Increased Metabolic Activity Highlighted by Positron Emission Tomography/Computed Tomography in the Wall of the Dissected Ascending Aorta in a Patient With Horton Disease

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Horton disease or giant-cell arteritis (GCA) is a chronic systemic vasculitis involving typically medium and large arteries. Giant-cell arteritis is a panarteritis characterized by a granulomatous inflammation, with lymphocytes, macrophages, and multinucleated giant cells related to autoimmune T-cell reactivity.1 Compared with conventional imaging tools (ultrasound, computed tomography (CT), MRI, and contrast angiography) that provide anatomic and morphological information, recent available imaging techniques such as positron emission tomography (PET)/CT provide metabolic assessment of the arterial wall. During the early 2000s, Sakalihasan et al2 observed a close correlation between clinically unstable abdominal aortic aneurysms and increased uptake of 18F-fluoro-2-deoxy-D-glucose (FDG) in the aneurysmal wall. A few years later, Hautzel et al3 studied the assessment of giant-cell arteritis with PET/CT. We describe a case of Horton disease involving the thoracic aorta and complicated with acute aortic dissection in a woman with a previous diagnosis of thoracic aortic aneurysm.

Case Presentation

In July 2011, a 66-year-old woman was referred to a cardiology center for evaluation of a recent mild hypertension related to use of high doses of corticosteroids. In December 2010, she had developed severe headache, rapid loss of weight, and elevation of sedimentation rate as high as 120 mm. Horton disease was diagnosed in April 2011 on temporal artery biopsy. During the first cardiologic examination, all features were normal, including revision of recent chest radiography, aortic root dimension (38 mm), and aortic valve function (except a minimal aortic regurgitation). Because of increased D-dimer level (4261 ng/mL) and the presence of limited deep venous thrombosis, a pulmonary V/Q single-photon emission CT/CT was performed to exclude pulmonary embolism. The low-dose CT part of this examination revealed a dilated arch and descending thoracic aorta. Contrast-enhanced CT confirmed the diagnosis of thoracic aortic aneurysm, with an estimated diameter of 60 mm for the aortic isthmus and for the descending aorta. (Figure 1A). The regular preoperative workup included routinely an FDG-PET/CT examination in all patients referred for aortic aneurysm, diagnosed initially by CT scan, and scheduled for surgery. This prospective translational study was designed and conducted within the framework of the European program Fighting Aneurysmal Disease. In this case, 18F-FDG PET/CT, which was performed 2 weeks later, showed an unexpected 12-mm increase in the size of the aneurysm (maximal diameter, 72 mm) and an asymptomatic dissection with an entry hole in the proximal ascending (A1) aortic segment. The FDG uptake was particularly high in the whole flap and maximal close to the entry hole in the proximal ascending aorta, as well as at the origin of the brachiocephalic and left carotid arteries (Figure 1B–1D).

During surgery to replace the aortic arch, pieces of dissected segments from the thoracic aorta, carotid artery, and brachiocephalic trunk were collected for histological analysis and to visualize smooth muscle cell organization in the media and phagocytic activity of heme iron (Figures 2 and 3).

Discussion

Thoracic aortic dissection is common in giant-cell arteritis and is associated with increased mortality.4 Subclinical aortic inflammation is often present in patients with giant-cell arteritis, and this inflammatory process may deteriorate the vessel wall, resulting in aortic aneurysm and dissection.5 In our patient, the histopathological analysis of the flap and dissected aortic wall, which were hypermetabolic on PET imaging, confirmed the presence of giant cells and lymphocytic granuloma, suggesting an association between the activation of these granuloma, FDG uptake, and the weakening of the vascular wall.
However, Perl+DAB (Di Amino Benzidine) staining was relatively intense in the external part of the media and the adventitia with regard to the hemothrombus (brachiocephalic trunk), providing evidence of phagocytic activities. Kato et al4 used FDG-PET/CT for predicting adverse outcomes in aortic dissection and showed that FDG uptake in the dissected aorta wall was significantly associated with an increased risk of progression and rupture. Horton disease can potentially lead to fatal complications, and our clinical observation provides the opportunity to underline the importance of an early detection of large-vessel pathology, which constitutes the principal threat. It is important to draw up a diagnosis before complications occur. Monitoring and aortic follow-up with PET/CT are warranted to perform a timely prophylactic and beneficial aortic surgery.

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References

Key Words: Horton disease • dissection • positron emission tomography

Figure 1. A, Computed tomography (CT) image shows the presence of the large thoracic aortic aneurysm without dissection. B, Left oblique 3-dimensional positron emission tomography (PET) image showing a dilated arch and descending thoracic aorta and a mostly clear hypermetabolism of the entire thoracic aorta and left subclavian artery. C, Coronal CT image showing the dissected ascending aorta. D, PET/CT fusion image clearly showing an increased 18F-fluoro-2-deoxy-D-glucose (FDG) uptake in the flap. Note that the focus with maximal FDG uptake (blue circle) is located at the proximal ascending segment, close to the entry hole. Note also the high uptake at the origin of the dissected brachiocephalic and left carotid arteries.

Figure 2. General organization of the different arterial walls. Different staining was performed on sections from the thoracic aorta (A, D, G, and J), the carotid artery (B, E, H, and K), and the brachiocephalic trunk (C, F, I, and L). By hematoxylin/eosin staining (H/E), the false channel was clearly observed in the 3 segments (A–C). A large inflammatory infiltrate was visualized in the thoracic aorta and seemed to be organized in adventitial tertiary lymphoid organs (arrow in A and at higher magnification in D). Arrows in E and F show giant cells. The elastic fiber density and organization in the media were maintained as seen by orcein staining in the 3 segments (G–I). Smooth muscle cells, visualized by an immunostaining with anti-α-smooth muscle actin (α-SMA, J–L), appeared less dense and rounded, having lost the lamellar infiltration along the false channel showed by arrows. The inset corresponds to a ×2.5 magnification of the original picture. Scale bar, 2 mm in A–C; 50 μm in D–F; and 500 μm in G–L. I indicates intima; and m, media.
Figure 3. Phagocytic activity in the different arterial specimens. Perl blue+DAB (di amino benzidine) staining showing phagocytic activity in the adventitia of the thoracic aorta (A and B), carotid artery (C and D), and brachiocephalic trunk (E–H). Note in E the DAB positivity of the hemothrombus present in the false channel and phagocytic activity of heme iron with regard to hemothrombus. Scale bar, 1 mm in A, C, E, and G; 25 μm in B, D, F, and H.
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