Primary mucinous cystadenomas are rare retroperitoneal tumors that are exclusively found in women. The histogenesis of these tumors is still uncertain, though mucinous metaplasia of the mesothelial lining cells is the most likely hypothesis. We report a 19-year-old pregnant female, initially believed to have a severe hydrenephrosis of left kidney, but was finally diagnosed as a retroperitoneal mucinous cystadenoma concomitant with an atrophic left kidney.

Key words: Atrophic kidney, Hydrenephrosis, pregnancy; retroperitoneal mucinous cystadenoma

Retroperitoneal mucinous cystadenomas are extremely rare, which was first reported in Europe [1] and only twenty-five cases had been reported in the literature [2]. Diagnosis cannot be established preoperatively by imaging modalities, since most primary retroperitoneal mucinous cystadenomas are usually mistaken for cystic lymphangiomas [3]. In Japan, aspiration cytology was tested in two cases of retroperitoneal mucinous tumor. The carciinoembryonic antigen levels in the cystic fluid were determined in order to increase diagnostic accuracy [4]. Surgery is the only treatment.

CASE REPORT

A 19-year-old woman, gravida 1, para 0, was diagnosed to have a retroperitoneal cystic lesion (Fig. 1) during a routine ultrasound examination at the twelfth week of gestation in a community hospital. The normal-shaped left kidney was not identified. She had a pertinent surgical history of resection of an ovarian tumor in her infancy. The nature of the tumor was not ascertained. She had no obvious clinical symptoms or other remarkable findings appearing in either physical examination or routine laboratory investigations.

She had been regularly followed up for three months until progressive enlargement of the homogeneous cystic lesion occupying the left retroperitoneum was noted on a conventional MRI study (Fig. 2). The normal-shaped left kidney was still not identified and no further contrast study was performed due to the pregnancy. The initial impression was severe hydrenephrosis of left kidney because of ureteropelvic junction obstruction. We performed an aspiration of the cystic lesion under ultrasonic guidance. Instead of urine some mucinous fluid had been aspirated.

The woman had both an uneventful pregnancy and a normal spontaneous delivery. The effective renal
plasma flow (ERPF) was 30.6 ml/min in the left kidney and 520.1 ml/min in the right one month after delivery. A retrograde pyelography (RP) showed an atrophic left kidney (Fig. 3) upward displaced by a retroperitoneal mass. Under the preoperative diagnosis of a retroperitoneal cystic tumor concomitant with an atrophic left kidney, she received surgery. Both the tumor and the atrophic left kidney were removed successfully. The primary site of the tumor remained unconfirmed. The tumor measured $14 \times 14 \times 4$ cm$^3$ and weighed 184 gm with small papillary enfolding. The left kidney measured $4 \times 2.5 \times 2$ cm$^3$ and weighed 33 gm. Pathological studies confirmed a mucinous cystadenoma with borderline malignancy (Fig. 4). The atrophic left kidney also had focal interstitial inflammation. The patient experienced a prompt recovery without evidence of tumor recurrence for 8 months.

**DISCUSSION**

The most popular hypothesis for the histogenesis

![Figure 1. Sonography. There is a huge cystic lesion with internal echoes in the left retroperitoneum.](image1)

![Figure 2. Coronal T2-weighted MRI (Fast spin echo 3337/129.6/1). There is a large cystic lesion (arrow) in the left retroperitoneum. Normal-shaped left kidney is not identified.](image2)

![Figure 3. Retrograde pyelography shows an atrophic left kidney (arrow).](image3)

![Figure 4. Histology (H&E, 200X) shows mucin-producing cells (arrow) mimicking gastrointestinal lining cells.](image4)
of retroperitoneal mucinous cystadenomas is metaplasia of the mesothelium [5]. Other possibilities include generation from teratomatous lesions [6] or ectopic ovarian tissues [7]. The later hypothesis may explain its exclusive occurrence in women. Although the presence of ovarian tissues is rarely documented, Rothacker et al found no difference in immunohistochemical examination between retroperitoneal mucinous cystadenomas and ovarian mucinous cystadenomas [7].

On the basis of the review of the literatures, there are three pathologic types of retroperitoneal mucinous cystadenoma. The most common type is characterized by a large unilocular or multilocular cyst [7, 8, 9]. In the second type, the tumors resemble ovarian cystadenoma and may contain foci of proliferative columnar epithelium [1, 4]. The third type is malignant mucinous cystadenocarcinoma [5], which warrants adequate sampling of the lining epithelium and careful microscopic examination for proper treatment.

Renal cysts, mesenteric cysts, and ovarian cysts had been reported as initial diagnoses of retroperitoneal mucinous cystadenomas [5, 9, 10, 11]. It would be difficult to recognize a cystic tumor, especially one concomitant with an atrophic kidney, in routine examinations such as sonography or conventional MRI. The case that we have reported here was believed to have a left ureteropelvic junction obstruction with a severe hydronephrosis until the retrograde pyelography was performed. MR angiography or contrast enhanced thin-slice CT may be useful in identifying the atrophic kidney through tracing of renal artery. Retroperitoneal mucinous cystadenomas concomitant with atrophic kidneys as presentation have not, to our knowledge, been reported in the literature. The pathogenesis of the atrophic kidney may be chronic obstructive nephropathy, caused by the large retroperitoneal tumor, or other miscellaneous insults.

Pregnant women are always concerned about radiation examinations, and, therefore, sonography and MRI are the modalities of choice for investigations. Other diagnostic tools, such as retrograde pyelography and cyst aspiration, may be required for differentiation of retroperitoneal cystic lesions.

REFERENCES

懷孕婦女後腹腔黏液性囊腺瘤合併腎臟萎縮：
病例報告

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原發性黏液性囊腺瘤為非常罕見的後腹腔腫瘤，僅發生於女性，其組織學來源仍未確定，
中胚層內襯細胞的黏液性不正常分化是最廣為人知的假說。我們報告一位 19 歲懷孕婦女，罹患
後腹腔黏液性囊腺瘤合併左腎萎縮，最初診斷懷疑左腎重度水腫。

關鍵詞：腎萎縮，水腫，懷孕，後腹腔黏液性囊腺瘤