Severe aortic regurgitation in a patient with left ventricular non-compaction cardiomyopathy

Introduction: Left ventricular non-compaction (LVNC) is a rare congenital cardiomyopathy characterized by excessive trabeculation and deep intertrabecular recesses most commonly affecting apical and mid-ventricular inferior and lateral segments. These patients are more prone to develop a typical triad of heart failure, thromboembolic events, and malignant arrhythmias. LVNC often coexists with other congenital or valvular heart diseases. Echocardiography is the standard imaging for the diagnosis of LVNC with several proposed criteria, mainly based on the ratio of non-compacted to compacted myocardial thickness1,2.

Case report: 75-year-old woman with a prior history of arterial hypertension and atrial fibrillation was hospitalized due to acute heart failure. Physical examination revealed holodiastolic precordial murmur, bilateral pulmonary crackles, and peripheral edema. Echocardiography showed severe regurgitation of tricuspid aortic valve based on the prolapse of the non-coronary cusp and volume overload – induced eccentric left ventricular (LV) hypertrophy with reduced ejection fraction (EF) of 35%. Moreover, a hypertrabeculation with intertrabecular recesses were observed in the inferolateral and apical region of LV fulfilling the echocardiographic diagnostic criteria for LVNC (Figure 1). Coronary angiography showed no signs of coronary artery disease. The patient was started on standard heart failure therapy and was referred to cardiac surgery for aortic valve replacement (AVR). Postoperative echocardiographic assessment one year after AVR showed no improvement in LV systolic function despite normally functional bioprosthetic aortic valve and standard of care heart failure therapy.

Conclusion: Symptomatic LVNC patients with LV systolic dysfunction generally have poorer prognosis1. Since the potential of recovery of myocardial function is questionable in patients with preexisting myocardial disease2, the postoperative outcome might not be satisfactory.

LITERATURE