Cor triventriculare with pericardial aplasia and mitral valve prolapse: a novel congenital dysplastic syndrome?

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A 37-year-old man was admitted to our hospital because of an apical abnormality, which was incidentally detected for the first time 10 years ago by echocardiography. The patient reported on normal physical activity and denied clinical signs of heart failure. His ECG showed anteroseptal ST-segment alterations.

For further assessment, magnetic resonance imaging was performed on a 1.5 T whole body scanner (Intera CV, Philips Medical Systems, Best, The Netherlands).

The four-chamber view displays an additional apico-lateral cavum, which impresses as a rudimentary third ventricle with abnormal trabecular and papillary structures (Panel A). The left ventricle is enlarged (end-diastolic volume 230 mL) and left ventricular function reduced (ejection fraction 47%) with the additional cavum appearing to be akinetic (see Supplementary material online, Movies S1 and S2). The mitral valve shows a prolapse of the posterior leaflet and an insufficiency jet towards the atrial wall.

In the different weighted turbo-spin-echo-sequences signal intensity was the same both in the apico-lateral cavum and in the normal myocardium. In addition, we found a local aplasia of the pericardium next to the additional cavum (Panel C). Scar images after admission of Gd-DTPA showed no pathological contrast enhancement in all segments of the heart (Panel D).

To our knowledge, this is the first published case of a third ventricle impressing as an apico-lateral aneurysm in a young asymptomatic person. The combination of this ‘cor triventriculare sinistrum’ with a posterior mitral valve prolapse, and the local aplasia of the pericardium has to be defined as a formerly unknown disease entity. These characteristic findings suggest a prenatal dysplastic process of the apico-lateral structures of the left ventricle including its neighbouring pericardium.

Panel A. The four-chamber view displays abnormal trabecular and papillary muscles (arrow) mimicking an additional septum and valvular structures.
Panel B. The mitral valve shows a moderate regurgitation jet (arrow, regurgitation fraction 18%) due to a slight prolapse of the posterior leaflet.
Panel C. Turbo-spin-echo in the four-chamber geometry reveals an aplasia of the pericardium (arrow) over the lateral left ventricular wall.
Panel D. Late enhancement analysis using gradient echo sequence with a 180° inversion recovery prepulse showed no pathological contrast enhancement.

Supplementary material is available at European Heart Journal online.