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Images in Rheumatology

Multiple embolic strokes in primary antiphospholipid syndrome

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A 58-year-old female was brought to our institution with an ischemic stroke. A holosystolic murmur in the fifth intercostal space in the mid-clavicular line was heard on cardiac auscultation. Cranial magnetic resonance imaging (MRI) revealed a sub-acute stroke in the posterior cerebral artery and multiple images related to previous embolic events (Figure 1). Treatment with atorvastatin and rivaroxaban was initiated for ischemic stroke. A transthoracic echocardiogram and a transesophageal echocardiogram (TEE) showed nodular thickening in both mitral leaflets and the sub-valvular apparatus with moderate mitral regurgitation. Moreover, small nodules were observed on the vascular side of the aortic valve. Mild tricuspid regurgitation was observed, and the pulmonary artery systolic pressure was normal (32 mm Hg). The patient had normal biventricular systolic function with a left ventricular ejection fraction of 65% and tricuspid annular plane systolic excursion of 19 mm (Figures 2 and 3). The laboratory examination revealed anemia (hemoglobin: 9.9 g/dL, hematocrit: 29.1%); no leukocytosis; and concentration of D-dimer: 10,000 ng/mL, protein C: 150 mg/dL, protein S: 80%, C3: 145.8 mg/dL, C4: 29.7 mg/dL, anti-cardiolipin IgA: 20 A phospholipids (APL), anti-cardiolipin IgM: 15 M phospholipids (MPL), anti-cardiolipin IgG: 19 G phospholipids (GPL), anti-B2GP1: positive, anti-dsDNA: 10 IU/mL, anti-nuclear positive and increased IgA: 505.5 mg/dL, and a heterozygous mutation in *MTHFT* (WT/C677T) was observed. At 12 weeks, increased levels of anti-cardiolipin IgG (50 GPL) and anti-cardiolipin IgM (29 MPL) were observed. A diagnosis of antiphospholipid syndrome (APS) and mitral valvulopathy was established. The New York Heart Association classification placed the patient in functional class II; she is currently receiving medical treatment (warfarin, atorvastatin, and aspirin).

The patient signed the informed consent that she agrees to participate in this study.

It is observed that 70% of patients with APS have at least 1 valvular lesion diagnosed through echocardiography (valvular thickening, stenosis, regurgitation, or non-infective vegetation). However, the most important manifestation is Libman-Sacks endocarditis (LSE), primarily in the mitral and aortic valves (1). TEE detects cardiac involvement in 75.9% to 82% of patients (2). The diagnosis requires the presence of non-infective vegetation on the echocardiogram and the exclusion of infective endocarditis (3). The prevalence of stroke in a patient with APS is 19.8% (2). Patients with APS and stroke should be evaluated using

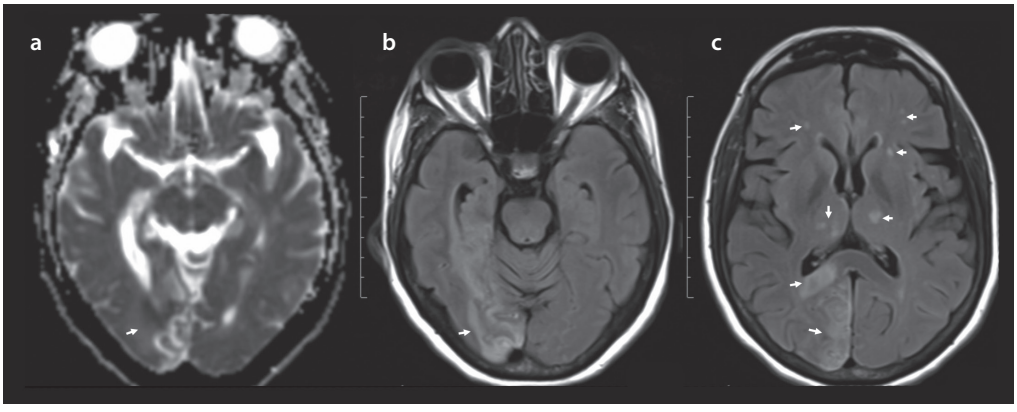

Figure 1. a-c. Cranial MRI. Hyperintense lesions in the right posterior cerebral artery territory that compromises the ipsilateral temporal region. T2-weighted MRI (a). FLAIR (b). Multiple axial hyperintense lesions in FLAIR within the cerebellum, and right occipital-cortical and frontoparietal midline thalamic regions (white arrows) (c).

FLAIR: fluid-attenuated inversion recovery; MRI: magnetic resonance imaging.

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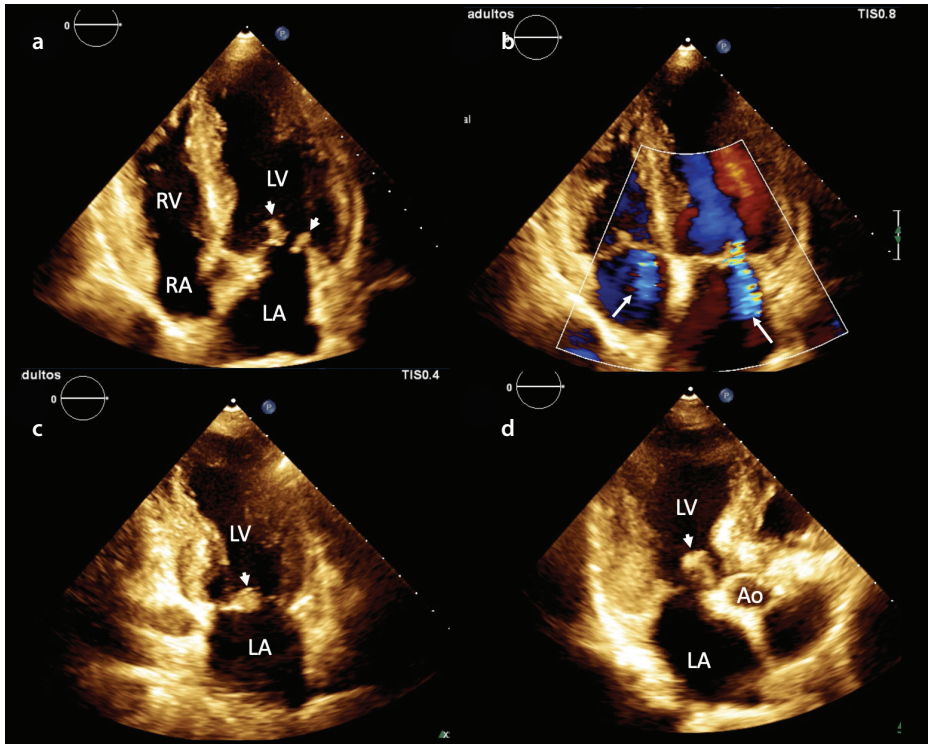

Figure 2. a-d. Bidimensional and color-flow transthoracic echocardiogram in apical views. Four-chamber view showing nodular thickening on the tip of both mitral leaflets (arrows) (a). Mild to moderate mitral regurgitation and mild tricuspid regurgitation (arrows) (b). Two- (c) and three-chamber (d) views showing nodular thickening of the mitral leaflets (arrows).
Ao: aortic valve; LA: left atrium; LV: left ventricle; RA: right atrium; RV: right ventricle.

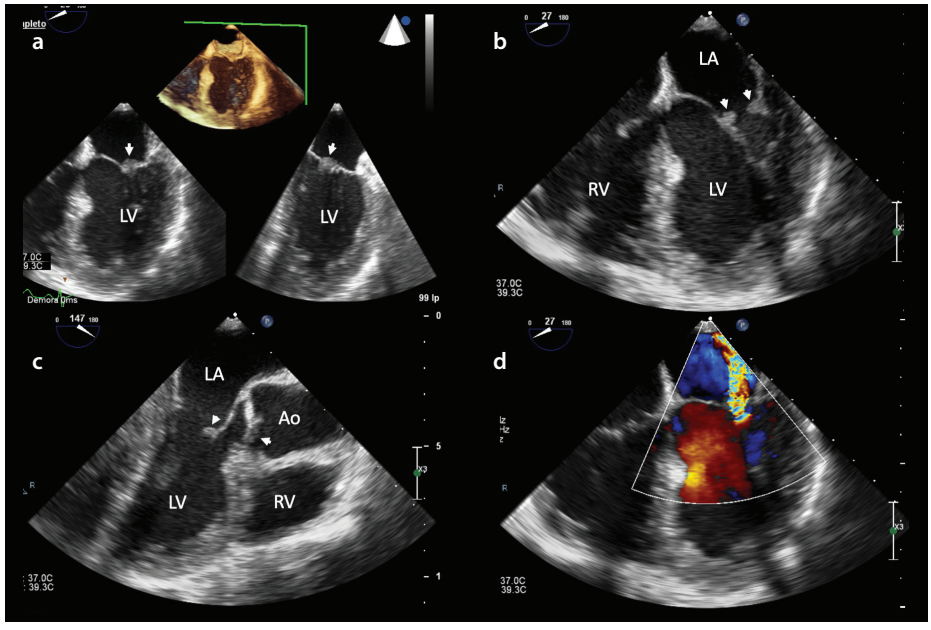

Figure 3. a-d. Two- and three-dimensional transesophageal echocardiograms in 12º (a) and 27º (b) showing nodules in the auricular side of mitral leaflets (arrows). The image at 147º (c) with nodules in mitral leaflets (arrows) and on the vascular side of the aortic valve (arrow). Doppler color with moderate mitral regurgitation (d).
Ao: aortic valve; LA: left atrium; LV: left ventricle; RA: right atrium; RV: right ventricle.

MRI, and intra- and extracranial vascular imaging (angiotomography). It is recommended to perform TEE in all patients with APS and stroke (4). Neuroimaging abnormalities have been reported in 35% to 90% of patients with APS. The most frequent findings were heart attacks, reported in 22% to 45.7% of patients, and white matter hyperintensities, found in 17% to 45% of patients with APS. Ischemic lesions can be small or large and involve superficial and deeper areas of the brain (2, 5).

This case is of great interest because in the sixth decade of life presented a cardioembolic stroke as the first manifestation of APS. The echocardiogram demonstrated non-infective vegetation that suggested the diagnosis of APS, which was confirmed by her immune profile.

Informed Consent: Written informed consent was obtained from the patient.

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