Light chain deposition disease (LCDD) is a rare plasma cell dyscrasia consisting of nonamyloidotic deposition of misfolded immunoglobulin light chain (LC) in tissues and organs. Heart involvement is a rare and late event in the course of the disease. Therefore, such a nonfibrillar deposition of LC in the myocardium has so far been described only in few endomyocardial biopsies. Potentially lethal ventricular arrhythmias and a nondiagnostic echocardiogram are 2 clinical hallmarks of the disease. In addition, no other in vivo imaging of the condition is presently available.

A 52-year-old man was admitted to the emergency department for syncope at rest. ECG showed a sinus rhythm with negative T waves in inferolateral leads alternating with ventricular bigeminism and episodes of torsades de pointes triggered by R-on-T phenomenon (Figure 1A). The previous history included serum IgGκ + κ monoclonal gammopathies and nephrotic syndrome. The bone marrow biopsy showed 60% clonal plasma cells, and the kidney biopsy examination by immunofluorescence and immunoelectron microscopy was consistent with LCDD. Light microscopic finding of kidney biopsy showed glomeruli with increased mesangial cellularity and nodular expansion of mesangial matrix; mesangial nodules were positive at periodic acid Schiff and trichrome stain (Figure 2A and 2B) and poorly argyrophilic; increased mesangial cellularity and patchy endocapillary proliferation were also seen. At electron microscopy, granular, nonfibrillar, moderately electron-dense material was present along the capillary basement membranes, within the mesangial nodules (Figure 2C) and along tubular basement membranes (Figure 2D).

The patient was treated with chemotherapy according to the bortezomib-dexamethasone regimen and, later on, tandem autologous stem cell transplantation was performed. The patient achieved a complete hematologic and renal remission. The disease relapsed because serum IgG/κ monoclonal gammopathy and κ serum-free light chain increased rapidly, and the patient was started on chemotherapy with lenalidomide. Two months later, the syncope with torsades de pointes occurred and the patient was admitted to the Cardiology Unit. On admission, continuous troponin I release (up to 24 μg/L) was detected. Despite a preserved systolic function, the patient underwent coronary angiography showing a myocardial bridge of the left main anterior descending coronary artery. The cardiac magnetic resonance (CMR) showed a left ventricle with regional hypokinesis and preserved ejection fraction, in the absence of wall hypertrophy (online-only Data Supplement Movie 1). Myocardial edema in the left ventricle inferolateral epicardial wall (with a nonischemic pattern) associated with a midwall septal stria was shown (Figure 1C through 1F). On postcontrast sequences, late gadolinium enhancement (LGE) in the same areas of edema with circumferential/epicardial pattern was also detectable (Figure 1G through 1L), with both nonischemic and nonamyloidotic patterns. On the basis of CMR, an endomyocardial biopsy was performed and showed aspecific findings on hematoxylin and eosin, with focal myocardial necrosis and fibrosis and no infiltration by inflammatory cells (Figure 2E); Congo Red (Figure 2F) and thioflavin T staining were negative (Figure 2G). Electron microscopy was performed and revealed granular electron-dense material in the interstitium, lacking the characteristics β-sheet fibrillary of amyloid being constituted at immunoelectron microscopy by κ LC deposition (Figure 2H). During hospitalization, the patient experienced repetitive nonsustained ventricular tachycardia (Figure 1B) and underwent implantable cardioverter defibrillator implantation. The diagnosis was a cardiac involvement by LCDD (κ). Lenalidomide chemotherapy goes on. After 3 months, the implantable cardioverter defibrillator was removed because of infection of the catheter. Before the implantation of a new device, a CMR was performed once again and revealed a significant reduction of myocardial edema but the persistence of the LGE. There are very few reports on cardiac involvement occurring in LCDD likely because of the rarity of the disease. The myocardial edema in the first CMR suggests acute cardiac injury probably attributable to the subacute deposition of LC...
in the interstitium and is likely the cause of the electric instability and arrhythmias. The type of LGE sparing the endocardium corresponds to deposits of LC found at autopsy. In conclusion, to the best of our knowledge, for the first time we have herein described the in vivo pattern of cardiac involvement in LCDD with tissue characterization both by CMR and by histological/immunoelectron microscopy findings. We have also shown that the pattern of CMR-LGE in this case is completely different from the pattern found in cardiac amyloidosis.

Disclosures

None.

References


Figure 1. Ventricular arrhythmias and cardiac magnetic resonance pattern in nonamyloidotic light chain cardiomyopathy. After the syncope, the ECG showed ventricular premature repetitive beats and episodes of torsades de pointes triggered by R-on-T phenomenon (A). During hospitalization, the patient experienced a repetitive nonsustained ventricular tachycardia with right bundle-branch morphology (B). On acute CMR (C through F), myocardial edema on T2-weighted images was detectable in the left ventricular lateral wall with epicardial pattern (C, short-axis view; D, long-axis view), white arrows associated with a midwall septal stria. On postcontrast sequences (E and F), LGE in the same areas of edema with circumferential/epicardial pattern (empty arrows) was also detectable with more pronounced involvement of the septum. At 3-month CMR (G through L), an important reduction of edema (G and H) with persistence of LGE was noted. CMR indicates cardiac magnetic resonance; and LGE, late gadolinium enhancement.
Figure 2. Renal (A through D) and endomyocardial (E through H) biopsies of nonamyloidotic light chain disease. The renal involvement (A through D): The periodic acid Schiff positive staining revealed nodular deposition in the mesangium (A, ×40). Masson Trichrome staining showed endocapillary hypercellularity (B, ×40). The electron microscopy identified a glomerular electron-dense deposit in the mesangium (C, ×7000) and in the subepithelial tubules (D, ×7000). The cardiac involvement: On hematoxylin and eosin, an aspecific myocardial pattern is shown, with focal myocardial necrosis without an inflammatory cell infiltrate and fibrosis (E, ×20). Congo Red staining (polarized light; F, ×20) and Thioflavin T (fluorescence; G, ×20) were negative. Electron microscopy revealed granular electron-dense material (H, ×7000), which lacked the characteristics of amyloid and which has been identified by immunoelectron microscopy as κ-light chain deposition.
Nonamyloidotic Light Chain Cardiomyopathy: The Arrhythmogenic Magnetic Resonance Pattern
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