Cor Triatriatum Sinister
– Source of Unusual Thrombogenesis in Mitral Stenosis –
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Figure 1. (A) Abdominal computed tomography showing a segmental infarction in the right kidney (arrowheads). (B) Transthoracic echocardiogram demonstrating an enlarged left atrium (LA) and linear structure (arrowheads) crossing the LA. (C) Transesophageal echocardiogram (TEE) showing 2 distinct left atrial chambers divided by a thin membrane (arrowheads). (D) Bicaval view showing a thrombus (arrow) in an accessory chamber. (E) Three-chamber view showing the thrombus (arrow), which took on the appearance of a barking dog on the membrane (arrowheads) of the septal side of the anteroinferior chamber. The size of the orifice of the membrane was 2 cm. (F) Three-dimensional TEE of the thrombus. IAS, interatrial septum; MV, mitral valve; RA, right atrium.
A 55-year-old man was admitted to the emergency room with right flank pain. Electrocardiogram showed atrial fibrillation (AF). Abdominal computed tomography (CT) indicated a segmental renal infarction in the right posterior portion (arrowheads; Figure 1A). Transesophageal echocardiogram demonstrated moderate grade rheumatic mitral stenosis. Mean diastolic pressure gradient was estimated to be 7.6mmHg and the left atrium (LA) was enlarged (anteroposterior diameter, 54 mm). Three-chamber view showed a linear structure crossing the LA (arrowheads; Figure 1B). Transesophageal echocardiogram was performed for better evaluation of the thromboembolic source, and showed that the LA was divided into 2 distinct chambers by a thin membrane (arrowheads; Figure 1C). The membrane was not elongated enough to cause any functional mitral stenosis, but a thrombus (arrow) was observed in an accessory chamber between the interatrial septum and membrane (Figure 1D). A thrombus was also observed on the membrane of the septal side of the anteroinferior chamber (Figures 1E,F). There was no visible thrombus in the LA appendage, despite the presence of spontaneous echo contrast. Cardiac CT, which was performed after anticoagulation for 2 months, showed (Figure 2A) a membrane (arrowheads) with an opening between the 2 LA chambers (arrowheads; Figure 2B).

Cor triatriatum sinister is a rare form of congenital heart disease, first described by Church in 1868.1 In cor triatriatum sinister, the LA is divided into 2 distinct chambers by a fibromuscular membrane: the posterosuperior chamber receiving blood from the pulmonary veins; and the anteroinferior chamber, which contains the appendage and behaves as the true LA, communicating with the mitral orifice.2 The opening area of the membrane does not change with age, and late presentation may be due to the development of complications such as mitral regurgitation and AF.3 In addition, because the hemodynamics of cor triatriatum can be similar to those of mitral stenosis,4 embolic events related to a state of blood stagnation have rarely been reported.4,5 Thrombi commonly develop in the LA appendage in patients with rheumatic mitral stenosis.6,7 In cor triatriatum, however, the geometric nature of the membrane leads to an unusual stagnation of the flow, and it can serve as a nidus for thrombus formation in an unexpected location, other than the LA appendage. Surgical removal of the membrane can be considered when the cor triatriatum causes functional mitral stenosis.8 Nevertheless, in cases in which the thrombus remains even after sufficient anticoagulation because of a structural problem, surgery may also be needed to avoid any recurrent embolic events despite the absence of functional mitral stenosis.

Disclosures

None.

References


Figure 2. Cardiac computed tomography, performed after anticoagulation for 2 months, showing (A) residual thrombus (arrow) on the opposite side of the left atrium (LA) appendage, and (B) a membrane (arrowheads) with an opening between the 2 LA chambers.