Unusual Presentation of Giant Cell Arteritis in 2 Patients: Uterine Involvement

To the Editor:

Giant cell arteritis (GCA) has been associated with extracranial involvement, usually affecting large- and medium-sized vessels. Involvement of the female genitourinary tract is rare, with < 30 cases reported in the literature[1,2,3]. Here, we discuss 2 cases of incidentally discovered uterine GCA ultimately found to also involve the aorta on positron emission tomography (PET) scan.

Ethics board approval was not required according to the authors' institution (Mayo Clinic, Jacksonville, Florida, USA). Both patients gave written permission to publish the material.

Case 1 involved a 73-year-old female with a history of polymyalgia rheumatica (PMR) 5 years prior and presented to rheumatology after surgical uterine pathology following hysterectomy for symptomatic uterine prolapse. The patient demonstrated GCA involving small and medium-sized vessels.

In the months preceding hysterectomy, she reported symptoms suggestive of PMR relapse, although she did not seek care. About 3 weeks after hysterectomy, her erythrocyte sedimentation rate (ESR) was elevated at 71 mm/h. She presented to us about 10 days later. She had no symptoms suggestive of cranial involvement.

Evaluation revealed symmetric blood pressures, normal pulses, and no bruits. The remainder of the physical examination was unremarkable. Her C-reactive protein (CRP) was elevated at 31.3 mg/l (normal level < 8.0 mg/l) and she was mildly anemic. ESR was normal. A PET scan identified hypermetabolic aortitis along the entire course of the vessel, highly suggestive of GCA with standardized uptake value (SUV) score ranging from 4.6 to 5.3 (Figure 1).

Case 2 concerned a 64-year-old female who was referred to rheumatology for an opinion regarding GCA involving the uterus after a hysterectomy was performed for uterine prolapse (Figure 2). ESR and CRP levels obtained 2 months after hysterectomy were within normal limits. She had no symptoms of GCA and no additional investigation was performed.

She presented to us 3 months later for a second opinion. She denied any classic symptoms of PMR or GCA; however, she reported about 40 lb unintentional weight loss over the past 20 months.

Evaluation revealed symmetric blood pressures, normal pulses and no bruits. The remainder of the physical examination was unremarkable. She had normal ESR and CRP. A PET scan noted moderate hypermetabolism along the entire course of the aorta (SUV score 3.1–3.4) and the major branches of the aortic arch were consistent with GCA.

Pathologic examination of the uteri in both cases revealed several arteries in the deep myometrium and parametrium with a transmural inflammatory infiltrate, including lymphocytes, histiocytes, and scattered giant cells. Often the intima of the arteries showed proliferation with intimal arteritis. No fibrinoid necrosis or fibrin thrombi were identified.

GCA involvement of the uterus is uncommon, although it could be speculated that many cases are unrecognized. The majority reported are associated with PMR features, with a small number describing temporal arteritis symptoms prompting biopsy. About half of the cases reported nonspecific inflammatory symptoms such as fever, malaise, or weight loss. In a series of 29 patients with gynecologic GCA by pathology, gynecologic symptoms included metrorrhagia (n = 6), uterine prolapse (n = 5), atypical cervical smear (n = 1), painful abdominal mass/pelvic pain (n = 3), and asymptomatic abdominal mass (n = 12)[1]. Two patients were asymptomatic. GCA of the male genitourinary tract has been reported in 1 patient with involvement of the epididymis, and another with prostate involvement[4,5]. Both our patients had incidental GCA findings on uterine pathology following hysterectomy for uterine prolapse. Our first patient had concomitant PMR symptoms with elevated CRP, and she responded well to low-dose corticosteroids. Our second patient exhibited only weight loss. Radiographic evidence for large-vessel vasculitis on PET scan was identified in both. The use of techniques such as PET scan may assist in the evaluation of these patients to identify occult areas of inflammation[6].

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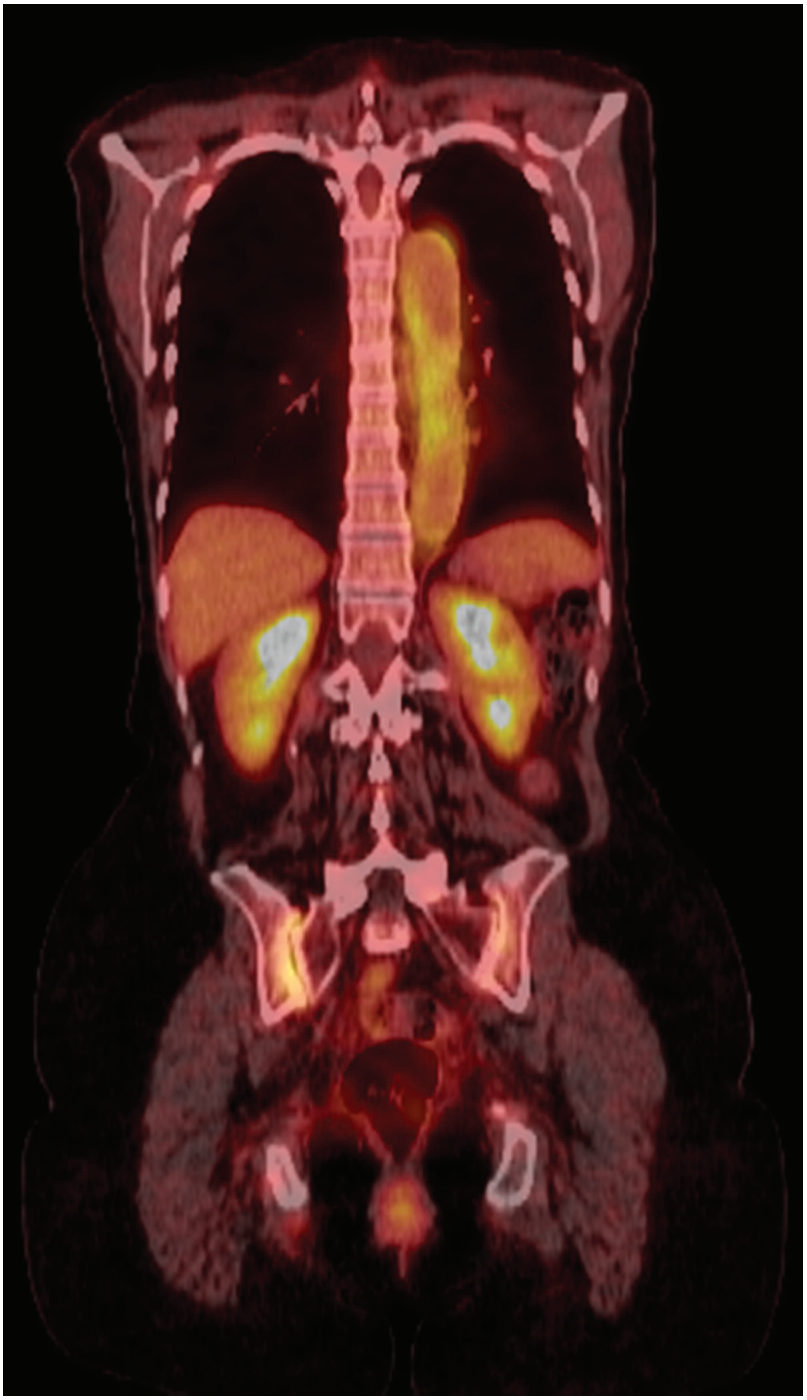

Figure 1. Diffusely increased FDG uptake in the wall of the thoracic aorta. This is most prominent in the descending thoracic aorta. FDG: 18F-fluorodeoxyglucose.

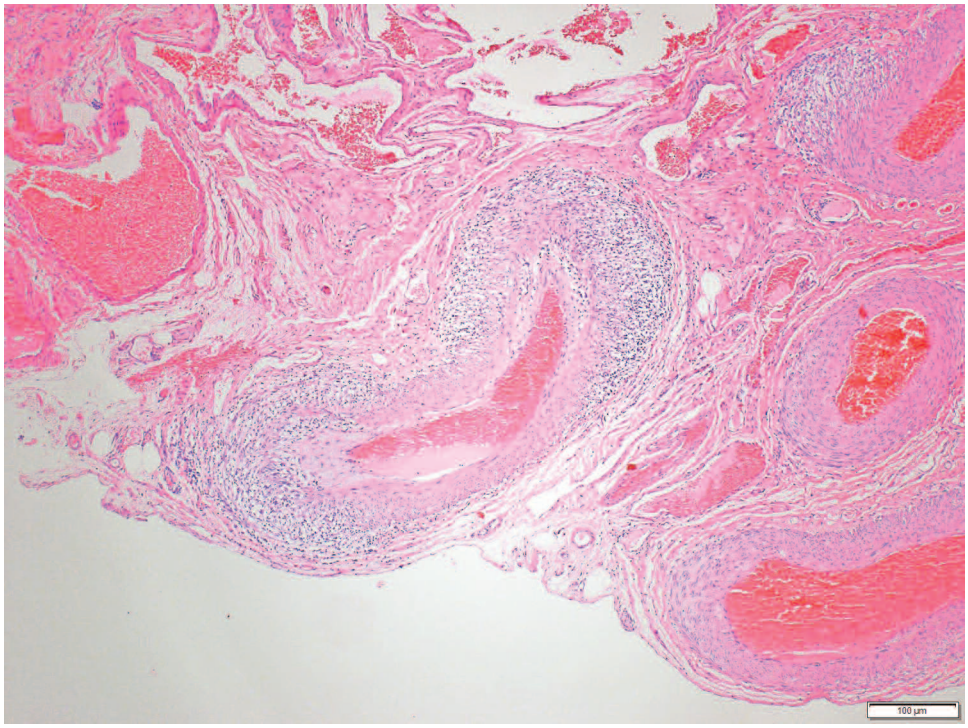

Figure 2. Overview of the uterine serosa, which demonstrates arteritis involving several vessels.