A 55-year-old woman was referred to our department for progressive dyspnoea. No personal nor familial history except a smoking habit was reported. Electrocardiogram showed a third-degree atrio-ventricular (A-V) block with ventricular rate at 35 b.p.m. (Panel A). Transthoracic echocardiography revealed a dilated myocardialopathy with ejection fraction estimated at 35%. Curiously, an abnormally thin aspect of the basal portion of the interventricular septum (IVS) was noted. Maximum thickness was measured at 4 mm (Panel B). There was no septal defect. Cardiac magnetic resonance found a pathologically thin septum with transmural delayed enhancement in that area (arrows) in favour of a localized septobasal fibrosis (Panel C).

Confronted to a complete heart block with poor left ventricular function, a triple-chamber pacemaker was implanted. Coronary angiography revealed the absence of septal branch arising from the left descending artery before the origin of the first diagonal (Panel D). Blood tests were normal including troponin T and Lyme disease serology.

Three months later, the patient is found symptomless and NYHA class I. Echocardiography confirms the same aspect of the IVS with ejection fraction slightly improved at 40%.

To our knowledge, this observation is the first to describe a third-degree A-V block relevant of a missing septal branch with localized septal thinning. Aetiology remains imprecise, no patent argument was found for an acute coronary syndrome and isolated agensis of a septal branch has never been noted. The origin of the myocardiopathy is also unknown and a prolonged A-V block could be incriminated.