movable and not tender. Doctor Stone removed the tumor by posterior excision after the coccyx was resected. The mass was dissected from the posterior rectal wall, to which it was closely attached. Microscopic examination of the cyst wall showed tall columnar epithelium of mucus-secreting type. The submucosa was composed of longitudinal and circular muscular layers covered by a layer of serosa. Diagnosis: enteric cyst.

The second case reported by McLanahan and Stone was in a one-month-old male infant with intestinal obstruction from a mass filling the rectum. The mass was thought to be an intussusception, but on exploration of the abdominal cavity, a fluctuant mass the size of a golf ball was found below the peritoneal reflexion. The abdominal wound was closed, and on the following day the tumor was aspirated by needle entering the skin one inch to the right of the

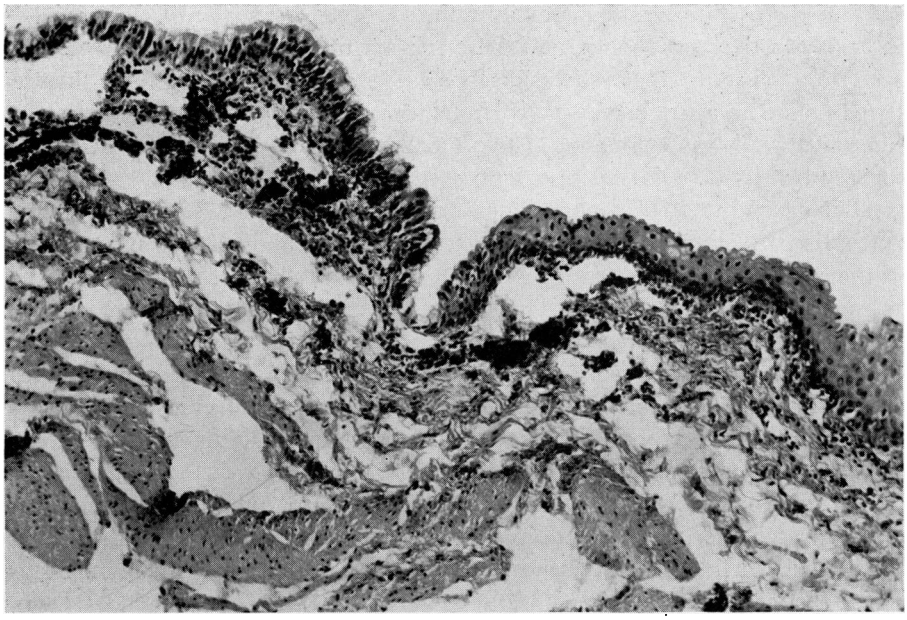

FIG. 3.—Metaplasia from columnar to squamous epithelium. Notice mucus droplets within the cytoplasm of columnar cells (proven by use of mucicarmin stain).

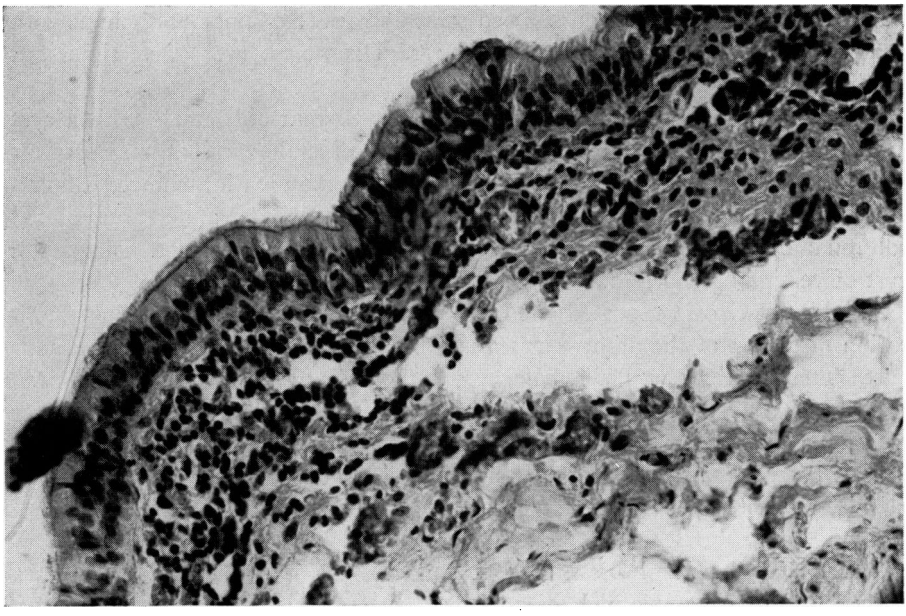

FIG. 4.—Cuticulum of columnar epithelium demonstrated by the use of iron-hematoxylin stain.

posterior margin of the anus. Twenty-five cc. of cloudy fluid were withdrawn, and the obstruction was temporarily relieved. The cyst was aspirated on five occasions in the next month, and the condition of the baby so improved that posterior excision of the cyst was performed. The microscopic sections showed a well-defined mucosa lined with columnar epithelium of mucus-secreting type. The outer wall was composed of two circular and longitudinal muscle layers covered by serosa. Diagnosis: enteric cyst.

Thomason,[4] in 1934, reported a case of sacro-coccygeal cyst of the postanal gut. His patient was a 34-year-old woman with a draining sinus near the tip of the coccyx which had been present since a mass in that area was incised and drained several years before. The sinus tract and the cyst were removed by posterior excision. Microscopic examination showed the wall of the cyst to be made up of columnar epithelium of the mucus-secreting type, Peyer's patches and two layers of smooth muscle. Diagnosis: postanal gut cyst.

Raven's[5] case, reported in 1935, was a 2½-month-old female who was acutely obstructed by a retro-rectal cyst. The condition was thought to be an intussusception, and laparotomy was performed. At autopsy the mass was removed and microscopic sections showed the cyst to be lined with several layers of epithelium, the innermost being ciliated columnar epithelium. Diagnosis: cyst of postanal gut.

Guis and Stout[6] reported two cases in 1938. The first was a 26-year-old female who complained of rectal pain and pressure. A palpable mass posterior to the rectum was thought to be an abscess. An intrarectal incision was made and the tumor wall marsupialized from within after removing a part of the wall. Pathologic examination showed stratified and columnar epithelium with mucin secretion and smooth muscle layers. Diagnosis: cyst of vestigial gut origin.

The second case of Guis and Stout[6] was a 36-year-old female with a cyst in the postero-lateral rectal wall found on routine rectal examination. Posterior excision of the cyst was done and the pathologic examination showed smooth muscle and a mucosa which was composed of stratified squamous and ciliated columnar epithelium tending to form glands. Mucin was present. Diagnosis: cyst of vestigial gut origin.

Ladd and Gross[7] reported one case in 1940. They prefer the term "duplication of a part of the alimentary canal," but also use the term enterogenous cyst. Their patient was a female, age 6 months, who had recurrent bowel obstruction due to a presacral mass. This tumor was removed by posterior excision. The sections of the cyst wall showed mucosa, submucosa and the muscle layers characteristic of intestine. The mucosa showed mixed types of glands, some of colon and some of gastric type. Diagnosis: duplication of rectum.

Custer, Kellner and Escue[8] report one case in 1946. Their patient was a 29-year-old man complaining of pressure in the pelvis, epigastric pain and a decrease in urinary bladder capacity. Rectal examination disclosed a cystic mass which filled the pelvis. This mass was removed through a low midline

incision. Its attachment was to the anterior rectal wall. Pathologic examination showed the cyst wall to be made up of tall columnar mucus-secreting epithelium and a well-defined layer of smooth muscle. Diagnosis enterogenous cyst.

ETIOLOGY

Three theories have been advanced to explain the presence of enterogenous cyst.

Middledorpf,[1] in reporting his case of enterogenous retro-rectal cyst in 1885, felt that the origin of the cyst was in the vestigial postanal gut. Five of the other authors reporting their cases chose to believe that this theory best explains the presence of the cyst in the midline between the coccyx and the rectum.

The diverticular theory of cyst origin is based on the work of Lewis and Thyng,[9] who demonstrated that diverticuli occur frequently in all portions of the digestive tube of the embryo. These may become occluded at the neck and the cyst results.

A third theory, known as the sequestration theory, suggests that a group of cells becomes pinched off from the primordial intestinal tube and undergoes subsequent development outside the lumen of the intestine.

PATHOLOGY

Although the term "enterogenous" or "enteric" cyst has become accepted when applied to these tumors, they are perhaps more accurately named "duplications of the alimentary tract" as suggested by Ladd and Gross.[7] Their term is actually the requirement for pathologic diagnosis of these cysts.

As Ewing[10] pointed out, the most variable component is usually the mucosal lining, which may show cylindrical, cuboidal, stratified or flat epithelium. The degree of tension within the cyst, together with the amount of inflammatory reaction, produce many variations in the type of mucosa. Ladd and Gross,[7] in reporting their case of presacral cyst, pointed out that the mucosa may be typical of that segment of the bowel to which it is adjacent or it may be typical of some bowel segment far removed from the cyst. The mucosa may be mixed, as in their case, showing large bowel and gastric mucosa.

The presence of intestinal mucosa, smooth muscle layers, nerve bundles and serosa establish the origin of the cyst to be enteric. The serosal layer is usually absent when the cyst is presacrally located.

TREATMENT

The treatment in reported cases has been aspiration, incision and drainage through the rectum, marsupialization and removal by intra-abdominal approach or posterior excision. All authors advise removal as a necessity in some cases and as a guard against malignant degeneration. The removal of the cyst usually can be accomplished by the removal of the coccyx in a posterior approach.

We wish to acknowledge our indebtedness to Dr. Phillip R. Rezek for the pathologic report in this case.

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DISCUSSION.—DR. G. V. BRINDLEY, Temple, Texas: I am sure all of you have enjoyed this excellent paper, for it is a most interesting presentation of one of the infrequent tumors which originate in and ventral to the sacrum. The caudal area is the location for many pathologic types of cysts, tumor, sinuses and fistulas which usually arise from vestigial embryonic tissue.

Some seven years ago, within a period of 14 months I encountered three neoplasms which originated ventral to the sacrum. It was my privilege to present these cases before this Association four years ago. One of these growths was a chordoma, another was an ependymal cell glioma, and the third was a large dermoid cyst. Since statistical data show that most patients with chordoma do not remain well, it was believed that it probably would be of interest to report at this time that the patient who had the chordoma is well and apparently free of disease. It is now eight years since the neoplasm was removed.

May I mention three facts worthy of emphasis pertaining to tumors of this region. The first is that diagnosis of most of these neoplasms depends upon a careful vaginal and/or rectal examination. Certainly no physical examination is complete unless these examinations are performed. Another fact deserving comment is that these tumors are cured only by complete removal. To attempt to cure a cyst of this region by incision and drainage usually results in secondary infection and a persisting sinus. Furthermore, adequate exposure is essential for removal of these lesions. The complete excision of a growth of this region is facilitated by the proper approach, which is obtained by an incision over the sacrum and coccyx extending to near the anus. The patient should usually be in the Kraske position. The coccyx and as much of the sacrum below the sacroiliac articulation as is necessary for adequate exposure of the neoplasm should be removed, together with the muscles and nerves of this region.

It should be appreciated that the segment of the sacrum below the sacroiliac articulation, together with the coccyx and all the sacral nerves except the first and second, can be removed without severe disability being incurred by the patient.

Again, let me congratulate the essayist upon this fine presentation.

DR. HARVEY STONE, Baltimore: I want to thank Doctor Perry for putting this case in the record, because it seems to me that, in view of his study of the literature to date, anyone encountering such a condition as this should record it, because of our scanty infor-