Case Report

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Retroperitoneal Enteric Duplication Cyst

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Enteric duplication cysts (EDCs) can occur in any portion of the alimentary tract, but are most commonly associated with the small bowel and esophagus. Retroperitoneal location is really unusual. This 19-year-old female was in excellent health, but a week's abdominal pain made her search for a doctor's help. After the detailed examination, surgical intervention was performed under the impression of cystic tumor of the retroperitoneum. A retroperitoneal cystic tumor, 13.0 × 8.0 × 3.5 cm in size, without any communication with the alimentary tract was noted during the operation. Finally, EDC was diagnosed after the pathologic examination of this resected cystic lesion. To our knowledge, there have been only 6 reported cases of EDC of the retroperitoneum in the English literature. This report concerns the seventh case of retroperitoneal EDC, in an adolescent, with different clinical presentation and histopathologic findings from the previous ones.

Key Words
alimentery duplication;
duplication;
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enteric duplication

In 1884, Reginald Fitz firstly used the word "duplication" to describe the remnants of the omphalomesenteric duct.[1] Since then, the terms "enteric cyst," "enterogenous cyst," "giant diverticulum" and "duplications of the alimentary tract" have been used to describe congenital cystic or tubular abnormalities of the gastrointestinal tract. In 1961, Mellish and Koop defined "enteric duplications" as spherical or tubular structures that possess a mucosal lining characteristic of 1 or more portions of the alimentary tract supported by muscular and serosal layers.[2] Enteric duplication cysts (EDCs) are uncommon lesions, accounting for approximately 15% of foregut cysts.[3] The majority are diagnosed within the first year of life. Approximately 80% are solitary and spherical in shape. The clinical symptoms depend on the location and type of mucosal lining. They are mostly associated with the esophagus or small intestine, and the retroperitoneum is an extremely rare site.

CASE REPORT

This 19-year-old female was hospitalized for abdominal pain in the past 1 week. The pain was initially dull but progressed to cramping. She had no history of nausea, vomiting, chills or jaundice. Her past history and family history were also unremarkable. Physical examination revealed nothing particular except left flank knocking pain, and laboratory results disclosed blood white blood cell (WBC) of 8.74×10^3 /μL, serum alkaline phosphatase

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(ALP) of 75 IU/L (normal, 41-133 IU/L), serum amylase of 49 U/L (normal, 20-110 U/L), urine red blood cell (RBC) of 6-8 per high power field (HPF) and urine WBC of 0-1/HPF. Chest roentgenograms showed no evidence of cardiac or pulmonary lesion. Sonography showed a lobulated thick-walled cystic lesion above the left kidney. Magnetic resonance T2-weighted coronal images demonstrated a hyperintense tubular fluid-filled folded cystic mass at left paraspinal region with downward displacement of the left kidney (Fig. 1). At operation, a large multiloculated retroperitoneal cyst with yellowish mucoid fluid content was removed (Fig. 2). Neither communication nor connection with the gastrointestinal tract was evident. All the feeding vessels were patent grossly. The cystic content was sent for analysis. It was composed of fibrinous exudate, moderate amounts of RBCs, polymorphonuclear leukocytes and some degenerative epithelial cells. The culture results for mycobacterium tuberculosis and other pathogens were all negative.

Macroscopic examination showed that it was a multilobulated tubular cyst. The outer surface was smooth. The inner surface was coated by thick yellowish brown mucoid material but partially showed dark red. Microscopically, the composition of the cystic wall was similar to that of the colon wall, including colon-type mucosa, submucosa with Meissner's plexus, muscularis propria with Auer-

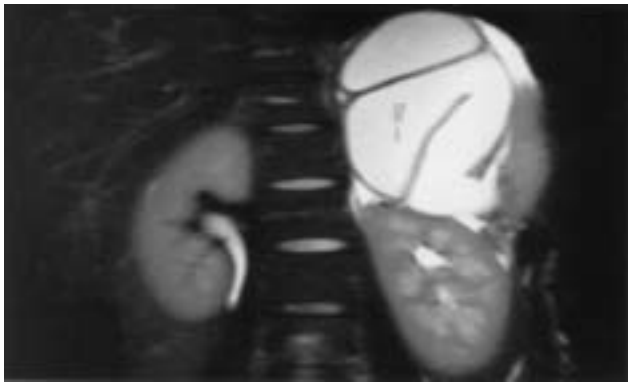

Fig. 1. MRI examination demonstrates the retroperitoneal enteric duplication cyst with an infolding tubular structure.

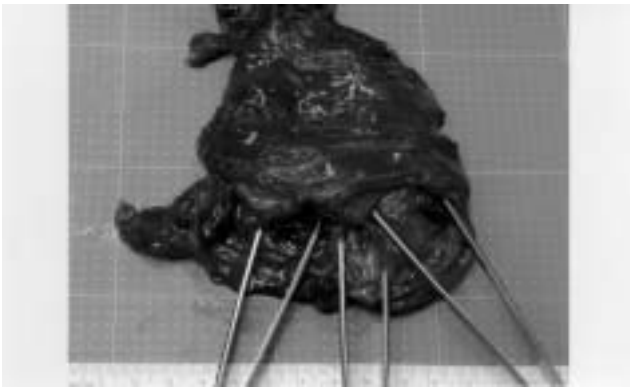

Fig. 2. At operation, a large multiloculated cyst with yellowish mucoid fluid content is noted.

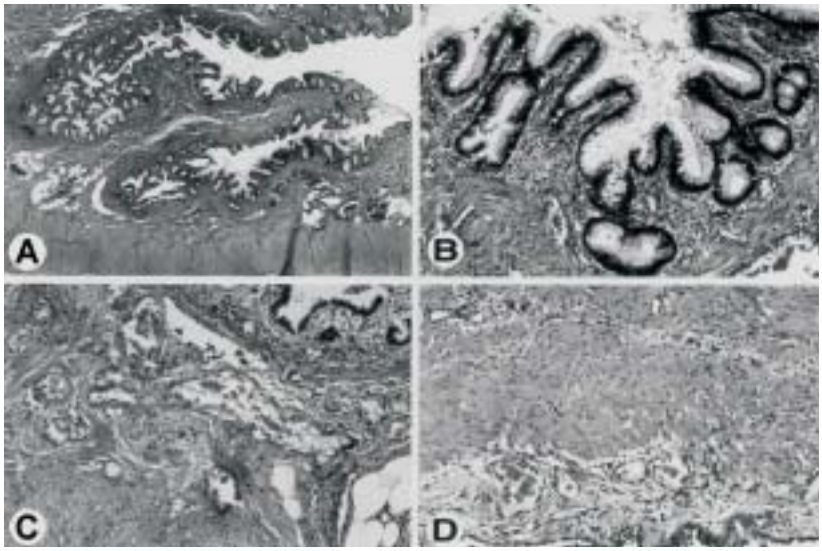

Fig. 3. (**A**) The cystic wall is similar to structure of intestinal wall and is composed of mucosa, submucosa, muscularis propria, subserosa and serosa. (H&E stain, original magnification x 40) (**B**) Mucosa is lined by colon-type epithelium. (H&E stain, original magnification x 100) (**C**) & (**D**) Meissner's plexus in the submucosa and Auerbach's plexus within the muscularis propria are identified. (H&E stain, original magnification x 100).

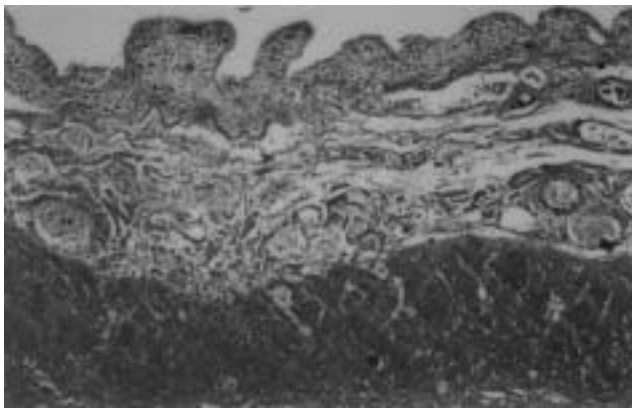

Fig. 4. Foci of enteric wall show acute ischemic necrosis of mucosa, and transmural infiltration of polymorphonuclear leukocytes. (H&E stain, original magnification x 40).

bach's plexus, subserosa and serosa (Fig. 3A-D). Foci of the enteric wall showed acute ischemic necrosis of mucosa, and transmural infiltration of polymorphonuclear leukocytes (Fig. 4). Neither vascular thrombus nor vasculitis was evident. There was no evidence of dysplasia or malignancy in the sections examined, either. The patient's recovery was uneventful, and the symptoms subsided.

DISCUSSION

The cause of EDCs is not definitely known; most are thought to be the result of some specific fault in embryological development and are defined by the presence of a cystic lesion with a coat of double muscular layers, neural elements, and an alimentary mucosal lining.[4,5] Embryologically, EDCs may occur during development of the dorsal foregut. When dorsal mesoderm representing the notochord splits in the middle, a defect is formed through which the ectoderm of the yolk sac herniates. Coexistent congenital anomalies, most frequently present as deformities of vertebrae and other bowel anomalies, may be present.[4,6] This is probably due to the cyst attaching to dorsal ectoderm and resulting in defects of vertebrae and neural tubes.[6] However, there was not any associated anomaly in our patient. EDCs are usually observed in early life, but occasionally are unsuspected until adulthood. They can occur in any portion of the alimentary tract, but the retroperitoneum is a highly unusual location.

Of the only 6 retroperitoneal cases reported in the English literature (Table 1),[1,4,5,7] 3 were prenatal diagnosis by ultrasonography,[5,7] while the others presented with variable clinical pictures including abdominal mass, jaundice and vomiting.[4,5] The ages ranged from 3 months to 31 years old.[1,4,5,7] Unlike EDCs at other sites, most of which were spherical in shape, the retroperitoneal ones had higher tendency to show a tubular configuration. The presentation of cramping pain was unique in our case, in which no clinical diagnosis of neurosis or renal colic had been impressed. Cramping pain is unusual in a

Table 1. Review of the reported retroperitoneal enteric duplication cysts in the literature

Reference	Age/Sex	Clinical feature	Location	Size (cm)	Histologic findings
Richard J. *et al*.(1978)	3 mos/ -	mass lesion	Retroperitoneum, left flank	8 cm	no data available
Richard J. *et al*.(1978)	4 mos/ -	mass lesion	Retroperitoneum, left flank	10 cm	no data available
Takiff H. *et al* (1992)	31 yrs/ F	vomiting, jaundice	lesser sac	5.0 cm cystic mass	EDC with massive mucosal necrosis
Duncan BM. *et al*. (1992)	newborn/ -	prenatal sonography diagnosis	retroperitoneum	3.5 × 2.0 cm	EDC with inflammation
Duncan BM. *et al*. (1992)	newborn/ M	prenatal sonography diagnosis	retroperitoneum	10.0 × 8.0 cm	EDC with intestinal mucosa lining
May DA. *et al*.(2000)	newborn/ F	prenatal sonography diagnosis	retroperitoneum, cross midline	total 6.5 cm in both lobes	no data available
present case (2001)	19 yrs/ F	left flank pain for a week	left suprarenal fossa	13.0 × 8.0 × 3.5 cm tubular mass	EDC with colonic mucosa lining and ischemic necriosis

cyst lesion. Probably, the pain can be explained by the intestinal loop-like structure of the EDC with acute ischemic necrosis resulting from the increasing pressure inside due to long-term mucus secretion. In the cases proved by histopathologic study, all were lined by columnar epithelium but were not otherwise specified.[1,4,5,7] The EDC of our patient was lined by colon-type mucosa. Morphologic feature of the ischemic necrosis with transmural polymorphonuclear leukocyte infiltration in our case might be correlated with the patient's abdominal pain.

Differential diagnosis should include any cystic lesions of the retroperitoneum. Although the final diagnosis was based on the histopathologic findings, in this case, the results of radiologic survey that reflected the gross pathologic appearance were of great help in differential diagnosis.

EDCs should be treated by complete excision whenever possible, because some complications may develop from the residual unresected cystic lining. All retroperitoneal cases, like most EDCs of other sites, were treated by surgical resection without any mortality or mobility.[6] Malignant transformation has been reported in rare cases of other sites,[6] but never in the retroperitoneal ones.

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