A 53-year-old male patient with chronic renal failure and biopsy-proven Immunoglobulin G4 (IgG4) renal involvement presented with complaints of chest pain and syncope. An ECG showed lateral ST segment depressions concerning for myocardial ischemia. The patient underwent coronary catheterization, which revealed multifocal aneurysms and severe stenosis. A cardiac computed tomographic angiogram was performed to further characterize aneurysms, which showed aneurysms involving left main, proximal left anterior descending artery, and proximal right coronary artery (Figures 1 and 2). Circumferential mural thickening with associated enhancement of the wall of the coronary arteries, particularly at the site of the stenoses, was concerning for vasculitis. A small pulmonary artery aneurysm was seen in the right upper lobe (Figure 3). Abdominal computerized tomography demonstrated abdominal aortic aneurysm as well as aneurysmal dilatation of dissected superior mesenteric artery (Figure 4). The patient underwent coronary artery bypass grafting surgery with the saphenous vein graft to the mid left anterior descending artery, ramus intermedius, and posterior descending artery. Intraoperative biopsies of the internal mammary artery, pericardium, and aortic adventitia were positive for lymphoplasmacytic infiltrate and IgG4 deposits.

Discussion

IgG4-related disease is a recently established, chronic fibro-inflammatory condition with multiorgan involvement, such as dacrtyoadenitis and sialadenitis, thyroiditis, type 1 autoimmune pancreatitis, nephritis, lymphadenopathy, and lung involvement.1 IgG4-related disease usually affects middle-aged to elderly patients with male preponderance. Pathologically, IgG4-related disease is characterized by tissue infiltration of IgG4-positive plasma cells, storiform fibrosis, obliterator phlebitis, and elevated concentration serum IgG4.2 Tissue biopsy is the gold standard for diagnosis of IgG4-related disease; however, imaging is an important part of the diagnostic workup of these patients. Arterial involvement in IgG4-related conditions usually affects the large arteries, including aorta and great vessels. However, involvement of medium-sized arteries, such as carotid and intracranial, coronary, pulmonary, mesenteric, and peripheral arteries, has been rarely described in the literature.3 IgG4-related arterial lesions are radiologically characterized by arterial wall thickening luminal dilatation or rarely stenosis, and homogeneous enhancement on the late phase of contrast-enhanced computerized tomography.4 The differential diagnosis of IgG4-related vasculitis includes giant-cell or Takayasu’s arteritis, infectious aortitis, Erdheim-Chester disease, and histiocytosis.2 Cross-sectional imaging techniques, such as coronary computerized tomographic angiography and magnetic resonance angiography, have an important advantage over catheter angiography, which allows direct visualization of vessel wall for wall thickening and mural enhancement, and this feature is useful for diagnosis of vasculitis and follow-up.

Disclosures

None.

References


Key Words: aortic valve = cardiac computed tomography = cardiac catheterization = coronary artery disease = vasculitis

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Figure 1. Catheter angiography (A) and correlative 3D volume-rendered images (B) demonstrate multifocal aneurysmal dilatation of the left main (LM) and left anterior descending (LAD) arteries with focal stenoses. Curved multiplanar reformatted (C) and oblique coronal plane (D) images of the LM and LAD showing diffuse circumferential mural thickening with associated enhancement of the wall (arrows).

Figure 2. Catheter angiography (A) and correlative 3D volume-rendered images (B) demonstrate diffuse ectasia of the right coronary artery (RCA) with significant proximal aneurysmal dilatation of the vessel. Again, curved multiplanar reformatted (C) and oblique axial plane (D) images of the RCA showing diffuse mural thickening of the RCA (arrows).

Figure 3. Axial minimum intensity projection (MIP) image through the chest at the level of the pulmonary valve demonstrates 1 cm enhancing lesion in the right upper lobe, communicating with a pulmonary artery branch, consistent with pulmonary artery aneurysm (arrow).

Figure 4. Coronal image of the abdomen shows aneurysmal dilatation of the abdominal aorta (asterisk) and common iliac arteries. Also noted aneurysmal dilatation of the superior mesenteric artery (arrowheads) with a dissection flap (arrow).
IgG4-Related Disease With Coronary Arteritis
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