A baby in the heart: ALCAPA with aortic arch fistula

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An 83-year-old female was transferred to our emergency department on Christmas Eve evening after a documented asystole and cardiopulmonary resuscitation. She had a history of chronic renal dysfunction stage III, mitral and tricuspid regurgitation grade II, heart failure, and paroxysmal atrial fibrillation. Chest CT revealed anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) with a retropulmonary ostium (Panel 1A and B; white arrow) and massive hypertrophy and calcification of the left main, left anterior descending artery, and circumflex artery (CX) (up to 2 cm in diameter—Panel 1A and B) and right coronary artery (Panel 1D). The CX was communicating with one aortic arch-feeding vessel (Panel 1B; white and black arrow) and forming a huge vessel convolute in the mid-mediastinum. She died on Day 7 in the hospital due to cerebral hypoxia and multi-organ failure. Ao, aorta; P, pulmonary trunk.

ALCAPA (or Bland–White–Garland syndrome) is a rare congenital condition (1/300 000 births) which usually manifests during the neonatal period as intractable heart failure and ischaemia. Late clinical manifestation is very rare. Early diagnosis with surgical treatment helps prevent an adverse outcome. Uniquely, our patient developed an extensive fistula of CX to the aortic arch due to chronic ischaemia of the left ventricle and chronic steal phenomenon over her lifetime, and surprisingly survived until the age of 83.

Such a case calls for CT-imaging in adult patients with heart failure of unclear aetiology in order to rule out rare grown-up congenital heart disease.

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