

Utility of Ultrasonography in Assessing Periaortitis Associated with Retroperitoneal Fibrosis: A Case Report

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Periaortitis is a rare vascular manifestation and is often associated with retroperitoneal fibrosis. Herein, we describe a case of periaortitis accompanied by retroperitoneal fibrosis in a patient who developed acute kidney insufficiency due to bilateral ureteral stenosis. Ultrasonography at presentation detected thickness of the outer layer of the bilateral common iliac artery and right internal and external iliac arteries, consistent with periaortitis. Moreover, follow-up ultrasound images revealed subsiding of the thickness of the arterial wall in response to treatment with corticosteroids. Because of its noninvasiveness and handiness, ultrasonography has become popular for the assessment of large vessels in clinical practice, particularly monitoring for affected lesions. Computed tomography, magnetic resonance imaging, and positron emission tomography are currently used for the diagnosis and monitoring of periaortitis, but in this case, ultrasonography was utilized in the diagnosis and monitoring of periaortitis as a supportive imaging modality, as the use of contrast agents was contraindicated because of renal insufficiency. (*J Nippon Med Sch* 2022; 89: 612–615)

Key words: periaortitis, retroperitoneal fibrosis, ultrasonography

Introduction

Periaortitis is an uncommon disease characterized by a retroperitoneal mass around the abdominal aorta and the iliac arteries¹. Local inflammation is involved in the disease process and includes idiopathic retroperitoneal fibrosis, perianeurysmal retroperitoneal fibrosis, and inflammatory abdominal aortic aneurysms¹. For diagnosis and monitoring of periaortitis, imaging modalities such as computed tomography (CT), magnetic resonance imaging (MRI), and positron emission tomography (PET) are widely used to detect vascular and perivascular lesions^{1,2}, but ultrasonography has not been widely used³. Herein, we report a case of a patient with periaortitis accompanied by retroperitoneal fibrosis leading to acute renal insufficiency, in which the use of contrast agents was contraindicated. The therapeutic response was successfully monitored with serial ultrasound evaluations. This case report suggests that ultrasonography is also a useful im-

aging tool in assessing and monitoring the therapeutic response of patients with periaortitis accompanied by renal insufficiency.

Case Report

A 66-year-old woman was admitted to our hospital with a 2-week history of nausea and vomiting. On admission, there were no significant findings on physical examination. The patient had stable vital signs and was afebrile without enlargement of superficial lymph nodes and salivary glands. Laboratory evaluations revealed normocytic anemia with hemoglobin of 8.8 g/dL, thrombocytopenia with platelet count of $10.9 \times 10^{10}/L$, elevated serum creatinine level (11.84 mg/dL), and elevated serum IgG, IgG4, and IgE levels (2,127 mg/dL, 796 mg/dL, and 1,573 U/mL, respectively). C-reactive protein was within the normal range, antinuclear antibody testing showed a borderline result with a titer of 1:40, and rheumatoid fac-

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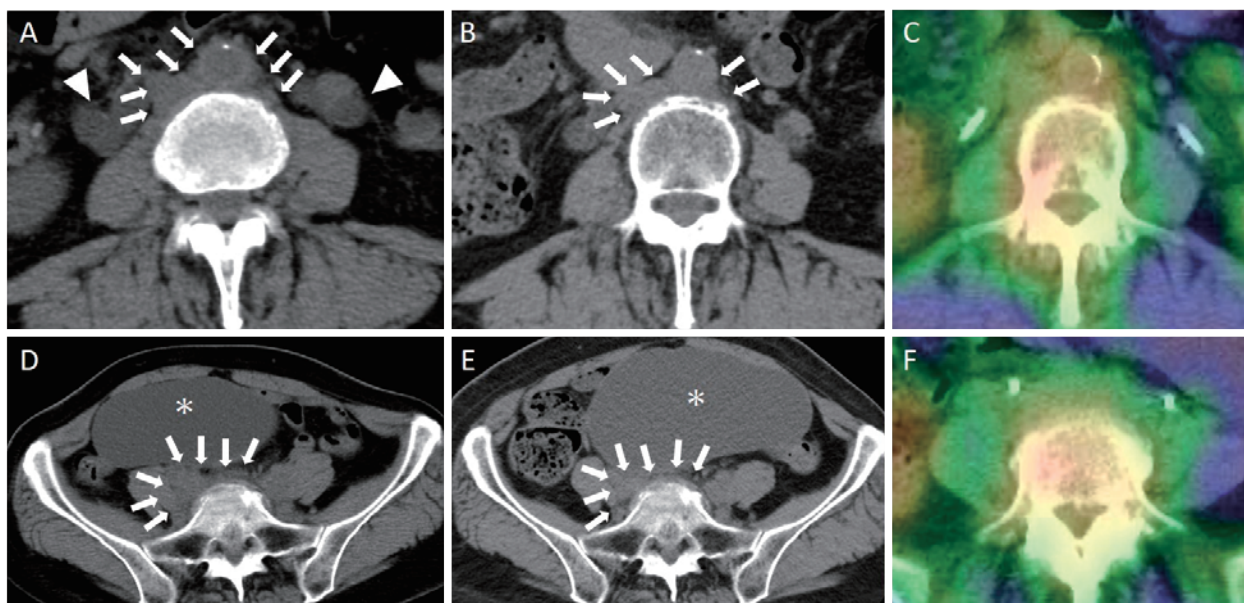


Fig. 1 Serial plain computed tomography scan images of the presented case before (A and D) and after (B and E) treatment with corticosteroids and gallium scintigraphy images before treatment (C and F). A retroperitoneal mass and thickening of the outer layer of abdominal aorta wall improved after the treatment. However, the common iliac arteries within the mass were poorly visualized. Gallium scintigraphy showed mild uptake in the periaortic and retroperitoneal regions surrounding of abdominal aorta and bilateral common iliac arteries (arrow: retroperitoneal mass, arrow head: dilated ureter, asterisks: ovarian cyst).

tor was 29 IU/mL. A series of autoantibodies, including anti-SS-A antibody, antineutrophil cytoplasmic antibodies against myeloperoxidase, and serine proteinase-3, were all negative. An interferon-gamma release assay for *Mycobacterium tuberculosis* was also negative. Despite apparent renal insufficiency, urinalysis with urinary sediment examination showed completely normal findings without proteinuria or hematuria. Urinary cytology was also negative for dysplastic cells. A plain whole-body CT scan revealed a massive retroperitoneal mass with bilateral hydronephrosis (Fig. 1A and D), leading to the diagnosis of acute renal insufficiency due to urethral stenosis associated with retroperitoneal fibrosis. Ureteral stents were immediately placed in the bilateral ureters, but biopsy of the retroperitoneal mass was halted because of prominent renal insufficiency and difficulties using a CT-guided approach. Additional evaluations for retroperitoneal fibrosis were conducted. Gallium scintigraphy showed mild uptake in the periaortic and retroperitoneal regions surrounding of abdominal aorta and bilateral common iliac arteries without significant uptake in other sites of the body (Fig. 1C and F). MRI and PET-CT were not tested. There was no evidence suggestive of conditions causing retroperitoneal fibrosis, including drugs, infection, malignancy, sarcoidosis, trauma, and surgical or radiation operation^{4,5}. A prominent elevation of serum

IgG4 suggested IgG4-related disease as a possible underlying condition, although no definitive diagnosis could be made due to the lack of histological confirmation⁶. Ultrasonography was conducted to evaluate vascular lesions. The ultrasonographic findings were intact in the following vessels: the abdominal aorta, celiac artery, superior mesenteric artery, splenic artery, bilateral renal arteries, left internal and external iliac arteries. In contrast, remarkable thickness of the outer layer of the bilateral common iliac arteries and right internal and external iliac arteries were found (Fig. 2A), indicating a final diagnosis of periaortitis accompanied by retroperitoneal fibrosis. The insertion of the ureteral stents resulted in renal function improvement. For retroperitoneal fibrosis, methylprednisolone pulse therapy (1,000 mg of methylprednisolone for 3 days), followed by oral prednisolone (20 mg daily) was initiated. A follow-up plain CT conducted 5 weeks later identified shrinking of the retroperitoneal mass and improvement of the hydronephrosis (Fig. 1B and E). In addition, thickening of the outer layer of the abdominal aorta wall improved after treatment with corticosteroids. The serial ultrasound images confirmed the improved thickened walls of the affected bilateral common iliac arteries and right internal and external iliac arteries without altering the luminal diameter (Fig. 2B).

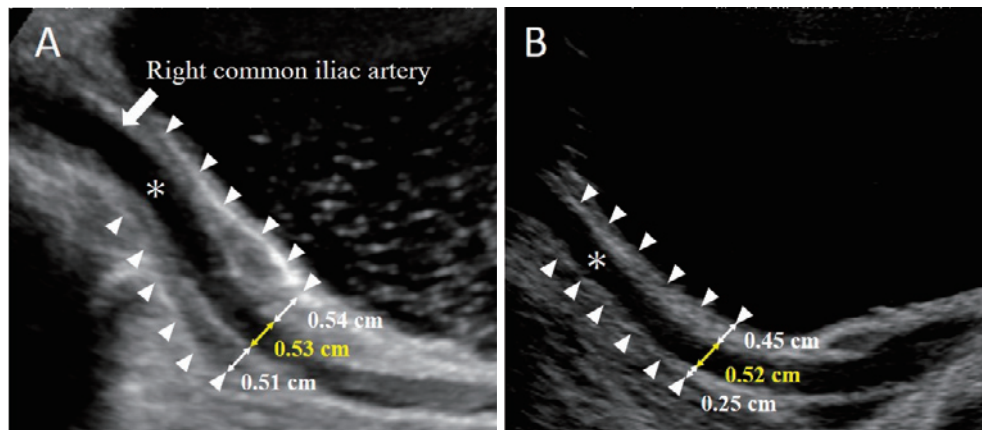


Fig. 2 Serial ultrasound images of the right common iliac artery in the presented case before (A) and after (B) treatment with corticosteroids. The outer layer of the common iliac artery remarkably improved after treatment, while the arterial lumen diameter remained unchanged (arrow head: the outer layer of the common iliac artery, asterisks: the arterial lumen).

Discussion

In this case, ultrasonography was useful in evaluating the arterial lesions for diagnosis and follow-up in a patient with periaortitis accompanied by retroperitoneal fibrosis. The thickening of the outer layer of the common iliac arteries was not visualized by a plain CT scan. Ultrasonography images should be utilized in the diagnosis and assessment of the treatment responses in patients with periaortitis, especially when the use of contrast agents is contraindicated because of renal insufficiency.

Ultrasonography is becoming popular in various medical fields due to its noninvasiveness, handiness, and low cost. Ultrasonography is widely used in the evaluation of large veins in the lower extremities and has recently started to be employed for the assessment of large arteries^{7,8}. In particular, ultrasonography has been recommended as the first imaging modality in patients suspected to have large vessel vasculitis, such as giant cell arteritis⁷. A “halo” sign encompassing the entire circumference of the hypoechoic thickened wall and intraluminal stenosis are indicative of large vessel vasculitis⁷. In contrast, in this case, thickening dominated the outer layer of the walls of the affected arteries, consistent with morphological feature of periaortitis^{4,9,10}. In addition, ultrasonography was able to illustrate the vascular lesions in major branches of the aorta and provide information on the primary affected lesion of the vascular wall (inner versus outer layers), while these were not evaluable in detail by a plain CT scan.

CT, MRI, PET, and gallium scintigraphy are currently used for the diagnosis and monitoring of periaortitis^{1,2,11}.

Table 1 summarizes the advantages and disadvantages

of each imaging modality in diagnosing and monitoring patients with periaortitis. Using contrast agents is desirable for assessment of periaortitis. Plain CT reveals soft tissue masses surrounding the abdominal aorta. In addition, contrast CT shows varying degree of enhancement of periaortic lesions². With regard to MRI, plain MRI enables assessment of the vessel wall. Furthermore, contrast-enhanced MRI is more useful to assess vessel wall inflammation with high resolution than plain MRI¹². However, in the presence of acute or chronic kidney disease, the use of contrast agents in CT and MRI is contraindicated. In this case, the resolution of imaging is degraded without the use of contrast agents. In this regard, periaortitis is commonly accompanied by retroperitoneal fibrosis, which often causes post-renal acute renal injury¹³. This is also true for patients with hypersensitivity to the contrast agents. In addition, monitoring of the disease activity of retroperitoneal fibrosis and periaortitis is crucial, since relapses are common^{9,14}. Ultrasonography enables to distinguish between periaortic and retroperitoneal lesions by detecting thickening of the vessel walls, and it is particularly useful in cases where contrast agents cannot be used due to renal dysfunction. Moreover, ultrasonographic assessment has advantages in terms of handiness, risk of radiation exposure, and cost-effectiveness. At the same time, ultrasonography has several disadvantages: image visualization depends on the scanning skills and experience of operators; fat tissues and excessive intestinal gas prevent clear vascular imaging; and there is no standardized protocol for the assessment of patients with periaortitis.

In conclusion, this case report suggests the usefulness

Table 1 Advantages and disadvantages of the imaging modalities in diagnosing and monitoring patients with periaortitis

Modality	Advantage	Disadvantage
US	No radiation exposure Easy accessibility Low cost No contrast agent required Handiness Distinction between periaortic lesion and retroperitoneal lesion	Skill-dependent Imaging limitation by artifacts No standardized protocol Inability in drawing thoracic and abdominal aorta
Plain/contrast-enhanced CT	Easy accessibility Short imaging time Standardized protocol available Whole body imaging	Radiation exposure High cost A contrast agent required for fine vascular imaging
MRI	No radiation exposure Standardized protocol available Whole body imaging	A contrast agent required for fine vascular imaging High cost Trapped in a confinement
PET-CT	Shows metabolic activity Standardized protocol No nephrotoxic contrast agents Whole body imaging	Lack of accessibility High cost Radiation exposure
Gallium scintigraphy	Shows metabolic activity Standardized protocol No nephrotoxic contrast agents Whole body imaging	Lack of accessibility High cost Radiation exposure Difficulty in distinguishing between periaortic lesion and retroperitoneal lesion

US, ultrasound; CT, computed tomography; MRI, magnetic resonance imaging; PET, positron emission tomography

of ultrasonography as a supportive noninvasive imaging tool for the diagnosis and monitoring of patients with periaortitis accompanied by retroperitoneal fibrosis.

Consent: A written informed consent for the publication of this report was obtained from the patient.

Conflict of Interest: The authors declare no conflict of interest.

References

- Vaglio A, Greco P, Corradi D, et al. Autoimmune aspects of chronic periaortitis. *Autoimmun Rev*. 2006;5:458–64.
- Zeina AR, Slobodin G, Naschitz JE, Loberman Z, Barmer E. Isolated periaortitis: clinical and imaging characteristics. *Vasc Health Risk Manag*. 2007;3:1083–6.
- Czihal M, Lottspeich C, Hoffmann U. Ultrasound imaging in the diagnosis of large vessel vasculitis. *Vasa*. 2017;46:241–53.
- Vaglio A, Salvarani C, Buzio C. Retroperitoneal fibrosis. *Lancet*. 2006;367:241–51.
- Tzou M, Gazeley DJ, Mason PJ. Retroperitoneal fibrosis. *Vasc Med*. 2014;19:407–14.
- Umehara H, Okazaki K, Masaki Y, et al. Comprehensive diagnostic criteria for IgG4-related disease (IgG4-RD), 2011. *Mod Rheumatol*. 2012;22:21–30.
- Dejaco C, Ramiro S, Duftner C, et al. EULAR recommendations for the use of imaging in large vessel vasculitis in clinical practice. *Ann Rheum Dis*. 2018;77:636–43.
- Schmidt WA, Nerenheim A, Seipelt E, Poehls C, Gromnica-Ihle E. Diagnosis of early Takayasu arteritis with sonography. *Rheumatology (Oxford)*. 2002;41:496–502.
- Vaglio A, Buzio C. Chronic periaortitis: a spectrum of diseases. *Curr Opin Rheumatol*. 2005;17:34–40.
- Parums DV. The spectrum of chronic periaortitis. *Histopathology*. 1990;16:423–31.
- Mizushima I, Inoue D, Yamamoto M, et al. Clinical course after corticosteroid therapy in IgG4-related aortitis/peri-aortitis and periarteritis: a retrospective multicenter study. *Arthritis Res Ther*. 2014;16:R156.
- Kato Y, Terashima M, Ohigashi H, et al. Vessel wall inflammation of Takayasu Arteritis detected by contrast-enhanced magnetic resonance imaging: association with disease distribution and activity. *PLoS One*. 2015;10:e0145855.
- Ezimora A, Faulkner ML, Adebisi O, Ogungbemile A, Marianna SV, Nzerue C. Retroperitoneal fibrosis: a rare cause of acute renal failure. *Case Rep Nephrol*. 2012;2012:645407.
- Raffiotta F, da Silva Escoli R, Quaglini S, et al. Idiopathic retroperitoneal fibrosis: Long-term risk and predictors of relapse. *Am J Kidney Dis*. 2019;74:742–50.

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