

Primary retroperitoneal mucinous cystadenoma adjacent to the kidney: report of two cases and review

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Summary

We report two cases of primary retroperitoneal mucinous cystadenomas mistaken as renal cysts and discuss the differential diagnosis and management of this tumor.

Key words: Retroperitoneal tumors; Mucinous cystadenoma; Renal cyst.

Introduction

Primary retroperitoneal mucinous cystadenomas (RMCs) are extremely rare tumors and the exact etiology is unclear. The diagnosis is never made preoperatively and preoperative abdominal imaging studies can only detect a cystic mass in the retroperitoneal space. A huge retroperitoneal mucinous cystadenoma adjacent to the kidney region always presents as a renal cyst.

Case Reports

Case 1: A 52-year-old woman was admitted to the Department of Urology for right lumbago of one month's duration. No quadrant mass was palpable at the abdominal examination. The percussion tenderness of the right kidney region was positive. Ultrasound (US) examination of the abdomen revealed a cystic mass measuring 14 cm in diameter associated with the right kidney. Intravenous urography (IVU) showed a huge soft tissue mass located below the right kidney and the right kidney was pushed up (Figure 1). Abdominal computed tomography (CT) revealed a 10 × 9.0 × 10.5 cm homogeneous cystic mass located in the inferior pole of the right kidney (Figure 2). The cystic mass was thought to be a right renal cyst. Therefore in May 26, 2006 laparoscopic marsupurization of the right renal cyst was performed. However the operation revealed a large retroperitoneal mass arising superiorly from the region of the lower pole of the right kidney, extending down into the right iliac fossa. The cystic retroperitoneal mass was completely removed and the cystic cavity was filled with pallide-flavens fluid. The pathologic examination showed a cyst measuring 10.0 × 8.5 cm. Its wall measured 0.2 cm in thickness and the luminal surface was smooth. The microscopic exam showed the wall of the cyst consisted of fibrous connective tissue lined by a single layer of benign mucinous columnar epithelium. There was no ovarian tissue in the submitted sections (Figure 3). The postoperative course was uneventful and the patient has remained asymptomatic after five years of follow-up.

Case 2: A 43-year-old woman was admitted to the Department of Urology due to findings of a retroperitoneal cyst during a health examination. No quadrant mass was palpable at the abdominal examination. The percussion tenderness of the

right kidney region was negative. US examination of the abdomen revealed a cystic mass measuring 6 cm in diameter adjacent to the kidney. Abdominal CT revealed a 6 × 5.0 × 5.5 cm homogeneous hypodense mass located in the middle pole of the right kidney (Figure 4). The cystic mass was thought to be a right renal cyst. In March 12, 2009 laparoscopic marsupurization of right renal cyst was performed. However the operation revealed that the retroperitoneal mass was separated from the right kidney. The cystic retroperitoneal mass was completely removed and the cystic cavity was filled with a pallide-flavens fluid. Pathologic examination showed a cyst measuring 5.0 × 4.5 cm. Its wall measured 0.1 cm in thickness and the luminal surface was smooth. Microscopic exam showed the wall of the cyst consisted of fibrous connective tissue lined by a single layer of benign mucinous columnar epithelium (Figure 5). The postoperative course was uneventful and the patient has remained asymptomatic after two years follow-up.

Discussion

The most common retroperitoneal mucinous cystadenomas (RMCs) are frequently ovarian tumors which share a histological similarity to ovarian mucinous cystadenomas but can arise in any location in the retroperitoneum without attachment to the ovary [1]. Primary RMCs are rare and occur only in female patients [2]. The histogenesis of primary mucinous cystadenomas of the retroperitoneum is not very clear, however some hypotheses have been proposed to explain the origin. The most plausible theory is that these tumors arise from inclusions of mesothelial cells with mucinous metaplasia [1, 3-5]. Pennel and co-workers that mucinous cystadenomas can arise from ectopic supernumerary ovaries [3, 6] or from teratomas [3].

To our knowledge, no more than 30 cases have been reported as primary RMCs in the literature. Since primary RMCs have the potential toward a phase of progression to malignancy, early diagnosis is very important [7]. With the use of US, CT and MRI the detection of retroperitoneal cysts is possible. However the diagnosis is difficult preoperatively since RMCs are often mistaken for ovarian cysts, cystic lymphangiomas, mesenteric cysts, hydatid cysts and renal cysts [3, 8].

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Fig. 1

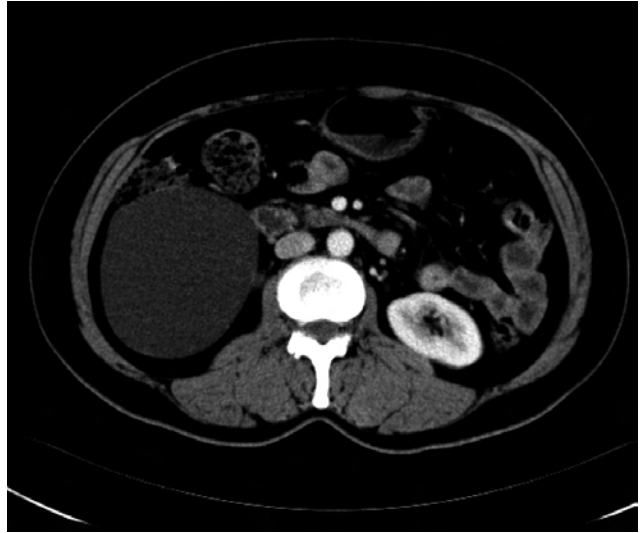


Fig. 2

Fig. 3

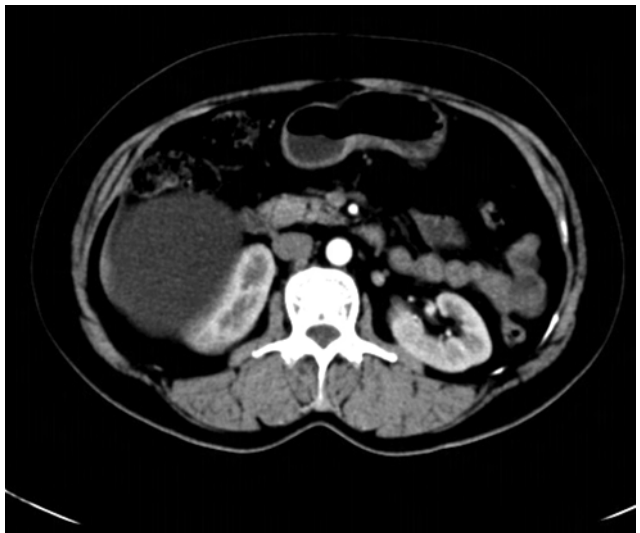
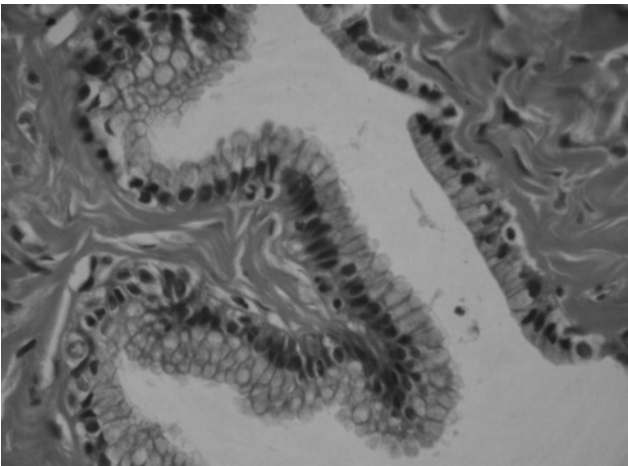


Fig. 4

Fig. 5

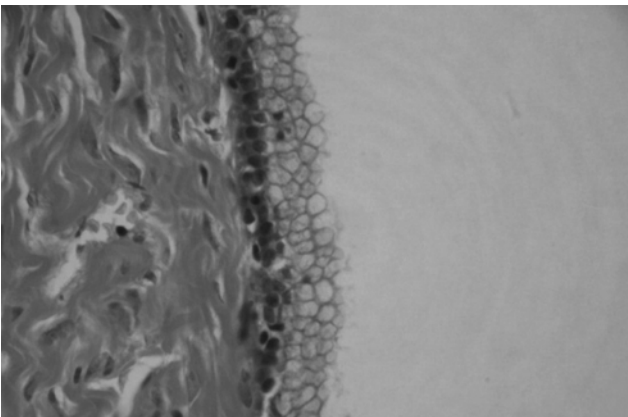


Figure 1. — IVU showed a huge soft tissue mass located below the right kidney and the right kidney was pushed up.

Figure 2. — Abdominal CT revealed a 10 × 9.0 × 10.5 cm homogeneous hypodense cystic mass located in the inferior pole of the right kidney.

Figure 3. — Wall of the cyst consisting of fibrous connective tissue lined by a single layer of benign mucinous columnar epithelium.

Figure 4. — Abdominal CT revealed a 6 × 5.0 × 5.5 cm homogeneous hypodense mass located in the middle pole of the right kidney.

Figure 5. — Wall of the cyst consisting of fibrous connective tissue lined by single layer of benign mucinous columnar epithelium.

A retroperitoneal mucinous cystadenoma associated with the kidney can easily be mistaken for a huge renal cyst. Interestingly, among these cases, two cases were considered as renal cysts preoperatively [3, 9-11]. The two cases we reported here both presented as renal cysts preoperatively.

Preoperative diagnosis of these tumors is very difficult. There is no relationship between the age of patients and the size of tumors. The symptoms are nonspecific and most of the patients complained of an asymptomatic mass or abdominal discomfort [12]. Aspiration is a good method to delineate the nature of the cyst, but cytology of the aspirated fluid frequently fails to reveal the cell type of the epithelial cells of the cyst lining. Progression to malignancy cannot be prevented, these it is not a very suitable method for diagnosis and treatment. As for the management of primary RMCs, complete surgical excision is recommended to eliminate the risk of infection, recurrence, and malignant degeneration [2, 13]. Today with the development of the laparoscopic technique, exploratory laparotomy with complete enucleation of the cyst is usually used for both diagnosis and treatment. Laparoscopic resection of primary RMCs was accomplished in our cases. The advancement of laparoscopic surgery offers the surgeon a useful option to remove a retroperitoneal cystic mass with further advantages including less postoperative pain, lower morbidity, shorter hospitalization, and an earlier recovery [5]. However when malignancy is suspected, laparoscopic excision may not be appropriate as decompression of the cystic mass is inevitable when it is removed through a trocar.

In conclusion, retroperitoneal mucinous cystadenoma adjacent to the kidney region always presents as a renal cyst. When confronted with a cystic mass in the retroperitoneum, a primary RMC should be included in the list of differential diagnoses.

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