A 69-year-old female patient was admitted to our hospital for further cardiac evaluation with symptoms of dyspnea (New York Heart Association class III) and peripheral edema. Her history revealed artificial mitral valve replacement due to mitral stenosis 8 years earlier, severe chronic pulmonary hypertension, and severe tricuspid valve incompetence.

Electrocardiogram showed normofrequent atrial fibrillation. Transthoracic echocardiography indicated moderate biatrial enlargement, although both ventricles had normal dimensions and normal global systolic function. The transmitral prosthetic valve gradient was 14/6 mm Hg. We documented severe tricuspid regurgitation and only mild mitral regurgitation. Nevertheless, echocardiography was difficult to perform because of lack of an adequate acoustic window and metal artifacts of the mitral valve prosthesis. Reflections at the epicardial surface close to the mitral valve were therefore interpreted as calcifications of pericardial layer, and constrictive pericarditis was expected to be the most likely cause of the patient’s symptoms and diastolic left ventricle dysfunction.

By right heart catheterization, severe postcapillary hypertension was confirmed (systolic/diastolic/mean pulmonary artery pressure, 65/23/43 mm Hg; mean pulmonary wedge pressure, 29 mm Hg). In all cardiac cavities, an end-diastolic pressure equilibration was measured (Figure 1). Pressure curves did not show a dip-plateau phenomenon. Therefore, constrictive pericarditis was excluded. However, in a 64-row multidetector computed tomography scan, the endocardial layer of the left atrium (LA) and the aortic arch were heavily calcified (Figure 2), whereas the pericardium itself did not show any relevant calcifications. As an additional finding, the patient showed tracheobronchopathia osteoplastica (Figure 2A and 2B). Cardiac MRI was performed to further assess ventricular and atrial function (Figure 3). Any acute or persistent cardiac inflammation or interstitial storage cardiomyopathy could be excluded by edema-sensitive T2-weighted short T1 inversion recovery (STIR) images and T1-weighted images early and late after intravenous Gd-DTPA according to the protocol previously described. The

Figure 1. End-diastolic pressure equilibration in all cardiac cavities. Pressure curves of left ventricle and left atrium (A), right ventricle and left ventricle (B), right atrium and left ventricle (C).
dynamic cine MRI study showed no relevant changes in LA volumes during the heart cycle, indicating a stiff LA due to the heavily isolated calcification of the endocardial surface of the LA (Figure 3).

The patient’s symptoms were explained as combination of elevated LA pressure and pulmonary hypertension. The singular calcification of the stiff LA led most likely to reduced filling of the LA itself. In combination with diastolic dysfunction and former mitral stenosis, it might be the main cause for backward failure and secondary pulmonary hypertension, tricuspid incompetence, and right heart failure. Therapeutic management included medication for diastolic dysfunction and decrease of afterload with diuretics (hydrochlorothiazide, furosemide), angiotensin-converting enzyme inhibitors and β-blockers. A surgical approach was abandoned because of low probability of success.

Discussion

Two rare conditions were found in 1 patient: a calcified LA and a tracheobronchopathia osteoplastica. The presence of LA calcification, also known as “porcelain atrium” or “coconut atrium,” has been reported as a result of extensive rheumatic pancarditis. The etiology of tracheobronchopathia osteoplastica, a mostly asymptomatic condition of the tracheobronchial tree with a probably benign course, is still unknown. Several other mechanisms may account for endo-
cardial LA calcification, including postoperative complication after mitral valve replacement due to hematoma and inflammation; atrial wall response to chronic strain forces present in the setting of mitral disease, idiopathic, or in the context of rheumatic fever in combination with tracheobronchopathia osteoplastica and calcification of the aorta; ischemic myocardial damage with dystrophic calcification in remodeling; or hypercalcemia and hyperphosphatemia (eg, in metastatic calcification or in combination with renal failure). In our patient, malignancies, electrolyte disturbances, or renal failure could be excluded. Although the patient was unable to recall a history of rheumatic disease, which may have been oligosymptomatic, it seems to be the most likely diagnosis.

Disclosures

None.

References

Endocardial Calcification of Left Atrium, Tracheobronchopathia Osteoplastica, and Calcified Aortic Arch in a Patient With Dyspnea
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