

Figure 1

Diagnosis of ALCAPA Syndrome in Adults

A 35-year-old woman with no cardiovascular risk factors consulted for dyspnea on heavy exertion over the preceding 6 months. The physical examination was normal. The electrocardiogram showed no abnormalities and echocardiographic study was normal. Stress scintigraphy using single-photon emission computed tomography (SPECT) disclosed a reversible perfusion defect in the anterior wall; hence, outpatient coronary angiography was indicated. The right coronary artery (RCA) (Figure, A) was seen to be dilated, ectatic, and tortuous, with a well-developed collateral network toward the left coronary artery (LCA) that drained into the pulmonary artery trunk. An imaging study with 64-slice multidetector computed tomography (MDCT) (Phillips Medical System) was performed, with a baseline rate of 52 bpm following 10 days of treatment with ivabradine 5 mg. The images showed an anomalous origin of the LCA (Figure, B and C) from the common trunk of the pulmonary artery (PA), receiving collaterals from a dilated RCA (Figure, B and D).

Anomalous left coronary artery arising from the pulmonary artery (ALCAPA syndrome) is uncommon, although the true incidence may be underestimated because of the difficulties in diagnosing the condition in the absence of symptoms in adults. The diagnosis has been based on coronary angiography findings, but now MDCT can clearly establish the diagnosis and provide additional anatomic information before surgery. The patient refused any type of invasive treatment and remains stable at the time of writing, 8 months later.

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