


CLINICAL VIGNETTE

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Helical form of hypertrophic cardiomyopathy: a new entity?

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A 73-year-old woman was admitted to the hospital because of recent onset of progressive dyspnea, fatigue, and malaise. On physical examination, an ejection systolic murmur (grade 2-3/6) split from S1 was audible at the base of the heart and S4 tone was also present. Lungs were clear and no increased venous jugular pressure was evident. She had well-controlled blood pressure under medications, no history of diabetes, coronary heart disease and of familial hypertrophic cardiomyopathy (HCM).

On 12-lead ECG, only leftward electrical cardiac axis deviation and minor abnormalities of the repolarization phase were present. On transthoracic echocardiography a significant asymmetric hypertrophy of basal anterior septum was described with systolic anterior motion of anterior mitral leaflet causing a peak dynamic gradient of 35 mmHg at rest. The patient was referred for cardiac magnetic resonance imaging (MRI).

On short-axis cine MRI using steady state free precession sequence, a peculiar pattern of left ventricle (LV) hypertrophy was seen. Starting from the anterior basal septum (Panel A), the hypertrophy progressively extents helically along the longitudinal axis involving the mid-basal inferior septum (Panels B and C), the mid-inferior wall (Panel D), and the inferior apical and lateral wall (Panels E and F). The maximal wall-thickness of 16.3 mm was measured at anterior basal septum. The non-involved LV regions had relative thin myocardial wall.

To the best of our knowledge, this is the first reported case of HCM showing a helical pattern of myocardial wall hypertrophy. In normal heart, clockwise and counter-clockwise spirally orientated cardiac fibres have been extensively described. Moreover, some authors have recently advocated the assembly of the myocyte fibres into a well-defined helical myocardial band, which twists and curls in two helical loops. Intriguingly in this case, the distribution of wall hypertrophy resembles the counter-clockwise (seen from LV base) helical loop of the helical myocardial band.

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