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Case Report

Two Rare Entities in One Patient: Mucinous Tubular and Spindle Cell Carcinoma of the Kidney and Peritoneal Adenomyomas ☆,☆☆

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ABSTRACT

Mucinous tubular and spindle cell carcinoma of the kidney is a rare subtype of renal cell carcinoma, that is believed to portend a favorable prognosis.

Adenomyomas are benign tumors that typically arise from the myometrium. Extrauterine adenomyomas are extremely rare, with only a few cases reported in the literature.

Here, we present an unusual case of a 46-year-old woman, with an incidentally detected bulky interpolar left kidney mass measuring 12 cm and multiple lobulated coalescent peritoneal nodules in the large epiploon suspicious for peritoneal carcinomatosis. A biopsy of the lesions revealed a mucinous tubular and spindle cell carcinoma of the kidney and extrauterine adenomyomas of the peritoneum. A left radical nephrectomy was performed and long-term hormone therapy with gonadotropin-releasing hormone agonists was prescribed.

The purpose of this article is to focus on these two rare lesions, review the current literature, illustrate their key imaging findings along with pathologic correlation, as well as to discuss the differential diagnosis and clinical management.

Introduction

Mucinous tubular and spindle cell carcinoma of the kidney (MTSCC-K) is a rare subtype of renal cell carcinoma (RCC) added to the World Health Organization (WHO) classification of renal neoplasms in 2004 [1]. MTSCC has a wide range of age distribution (from 13 to 82 years old) and a female predilection [2]. The tumor is mostly found incidentally as other renal tumor subtypes [3]. If the tumor size is large, patients may complain of gross hematuria, flank pain, or lumbar mass [2,3]. The precise origin is unclear, although it has been hypothesized that it arises from the distal nephron segments [1,4]. This

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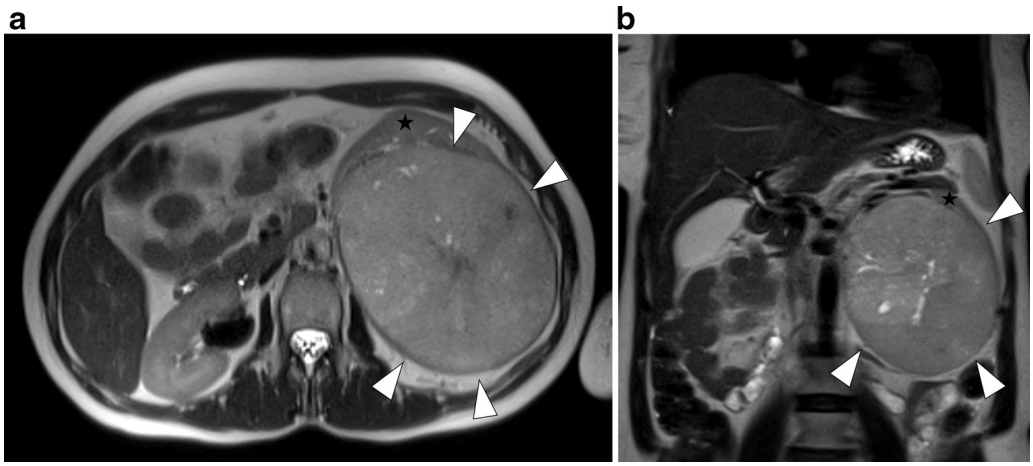

Fig. 1 – Pelvic MRI. Axial (a) and coronal (b) T2-WI show a hyperintense bulky left kidney mass. *White arrowheads*: renal cell carcinoma. *Black star*: renal parenchyma.

subtype of renal carcinoma is believed to portend a favorable prognosis when compared to other malignant renal tumors [2], making the correct classification of clinical relevance.

Adenomyomas are benign tumors composed of smooth muscle cells, endometrial glands, and endometrial stroma, that typically arise from the myometrium [5]. Adenomyomas presenting outside the uterus are extremely rare, with only a few cases reported in the literature. The ovary is the most common site of extrauterine adenomyoma [6]. The most common clinical presentation is lower abdominal pain and abnormal menstruation [6,7]. The pathogenesis behind extrauterine adenomyomas is not well understood, in part because of the rarity of the condition. Several reported cases consider a history of gynecological surgery as a contributing factor [8-14].

Case Presentation

We present a 46-year-old woman, G0P0, with complaints of menometrorrhagia and dysmenorrhea for two years. She resorted to our institution with the diagnosis of a bulky left kidney mass discovered incidentally in a pelvic magnetic resonance imaging (MRI) following routine exams (Fig. 1). There were no symptoms of left flank pain, hematuria, anorexia, or weight loss. She had a history of endometriosis, uterine leiomyomas, and adenomyosis. She underwent a laparoscopic myomectomy 10 years prior. The physical examination was unremarkable. Blood tests were within normal limits.

After the MRI, the patient performed an abdominal and pelvic computed tomography (CT) scan which showed a marked increase in left renal dimensions, conditioned by the presence of a massive solid interpolar ovoid lesion measuring 12 x 9,7 x 9,4 cm with well-defined limits and regular contours, predominantly intraparenchymal without the involvement of the renal pelvis (Fig. 2). The tumor had lower mean attenuation in all phases compared to the renal parenchyma but tended to show maximum enhancement in the latest phase of the study. There were no calcifications, macroscopic fat, or cystic changes within the tumor. There was no evidence of perinephric extension or ipsilateral renal vein involvement. There were no synchronous renal neoplasms or suspicious lymph nodes present. Concomitantly, there were multiple lobulated iso to hypodense coalescent peritoneal nodules with more expression in the greater omentum that showed moderate enhancement (Fig. 3), suggestive of peritoneal carcinomatosis.

The patient was discussed in a multidisciplinary meeting and it was decided to perform a biopsy of the lesions of the left kidney and the peritoneum, under imaging guidance. The pathology report revealed a mucinous tubular and spindle cell carcinoma (MTSCC) and extrauterine adenomyoma, respectively. With this diagnosis, the patient underwent laparoscopic radical resection of the left kidney. During the surgery, there were innumerable small nodules in the peritoneum, as described in the CT scan. Samples of the peritoneal nodules were taken and sent for histopathological evaluation. The conclusion of the final pathology report was similar to the biopsy performed preoperatively.

The gross examination of the kidney revealed a well-circumscribed grayish-white tumor, measuring 12 cm, confined to the hypertrophied left kidney (size: 17 cm weight: 1194 g). No areas of hemorrhage or necrosis were identified. The residual renal parenchyma and ureter were normal. There was no involvement of the renal pelvis, hilar vessels, or surrounding perinephric fat by the tumor. Lymph node metastases were not detected. Histological examination showed spindle cells arranged in tubular patterns, within a mucinous stroma (Fig. 4). No atypical histological features were present. Immunohistochemically, the tumor was positive for low molecular weight cytokeratin 7 (CK7), PAX2, alpha-methylacyl-coenzyme A racemase, Alcian blue, Periodic acid Schiff (PAS), while negative for CD10. The diagnosis was MTSCC of the kidney Fuhrman grade 2, and the tumor stage was pT2b (AJCC/TNM 2017).

The peritoneal nodules on gross examination appeared pale, lobulated, and firm, measuring 2,1 x 1 cm. On

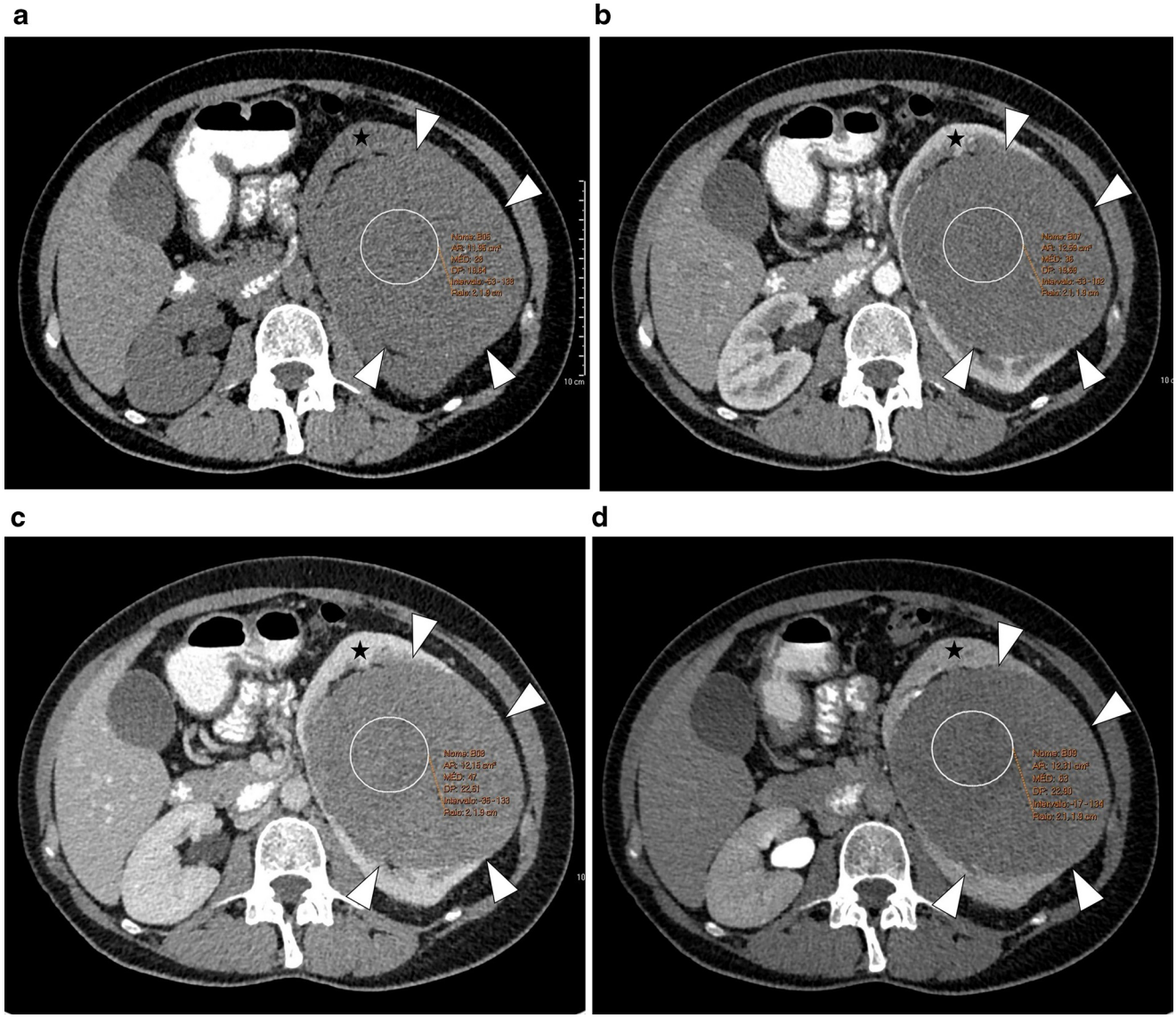

Fig. 2 – CT scan of the abdomen and pelvis illustrates a marked increase in left renal dimensions, conditioned by the presence of a massive solid interpolar ovoid lesion. On unenhanced CT (a), the baseline attenuation value of the lesion was 28 Hounsfield units (HU). With contrast injection, the mean attenuation was 36 HU at the cortico-medullary phase (b), 47 HU at the nephrographic phase (c), and 63 HU at the excretory phase (d). *White arrowheads*: renal cell carcinoma. *Black star*: renal parenchyma.

histopathological examination, they were composed of whirling bland smooth muscle tissue with benign endometrial glands randomly distributed surrounded by a rim of endometrial stroma (Fig. 5). There was no significant atypia, mitotic activity, or necrosis. Immunohistochemistry showed positive staining for PAS, CD10, smooth muscle actin, desmin, estrogen and progesterone receptors (ER, PR), CK7, and PAX8, while negative for CDX2 and CAIX. These features lead to the diagnosis of adenomyoma.

After the surgery, the patient began monthly gonadotropin-releasing hormone (GnRH) agonist therapy and is asymptomatic at 12-month follow-up. On periodic examinations, there is no evidence of recurrence or metastasis and the peritoneal disease is stable.

Discussion

On imaging, MTSCC usually presents as a solitary mass, with an expansive growth pattern and a spherical or oval shape, well-demarcated from the surrounding renal parenchyma [15]. On an unenhanced CT scan, the tumor is homogeneous. The pattern of tumor enhancement is slow and progressive, with a plateau in the latest phases of the study [3, 15]. Tumors larger than 5 cm often show a heterogeneous enhancement pattern [3]. Despite the paucity of data about the MRI appearance of MTSCC, there are some specific imaging features such as its iso-to-hyperintensity on T2-weighted images (WI) due to the mucinous stroma component [15]. After gadolinium, the

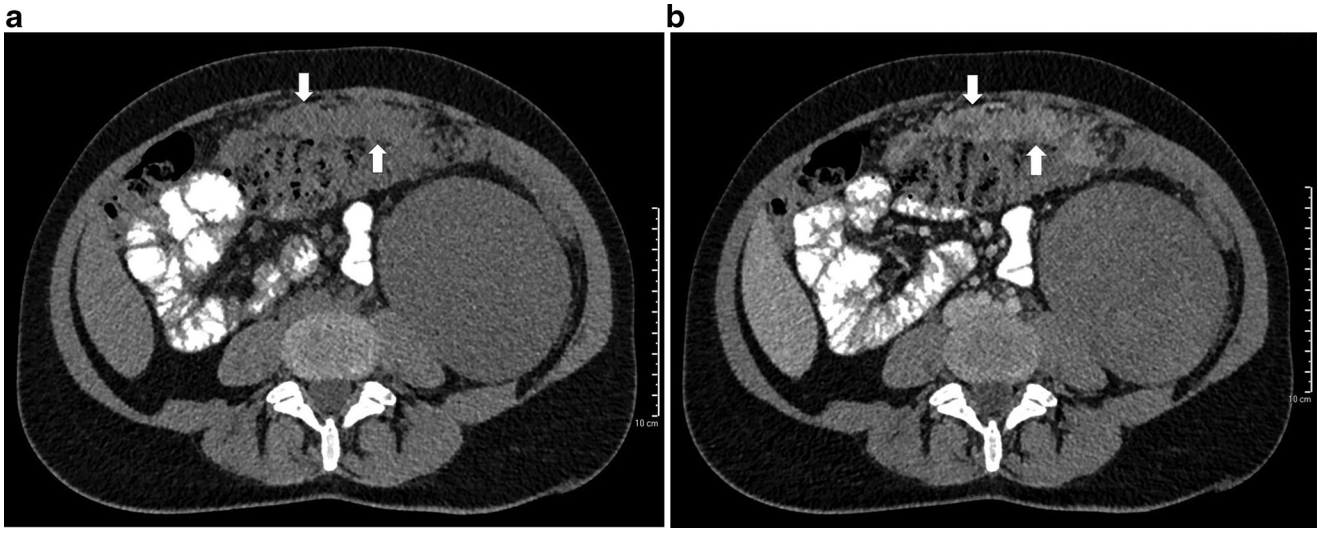

Fig. 3 – CT scan of the abdomen and pelvis demonstrates multiple lobulated iso to hypodense coalescent peritoneal nodules with moderate enhancement. *White arrows*: peritoneal nodules.

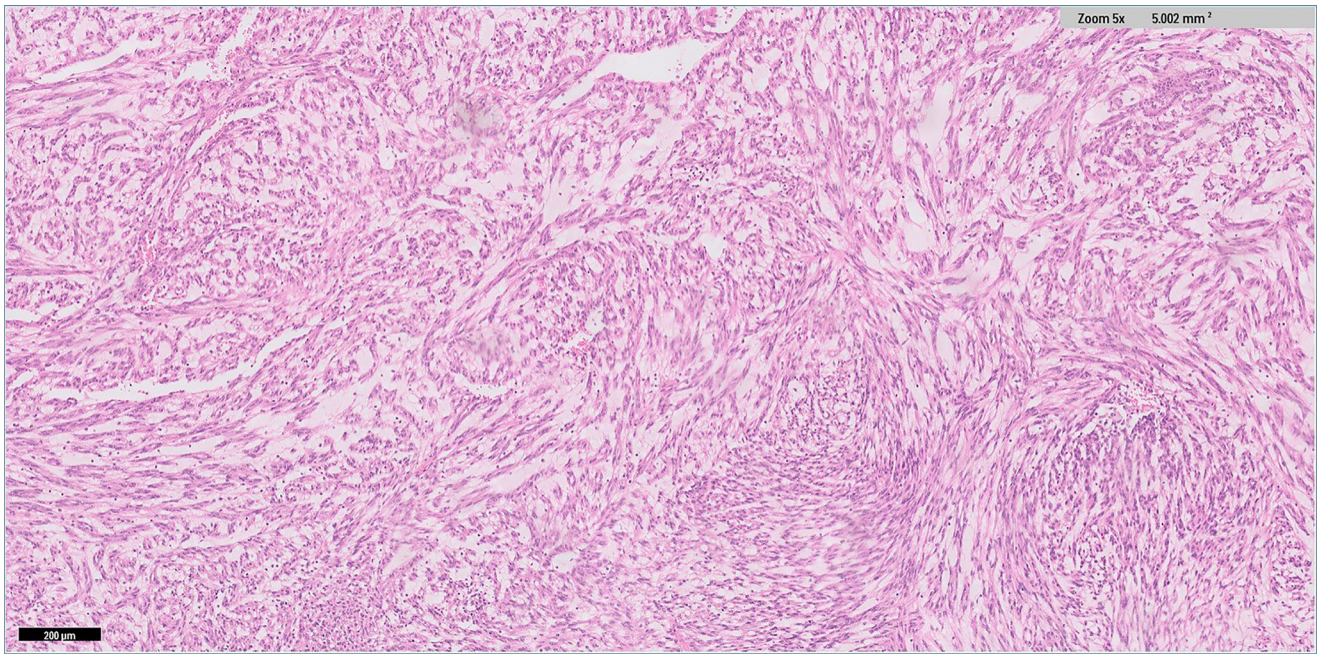

Fig. 4 – Microscopic pathology of the mucinous tubular and spindle cell carcinoma of the kidney composed of tubular and spindle cell components separated by mucinous stroma (hematoxylin-eosin stain, 5x).

tumor exhibits the same progressive enhancement pattern as seen on contrast CT [15]. The characteristics present in our case are consistent with the literature.

Papillary RCC is the most important entity in the differential diagnosis as they are both hypovascular tumors [3,15]. However, papillary RCC tends to be small (less than 2 cm) and simultaneously occurs in multiple distributions or bilaterally [16]. On MRI, papillary RCC generally shows low signal intensity on T2-WI [15-17].

Although a combination of CT and MRI features may suggest the diagnosis and help differentiate it from other renal tumors, a biopsy should be performed. Macroscopically, the tumors are usually well-circumscribed with a wide size range (from less than 1 cm to greater than 18 cm). Histologically, MTSCC consists of tightly packed elongated tubules, lined by spindle cells, separated by mucinous stroma [1]. Regarding immunohistochemistry, our results are similar to previous studies, and CD10 is a helpful marker that allows distinguishing MTSCC from type-1 papillary RCC, as it is less likely to be reactive in MTSCC [2,18,19].

This tumor is generally of low pathological stage (pT1, pT2) at diagnosis and is usually treated with partial or radical

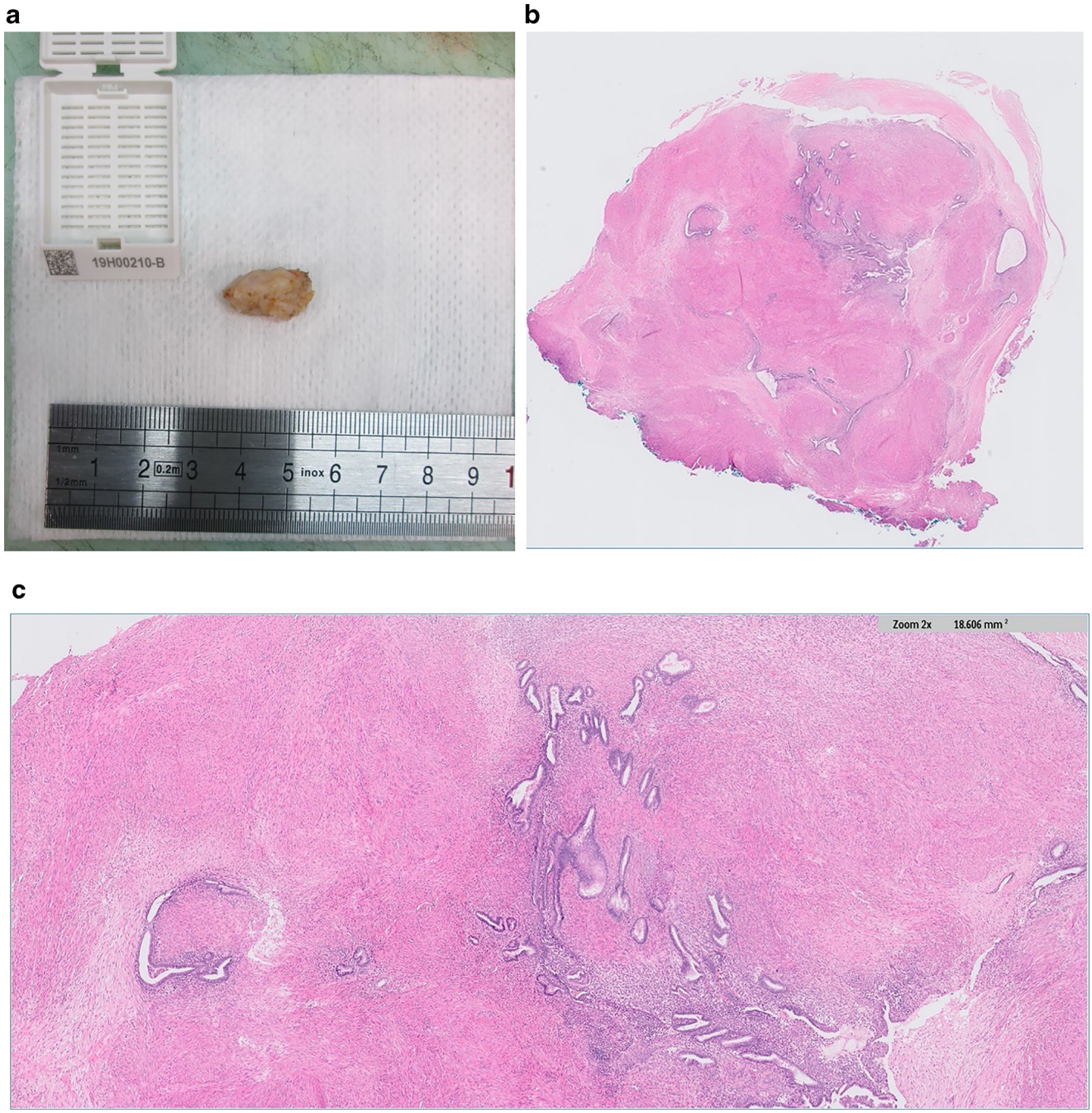

Fig. 5 – Gross photograph of the excised omental lesion (a) shows a circumscribed tan-white nodule. Microscopic pathology shows smooth muscle tissue with benign endometrial glands surrounded by a rim of endometrial stromal, compatible with adenomyoma (b: hematoxylin-eosin stain, 0.25x; c: hematoxylin-eosin stain, 2x).

nephrectomy. As the tumor was 12 cm in size, our patient had a radical nephrectomy. This case supports previous findings of MTSCC as an indolent subtype, nevertheless, a close follow-up is recommended for all MTSCC as metastases have been described [2,3,20].

Given the paucity of literature on the radiological appearance of extrauterine adenomyomas, they are often initially mistaken for peritoneal carcinomatosis, which is far more common [13,21]. However, the lack of omental fatty infiltration of tumor, significant ascites, or solid organ metastases can help differentiate this entity from malignant conditions. Nonetheless, a definitive diagnosis is only established after histopathological examination of the resected specimens [7,21].

Differential diagnoses include endometriosis with a smooth muscle component and leiomyoma with entrapped benign endometrial tissue [5,11,12]. However, the smooth muscle within endometriosis is typically focal and minor,

in contrast to the dominant smooth muscle component of the lesions that we describe. In leiomyoma with entrapped endometrial glands, these are usually seen at the periphery and not surrounded by endometrial stroma, while in adenomyomas endometrial glands are scattered within the smooth muscle tissue and surrounded by endometrial stroma.

Positivity for ER and PR suggests that the nodules are responsive to hormonal stimulation. Long-term therapy with monthly GnRH agonists appears effective in keeping the disease stable [5,9].

This case raises the question of whether the previous laparoscopic myomectomy facilitated the seeding of myometrial fragments containing adenomyotic tissue in the peritoneal cavity [13]. These fragments can attach to the peritoneum, become vascularized, and grow under the influence of hormones to form iatrogenic adenomyomas [22]. Due to the lack of a typical pattern of presentation, it is difficult to make this diagnosis with the data available so far, therefore more cases need to be reviewed.

Statement of Ethics

The patient has given written informed consent to publish this case and the identity of the patient has been protected.

REFERENCES

[1] Lopez-Beltran A, Scarpelli M, Montironi R, Kirkali Z. 2004 WHO Classification of the Renal Tumors of the Adults. Eur Urol 2006;49(5):798–805. doi:10.1016/j.eururo.2005.11.035.

[2] Zhao M, He X, Teng X. Mucinous tubular and spindle cell renal cell carcinoma: a review of clinicopathologic aspects. Diagn Pathol 2015;10(1). doi:10.1186/s13000-015-0402-1.

[3] Kenney P, Vikram R, Prasad S, et al. Mucinous tubular and spindle cell carcinoma (MTSCC) of the kidney: a detailed study of radiological, pathological and clinical outcomes. BJU Int 2015;116(1):85–92. doi:10.1111/bju.12992.

[4] Fleming S. Distal nephron neoplasms. Semin Diagn Pathol 2015;32(2):114–23. doi:10.1053/j.semdp.2015.02.004.

[5] Huanwen W, Hui Z, Xiaowei X, Zhaohui L. Extrauterine adenomyoma of the liver with a focally cellular smooth muscle component occurring in a patient with a history of myomectomy: case report and review of the literature. Diagn Pathol 2013;8(1). doi:10.1186/1746-1596-8-131.

[6] NA K, PARK S, KIM H. Clinicopathological Characteristics of Primary Ovarian Adenomyoma: A Single-institutional Experience. Anticancer Res 2017;37(5):2565–74. doi:10.21873/anticanres.11600.

[7] Paul P, Gulati G, Shintre H, Mannur S, Paul G, Mehta S. Extrauterine adenomyoma: a review of the literature. European Journal of Obstetrics & Gynecology and Reproductive Biology 2018;228:130–6. doi:10.1016/j.ejogrb.2018.06.021.

[8] Redman R, Wilkinson EJ, Massoll NA. Uterine-like mass with features of an extrauterine adenomyoma presenting 22 years after total abdominal hysterectomy-bilateral salpingo-oophorectomy: a case report and review of the literature. Arch Pathol Lab Med 2005;129(8):1041–3. doi:10.1043/1543-2165(2005)129[1041:UMWFOA]2.0.CO;2.

[9] Carinelli S, Motta F, Frontino G, Restelli E, Fedele L. Multiple extrauterine adenomyomas and uterus-like masses: case reports and review of the literature. Fertil Steril 2009;91(5):1956 e9-1956.e11. doi:10.1016/j.fertnstert.2009.01.076.

[10] Khurana A, Mehta A, Sardana M. Extrauterine adenomyoma with uterus like features: A rare entity presenting 17 years post hysterectomy. Indian Journal of Pathology and Microbiology 2011;54(3):572. doi:10.4103/0377-4929.85096.

[11] Moghadamfalahi M, Metzinger D. Multiple Extrauterine Adenomyomas Presenting in Upper Abdomen and Pelvis: A Case Report and Brief Review of the Literature. Case Rep Obstet Gynecol 2012;2012:1–3. doi:10.1155/2012/565901.

[12] Tandon N, Showalter J, Sultana S, Zhao B, Zhang S. Extrauterine Adenomyoma of the Liver in a 50 year old Female with Pelvic Endometriosis. Ann Clin Lab Sci 2017;47(2):208–12.

[13] Belmarez J, Latifi H, Zhang W, Matthews C. Simultaneously occurring disseminated peritoneal leiomyomatosis and multiple extrauterine adenomyomas following hysterectomy. Baylor University Medical Center Proceedings 2019;32(1):126–8. doi:10.1080/08998280.2018.1520555.

[14] Won Y, Woo J, Byun J, Hong M. Extrauterine Adenomyoma of the Liver Mimicking a Hepatic Adenoma: A Case Report. Journal of the Korean Society of Radiology 2020;81(1):197. doi:10.3348/jksr.2020.81.1.197.

[15] Cornelis F, Ambrosetti D, Rocher L, et al. CT and MR imaging features of mucinous tubular and spindle cell carcinoma of the kidneys. A multi-institutional review. Eur Radiol 2016;27(3):1087–95. doi:10.1007/s00330-016-4469-1.

[16] Roy C, Sauer B, Lindner V, Lang H, Saussine C, Jacqmin D. MR Imaging of papillary renal neoplasms: potential application for characterization of small renal masses. Eur Radiol 2006;17(1):193–200. doi:10.1007/s00330-006-0271-9.

[17] Cornelis F, Tricaud E, Lasserre A, et al. Routinely performed multiparametric magnetic resonance imaging helps to differentiate common subtypes of renal tumours. Eur Radiol 2014;24(5):1068–80. doi:10.1007/s00330-014-3107-z.

[18] Paner G, Srigley J, Radhakrishnan A, et al. Immunohistochemical Analysis of Mucinous Tubular and Spindle Cell Carcinoma and Papillary Renal Cell Carcinoma of the Kidney. Am J Surg Pathol 2006;30(1):13–19. doi:10.1097/01.pas.0000180443.94645.50.

[19] Sarsik B, Simsir A, Karaarslan S, Sen S. Mucinous tubular and spindle cell carcinoma of kidney and problems in diagnosis. Turkish Journal of Pathology 2011;27(2):116. doi:10.5146/tjpath.2011.01059.

[20] Thway K, du Parcq J, Larkin J, Fisher C, Livni N. Metastatic renal mucinous tubular and spindle cell carcinoma. Atypical behavior of a rare, morphologically bland tumor. Ann Diagn Pathol 2012;16(5):407–10. doi:10.1016/j.anndiagpath.2011.04.001.

[21] Gruttadauria M, Wen X, Burke W. Extrauterine adenomyomas presenting in a 47 year old woman with a previous cesarean section. Gynecol Oncol Rep 2020;31:100524. doi:10.1016/j.gore.2019.100524.

[22] Donnez O, Squifflet J, Leconte I, Jadoul P, Donnez J. Posthysterectomy pelvic adenomyotic masses observed in 8 cases out of a series of 1405 laparoscopic subtotal hysterectomies. J Minim Invasive Gynecol 2007;14(2):156–60. doi:10.1016/j.jmig.2006.09.008.