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Retroperitoneal Fibrosis Confined to the Pelvic Cavity

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Aim : To find whether there were characteristic clinical or imaging findings in retroperitoneal fibrosis (RPF) confined in the pelvic cavity.
Methods : Literature review was performed using MEDLINE with key words of [retroperitoneal fibrosis and pelvis] and [pelvic fibrosis].
Results : In addition to our 68-year-old Japanese woman, 13 cases were found, in which computed tomography (CT) or magnetic resonance imaging (MRI) confirmed that the mass lesions were confined in the pelvic cavity. The pelvic RPF (age range, 29-69 y.o. ; seven male) showed variable symptoms and clinical presentation suggestive of gynecological, urological or colonic diseases. CT usually revealed an ill-defined well-enhanced mass lesion, which may be associated with fascial thickening. On MRI the pelvic masses showed low signal intensity on T1-weighted images and high or mixed signal on T2-weighted images. In nine of 14 patients the presacral or retrorectal spaces were involved, and the remaining patients had RPF around the bladder. In seven patients, imaging-guided needle biopsies were employed. Good prognoses were reported in all 11 patients, in which corticosteroid therapy were given.
Conclusion : Pelvic RPF may show different symptoms and clinical features from typical paraaortic RPF, but shows similar imaging findings. The imaging-guided biopsy is the choice for histopathological diagnosis. (Kitakanto Med J 2005 ; 55 : 87～90)

Key words : Retroperitoneal fibrosis, Pelvis, Pelvic fibrosis, Computed tomography (CT), Magnetic resonance imaging (MRI)

Introduction

Retroperitoneal fibrosis (RPF) is a well-described clinical entity, which is characterized by replacement of the normal tissue of the retroperitoneum with fibrosis and/or chronic inflammation. RPF most commonly involves the paraaortic area extending laterally to envelope the ureters, and RPF confined in the pelvic cavity is rare.[1-3] We report a case of pelvic RPF, which was entirely confined to the pelvic cavity and no paraaortic mass lesions were associated. We also performed literature review to find whether there were characteristic clinical or imaging findings in pelvic RPF.

Materials and Methods

We reported a case of pelvic RPF with clinical and imaging findings.

Literature review was also performed using MEDLINE with key words of [retroperitoneal fibrosis and pelvis] and [pelvic fibrosis].

Results

Case Report

A 68-year-old Japanese woman was admitted to our hospital with dull pain in the right flank for several weeks. Clinical findings were grossly normal and no weight loss or fever was reported. She had an appendectomy at age 20 and cholecystectomy at age 51. Urinalysis and blood count were normal. Abdominal ultrasonography (US) showed right hydronephrosis and probable hydroureter, thus enhanced computed tomography (CT) of the whole abdomen was performed.

CT demonstrated a homogeneously-enhanced infiltrating mass lesion in the presacral space, involving the right distal ureter without any bony abnormality (Fig. 1). This mass lesion was confined to the pelvic cavity, although the cystoscopy and colonoscopy did not indicate any pathology. On magnetic resonance

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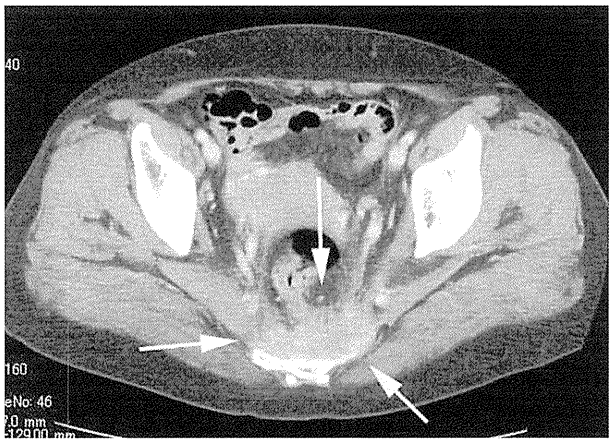

Fig.1 Contrast-enhanced CT demonstrates a homogeneously-enhanced infiltrating mass lesion in the presacral space (arrows).

imaging (MRI), the pelvic mass lesion showed low signal intensity on T1-weighted images, and slightly high intensity on T2-weighted images. After intravenous administration of contrast material (Gd-DTPA), inhomogeneous enhancement was observed (Fig. 2).

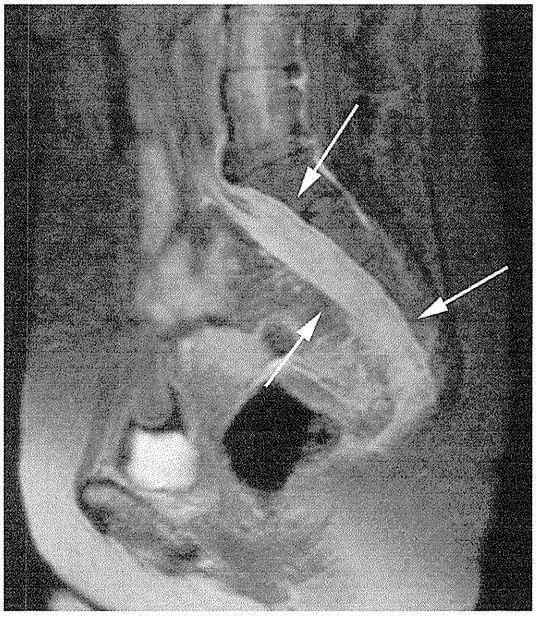

Fig.2 Parasagittal enhanced T1-weighted image with fat suppression shows an enhanced mass lesion (arrows) in the presacral space.

CT-guided biopsy was performed using 18-gauge needle in prone position (Fig. 3), and three samples were obtained from the different parts of the mass lesion. The histological specimen showed an extensive fibroblastic proliferation with abundant collagen deposition and inflammatory infiltrate. There was no evidence of malignancy, and the diagnosis of RPF was made. Corticosteroid therapy was started, and the presacral mass lesion had dramatically decreased in size, and the dilatation of the right urinary tract had disappeared (Fig. 4).

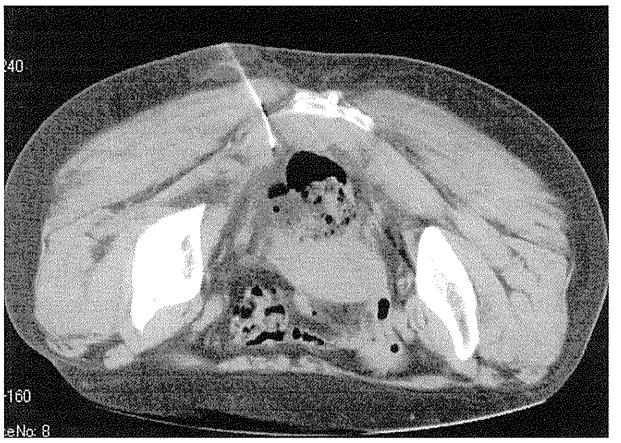

Fig.3 CT-guided biopsy of the presacral mass was performed using 18-G biopsy needle, and three samples were obtained from the different parts of the mass lesion.

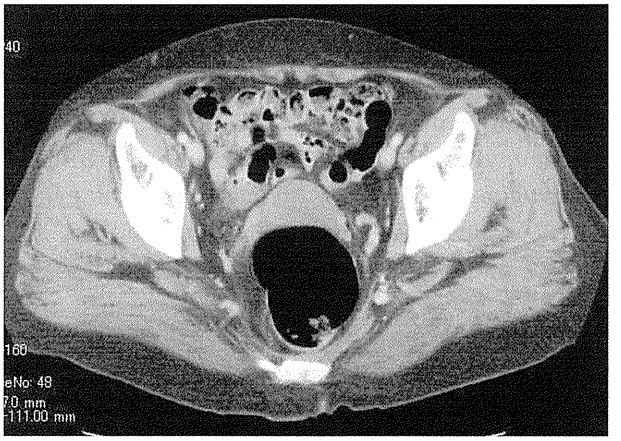

Fig.4 Contrast-enhanced CT shows a dramatic decrease in size of the presacral mass after corticosteroid therapy.

Literature review

We found 13 patients with RPF, in which CT or MRI confirmed that the mass lesions were confined in the pelvic cavity.[1-10] The details of these 13 patients and our patient are summarized in Table 1 (age, 53.3+/−11.3 y.o.; range, 29-69 y.o.; seven male and seven female). The symptoms of the pelvic RPF were variable and nonspecific. Colonic symptoms were reported in three patients and the rectal examinations suggested gynecological diseases in four patients. Three of 14 patients received medications which may induce RPF (ergotamine, acyclovir and anticonvulsive medication, each).

CT usually revealed an ill-defined well-enhanced mass lesion, which may be associated with fascial thickening. In nine of 14 patients, the presacral or retrorectal spaces were involved, and the remaining five cases had RPF around the bladder. On MRI, the pelvic masses showed low signal intensity on T1-weighted images and high or mixed signal intensity on T2-weighted images, and were well enhanced with intravenous contrast material. No calcifications or

Table 1 Reported cases with RPF confined in the pelvic cavity.

Age/ Sex	Symptoms and clinical presentations	CT and/or MRI findings	Ureter involvement	Pathological diagnosis	Past history and complications	Treatment
55/F[4]	Incidentally discovered hydronephrosis on US.	Mass between the bladder and rectum.	Right	US-guided transvesical biopsy.	35y.o. orbital pseudotumor (multifocal fibrosclerosis?).	Steroid (effective).
55/M[5]	Poor general condition and bilateral exopthalmos. A frozen pelvis in the rectal examination.	Mass surrounding the bladder, seminal vesicles, distal ureters and the anterior rectal wall, and fanning out to the lateral pelvic wall.	Bilateral	US-guided transrectal biopsy.	Bilateral orbital pseudotumors (limited Wegener's granulomatosis?).	Nephrostomy and steroid (effective).
54/F[6]	Incidentally discovered hydronephrosis on US.	Mass posterolaterally to the bladder.	Right	Laparotomy.	No	Surgical resection.
60/M[7]	Constipation, pelvis pain, and weight loss.	Presacral mass.	Right	Laparotomy.	Six months previously, treated with acyclovir for generalized herpes zoster.	Subtotally resected, followed by tamoxifen (not effective).
54/F[8]	Anorexia, nausea, pretibial edama and weight gain.	Presacral mass.	Bilateral	No pathological diagnosis.	Hypertension and systemic lupus erythematosus.	Steroid (effective).
42/F[1]*	NR	Presacral mass, encasing the bladder, the ureters and rectal wall.	NR	NR	Ergotamine for migraine.	Steroid (effective).
69/M[1]*	NR	Presacral mass, encasing the bladder, the ureters and rectal wall.	NR	NR	Arterial hypertension.	Steroid (effective).
65/M[2]	Abdominal fullness and difficulty in defaecations.	Presacral mass, extending to the lateral pelvic wall with irregular thickening of perirectal fascia.	NR	CT-guided biopsy & laparotomy.	Six years previously, gastrectomy for gastric carcinoma ; eight months previously appendectomy ; a long history of diabetes mellitus.	NR
46/M[3]	Dysuria and pollakisuria. An induration of the anterior rectal wall in the rectal examination.	Diffuse infiltration around the iliac vessels and in the perivesical and retrovesical soft tissue.	Bilateral	CT-guided biopsy.	NR	Steroid (effective).
56/M[3]	Constipation. A plane induration of the posterior rectal wall in the rectal examination.	Diffuse infiltration in the retro-and pararectal space, and around the iliac vessels.	Bilateral	US-guided transrectal biopsy.	NR	Steroid (effective).
38/M[9]	Lower abdominal pain and weight loss. Tenderness in the lower pelvis.	Presacral mass infiltrating into the perirectal space.	No	US-guided transrectal biopsy.	NR	Steroid (effective).
55/F[3]	Right flank pain.	Presacral mass.	Right	Laparotomy.	Anticonvulsive medication for temporal lobe epilepsy for 15 years.	Steroid (effective).
29/F[10]	Abdominal pain. Anterior firmness in the rectal examination.	Irregular bladder wall thickness at the right side and an infiltrative mass extending to the adjunctive structures and the iliac crest.	Right	Laparotomy.	NR	Empirical ciprofloxin and indomethazin (non-effective?) Steroid (effective).
68/F**	Right flank pain	Presacral mass, involving the right ureter.	Right	CT-guided biopsy.	Appendectomy and cholecystectomy.	Steroid (effective).

*Vivas et al.[1] Ireviewed 30 patients with retroperitoneal fibrosis, and reported that four of them were localized in the presacral spaces and two in the retrovesical regions. Two of them were reported in detail.

**Present case.

NR : not reported.

necrotic components were associated in any patients. Right-side ureters may be predominantly involved than the left ones, but it was unclear if this difference was significant.

In seven of 14 patients, imaging-guided needle biopsies were employed (four by US-guided and three by CT-guided), avoiding laparotomy to obtain histopathological diagnoses. Good prognoses were reported in all 11 patients in which corticosteroid therapy were given.

Discussion

Clinical signs and symptoms of typical paraaortic RPF are usually related to entrapment and compression of retroperitoneal structures, and may be associated with renal failure.[11,12] The CT and MRI findings of RPF are well known : a paraaortic soft tissue mass of variable size enveloping the lower abdominal aorta, extending laterally to involve the ureters. A paraaortic mass lesion shows a variable degree of enhancement on CT. On MRI, the mass is low intensity on T1-weighted images, but the signal intensity on T2-weighted images can vary according to the state of disease.[2]

However, when RPF appears in pelvic cavity, the symptoms and clinical presentation can vary from gynecological, urological to colonic symptoms. Colonic symptoms and suspected gynecological diseases in the rectal examinations are uncommon in the typical paraaortic RPF. Because of atypical location of the mass lesion the differential diagnosis increases considerably,[1-3] such as lymphomas, infiltrating carcinomas, sarcomas, desmoid tumors, radiation fibrosis and granulomatous diseases (e.g. tuberculosis, sarcoidosis etc.). CT and MRI are clearly useful to demonstrate anatomical detail, although the definitive

diagnosis must be established by means of histopathological evaluation. Although CT-guided biopsy of deep pelvic masses remains challenging because vital structures often obstruct the projected needle path, familiarity with normal cross-sectional pelvic anatomy facilitates planning of a safe access route and helps avoid injury to adjacent structures.[13] For the presacral and perirectal mass lesions the transgluteal approach is suitable, and in three of 14 patients CT-guided needle biopsies were succeeded without any complications. Since there may be relatively few neoplastic cells within the malignant RPF mass, multiple biopsy specimens are required to make a correct diagnosis of benign RPF.

It has been known that about two-thirds of RPF is idiopathic,[12] and the remainders may be induced by drugs, malignancy and abdominal aortic aneurysm.[13] Three of 14 pelvic RPF patients in our literature review might be induced by drugs, and the others were suspected to be idiopathic. RPF may be associated with numerous immune-mediated connective tissue diseases.[12] In two of 14 patients with pelvic RPF, the associations with systemic lupus erythematosus and Wegener's granulomatosis were reported. In a small proportion of patients, the RPF is the results of an exuberant desmoplastic response to metastatic tumor cells within the retroperitoneum, and prognosis of these patients is poor.[12] However, no cases of malignant fibrosis were reported in pelvic RPF.

In cases of benign FPF, corticosteroid therapy has been reported to be effective and the prognosis is usually excellent. In all 11 patients, in which corticosteroid therapy were given for the pelvic RPF, the pelvic mass lesions have been decreased in size followed by good prognoses.

In conclusion, RPF confined to the pelvic cavity is rare, and may show different symptoms and clinical features from those of typical paraaortic RPF. However, pelvic RPF shows similar imaging findings (except for anatomical location), majority of cases are idiopathic, and corticosteroid therapy is effective. RPF should be considered in the differential diagnosis of the presacral and paravesical masses, and the imaging-guided biopsy is the choice for histopathological diagnosis.

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