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Case Report

Retroperitoneal primary adenocarcinoma of Mullerian origin: case report with radiology review ☆

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ABSTRACT

Retroperitoneum Mullerian neoplasms are extremely rare malignancies. We report a case of a 35-year-old woman who presented with a 12-year history of lower abdominal cystic lesion, presumed of renal origin and benign, as such was not followed for 10 years. Prior to pregnancy, the patient received additional imaging and the lesion was again redemonstrated and questioned to be of ovarian origin. As such, the patient underwent laparoscopy for planned cystectomy and was found to have normal ovaries. After pregnancy, the lesion had increased in size and surgical excision revealed that the cystic mass was retroperitoneal in origin. The histopathology was reported as microinvasive mucinous adenocarcinoma with Mullerian origin. Positron emission tomographic scanning, colonoscopy, and endoscopy were unrevealing. Tumor markers were followed and follow-up scans demonstrated no recurrence.

The preoperative diagnosis of primary retroperitoneal adenocarcinomas of Mullerian origin is difficult, and a definitive diagnosis cannot be made without postsurgical histopathological analysis. However, it is important for radiologists to recognize imaging features of this entity and include it in the differential diagnosis. Here we report a case and review imaging features reported for retroperitoneal primary adenocarcinomas of Mullerian origin.

Introduction

Retroperitoneal primary adenocarcinomas of Mullerian origin are extremely rare entities. Majority of cases have been reported in females, but some have also been reported in males.

We present a case of primary retroperitoneal adenocarcinomas of Mullerian origin in a 35-year-old woman. We provide a case review of radiologic features seen in this entity. A comprehensive analysis of primary retroperitoneal Mullerian adenocarcinoma radiologic features will aid to improve the current understanding of this rare malignancy and help

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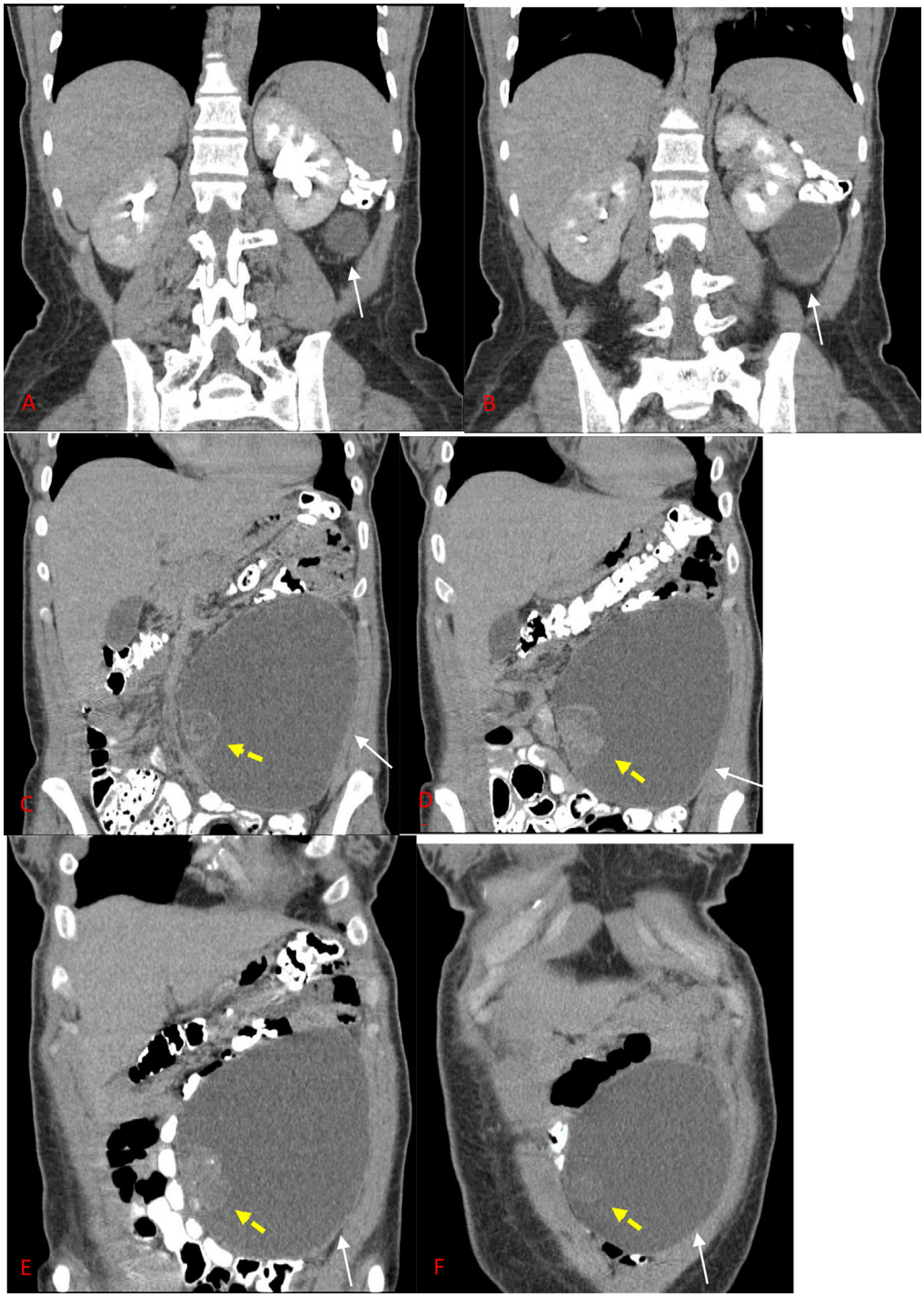

Fig. 1 – Demonstrates a large cystic and solid mass (white arrow) located within the left hemiabdomen with the superior portion abutting the lower pole of the left kidney. The solid component is mildly enhancing (yellow arrow). The mass extended into the pelvis and resulted in outward bulging of the left abdominal wall. The lesion resulted in displacement of the left colon medially, as well as, displacement of the aorta and IVC to the right. There was additional compression of the left ureter causing moderate hydronephrosis. The lesion measured 17 x 14.5 x 9.8 cm and was predominantly cystic, however, there was a 5.3 x 3.2 x 5 cm solid enhancing component. Axial pre-contrast (G), postcontrast (H), and 8-minute delay postcontrast (I) further shows the mass (white arrows) flattening the left psoas muscle (red asterisk). There is enhancement of the solid component (yellow arrow) on postcontrast images and persistent enhancement (yellow arrow) on delayed imaging.

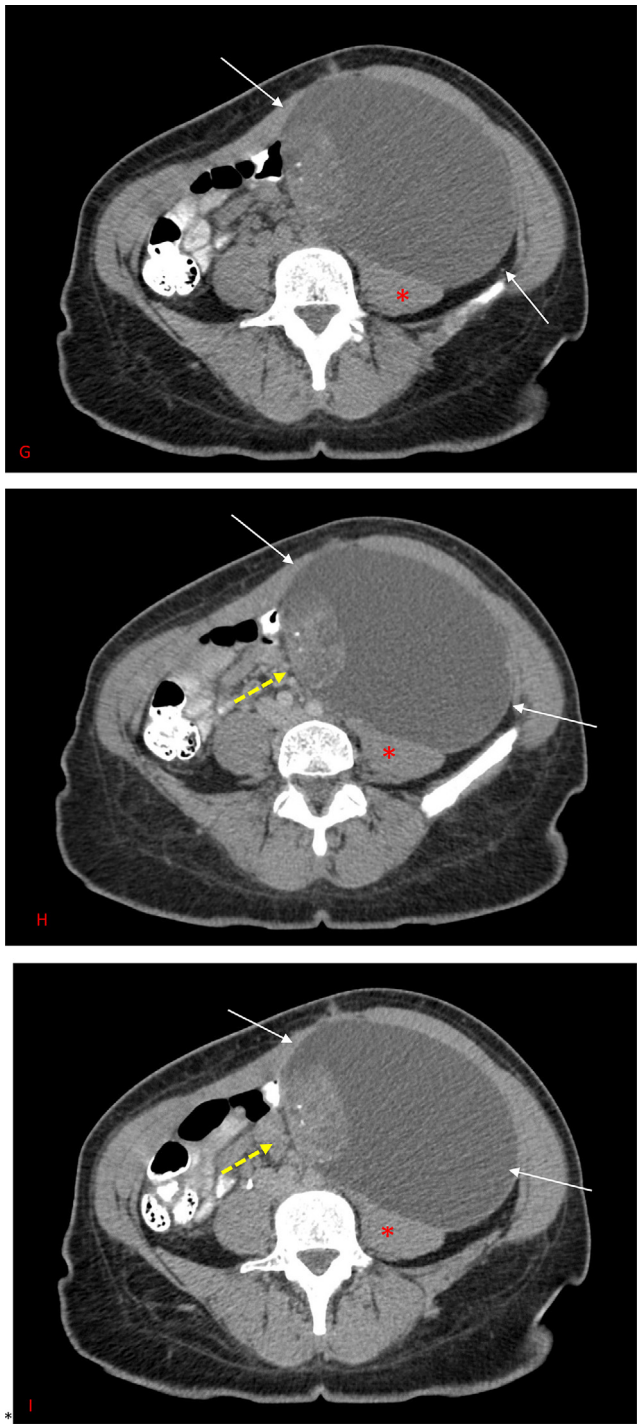

Fig. 1 – Continued

radiologists to better recognize and narrow the differential diagnosis based on imaging features.

Case

A 35-year-old female G1, P1 with a history of a presumed left renal cyst incidentally found 10 years prior presents with a palpable left abdominal mass for the past several months.

On initial presentation, patient received a CT abdomen and pelvis exam to rule out appendicitis. It was reported that there was an 8.5 × 7.1 cm hypoattenuating lesion with HU of 5 anterior to the lower pole of the left kidney, displacing the left colon anteriorly. The lesion was presumed to be a simple cyst at that time. Ten years later, the patient reported having follow-up imaging prior to pursuing pregnancy and mentioned the cystic lesion was located more towards her ovary and laparoscopic cystectomy was recommended. She underwent diagnostic laparoscopy and the ovaries appeared normal, with the

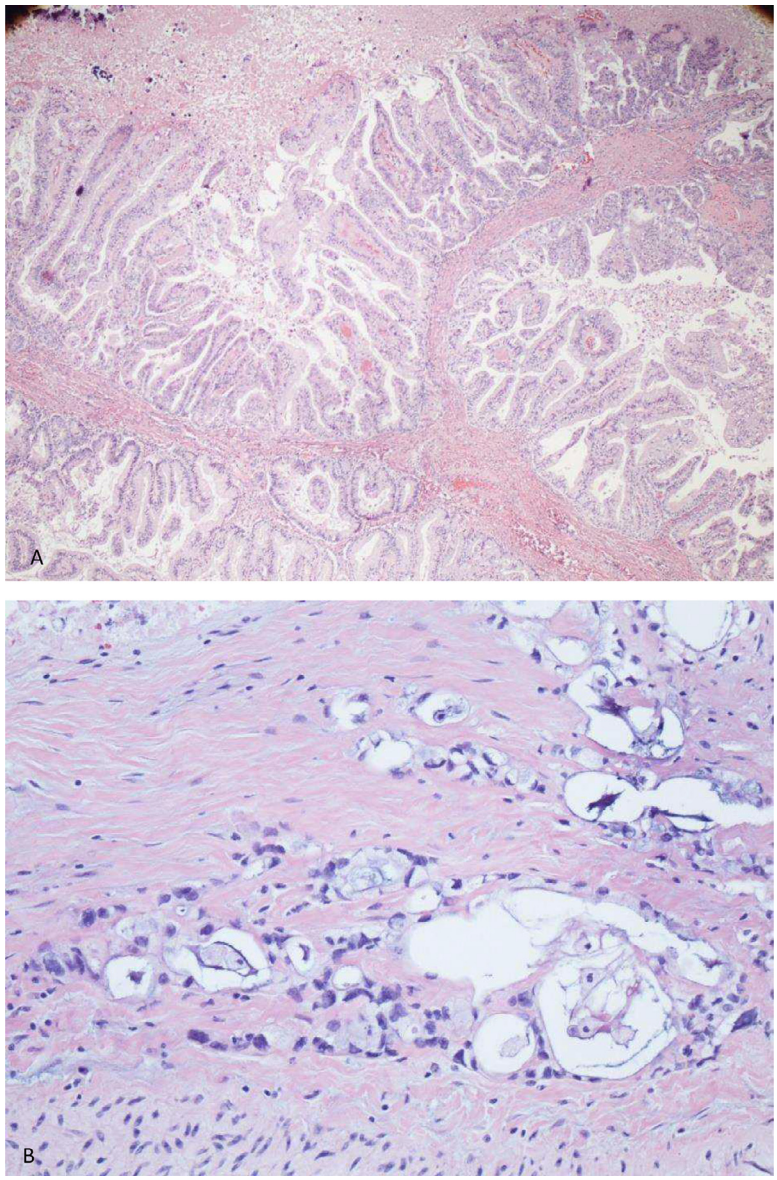

Fig. 2 – Images A (Low Power Photomicrograph at 20× magnification) and B (High Power Photomicrograph at 40× magnification) with hematoxylin and eosin stain. Photomicrograph demonstrates mucinous neoplasm with cytologic atypia, complex papillary growth, and multiple infiltrative foci of invasive carcinoma

mass appeared to be retroperitoneal. She then pursued pregnancy and at 22-week pregnant, she had a retroperitoneal ultrasound and was found to have a 16.7-cm exophytic cyst appearing to arise from the lower pole of the left kidney. Her pregnancy did not have any complications and she delivered a boy. After delivery of her son and weight loss, she then began to feel left abdominal swelling and pursued repeat imaging, which revealed a large cystic and solid mass located within the left hemiabdomen with the superior portion abutting the lower pole of the left kidney (Fig. 1 B). The mass extended into the pelvis and resulted in outward bulging of the left abdominal wall (Fig. 1 G and H). The lesion resulted in displacement of the left colon medially and flattened the left psoas muscle, as well as displacement of the aorta and IVC to the right (Fig. 1 A-H). There was additional compression of the left ureter causing moderate hydronephrosis. The lesion measured 17 × 14.5 × 9.8 cm and was predominantly cystic, however, there was a 5.3 × 3.2 × 5 cm solid enhancing component (Fig. 1 H and I). The patient was referred to urology and was recommended to undergo an exploratory laparotomy with excision.

During exploratory laparotomy, the mass was seen to be freely mobile and demonstrated small investing vascularity. It did not attach to the kidney, ureter, or psoas muscle. It appeared most attached to the mesentery to the descending colon. The mass was excised whole in its entirety and without rupture and was sent for pathology which revealed microinvasive mucinous adenocarcinoma with Mullerian origin. The postoperative period was uneventful, and the patient was discharged home the following day after surgery.

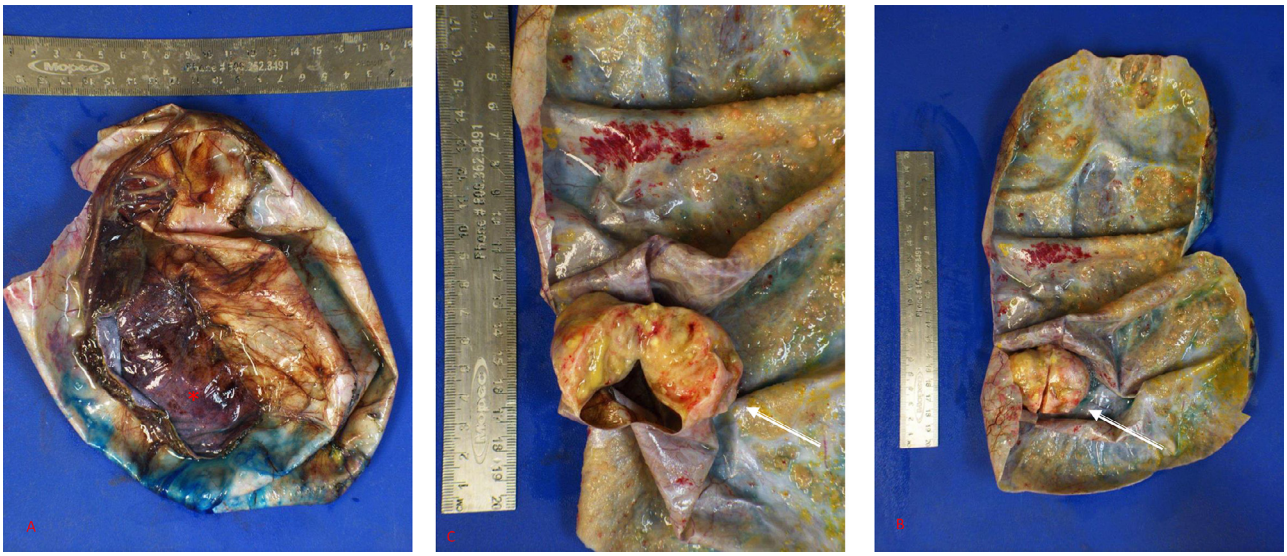

Fig. 3 – Shows thin-walled cystic structure measuring 18.5 x 15.5 x 12.5 cm, focally covered by pink, red congested fibro-membranous tissue consistent with peritoneum measuring 13.0 x 7.0 cm in surface area. The cyst contains brown, semi-cloudy fluid (red asterisk). The cyst lining demonstrates diffusely scattered areas of pink tan and is granular to nodular appearance, with the largest nodule measuring 6.0 x 5.0 x 2.5 cm (white arrow).

The gross pathological findings showed an intact and thin-walled cystic structure measuring 18.5 × 15.5 × 12.5 cm and weighing 1804 g, focally covered by pink, red congested fibro-membranous tissue consistent with peritoneum measuring 13.0 × 7.0 cm in surface area. Upon opening, the cyst contained brown, semi-cloudy fluid. The cyst lining was predominantly smooth with diffusely scattered areas of pink tan, granular to nodular appearance, with the largest nodule measuring 6.0 × 5.0 × 2.5 cm. Upon sectioning, the nodule has a pink-tan, fleshy, solid, and heterogeneous cut surface with a cystic degenerated component and was grossly noted to abut the outer surface. The remaining cyst wall measures 0.1 cm in maximum thickness (Figure 3 A-C). Microscopically, the solid and cystic mass consisted of a mucinous neoplasm with cytologic atypia, complex papillary growth, and multiple infiltrative foci of invasive carcinoma, with each invasive focus measuring less than 5 mm (Figure 2 A and B).

Immunohistochemical profile demonstrated positive ER and PR immunostains within some stromal cells, consistent with an ovarian-type stroma. In addition, the tumor cells were positive for cytokeratin 7 and cytokeratin 20. The cells were negative for CDX2, SATB2, gAtA-3, and TtF1. The overall findings were most consistent with Mullerian origin, however, clinical correlation was recommended to exclude a gastrointestinal primary.

Three weeks after surgery, patient underwent a PET scan which demonstrated no abnormal FDG uptake to suggest a FDG avid tumor. The patient subsequently had a colonoscopy and endoscopy to further exclude gastrointestinal primary which demonstrated no abnormalities. A month later, she had a retroperitoneal ultrasound which was documented to show no abnormalities. CA 19-9, CEA, and CA-125 markers were obtained 3 months after surgery and resulted as 17, 1.1, and 25.1 U/mL respectively. Two months follow-up for lab markers were relatively stable and resulted 15, 1.2, and 26.1, respectively. A follow-up CT abdomen and pelvis 5 months after surgery demonstrated no evidence of residual tumor or metastatic disease. Patient desired continued fertility and close observation was recommended and patient was encouraged not to resume fertility efforts for 1 year.

Discussion

Adenocarcinomas of Mullerian origin are malignancies that rarely occur in the retroperitoneum. Clinical findings are nonspecific, and patient may present with abdominal pain, swelling, and/or palpable abdominal mass.

There are several theories of the origin of these tumors including ectopic, supernumerary ovarian tissue, and endometriosis implants [2,8]. In addition, other theories postulate metaplasia of celomic epithelial cells which have deposited in the retroperitoneal area and Mullerian differentiation of peritoneal epithelium [2].

There are no specific established radiologic findings for adenocarcinomas of Mullerian origin. As in our case, a previously reported case on CT demonstrated a large hypoattenuating mass with mildly enhancing solid component [2,3,10,12,16]. Further, the solid component was seen to be in the cystic wall [2,12,16]. In addition, the solid components demonstrate progressive enhancement in our case and had thin calcifications [12]. However, it is important to note, in our case, when the patient initially presented years prior, the mass was found as an incidental finding and did not demonstrate a solid component and previous reported cases also did not demonstrate solid components [1,4,5,7,9,13,14].

On MRI, adenocarcinomas of Mullerian origin have been reported as a large cystic mass which demonstrate T1 low-signal intensity and T2 high-signal intensity [15]. The solid compo-

nent demonstrated T1 low-signal and mixed-signal on T2. The solid component also demonstrated restricted diffusion [16]. Another study also demonstrated a cystic mass with an enhancing solid portion and partial water content [12]. An additional study demonstrated a cystic lesion without solid component [7].

Sonographically, adenocarcinomas of Mullerian origin may present as an anechoic to hypoechoic cystic mass with or without an echoic solid component [3,6,8,12,17].

Treatment includes laparotomy and resection of the mass. During surgery, it is recommended that the mass must be resected entirely without breaking the tumor as there is an increased risk of peritoneal implantation [12]. For patients who no longer want to pursue fertility, it has been proposed to also have total hysterectomy with bilateral salpingo-oopherectomy. However, there is no strong established evidence which justifies hysterectomy and oophorectomy [2,11]. In addition, there is no established evidence that demonstrates efficacy of adjuvant chemotherapy [11]. In our case, the patient wanted to preserve fertility and was followed with tumor markers to detect if there was any recurrent disease. Further, the patient had follow-up imaging to confirm that there is no tumor recurrence.

Further studies will be needed to determine the most appropriate treatment, management for long-term follow-up, and prognosis of this disease.

Patient consent

The patient gives consent to be included as the subject of a case report.

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