

Original Article

Enterogenous cyst in the retroperitoneal region: clinical and radiological analysis of two cases

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Abstract: We reported two enterogenous cyst (EC) cases that incidentally discovered in retroperitoneal space. All patients eventually treated with surgery and had a good result. Pathological examination after surgical extirpation revealed enterogenous cysts. We present the radiological and pathological findings of this rare congenital malformation in order to be included in the differential diagnosis of other cystic mass.

Keywords: Enterogenous cyst, congenital abnormality, computed tomography

Introduction

Enterogenous cyst (EC) is a congenital abnormality resulting from embryogenetic error during the third embryonic week of notochord development. EC mostly locates in the spinal canal, intracranial site and mediastinum, it is rarely reported in the retroperitoneal region. Their pathogenesis is not well understood and several theories have been proposed to explain [1]. Here, we presented two rare cases of EC in the retroperitoneal space and treated with surgery successfully, without recurrence during long lasting follow-up.

Cases presentation

Case 1

A 4-year-old girl was referred to our hospital for evaluation of a mass which was incidentally discovered in the retroperitoneal region by ultrasonic scan. She had no history of trauma and routine physical examination was negative. Moreover, laboratory results were normal. Ultrasound scan showed an anechoic mass measured about 44 mm in diameter adjacent to the pancreas. Abdominal computed tomography (CT) scan demonstrated a well-circumscribed homogeneous cystic mass measured

about 4.5×4×4.5 cm located between the left kidney and tail of the pancreas (**Figure 1A**). The cystic lesion was not enhanced on contrasted CT scan (**Figure 1B**). She was diagnosed for retroperitoneal cystic mass and underwent laparoscopic exploration. On operation, a cystic and encapsulated lesion with smooth and clear boundary was found to be located in the posterior wall of the pancreas tail, closely adherent to the pancreas and the left kidney. The mass was totally excised with an uneventful recovery. Grossly, the cyst was unilocular and well-circumscribed measuring about 4 cm in diameter, it contained thick and white mucoid fluid. The cystic wall varied in thickness from 2 mm to 4 mm and the inner lining appeared smooth with some yellow-white thickened plaques. Microscopically, a thin fibrous cyst wall lined by a mucus-secreting columnar epithelium which was finally diagnosed as an enterogenous cyst (**Figure 2**). The patient made an uneventful postoperative recovery and discharged 7 days after surgery. During the two years follow-up, there was no evidence of recurrence.

Case 2

A 19-year-old man presented to our outpatient department with a history of progressively left flank pain for one month. Multiple masses

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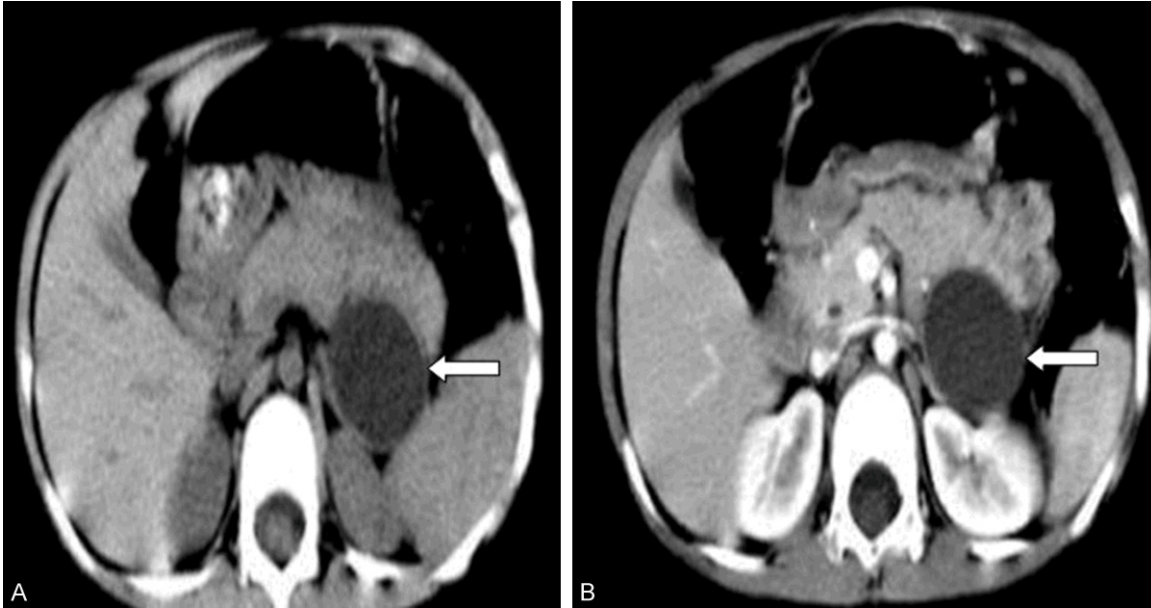


Figure 1. A. CT examination demonstrated a well-circumscribed 4.5×4×4.5 cm homogeneous cystic mass located between the left kidney and tail of the pancreas. B. Contrast CT scan demonstrated the cystic lesion was not enhanced.

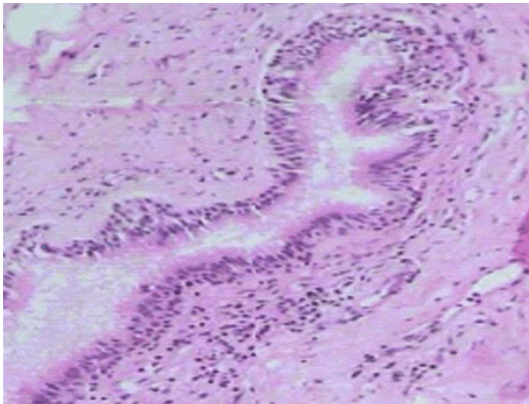


Figure 2. Microscopically showed a thin fibrous cyst wall lined by a mucus-secreting columnar epithelium, which is consistent with an enterogenous cyst.

located in the left retroperitoneal space were detected by ultrasonography. He had a medical history of hypertension for 3 years and treated with oral intake of drugs to control blood pressure. Physical examination revealed flank pain during percussion in the left side. Routine analysis of blood, hepatic and renal functions were all normal. Sonography showed multiple anechoic masses with thick, ill-defined echogenic wall occupying the left retroperitoneal region. Unenhanced abdominal CT scan revealed multiple well-circumscribed solid nodosity lesions

with high density in the left retroperitoneal space, adjacent to the left kidney and posterior to the pancreatic tailor (**Figure 3A**). Following contrast enhanced CT revealed no enhancement (**Figure 3B**). Coronal CT demonstrated three separated solid masses abutting the pole of the left kidney (**Figure 3C**). Radiological findings suggested the presence of multiple retroperitoneal masses and renal dysplasia. Laparoscopic exploration was performed. At surgery, several unilocular cystic masses were found located in retroperitoneal space and adjacent to the upper pole of the left kidney. All cystic masses were excised, totally; no evidence of retroperitoneal lymphadenopathy was noted. Grossly, the specimen showed a dark multilocular cysts measuring about 4×7×3 cm. Histopathologic examination revealed a multilocular cyst filled with yellowish and viscous fluid, the cyst was lined by stratified squamous epithelium and gastrointestinal mucosa with lamina propria and muscularis propria. The cyst wall was smooth and filled with hemoglobin deposition with variable thickness (approximately 0.4 cm). There were multilocular non-communicating small cysts, cystic component contained blood. The solid component mainly consisted of fibrous tissue and haemorrhage. Histopathologically, the cyst wall was composed of loosely textured vascular fibrous stroma and a few spe-

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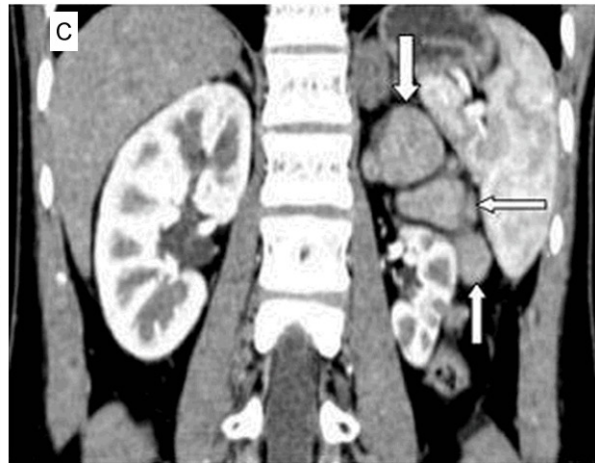
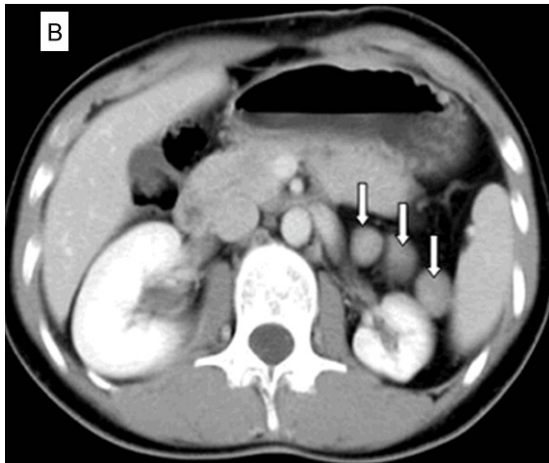
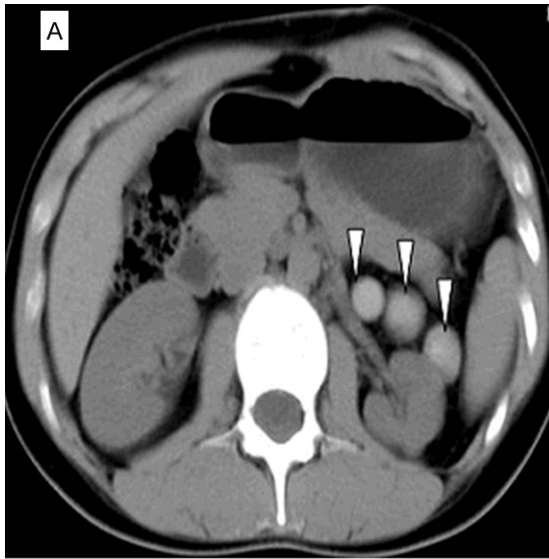


Figure 3. A. CT revealed multiple well-circumscribed solid nodosity lesions with homogeneous high density contents in the left retroperitoneal space and adjacent to the left kidney, posterior to the pancreatic tailor. B. Contrast enhanced CT revealed no enhancement of the mass. C. Coronal CT demonstrated three separated solid masses abutting the pole of the left kidney.

cialized gastric glands (**Figure 4**). The final histological diagnosis was enterogenous cyst. The postoperative recovery was uneventful and the patient was discharged 7 days later. With a follow-up for 14 months, the patient is asymptomatic and there was no evidence of recurrence.

Discussion

Enterogenous cysts, especially in the retroperitoneum, are rare congenital malformations which originate from misplaced epithelium of respiratory tree or intestinal tract and are usually lined by mucus secreting epithelium resembling that of the gastrointestinal tract. Pathology defines them as "cysts lined by columnar epithelium of presumed endodermal derivation". They usually originate during the third or the fourth week of the embryonic development. These lesions can be found in all age groups, but they are most commonly diagnosed in chil-

dren and young adults. General localizations of these cysts are in the vertebral canal, posterior mediastinum, intracranial region [2, 3]. Sometimes EC can be found in rare locations such as pancreas, testis and subcutaneous tissue [4-6]. A retroperitoneal location is exceptionally rare and the exact mechanism is not known. It speculates that the migration from the foregut to the retroperitoneal space during embryo development must play an important role. The nomenclature and classification are controversial [7]. Bronchogenic and foregut cyst are other names used for these lesions [8]. Ciliated epithelium and mucinous epithelium, both potentially of endodermal origin, are observed in EC.

EC has been reported to occur in a wide age, most of these cysts present in infancy or childhood and have been identified on the left side of the retroperitoneal region just as we report-

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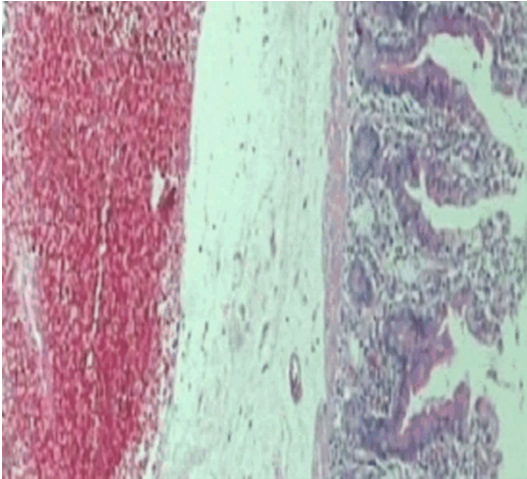


Figure 4. Histopathology showed the cysts' wall was composed of loosely textured vascular fibrous stroma and a few specialised gastric glands.

ed. To the best of our knowledge, there are less than ten ECs with a retroperitoneal location reported in the English literature. Clinical signs of retroperitoneal ECs are usually asymptomatic and non-specific, most cases have been diagnosed incidentally. Symptoms depend on location and size of the lesion, complications of cystic lesions (such as haemorrhage or infection) leading to swelling. Predominating signs are local pain, and the secretion of noxious and irritant cyst content may lead to inflammation, extension of the cyst may cause gastrointestinal symptoms and compression pain. Occasionally, EC simulates a slow-growing tumor, causing a progressive compression of adjacent structures. Moreover, three cases of adenocarcinomas have been reported as malignant degeneration in retroperitoneal enterogenous cyst [9-11].

It was controversial about embryology, histopathology and genetics of ectopic EC. EC may be diagnosed composed of tissues originating from 1, 2 or 3 embryonic layers. In early study, the pathological criteria proposed for the diagnosis of ECs include: the presence of alimentary mucous membrane lining, gastrointestinal mucosa is predominant in EC, a smooth muscle coat, and intimate attachment to some portion of the gastrointestinal tract [12]. Nevertheless, other research described EC lined by respiratory-type mucosa [13]. In our cases, there was no respiratory-type mucosa and no attachment to the digestive tract. A better understanding and further investigation are desired.

Preoperative diagnosis especially in the retroperitoneum is a challenge, because the lesions occasionally have overlapping clinical, radiological, and histologic features and can be mistaken for other cystic lesions such as cystadenoma, cystic teratoma, lymphangioma, hemangioma, Mullerian's tube cyst and other congenital cysts. To the best of our knowledge, although several histologically proven cases have been described in the literature, there was not a report on analysis of the clinico-radiological aspects of EC.

Radiological distinctions from those cystic lesions pose a diagnostic challenge. These unusual cysts should be further investigated radiologically. Imaging modalities available to evaluate suspected cases include to assess cysts and associated complications. When there is a diagnostic uncertainty, additional radiological investigations of CT scan or MRI can be of value and may guide the operative approach. CT scan may show better definition and characterization. An abdomen CT especially contrast enhancement forms the gold standard because of the ability to assess the cystic content and the surrounding structures. CT demonstrates a well-circumscribed, hypoattenuating homogeneous, cystic lesion while some may be in close proximity to the gastrointestinal tract. Those attachments to adjacent digestive tract organs can be falsely interpreted as stomach or a pancreatic pseudocyst. While secondary infection, hemorrhage, thick proteinaceous secretion and malignant transformation may demonstrate solid masses which lead to mistake for tumors. A false-negative diagnosis can be made when EC is complicated by infection, because the cystic wall might be irregularly thickened. In our study, the first case showed low attenuation of 4HU which corresponding to the serosity content of surgical specimen. In our second case, the multiple and predominantly iso-high attenuation EC mimicking solid masses, the CT value was higher and inhomogeneous, which was related to hemorrhage and inspissated mucoid material. MRI is another alternative modality, it can define the anatomical edge clearly. The cyst fluid will demonstrate mixed signal intensity on T1- and T2-weighted images or hypointense at T1 and hyperintense at T2, with signal uniformity similar to fluid [14]. These findings might help to differentiate EC from other cystic lesions. No enhancement found in the solid component

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would reflect hemorrhage. Consequently, directly guide appropriate surgical procedures may be required to make an exact diagnosis.

Because of the benign nature, some ECs are advised to take conservative treatment. Nevertheless, at least three malignant tumors arising from EC had been reported. In our opinion, the ideal treatment of ectopic lesions especially in retroperitoneum space should be resected totally for preventing infection or malignant transformation, minimally invasive approach such as laparoscopy had been proposed. In cases of difficulties to individualize the neighboring anatomical structures preoperatively, CT with 3D reconstruction is a helpful approach to determine the surgical strategy. The location, size and peculiarities of tumors are all determinative factors. At the time of surgery, total removal of the cystic wall is important to prevent cyst relapse with favorable long term prognosis [9].

In conclusion, the EC in the retroperitoneal region could lead to a mistaken and delayed diagnosis. The risk of EC causes inflammation or malignant transformation eventually requires total resection, the recognition of its radiological findings could facilitate timely diagnosis and treatment.

Disclosure of conflict of interest

None.

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