Ventricular Tachycardia as an Initial Manifestation of Randall-Type Monoclonal Immunoglobulin Deposition Disease: A Case Report and Literature

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Abstract

Monoclonal immunoglobulin deposition disease (MIDD) is a rare complication of B-cell clonal disorders, defined by Congo-red negative deposits of monoclonal light chain (LCDD), heavy chain (HCDD), or both (LHCDD). MIDD is a systemic disorder with prominent renal involvement but little attention has been paid to the description of extra-renal manifestations. Moreover, mechanisms of pathogenic immunoglobulin deposition and factors associated with renal and patient survival are ill-defined. Extra-renal involvement (most commonly cardiac) is seen in up to 10% of cases [1]. The frequency of cardiac manifestations is increasing with the duration of evolution of the disease. They manifest themselves by restrictive hypertrophic cardiomyopathy, close to the amyloid heart disease, with disturbances in rhythm or conduction, associated with electrocardiographic micro voltage, and subsequently heart failure congestive, sometimes revealing the disease [1]. Our case describes an unusual presentation of Randall-type monoclonal immunoglobulin deposition revealed by rhythm disorder (VT).

Keywords: Monoclonal immunoglobulin deposition disease, hypertrophic cardiomyopathy, Randall.

INTRODUCTION

Monoclonal immunoglobulin (Ig) deposition disease (MIDD) is a rare nephropathy (<1% of renal biopsies). The cornerstone of diagnosis is immunofluorescence study of the kidney biopsy, showing linear deposits of the involved monoclonal immunoglobulin predominantly along tubular basement membranes (TBM), and usually in the mesangium, along glomerular basement membranes (GBM) or around arteriolar myocytes.

The frequency of Randall's syndrome is poorly known and probably underestimated. The main series of MIDD patients indicate that the disease is more common in men, mainly around 60 years.

Nevertheless, the age of discovery is very variable (from 30 years old). It is a systemic disease, which involves almost all organs, but the extra-renal locations, are often asymptomatic [1].

The liver, heart and peripheral nervous system are the most common. It's associated with a poor prognosis and an unsatisfactory response to chemotherapy. Our case report is characterized by a first unusual presentation: isolated Ventricular tachycardia.

CASE REPORT

A 57-year-old patient, with no past medical history, presented at the emergency department with palpitation, and chest pain. Initial vital signs were as follows: blood pressure 80/51 mmHg, heart rate 160 bpm, respiration rate 30/min, temperature 36.5°C, O2 saturation 92%. Electrocardiogram quickly realized in the ER has shown a ventricular tachycardia. The patient was defibrillated by an automated external defibrillator, then the heartbeat became sinus and the hemodynamic state has become stable again. On physical examination the patient presents no signs of heart failure, except a hepatomegaly.

- Chest x-ray: reveals a generalized cardiac enlargement
- Electrocardiogram shows ventricular tachycardia 160 b/mn. (Figure 1)
- Transthoracic echocardiography revealed:
Two-dimensional (2D) echocardiographic confirm Hypertrophy involving the interventricular septum in the basal LV segments extends into the posterolateral wall.

- Septal/posterior wall thickness: 19/23 mm
- Septal/posterior wall thickness ratio: 0.87
- Absence of latent obstruction- LVOTO
- Absence of Systolic anterior motion - SAM
- Mitral regurgitation grade I
- Left atrial enlargement VOG: 38 ml/m2
- Ejection fraction is preserved LVEF: 57%
- Diastolic dysfunction grade II:
  - e/a: 0.8  e'/e: 14  e' septal: 3 cm/s  e' lateral: 4 cm/s
- Left atrial volume: 38 ml
- Blood test: renal failure eGFR 35 ml/mn and moderate elevation of alkaline phosphatase
- Electrophoresis of serum proteins: Hypoalbuminemia with hyperalpha1-globulinemia (figure 2)
- Proteinuria of BenceJones: presence of kappa light chains without free component (figure 3)
- Myelogram: rich marrow showing many megakaryocytes absence of significant cytological abnormalities (figure 4)
- Renal biopsy: On histological examination, it is a renal parenchyma with ten glomeruli. The latter are most often in sealed bread and bordered by fibrosis of varying thickness. The tubes are cystized in places and lined by a simple cubo-cylindrical epithelium. The interstitial tissue is fibrous, seat of an inflammatory infiltrate made essentially of lymphocytes and plasma cells. The capillaries are regular thickened wall in places. PAS staining revealed continuous linear eosinophilic deposition at the basement membranes of the tubes and perivascularly. Congo red staining showed no amyloid deposits. Concluding to a morphological aspect of a nephropathy consistent with Randall syndrome.
- Thyroid Echo: nodules + cysts classified Eu tirads 2 (figure 5)
- Abdominal Echo: Homogeneous hepatomegaly, kidneys of normal size and shape
DISCUSSION

Monoclonal immunoglobulin deposition disease Randall-type (MIDD) is a rare complication of plasma cell proliferations [2]. It was known from the late 1950s that nonamyloidotic forms of glomerular disease that resemble the lesion of diabetic glomerulosclerosis could occur in multiple myeloma. The presence of monoclonal light chains (LC) in these lesions first was recognized in 1973 by Antonovych et al. and confirmed by Randall et al. who published in 1976 the first description of light-chain deposition disease [2].

Clinical Manifestations MIDD is a systemic disease with Ig-chain deposition in a variety of organs, leading to various clinical manifestations, but visceral Ig-chain deposits may be totally asymptomatic and found only at autopsy. Renal involvement is a constant feature of MIDD, and renal symptoms, mostly proteinuria and renal failure, often dominate the clinical presentation.

In 18 to 53% of patients with LCDD, albuminuria is associated with the nephrotic syndrome. However, in approximately one quarter, it is 1 g/d, and these patients exhibit mainly a tubulointerstitial syndrome [4]. Which is the case of our case report?

Liver and cardiac involvement occurs in approximately one quarter of patients with LCDD. Liver deposits are constant. Hepatomegaly with mild alterations of liver function tests are the most usual symptoms, but patients also may develop life-threatening hepatic insufficiency and portal hypertension [4].

Myeloma is diagnosed in approximately 50% of patients with LCDD or LHCD and in approximately 25% of those with HCDD. MIDD, like AL-amyloidosis, often is the presenting disease that leads to the discovery of myeloma at an early stage [4].

Cardiac involvement may be responsible for cardiomegaly and severe heart failure. Arrhythmias, conduction disturbances, and congestive heart failure are seen. Echocardiography and catheterization may reveal diastolic dysfunction and reduction in myocardial compliance similar to that found in cardiac amyloid [4].

To compare cardiac dysfunction In LCDD and light chain amyloid (AL) disease, Buxbaum et al. Hypothesized that, despite differences in physical properties, nonamyloidotic light chain deposition in the myocardium could produce similar clinical and physiological abnormalities. Cardiac tissue from five patients with LCDD and cardiac dysfunction was examined by immunohistochemical and electron microscopic techniques. Hospital charts, electrocardiograms, echocardiograms and cardiac catheterization results were reviewed. As a result: In four with adequate clinical documentation, the diastolic dysfunction and conduction abnormalities were similar or identical to that described in cardiac AL disease [3].

The diagnosis of MIDD must be suspected in any patient with the nephrotic syndrome or rapidly progressive tubulointerstitial nephritis or with echocardiographic findings indicating diastolic dysfunction and the presence of a monoclonal Ig component in the serum and/or the urine. The same combination also is seen in AL-amyloidosis, but this more often is associated with the LC isotype. Because sensitive techniques including immunofixation fail to identify a monoclonal Ig component in 10 to 20% of patients with LCDD/LHCD and approximately 40% of patients with HCDD, renal biopsy plays an essential role in the diagnosis of MIDD and of the associated dysproteinemia. The definitive diagnosis is made by the immunohistologic analysis of tissue from an affected
organ, in most cases the kidney, using a panel of Ig chain-specific antibodies, including anti-K and anti-λ LC antibodies to stain the non-Congophilic deposits [4].

The treatment of MIDDs is based on the removal of secretion of monoclonal Ig nephrotoxic, and therefore on chemotherapy adapted to the nature of the underlying clone. Some data from the literature argue in favor effectiveness of a combination of bortezomib-cyclophosphamide-dexamethasone in MGM, and in particular AL amyloidosis with serious cardiac involvement but Cohen et al. did not evidence of superiority of TBB-based triple therapies compared to schema bortezomib-dexamethasone on a retrospective study out of 49 patients [2].

The outcome of MIDD remains uncertain, mainly because extrarenal deposits of LC can be totally asymptomatic or cause severe organ damage that leads to death. Survival from onset of symptoms varies from 1 mo to 10 yr. In the largest series (34), 57% of the patients reached uremia and 59% died during follow-up (mean 27.5 mo), and patient survival was only 66% at 1 yr and 31% at 8 yr, although 86% of the patients received chemotherapy. The only variables that were independently associated with renal survival were age and degree of renal insufficiency at presentation or the time of renal biopsy. Variables that were independently associated with a worse patient survival were age, initial creatinine, associated multiple myeloma, and extrarenal LC deposition. Survival of the uremic patients who were treated with dialysis was not different from that of the patients who did not reach uremia [4].

CONCLUSION

MIDD is a rare systemic disease that is characterized by severe renal failure as a result of the deposition of a monotypic LC and/or HC of Ig. Glomerular lesions are so similar to diabetic nephropathy that MIDD may serve as a model for the understanding of this common disease.

However the involvement of other organs including the heart can precede the renal manifestations, as the case of our patient. Knowing this pathology can significantly improve the prognosis by early diagnosis.

REFERENCES