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## Another case of typical hypertrophic cardiomyopathy?

### Case report

A 51-year-old male patient developed rapidly progressive shortness of breath on exertion and prelung oedema. His past history was notable for right kidney agenesis, but negative for arterial hypertension. Also, the family history was negative for sudden cardiac deaths. The ECG on admission is shown in figure 1.

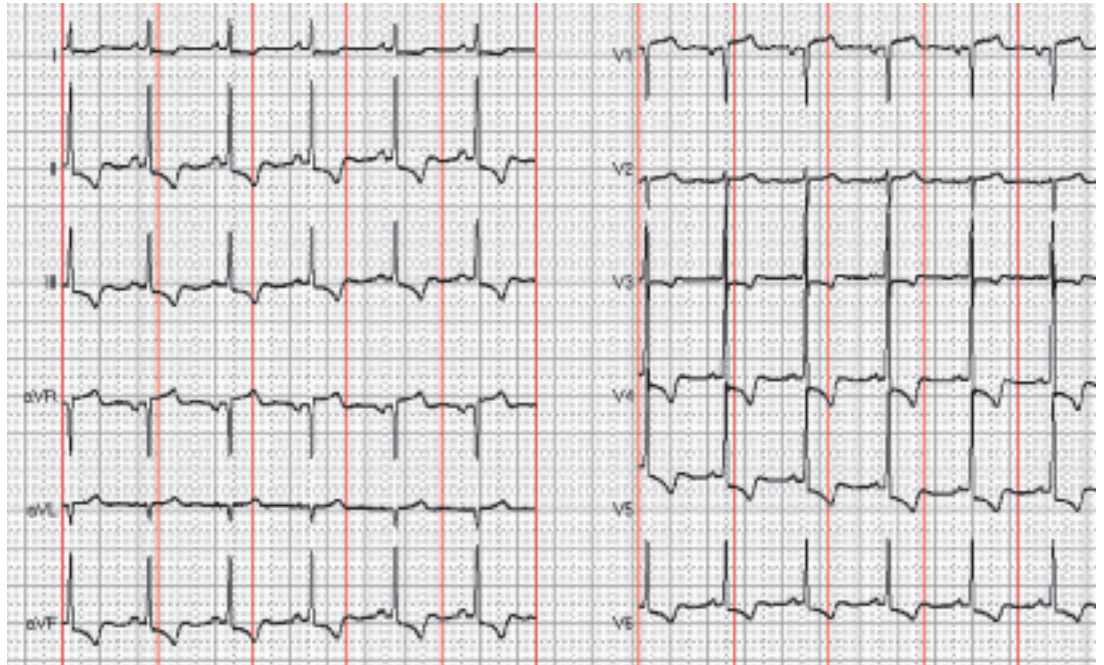
(1.) What is your diagnosis: hypertrophic cardiomyopathy, hypertensive cardiomyopathy or something else?

(2.) Which examination will help to establish the diagnosis?

### Explanatory answers

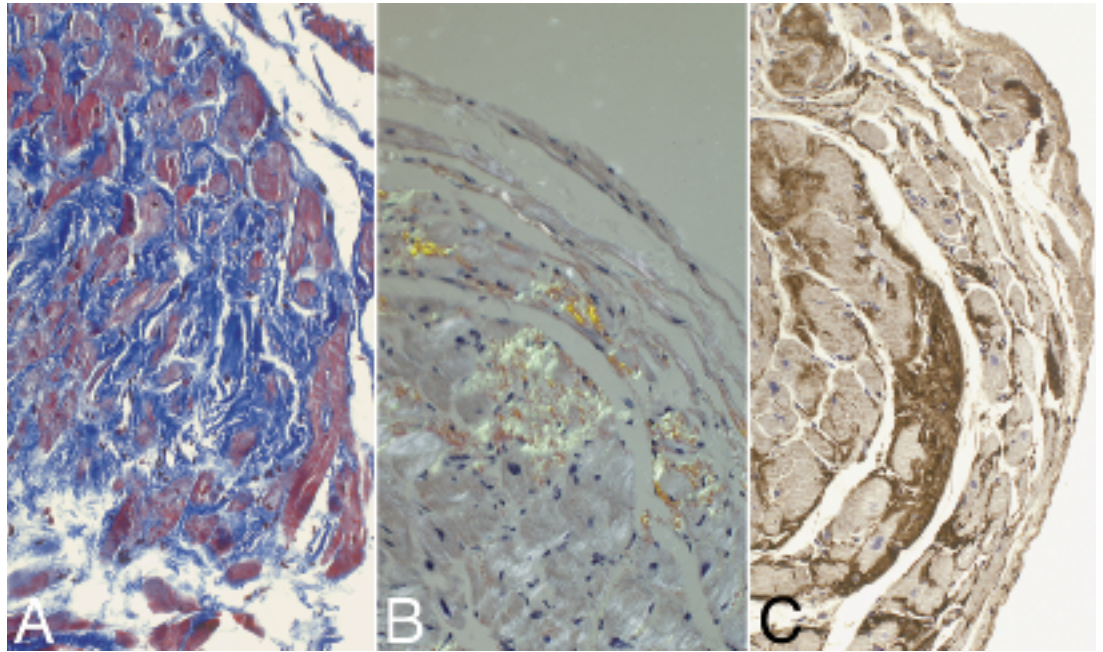
(1.) The 12-lead-ECG is highly suggestive for left ventricular hypertrophy (LVH) with a typical strain pattern as well as signs of left atrial hypertrophy. However, there is no left axis deviation as one would expect in a case of severe LVH. PQ and QTc intervals are within normal limits (fig. 1). Transthoracic echocardiography revealed severe left ventricular thickening of the interventricular septum (19 mm) with involvement of the apex and the posterior wall (12 mm) without LVOT obstruction. Systolic left ventricular function was preserved, but diastolic dysfunction was present. There was a tendency towards hypertrophy of

**Figure 1**  
12-lead-ECG at rest.  
See text for details.



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**Figure 2**  
Specimen of right ventricular endomyocardial biopsy.  
See text for details.



the right ventricular free wall as well, however, pulmonary hypertension could be ruled out.

(2.) Since clinical and echo findings could not rule out any infiltrative disease, catheter-guided cardiac biopsy was performed, which revealed severe amyloidosis of the heart (fig. 2). Histology (fig. 2A) showed interstitial deposition of amyloid with additional replacement of muscle fibers by amyloid (CAB staining). In panel 2B, the amyloid depositions were visualised in the polarised light (Kongo red staining), and in fig. 2C, by immuno-histochemic staining with antibodies against light chain lambda proteins.

### ECG findings

In familial hypertrophic cardiomyopathy, apart from voltage criteria for left ventricular hypertrophy similar to those in hypertensive cardiomyopathy, additional more specific signs may be found depending on the localisation of hypertrophy. In asymmetric septal hypertrophy, typically Q-waves in leads I, II, aVF, aVL,

V<sub>4</sub>-V<sub>6</sub> may be seen, whereas in the apical form (type Yamaguchi), giant negative T-waves predominantly in the inferolateral leads may be hallmarks of the disease. The typical ECG findings of cardiac amyloidosis are usually low-voltage amplitudes in the peripheral leads, and depending on the degree of His-Purkinje involvement, also atrio-ventricular block, which both were not present in our patient. Rarely, anteroseptal Q-waves similar to those observed in familial hypertrophic obstructive cardiomyopathy or in patients with status post anterior wall myocardial infarction may be observed [1, 2]. The ECG pattern of our case, however, is not described as typical for amyloidosis.

### References

- 1 Braunwald's Heart Disease: A Textbook of Cardiovascular Medicine. Zipes DP, Libby P, Bonow RO, Braunwald E (eds). Seventh edition, ISBN 0-7216-0479-X. Philadelphia, Pa: Elsevier Saunders; 2005. pp 1683-5.
- 2 The ECG: A Two Step Approach to Diagnosis. Marc Gertsch (ed.). Heidelberg: Springer Verlag; 2004. pp. 268, 278.