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Case Report

Renal Endometriosis Tends to Be Misdiagnosed as Renal Tumor: A Rare Case Report

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Renal endometriosis is a rare disease for which the mechanisms of pathogenesis are still unclear. As such, early diagnosis and an appropriate treatment are often delayed because of the tendency to be misdiagnosed as a renal tumor. In October 2013 we performed a radical nephrectomy for a 37-year-old woman with renal endometriosis who was preoperatively misdiagnosed as having a right renal tumor. Avoiding the misdiagnosis of renal endometriosis requires a detailed case history, especially regarding whether the cyclicity of lumbodorsal pain and hematuria correlates with patients' menstrual cycles. Imaging examinations are commonly helpful for localization, whereas relieving symptoms with drugs to create a hypoestrogenic state is useful for clinical diagnosis. However, a final diagnosis for renal endometriosis still must depend on histopathologic examination.

Key words: Renal endometriosis – Renal tumor – Radical nephrectomy – Misdiagnosis

Endometriosis is a condition characterized by ectopic endometrial tissue growth in extrauterine sites. The incidence rate is about 8% to 10% in women of reproductive age.[1] The ectopic focus is most common in the pelvic organs and peritoneum, but it seldom occurs at other distant sites of the body. Urinary tract endometriosis is less common, accounting for only 1.2% of all cases. Endometrial lesions involving the bladder are found in about 85% of cases and are localized to the ureter in about 15%, whereas lesions involving the kidneys and urethra are exceedingly rare, accounting for less than 1% of cases with urinary tract endometriosis.[2] Because renal endometriosis is an extremely rare manifestation of this disease, it has only occasionally been reported in the past.[3–6] In July 2013, we performed a radical nephrectomy for a woman with renal endometriosis who was preoperatively misdiagnosed as having a renal tumor. We report our experience to raise awareness that cyclical manifestations of lumbodorsal pain and hematuria should be approached with a high index of suspicion to avoid misdiagnosing renal endometriosis in the future.

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Case Report

History

In September 2013, a 37-year-old married mother of two daughters came to our hospital seeking medical attention and complained of paroxysmal dull pain in the right lower back. The pain had been aggravated by menstrual periods for a 6-month duration without any obvious causative factors. There was no history of abdominal pain or gross hematuria. Otherwise, the menstrual pattern and leucorrhea were normal, and there was no history of dysmenorrhea and sexual behavior during the menstrual period. There were no reports of any pelvic trauma or surgery.

Five months ago, the ultrasonography of the patient's abdomen in a local clinic showed right hydronephrosis and several calculus in the right kidney and ureter. These findings were replicated using unenhanced abdominal X-ray. After the patient was treated with calculus-dissolving drugs and extracorporeal shock-wave lithotripsy in a local hospital, there was only temporary relief from repeated right lumbodynia. The patient was hospitalized in October 2013 for further treatment.

Preoperative examinations

Intravenous pyelography displayed two radiopaque calculus images and compressed deformation of the middle and lower calyces in the right kidney (Fig. 1A). Plain computed tomography (CT) scanning of the abdomen showed an inhomogeneous (10–36 Hounsfield units) mass of $7.5 \times 5.0 \times 3.5$ cm in the lower pole of the right kidney, with multiple cysts and small calcifications (Fig. 1B). The mass can only be enhanced slightly in contrast-enhanced CT images, with a low-density central area consistent with necrosis or cystic change. Furthermore, the CT scanning did not display any other evidence of retroperitoneal lymphadenectasis, involved pelvicaliceal system, renal vein or inferior vena cava (Fig. 1C–1E). Based on the imaging results we considered the mass in the right kidney to be a renal tumor with the clinical stage of T2a, N0, M0. We did not carry out preoperative percutaneous biopsy for this 37-year-old woman, who had a long life expectancy and a clearly suspicious tumor lesion, in order to avoid the potential risk of tumor implantation via needle tracks. Finally, we chose radical nephrectomy as the surgical method for this case because of the relatively large tumor volume and the possibility of cystic renal cell carcinoma (RCC).

Operative and histopathologic findings

After successful administration of general anesthesia, the exploration of the right renal region via the 11th intercostal incision revealed a mass situated at the lower pole of the right kidney without any surface anomalies. The mass did not show any sign of invading the perirenal fat and surrounding organs, including the right ureter. The radical nephrectomy was completed successfully.

The operative specimen measured $11.0 \times 6.5 \times 5.5$ cm^3. After the specimen was longitudinally dissected, we discovered several capsular spaces containing brown, serumlike fluid in the lower pole of the right kidney, and the cut surface of the mass was hoar with intact capsule.

Finally, histopathologic examination found ectopic endometrium and abundant fibrous tissue in both the renal cortex and medulla, confirming the final diagnosis of the case as endometriosis of the right kidney (Fig. 1F).

Discussion

The pathogenesis of endometriosis is still not well established. Several hypotheses have been proposed, such as ectopic transplantation theory, metaplasia of coelomic epithelium, autoimmunity, blood-lymphatic embolism, embryonic theory, and others.[7] It is universally acknowledged that ectopic transplantation plays a role in the development of endometriosis, so it follows that the pathogenesis of urinary tract endometriosis may involve similar mechanisms.[8]

The basic histopathologic change of renal endometriosis is that the ectopic endometrium cyclically thickens and sheds in response to the changing levels of ovarian sex hormones. It results in the growth of surrounding fibrous tissue and the formation of endometrioma in the kidney. Repeated periodic bleeding makes accumulated blood cysts gradually increase in the renal tissue, and intracapsular blood and the deciduous ectopic endometrium form gross hematuria when the lesions break into the renal calyces. If the lesions invade the renal capsule, patients often experience dull lumbodorsal pain. In addition, blood clots and deciduous endometrium may cause ureteral obstructions, which may then evoke renal colic.[9] Continued proliferation of fibromuscular tissue around lesions results in trabs and nodules, which finally form variously sized scleroma called endometrioma. The endometrioma distort the shape of the kidney by drawing in adjacent renal parenchyma.[10]

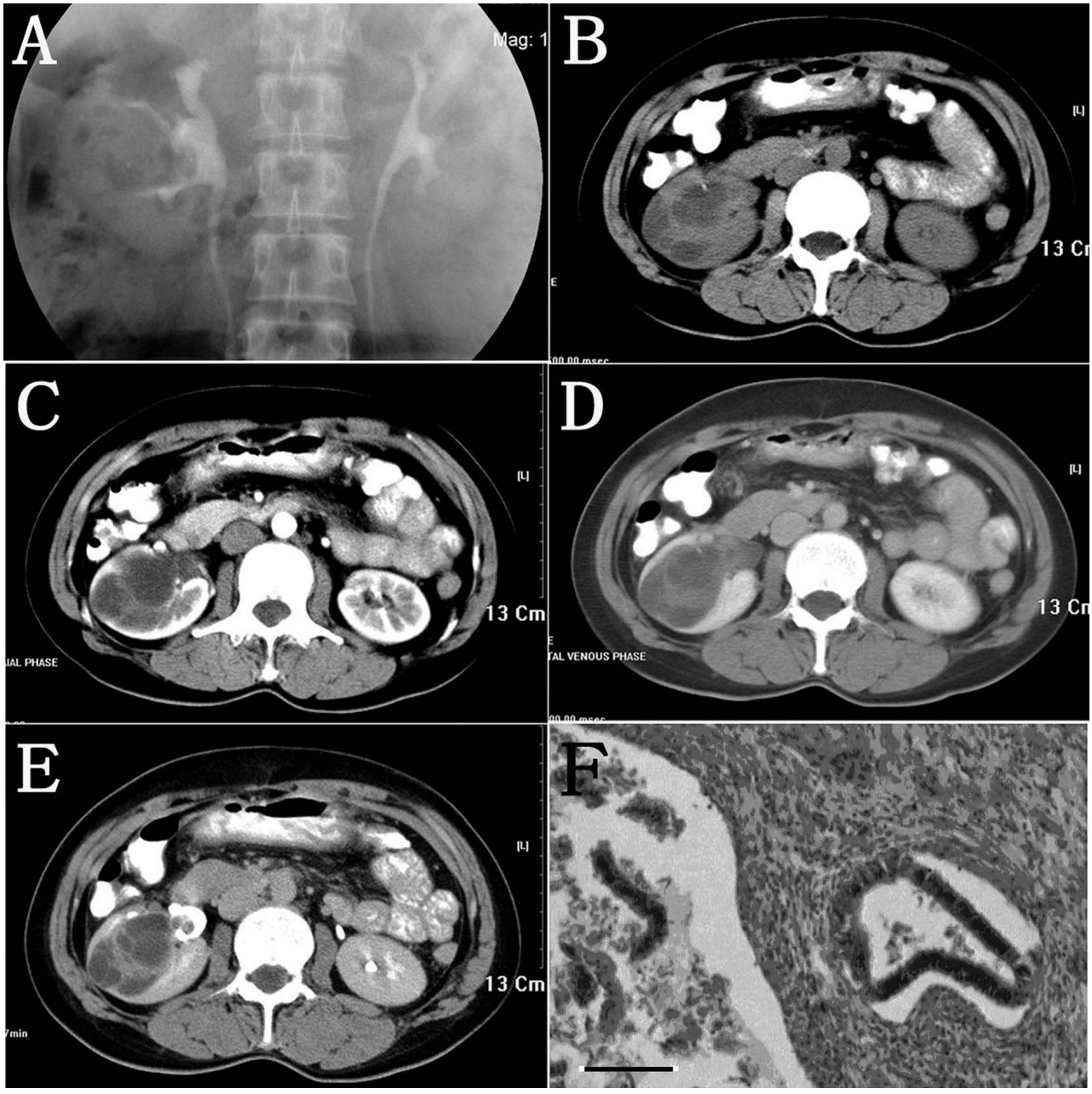

Fig. 1 Imaging examinations (A–E) of the patient showed an inhomogeneous mass of 7.5 × 5.0 × 3.5 cm, with multiple cysts and small calcifications in the lower pole of the right kidney, which was only slightly enhanced and had a low-density central area consistent with necrosis or cystic change. The mass led to compressed displacement of the right renal pelvis and calices. Histopathologic examination (F) found ectopic endometrium in both the renal cortex and medulla. (A) The 15th-minute image of a total of 90 minutes of intravenous pyelography. (B) Unenhanced CT scanning of kidneys. (C) The renal cortical phase of enhanced CT scanning. (D) The renal corticomedullary phase of enhanced CT scanning. (E) The renal excretory phase of enhanced CT scanning. (F) The histopathologic picture of the excisional specimen (bars = 100 μm).

The typical triad of RCC characteristics, a palpable lumbodorsal mass, pain, and gross hematuria are also common in renal endometriosis, thus causing the tendency to misdiagnose renal endometriosis as RCC. One defining characteristic specific to renal endometriosis is the cyclicity of lumbodorsal pain and hematuria, which correlates with patients' menstrual cycles. In the present report, the patient

complained about a paroxysmal pain of the right lower back that became aggravated during her menstrual period, but without gross hematuria. Regretfully, the diagnostic significance of the patient's aggravated lumbodorsal pain during her menstrual period did not come to our attention.

The imaging manifestations of renal endometriosis do not facilitate accurate diagnosing, thus it is commonly misdiagnosed before the operation as a tumor, inflammation, congenital anomaly, etc. Imaging examinations by CT scanning are commonly helpful for localization.[10] Unenhanced CT scans show low-density oval lesions less than 3 cm with clear, irregular boundaries. Lesion density is usually uniformly hypodense, but it fluctuates with the menstrual cycle. When ectopic endometrium bleeding occurs during the menstrual period, the density of lesions may increase or appear uneven. CT enhancement scanning often fails to show enhancement in the central parts of lesions and uneven enhancement in the circumjacent proliferative fibromuscular tissue.[6,9]

Compared with CT, magnetic resonance imaging (MRI) shows superior spatial resolution. The typical MRI of renal endometriosis appears as a single-room or multiroom cystic mass with blood. The solid components of renal endometrioma usually show hypointensity or isointensity on T1-weighted images and hypointensity on T2-weighted images.[11] We did not carry out MRI examination for the patient because plain and enhanced CT scanning was the preferred modality for imaging the working diagnosis of renal tumors or RCC. Intravenous pyelography examination provides little diagnostic information to help identify renal endometriosis, whereas type B ultrasound is useful only for confirming the existence of solid-cystic lesions rather than the diagnosis of renal endometriosis. Using drugs (such as norethindrone) to create a hypoestrogenic state is extremely useful for clinical diagnosis of renal endometriosis if the symptoms of lumbodorsal pain and gross hematuria can be relieved by this treatment.

Preoperative biopsy via fine-needle aspiration can greatly help to determine the nature of the lesions and avoid the misdiagnosis, but it has not been recommended by the guidelines of the European Association of Urology on RCC because of the high diagnostic accuracy of current abdominal imaging findings and the potential risk of tumor seeding via needle tracks.[12,13] Nephron-sparing surgery and intraoperative frozen-section examination also could not be carried out for this patient because of the preoperative stage of cT2a, N0, M0, according to imaging results. The European Association of Urology guidelines for RCC suggest that radical nephrectomy is the standard of care for patients with T2 tumors and those larger-size (>7 cm) renal masses were not treatable by nephron-sparing surgery because of an increased risk of intrarenal recurrences.[12,13] Another important reason for choosing radical nephrectomy instead of partial nephrectomy is that the possibility of cystic RCC cannot be excluded. Partial nephrectomy needs to cut into the kidney and has a far higher risk of breaking the cyst walls of the lesion and causing tumor dissemination through the flow of hydatid fluid than radical nephrectomy, which is performed only outside the renal adipose capsule (Gerota fascia). Therefore, the misdiagnosis finally resulted in the overtreatment of nephrectomy.

In short, renal endometriosis is an orphan disease. The causation and pathogenesis are still unclear, and now no specific diagnostic method can be acquired. All of these things hinder the early diagnosis of renal endometriosis and even cause it to tend to be misdiagnosed as renal tumors. Thus, to avoid misdiagnosis it requires clinicians to inquire about case history in detail, especially whether the cyclicity of lumbodorsal pain and hematuria is correlative with patient's menstrual cycles. Basic diagnostic techniques, such as history taking and physical examination, should not be overshadowed by high-tech imaging. A detailed case history, careful consideration of all investigation results, joint consultation with senior colleagues, and an open mind are all crucial elements for a good medical practice. If all of these basic principles have been executed, this case may end up with a fine-needle aspiration biopsy, followed by the correct diagnosis of renal endometriosis and possibly just hormonal treatment instead of a radical nephrectomy.

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