Chronic Periaortitis

Nicolò Pipitone, MD, PhD; Alessandra Ghinoi, MD; Annibale Versari, MD; Augusto Vaglio, MD; Alessandra Palmisano, MD; Carlo Salvarani, MD

A 60-year-old woman presented in 2004 with fatigue, anorexia, and weight loss. Her erythrocyte sedimentation rate and C-reactive protein levels were elevated at 114 mm/h and 45 g/dL (normal values <6 g/dL), respectively, whereas autoimmune serology, including antinuclear antibodies, antibodies to extractable nuclear antigens, and antineutrophil cytoplasmic antibodies, and tests for common infectious diseases, including tuberculosis, were normal or negative. Occult neoplasm was suspected and computerized tomography (CT) of the abdomen and F18-fluorodeoxyglucose positron emission tomography (PET) were scheduled. CT showed concentric wall thickening of the aortic arch, ascending thoracic aorta, and proximal left internal carotid arteries, with a perivascular cuff around the abdominal aorta. PET disclosed increased tracer uptake at the ascending and abdominal aorta and, to a lesser extent, in the common iliac arteries. Chronic periaortitis (CP) was diagnosed on the basis of these symptoms and findings, and treatment with prednisone 1 mg $\cdot$ kg^{-1} $\cdot$ d^{-1} was begun, with marked clinical improvement and normalization of erythrocyte sedimentation rate and C-reactive protein level. A repeat CT scan demonstrated a reduction in the size of the abdominal periaortic cuff, and a second PET scan documented reduced tracer uptake in the ascending and abdominal aortas with unchanged iliac artery uptake. However, attempts to taper the glucocorticoid dose resulted in 3 flares over the subsequent 3 years that required retreatment with high-dose glucocorticoids in combination with methotrexate. At her last visit in 2007, the patient complained of fatigue. Her erythrocyte sedimentation rate was 72 mm/h and her C-reactive protein level was 7 g/dL. A PET/CT scan revealed increased tracer uptake in the aortic arch, ascending thoracic aorta, and abdominal aorta with an abdominal perivascular cuff, consistent with active vasculitis and CP, respectively (Figures 1 through 3). Magnetic resonance angiography of the lower thoracic and abdominal aortas disclosed narrowing of the lumen of the aorta below the renal arteries and of the proximal iliac arteries, and short tau inversion recovery sequences showed increased signal (consistent with inflammatory edema) of the vessel wall (Figure 4). A chest x-ray was unremarkable apart from an enlarged left hilum and a small retrosternal nodule that had previously been documented (Figure 5). ECG showed 1-mm ST-segment depression in leads V5 through V6 (Figure 6). Transthoracic echocardiography confirmed the presence of a thickened ascending aortic wall, with a periaortic hypoechoic halo consistent with periaortic inflammatory tissue (Figure 7).

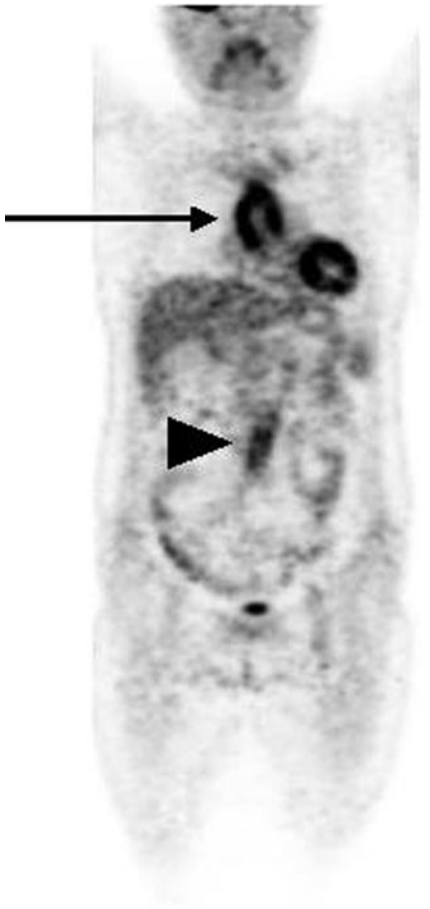

Figure 1. F18-Fluorodeoxyglucose positron emission tomography (coronal view) showing increased tracer uptake in the ascending (arrow) and abdominal (arrowhead) aorta.

CP is a rare, mostly idiopathic, fibroinflammatory condition that ranges from inflammatory aneurysm of the abdominal aorta with or without perianeurysmal fibrosis to isolated retroperitoneal fibrosis.[1] The clinical picture is usually characterized by constitutional symptoms and by dull abdominal

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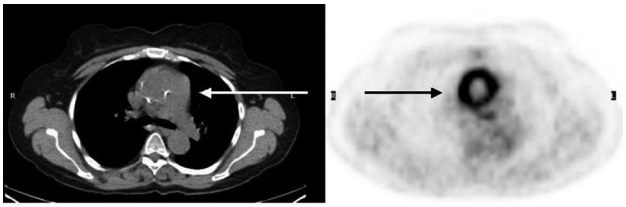

Figure 2. Left, CT (axial view) of the chest showing concentric wall thickening (white arrow) of the ascending aorta. Right, F18-Fluorodeoxyglucose positron emission tomography (axial view) showing increased tracer uptake of the wall of the ascending thoracic aorta (black arrow).

and lower back pain. Manifestations related to extrinsic compression of the ureters and of retroperitoneal lymphatics and veins may also occur.

On an unenhanced CT scan, CP usually appears as a homogeneous, muscle-isodense plaque surrounding the lower abdominal aorta and the iliac arteries, sometimes encasing the ureters and/or the inferior vena cava.[1] On magnetic resonance imaging, CP is hypointense in T1-weighted sequences and hyperintense in T2-weighted images in the early and active stages of disease because of inflammatory edema, whereas low-intensity signal is commonly seen in the late fibrotic stages.[1] PET is not specific for the diagnosis of CP, but it is sensitive in evaluating the metabolic activity of the retroperitoneal mass[2] and also allows whole-body large-vessel imaging. Finally, combined PET/CT enables both metabolic and morphological imaging to be performed in a single session.[3] We have recently demonstrated[4] involvement of the thoracic aorta and of its branches in 43% of CP patients. This suggests that CP may represent, at least in some patients, a systemic vasculitis, rather than a localized inflammatory reaction to antigens in the atherosclerotic plaques of the abdominal aorta as previously thought.[5] The hypothesis that CP may represent a systemic vasculitis is also supported by the presence of constitutional symptoms and by the evidence of raised inflammatory markers.

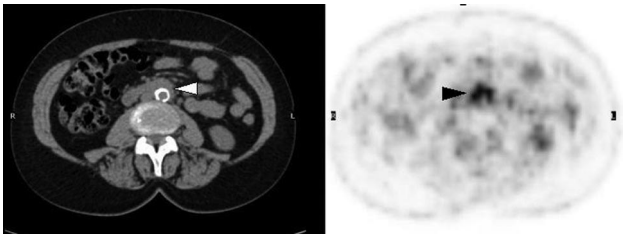

Figure 3. Left, CT (axial view) of the abdomen showing a periaortic fibroinflammatory cuff (white arrowhead). Right, F18-Fluorodeoxyglucose positron emission tomography (axial view) showing increased tracer uptake of the wall of the abdominal aorta and of the periaortic fibroinflammatory cuff (black arrowhead).

Disclosures

None.

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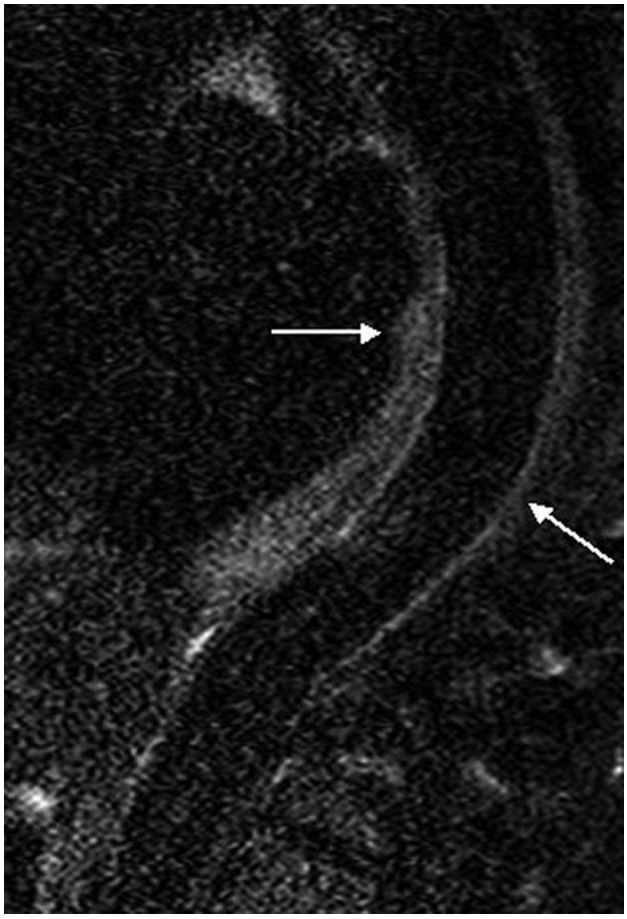

Figure 4. Magnetic resonance of the lower thoracic and of the abdominal aorta. Short tau inversion recovery image showing increased signal (arrows) of the aortic wall, consistent with inflammatory edema.

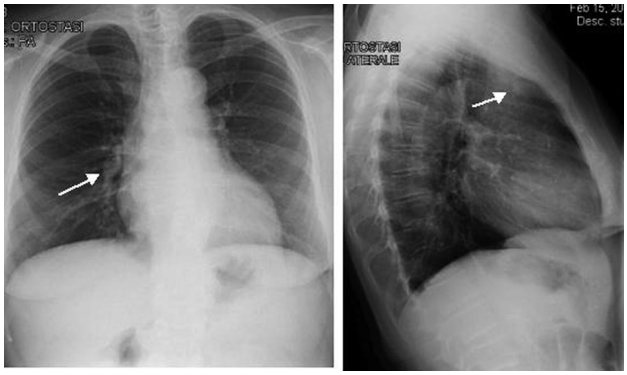

Figure 5. Chest x-ray of the patient, posteroanterior (left) and lateral (right) views. Apart from an enlarged left hilum (arrow, left) and a small retrosternal nodule (arrow, right) that had previously been documented, there were no significant abnormal findings. In particular, there were no parenchymal lesions or pleural effusion, and the heart shadow was not enlarged.

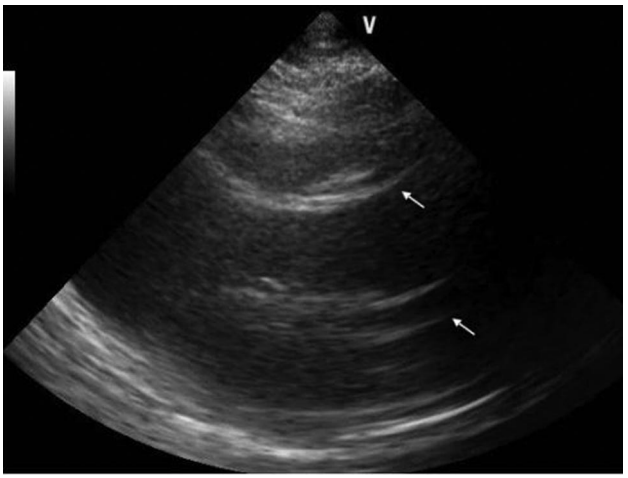

Figure 7. Echocardiography showing thickening of the ascending aortic wall (arrows), with periaortic hypoechoic halo consistent with periaortic inflammatory tissue.

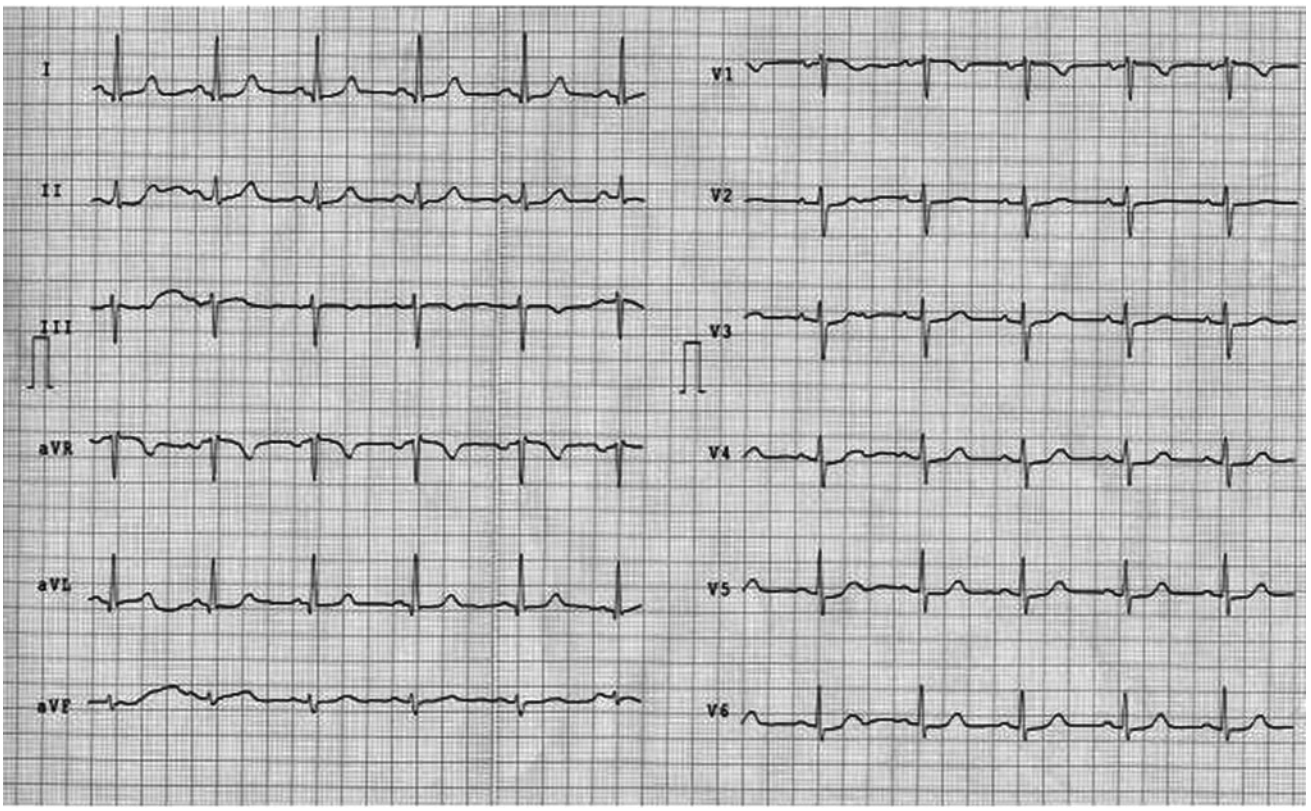

Figure 6. ECG showing sinus rhythm, heart rate 73 bpm, axis in normal range (0 degree), normal PR, QRS complex, and QT intervals, and 1-mm ST-segment depression in leads V5 through V6.

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