INTRODUCTION

Retroperitoneal fibrosis (RPF) is an uncommon fibrotic reaction in the retroperitoneum. There is an idiopathic form and a secondary form associated with IgG4-related disease, drugs (methysergide, pergolide, bromocriptine, ergotamine, methyl dopa, hydralazine), infections (tuberculosis, histoplasmosis, actinomycosis), malignancies (carcinoid tumor, Hodgkin’s and non-Hodgkin’s lymphomas, sarcomas, carcinomas of the colon, prostate, breast, and stomach), prior surgeries, and radiation therapy. The idiopathic form accounts for more than two-thirds of cases and it was thought to result from a local inflammatory response to antigens in the atherosclerotic plaques in the abdominal aorta, combined with auto-immunologic factors. Thus, the fibroinflammatory tissue usually encases the infrarenal portion of the abdominal aorta, inferior vena cava, and ureters (1-3). Here, we report two rare cases of idiopathic RPF involving only a unilateral renal pelvis and sinus; thus mimicking renal pelvic cancer.

CASE REPORT

Case 1

A 66-year-old female presented with an incidental finding at computed tomography (CT) screening. Her previous medical history revealed that she had undergone hysterectomy for an unknown cause 10 years ago. Contrast-enhanced genitourinary CT showed a homogeneously enhancing soft tissue mass replacing the left renal pelvis, which was suspicious for renal pelvic cancer. The diagnosis of idiopathic RPF was surgically confirmed. To the best of our knowledge, a few cases of idiopathic RPF presenting with features of a localized unilateral renal pelvic mass mimicking renal pelvic cancer have been reported.
also yielded unsatisfactory results. She underwent laparoscopic radical nephrectomy. Histopathologic findings of the resected kidney were negative for malignancy. Fibrosis and chronic inflammation with myofibroblast proliferation and lymphoplasma cell infiltration were observed. The lesion was mainly located in the renal sinus fat tissue around renal vessels, suggesting an inflammatory or immunologic process such as idiopathic RPF (Fig. 1C). Immunochemistry of IgG and IgG4 was performed to confirm the presence of IgG4+ plasma cells in the tissue. The ratio of IgG4 to IgG plasma cells was within the normal range (<5%), and these results were inconsistent with the diagnosis of IgG4-related disease, which shows an IgG4 to IgG ratio of more than 30-40%. Pathology report was compatible with idiopathic RPF.

Case 2

An 80-year-old man presented with fever and cough for one week. His past history was nonspecific except for hypertension. There was no abnormal finding on physical examination. The results of peripheral blood smear examination showed slightly elevated C-reactive protein (2.93 mg/dL). On chest CT, mild bronchitis and the incidental finding of a left renal pelvic mass were detected. Additional contrast enhanced genitourinary CT was performed for further evaluation. CT showed an enhancing soft tissue mass in the left renal pelvis and sinus with mild pelvic dilatation. Periureteric fat infiltration in the left proximal ureter without definite wall thickening was observed (Fig. 2A, B).

Two urine cytologies of washed urine from the left renal pelvis were negative, and on RGP, there was no filling defect but mild luminal narrowing of the renal pelvis and mild calycectasia were detected (Fig. 2C). He underwent laparoscopic radical nephrectomy. Histopathologic findings were consistent with idiopathic RPF (Fig. 2D). Stromal fibrosis with lymphoplasma cell infiltration mainly into the renal sinus was observed. On immunohistochemistry of the tissue, the ratio of IgG4 to IgG plasma cells was less than 5%, which was within the normal range, and on laboratory examination, serum levels of IgG and IgG4 were within their normal range [1056.6 mg/dL (range, 700–1600 mg/dL) and 1020.0 mg/L (range, 30–2010 mg/L), respectively].

DISCUSSION

RPF is a rare autoimmune, inflammatory disorder that develops in about one in 200000 people (4). It is most common in individuals 40 to 60 years of age (5).

Men are affected by this disorder two to three times more often than women (1). In typical RPF, a mass of fibrous tissue develops around the abdominal aorta with an epicenter at the L4–5 level and it also encases the inferior vena cava and the ureter. Ureteral involvement is reported in 80–100% of cases at presentation (4). At presentation, such ureteral involvement is often bilateral, but in patients with an apparently unilateral obstruction, contralateral disease can develop even within a short period (1).

CT is the modality of choice for imaging diagnosis of RPF and for evaluating the extent of the process. On CT scan, RPF usually
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appears as a homogeneous plaque, which is isodense to the adjacent muscle, surrounding the lower abdominal aorta and the iliac arteries, and often enveloping the ureters and the inferior vena cava (4). The degree of contrast enhancement is related to inflammation and process activity.

Intense enhancement indicates fibrous tissue in an initial phase with associated inflammation (6). The advantage of MR imaging is avoidance of nephrotoxicity of iodinated contrast media in patients with compromised renal function and better definition against the surrounding tissues, mainly when fat-saturation images are used.

Idiopathic RPF is hypointense in T1-weighted MR images. In T2-weighted MR images, its intensity is variable; high signal intensity in the active inflammatory stage because of tissue edema and hypercellularity and low signal intensity in the late fibrotic stage (5-7).

In rare cases, RPF presents as an asymmetric poorly circumscribed retroperitoneal mass with atypical localization, and thus it is not characterized by aortic involvement as observed in our cases, and differentiation from a primary retroperitoneal tumor or malignant lymphadenopathy is very difficult. Atypical sites of RPF involvement include the small bowel mesentery, duodenum,

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Table 1. Previous Reports of Perirenal Extension of RPF

| Year   | Authors (Ref. No.) | Age | Sex | Perirenal Involvement | Periaortic, Pericaval Involvement |
|--------|--------------------|-----|-----|------------------------|-----------------------------------|
| 1937   | Erischar           | -   |     | Bilateral              |                                   |
| 1941–1943 | Lebbin            | -   |     | -                      |                                   |
| 1988   | Yancey             | 35  | F   | Bilateral              | No involvement                    |
| 1992   | Rominger           | 60  | F   | Unilateral             | Involvement                       |
| 1995   | Barret             | 68  | M   | Unilateral             | Involvement                       |
| 1990   | Brooks             | 14  | F   | Unilateral             | Involvement                       |
| 1996   | Bechtold           | -   |     | Bilateral              | No involvement                    |
| 1999   | Ayuso et al. (7)   | 53  | F   | Bilateral              | No involvement                    |
| 2002   | Triantopoulou      | 51  | F   | Unilateral             | Involvement                       |
| 2005   | Ergen et al. (6)   | 51  | M   | Unilateral             | Involvement                       |
| 2012   | Ellimoottil        | 62  | F   | Bilateral              | No involvement                    |
| 2013   | Sofiane            | 64  | M   | Bilateral              | No involvement                    |
| 2013   | Yoshino et al. (8) | 71  | M   | Unilateral             | No involvement                    |
| 2014   | Weinig             | 51  | M   | Unilateral             | No involvement                    |
| 2015   | Lang et al. (9)    | 45  | M   | Unilateral             | No involvement                    |

RPF = retroperitoneal fibrosis
colon, bladder, and periduodenal, peripancreatic, pelvic, perirenal and epidural space (1, 4).

Perirenal extension of RPF is rarely reported (6-10), whereas in only a few cases, the disease was unilaterally localized. To the best of our knowledge, three cases of idiopathic RPF presenting with features of a localized unilateral renal pelvic mass mimicking renal pelvic cancer (8, 9) without periaortic or pericaval involvement have been reported (Table 1).

IgG4-related kidney disease is an important part of the spectrum of IgG4-related sclerosing disease, which manifests as a secondary form of RPF. IgG4-related kidney disease is usually accompanied by other organ involvement, and isolated IgG4-related kidney disease without other organ involvement is very rare; only 6% of cases have been reported as renal lesions alone (10).

Although CT and MR imaging have an important role in diagnosis of RPF, histological tissue examination is needed, especially in cases with atypical location of a retroperitoneal mass to rule out malignancy. In addition to malignancy (urothelial carcinoma), differential diagnosis includes retroperitoneal fibromatosis, which is characterized by uniform proliferation of fibroblasts and is associated with Gardner’s syndrome. Another differential diagnosis is inflammatory pseudotumor, which mainly affects children, and its appearance is that of a huge mass with infiltrative borders. It is histologically characterized by myofibroblast proliferation with myxoid and inflammatory areas (1).

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일측 신장 신우만 침범한 특발성 복막후섬유증: 증례 보고와 문헌 고찰

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특발성 복막후섬유증은 드문 질환으로 그 원인은 아직 잘 알려져 있지 않다. 저자들은 66세 여자 환자와 80세 남자 환자에서 우연히 발견된 좌측 신장 신우만을 침범한 종괴 형태로 발현된 비전형적 특발성 복막후섬유증을 보고한다. 전산화단층촬영(computed tomography)에서 좌측 신장의 신우를 대치하는 연조직 음영으로 신장암을 의심하게 하는 종괴였고 수술로 특발성 복막후섬유증으로 확진되었다. 이와 같이 일측 신장 신우만을 침범하여 신장암과 유사한 형태로 발현되는 특발성복막후섬유증은 매우 드문 형태로 문헌 고찰과 함께 보고한다.

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