Uncommon insertion of papillary muscles and abnormal cardiac rotation

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A 43-year-old man was admitted with acute episode of chest pain unrelated to exertion. His electrocardiogram showed increased complex voltages with extensive repolarization changes (A). A transthoracic echocardiogram revealed a dilated left ventricle (LV) with normal wall thickness at the base, good overall systolic function, and an undefined mass vs. thickening at the apex (B). Left ventricular opacification by contrast echocardiography enabled to exclude apical LV hypertrophy and fibrotic obliteration of the apex (C), and confirmed the diagnosis of anomalous apical insertion of papillary muscles (PM). Both PM were displaced distally, exhibited significant calcifications at their peaks and were asymmetrically hypertrophied, with the anterolateral PM being larger. The mitral valve leaflets were not involved in this anomalous mitral apparatus. Additionally, images were analysed by speckle-tracking technique (2D strain EchoPac, version 7.0.1, GE Healthcare, Norway) which showed abnormal cardiac twist, with basal-like rotation (clockwise as viewed from the apex) throughout the LV (D). This pattern utterly differed from the normal basal clockwise, mid neutral, and apical counter-clockwise rotation of the heart. The patient also underwent a coronary angiogram which was normal, and a 24 h Holter monitor showed no evidence of ventricular arrhythmias. After 2 years follow-up, the patient remains clinically well.

Anomalous apical insertion of the PM is a rare heart condition of congenital origin and unknown outcome. The unidirectional cardiac rotational pattern has been only reported in non-compaction cardiomyopathy and is thought to be related to altered myofibre architecture. Both electrocardiographic features and abnormal twist cardiac motion in the present case are highly suggestive of underlying myocardial disease. Nonetheless, the association between abnormally arranged PM at the LV apex and cardiomyopathy is unknown. To our knowledge, this is the first case of anomalous apical insertion of PM reported in the literature.