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Laparoscopic excision of primary retroperitoneal mucinous cystadenoma and malignant predicting factors derived from literature review

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ABSTRACT

INTRODUCTION: Primary retroperitoneal mucinous cystic neoplasm is very rare and its histogenesis is unclear.
PRESENTATION OF CASE: This paper presents the case of a 31-year-old female in whom an incidentally detected retroperitoneal cystic mass, 6.5 cm in size, was successfully resected through laparoscopic approach. Pathologic examination revealed a mucinous cystadenoma. The postoperative course was uneventful, and the patient remained free of recurrence six months after surgery.
DISCUSSION: Literature review showed that male sex ($p = 0.019$), and solid nodules in cysts ($p < 0.001$) were both significantly associated with malignancy.
CONCLUSION: When confronted with a cystic mass in the retroperitoneum, a primary mucinous cystic neoplasm should be considered and complete surgical removal of the tumor without spillage is recommended.

1. Introduction

Primary retroperitoneal mucinous cystic neoplasm is very rare. Although mucinous cystic neoplasm is common ovarian tumor and is grossly and microscopically similar to retroperitoneal mucinous cystic neoplasm [1], because of the limited number of reported cases, the histogenesis of primary retroperitoneal mucinous cystic neoplasm is unclear and establishing a preoperative diagnosis is very difficult. We report a case of primary retroperitoneal mucinous cystadenoma and further provide a literature review.

2. Presentation of case

A 31-year-old female was admitted due to pain in the right upper quadrant. She had an unremarkable medical history. Physical examination revealed that her BMI was in the normal range (height 163 cm, weight 50 kg) with a blood pressure of 110/60 mmHg, a pulse of 80 beats/min, and a body temperature of 36.4 C. Her abdomen was tender in the right upper quadrant but no guarding and rebound tenderness were noted. Laboratory findings including liver function test and serum tumor markers (CEA, CA125, and CA19-9) were within the normal limits. Computed tomography revealed multiple tiny gallstones with segmental gallbladder wall thickening and a 6.5 cm sized multiloculated cystic mass in the left retroperitoneal space just below the spleen (Fig 1). The preoperative diagnoses were either retroperitoneal tumor such as cystic mesothelioma, mucinous cystadenoma, or cystic lymphangioma and symptomatic gallstone with chronic cholecystitis. We proceeded to perform laparoscopic excision of the cystic mass and then cholecystectomy. The patient was placed on the operating room table in the right semi-lateral decubitus position (to a 45-degree angle to the operating table). An 11 mm trocar was inserted in the infraumbilical region and two more 5 mm trocars were inserted 3 cm below the costal margin at the left anterior axillary line and 4 cm below the xyphoid process. Laparoscopy revealed a large, well-demarcated cystic mass behind the inferior pole of the spleen that seemed to have no apparent attachment to the adjacent structures such as the pancreas, spleen, colon, and etc. The tumor was completely excised without spillage of cystic fluid. Laparoscopic cholecystectomy was then performed in the usual manner with one more 5 mm trocar 3 cm below the right costal margin at the mid-clavicular line. The mass and gallbladder were placed in a Lap-bag (Sejong Medical, Paju, Korea) and were retrieved via the umbilical port site with extension of incision upto 2 cm. The total operation time was 80 minutes. Upon

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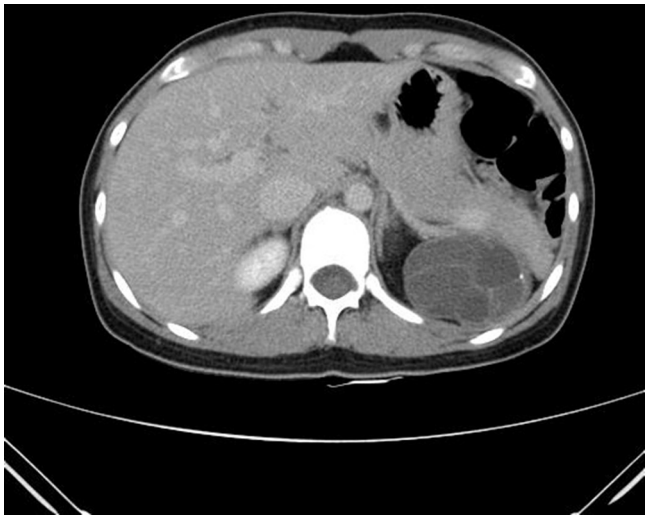

Fig. 1. Abdominal computed tomography shows a 6.5 cm sized multiloculated cystic mass in left retroprertoneal space just below spleen.

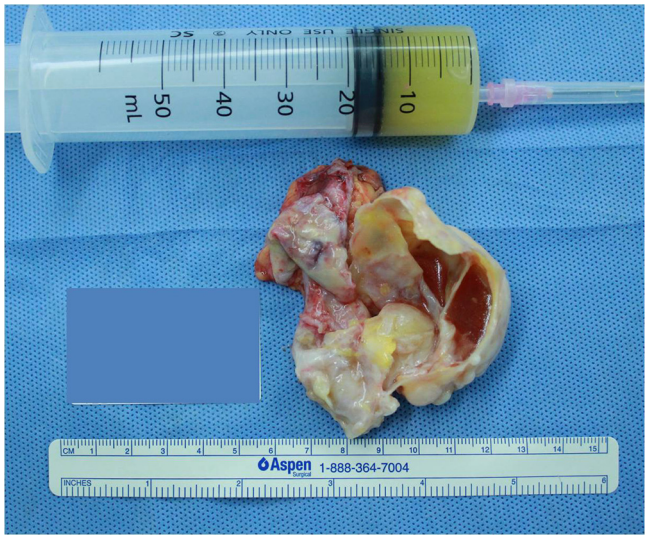

Fig. 2. Glossly it shows a 6.5 × 5.5 × 0.4 cm unilocular cyst with smooth inner surface containing yellowish viscous fluid.

gross inspection, the specimen was a thick-walled, multilocular cyst with a smooth and glistening internal surface (Fig 2). It was filled with slightly turbid, mucoid, and yellowish fluid. When microscopically examined, the lining of the cyst wall consisted of a single layer of columnar epithelium with abundant intracellular mucin and basally arranged small nuclei (Fig 3). Ovarian-like mesenchymal stroma was not seen. There was also no evidence of a malignant or borderline lesion, and the final diagnosis was thus primary retroperitoneal mucinous cystadenoma. The postoperative course was uneventful and the patient was discharged on postoperative day 5. During the six month follow-up period, the patient remained completely free of symptoms and without evidence of recurrence.

3. Discussion

Primary retroperitoneal mucinous cystic neoplasm is a rare cystic lesion for which only single case reports [1–32] and small series [33–36] have been published. This neoplasm shares a histological similarity to ovarian mucinous cystic neoplasm but can arise at any

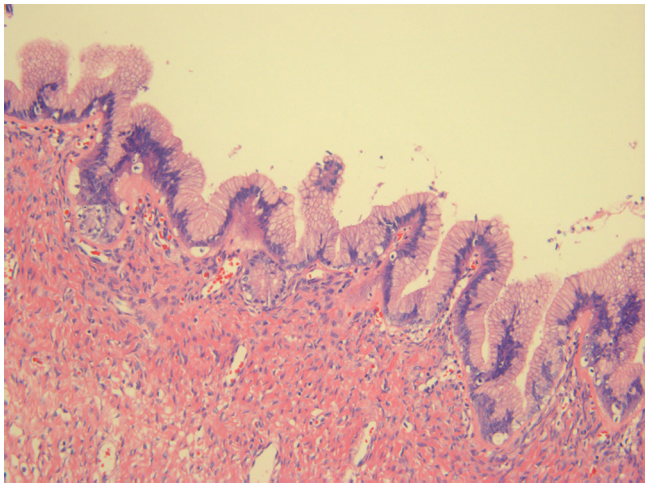

Fig. 3. Multilocular cyst lined by a single layer of tall columnar cells. The stroma consisted of fibrocollagenous tissue with no pancreatic or ovarian components (H&E, ×200).

location in the retroperitoneum without attachment to the ovary [1].

The histogenesis of primary retroperitoneal mucinous cystic neoplasm remains unclear; however, four main theories have been proposed to explain its potential origin. First, these tumors may arise from teratoma, in which the columnar epithelium has overridden all other components to survive as the single cell component [1,2]. Another possibility is that they arise from ectopic ovarian tissue because they have resemblances to ovarian mucinous cystadenomas, which may explain why they occur mainly in women [3]. Until now only two cases of primary retroperitoneal mucinous cystic neoplasm in men have been reported in the literature [12,13]. However, ovarian remnants have not been identified in the wall of the cysts [4,5]. A third theory is that they arise from remnants of the embryonal urogenital apparatus [6–8]. However, the most plausible theory is that mucinous cystic neoplasm arises from invaginations of the peritoneal mesothelium, with subsequent endocervical metaplasia [1,2]. To support this theory, in these neoplasms the immunohistochemical staining for estrogen or progesterone is negative and there has been no gut-like muscle found in the pathologic specimens.

We performed a literature review using Medline searching from 1970 to 2013, and including the present study, a total of 56 cases [1–37] of primary mucinous cystic neoplasm of the retroperitoneum were identified and reviewed in terms of their clinicopathologic characteristics. Among these 56 cases, 54 cases were women and only two cases [12,13] were men. The median age of the patients was 39 years with wide range from 14 to 85 years. The size of the reported tumors ranged from 5 cm to 30 cm (median 12 cm). Most patients ($n = 24$, 51.1%) complained of abdominal mass. However, 14 patients (29.8%) complained of abdominal pain and 9 cases (19.1%) were detected incidentally. Postoperative pathology showed 31 cases (55.4%) of cystadenoma, 10 cases (17.9%) of moderate dysplasia, and 15 cases (26.8%) of cystadenocarcinoma. Both of the two male patients had cystadenocarcinoma and 9 (60.0%) of the 15 cystadenocarcinomas showed solid nodules in cysts on the preoperative images and in the gross specimen. Univariate analyses showed that male sex ($p = 0.019$), and solid nodules in cysts ($p < 0.001$) were both significantly associated with malignancy. In multivariate analysis, the presence of solid nodules in the cysts was the only significant predictive factor of malignancy ($p < 0.001$). In most cases of cystadenocarcinoma, the prognosis seemed to be not so bad. Among the 15 cystadenocarcinoma cases, follow-up data was available for 10 cases. In two