Non-compaction cardiomyopathy with diffuse left coronary artery fistulae as a rare cause of congestive heart failure

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A 46-year-old woman was admitted to hospital due to dyspnoea. Chest X-ray, serological parameters, and spirometry ruled out pulmonary reasons or infection as its cause.

Transthoracic echocardiography revealed a moderately impaired left ventricular (LV) contractility (ejection fraction 42% calculated according to Simpson’s rule); there were no relevant valvular pathologies. In the apex of the LV, prominent trabeculae were detected (Panel A); colour Doppler showed perfusion of the intertrabecular spaces from the LV cavity. This raised the suspicion of non-compaction cardiomyopathy (NCCM). Coronary angiography was performed, which ruled out coronary artery disease. Surprisingly, after injection of contrast agent (CA) into the left coronary artery (LCA), CA was rapidly detected within the LV, indicating diffuse fistulae from the LCA to the LV (Panel C: angiogram of the LCA, Panel D: beginning inflow of CA from the LCA into the LV, Panel E: opacification of the LV by the CA, Panel F: magnification of the marked area showing CA passed over from the LCA to the LV). For further diagnosis, magnetic resonance tomography was performed. This excluded a myocarditis, but confirmed non-compaction in the region, where the coronary fistulae were detected (Panel E).

Non-compaction cardiomyopathy is a rare cause of congestive heart failure, with an incidence of 0.05–0.25% in the whole population. During embryonic development, disturbed compression of the trabeculated myocardium leads to non-compaction with impaired contractility. Theoretically, this process may lead to coronary fistulae, too. Nevertheless, the co-incidence of non-compaction and coronary fistulae has been described only a few times before.