CASE REPORT

Retroperitoneal Enteric Duplication Cyst Presenting as a Pancreatic Cystic Lesion. A Case Report

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ABSTRACT

Context Retroperitoneal enteric duplication cysts (EDC) are rare lesions and its presentation during adulthood is a diagnostic challenge for clinicians. The diagnosis of this condition is established following histopathological analysis, which often requires surgical intervention.

Case report We report a case of a retroperitoneal enteric duplication cysts, presenting as a cystic lesion of the pancreas in a 19-year-old woman.

Conclusions We recommend surgical intervention for retroperitoneal EDCs due to its potential local complications including pressure effects on surrounding structures and neoplastic change.

INTRODUCTION

Enteric duplication cysts (EDCs) are rare congenital intra-abdominal cystic or tubular lesions composed of gastrointestinal mucosa with muscular and serosal layers. EDCs are more commonly associated with the oesophagus and small intestine [1, 2], but can occur throughout the gastrointestinal tract [1, 2, 3].

Retroperitoneal EDCs have been described but are extremely rare. Most previous cases have described prenatal detection with ultrasound [4, 5]. Reports of retroperitoneal EDCs diagnosis in adulthood are exceptionally uncommon [6, 7], and far from straightforward.

We present a case of a retroperitoneal EDC in a 19-year-old woman.

CASE REPORT

A 19-year-old Asian lady presented with a two-day history of sudden onset epigastric pain and vomiting. Past medical history was unremarkable other than involvement in a low impact road traffic accident five months earlier with no reported injuries. Clinical examination revealed epigastric tenderness and laboratory tests, including amylase, were all within normal limits.

Ultrasound scan of the abdomen highlighted a uniform level echo collection in the left upper quadrant, adjacent to the spleen. An abdominal computer tomography revealed an 11x5 cm retroperitoneal cystic lesion, which centred in the region of the pancreatic tail and extended along the greater and lesser curves of the stomach. Magnetic resonance imaging of the pancreas demonstrated a cystic mass arising from the pancreatic tail (Figure 1).

Endoscopic ultrasound guided biopsy was
unsuccessful. Overall, radiological diagnosis favoured a cystic lesion of the pancreas rather than a haemorrhagic pseudocyst secondary to trauma.

At operation, a large dumbbell shaped retroperitoneal cystic lesion arising anterior to, but separate from the left kidney and adrenal gland was identified. The cystic lesion extended posteriorly towards the spleen and pancreas, above the level of the diaphragmatic crus. Although the cystic lesion required dissection from the posterior wall of the stomach, there was no obvious communication with the gastrointestinal tract. There was no evidence of malignancy. Post-operatively, the patient made an uneventful recovery and was asymptomatic at six months follow-up.

The surgical resection specimen consisted of an 11x6.5x5 cm unilocular cystic tumour with a smooth inner surface and a viscous semi-solid content (Figure 2). Microscopically, the cyst wall showed a thin and a thicker smooth muscle, separated by a layer of loose fibrous tissue, consistent with the muscularis mucosae, submucosa and muscularis propria of the gastrointestinal tract. There was patchy transmural chronic inflammation. The mucosal lining varied from flattened cuboidal to ciliated or mucinous high columnar epithelium. In addition, there were areas of squamous epithelial lining and foci with formation of gastric type mucosa that included oxyntic glands (Figure 3). There was no evidence of dysplasia or malignancy.

DISCUSSION

Retroperitoneal EDCs are rare cystic congenital lesions. They are usually detected early in life [4, 5] while diagnosis in adulthood is extremely rare [6, 7]. EDCs are believed to result from disturbances in embryonic development. Although several pathogenic mechanisms have been postulated [8, 9, 10] for the formation of EDCs at various anatomical sites, none have been fully satisfactory. Environmental stresses such as intra-uterine hypoxia or trauma have been recently implicated as important contributory factors [11]. In retrospect, trauma is unlikely to be a contributory factor in our case. EDCs are generally spherical or nearly spherical in shape and 18% show a tubular configuration [12]. Retroperitoneal EDCs have a higher tendency to be tubular [7]. The cystic lesion in our case was of a dumbbell shape.
configuration. It is believed that this shape is due to fixation of the cyst to both the bowel and retroperitoneum, and that the unusual position is acquired during bowel rotation in the first trimester [4].

Clinical features are determined by the location, growth and mucosal lining of the EDC [7]. In the present case, the epigastric pain and vomiting were the result of compression of the stomach by the cystic lesion.

In the absence of obvious underlying pathology to account for reported symptoms, ultrasound scan of the abdomen is an ideal, non-invasive first-line investigation, followed by computer tomography and magnetic resonance imaging studies. In tandem with other reports, imaging studies used in the present case were able to delineate the mass, but failed to identify the nature of the cystic lesion. Malignant transformation has been reported in cases of EDCs [1], although there have been no such reports concerning retroperitoneal EDC.

The differential diagnosis in these cases includes all cystic neoplasms of the pancreas and tumour-like lesions with cystic features, as suggested by Kosmahl et al. [13]. This extensive pathological classification includes neoplastic and non-neoplastic lesions of epithelial and non-epithelial origin such as pancreatitis-associated pseudocyst, intraductal papillary-mucinous neoplasm, cystadenocarcinoma and lymphangioma [13]. Management of a retroperitoneal cystic lesion is determined by the underlying pathology. Retroperitoneal cystic lesions, malignant or benign, can be technically difficult to excise because of the proximity to major vessels or other organs. In cases where an underlying pathology cannot be elucidated on radiological modalities, i.e. where suspicion of cystic malignancy remains, careful pre-operative mapping and diligent surgical clearance is appropriate. In all reported cases of retroperitoneal EDCs, there was no mortality or morbidity associated with surgical resection [4, 5, 6, 7].

**CONCLUSION**

Based on literature, EDCs represents a rare clinical condition whose diagnosis can only be established following histopathological analysis, and which often requires radical
surgical intervention. The differential diagnosis includes a number of other rare cystic lesions, and detailed imaging is crucial to investigate and delineate the underlying pathology and appropriate management strategies. We recommend surgical intervention for retroperitoneal EDCs due to its potential local complications including pressure effects on surrounding structures and neoplastic change.

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Abbreviations  EDC: enteric duplication cyst

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References