The patient is a 54-year-old woman with no significant medical history who was referred for cardiovascular magnetic resonance imaging due to physical and electrocardiography findings. Sequences were performed synchronized to the patient’s heartbeats (cardiac gating) in black blood (double IR), white blood (vascular multislice), and cine sequences following the cardiac axes. On all slices a heart integrally located in the left hemithorax was observed (Figures 1 and 2) due to left pulmonary hypoplasia. The latter involved mediastinal displacement toward the left, with herniation of the right lung into the retrospinal space. On magnetic resonance angiography extreme hypoplasia of the left pulmonary artery was observed (Figure 3), and developed left pulmonary veins were not visualized. The rest of the vessels were normal in size, structure, and position. The size, volume, mass, and function of both ventricles were also normal.

This, then, is a case of extreme levocardiia determined by hypoplasia of the left lung, where the visceroatrial, atrioventricular, and ventriculoatrial connections, as well as the ventricular opening, are all normal. This should not be confused with the amalgam of syndromes of the heart and badly positioned tip that bring with them complex cardiac and extracardiac anomalies due to defects in the rotation and alignment of the cardiac tube during embryogenesis. The syndromes of asplenia, polysplenia, isolated levocardiia (correct position of the heart but abnormal position of the viscera), transposition of the large vessels, etc., are examples of the latter.

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