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Light Chain Deposition Disease Associated With Multiple Myeloma Developing in Late Pregnancy

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Preeclampsia is the most common cause of proteinuria with hypertension during pregnancy. Primary kidney disease and kidney disease secondary to systemic disorders may rarely occur during pregnancy, resulting in proteinuria. A 34-year-old woman was admitted to our hospital with abdominal distention and lower extremity edema. The pregnancy was terminated at the 24th week of gestation due to preterm labor. Even after the delivery, proteinuria and renal deterioration continued to progress. The M-peak was not found on serum and urine protein electrophoresis. The serum free light chains assay showed absolute elevation of lambda chains at 1013.9 mg/L with a decreased kappa to lambda ratio of 0.05. Kidney biopsy revealed light chain deposition disease with lambda light chain deposits on immunofluorescence. Bone marrow examination was compatible with multiple myeloma. To our knowledge, this is the first reported case of light chain deposition disease associated with multiple myeloma during pregnancy.

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Case Report

INTRODUCTION

Light chain deposition disease (LCDD) is a rare disease characterized by monotypic immunoglobulin depositions in various organs including the liver, the heart, and especially the kidney.[1] Renal involvement in LCDD presents with proteinuria, hypertension, and kidney function impairment.[2-4] Greater than 50% of LCDD cases are associated with multiple myeloma (MM).[5] However, the rate of LCDD might be underestimated because kidney biopsy is not generally performed in cases of MM.[6] Forty-two cases of MM in pregnancy have been reported in the literature; however, LCDD diagnosed by kidney biopsy during pregnancy has not been described.[7] Here, we describe a case of a 34-year-old woman with LCDD-associated MM developed in late pregnancy.

CASE REPORT

A 34-year-old woman was admitted to our hospital with abdominal distention and lower extremity pitting edema. No other abnormalities had been detected during her first and second pregnancy. At the 20th week of gestation, routine check-up revealed new-onset proteinuria (2+) on a urine dipstick test and total urinary protein of 7.9 g/24 h. At the 24th week of gestation, she delivered a still-born baby due to preterm labor.

Even after the delivery, proteinuria (random spot urine protein-creatinine ratio, 12000 mg/g), and renal deterioration (serum creatinine, 1.18 mg/dL) continued to progress. She had no history of general weakness, weight loss, and bone pain. On admission, her blood pressure was 100/60 mm Hg and the laboratory measurements were as follows: leukocyte count, 11.88 × 10^9/L; erythrocyte count, 3.93 × 10^{12}/L; hemoglobin, 12.1 g/dL; platelet count, 65.8 × 10^9/L; blood urea nitrogen, 33.9 mg/dL; serum creatinine, 1.18 mg/dL; aspartate aminotransferase, 72 IU/L; alanine

aminotransferase, 29 IU/L; serum total protein, 4.3 mg/dL; serum albumin, 2.2 mg/dL; serum calcium, 8.4 mg/dL; serum phosphate, 3.8 mg/dL; total cholesterol, 413 mg/dL; triglyceride, 185 mg/dL; erythrocyte sedimentation rate, 66 mm/h; C-reactive protein, 0.16 mg/dL.

Urinalysis revealed proteinuria of 3+ and 11 to 15 erythrocytes per high-power field. There urine was negative for glucose. A random spot urine protein-creatinine ratio was 12000 mg/g. The serum free light chains assay showed absolute elevation of lambda chains at 1013.9 mg/L with a decreased kappa to lambda ratio of 0.05. Echocardiography showed increased ventricular septal wall thickness and a granular sparkling appearance of the myocardium. A kidney biopsy was performed on the 2nd day of admission. Light microscopic examination showed amorphous homogeneous nodular deposition in the mesangium (Figures 1 and 2). Congo red stain was negative. Electron microscopy showed fibrillary deposits in the mesangium (Figure 3). Immunofluorescence microscopy revealed deposits of lambda type light chains (3+) in the mesangium (Figure 4). Bone marrow examination showed an elevation of plasma cells (10%) with no evidence of amyloid deposition. Flow cytometry demonstrated monotypic plasma cells expressing intracytoplasmic lambda light chain restriction.

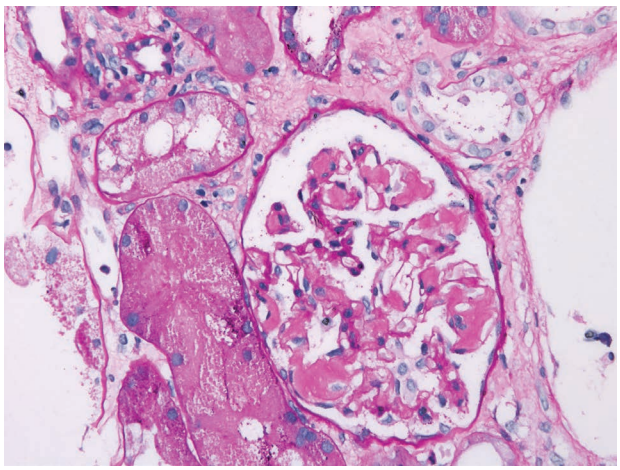

Figure 1. The glomerulus shows mesangial nodules (hematoxylin and eosin, × 400).

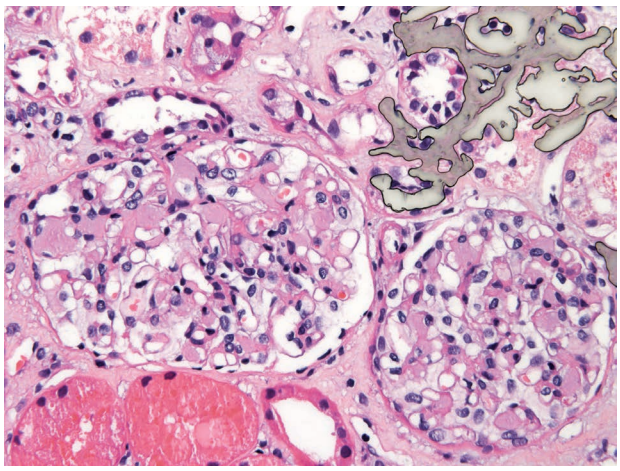

Figure 2. Periodic acid-Schiff staining shows amorphous homogeneous nodular deposition in the mesangium (× 400).

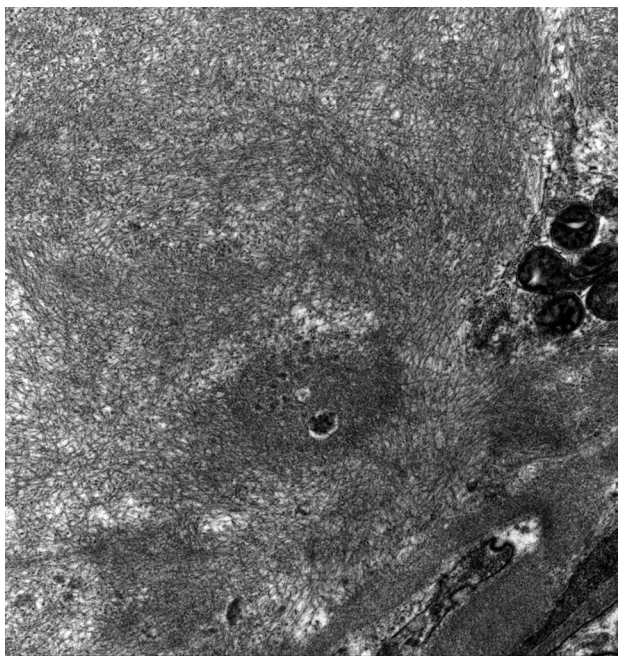

Figure 3. Electron micrograph shows fibrillary deposit in the mesangial matrix with foot process effacement (× 3000).

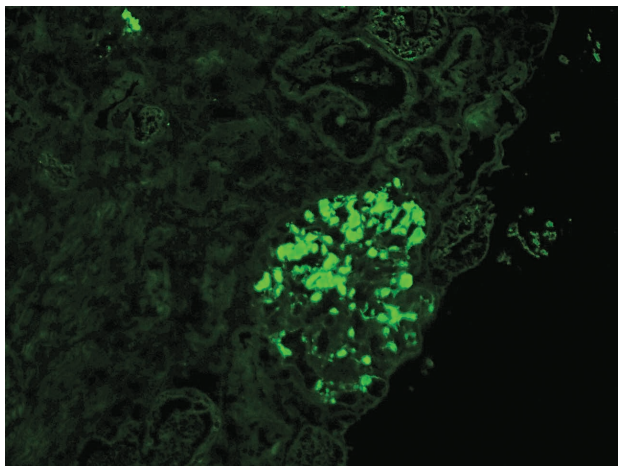

Figure 4. Immunofluorescence shows lambda chain positivity in the glomeruli.

Based on these findings, a diagnosis of LCDD associated with MM was made. The patient was started on a regimen of bortezomib, thalidomide, and dexamethasone for 4 cycles. After 10 days of chemotherapy, serum free light chains assay showed decreased lambda chains of 77.64 mg/L, increased kappa to lambda ratio of 0.32, decreased spot urine protein-creatinine ratio from 44000 mg/g to 16200 mg/g, and decreased serum creatinine level from 4.08 mg/dL to 2.14 mg/dL. Chemotherapy reduced the light chain burden; however, the patient suddenly died due to cardiac arrest.

DISCUSSION

Light chain deposition disease is an unusual disease that is rarely composed of lambda-light chains rather than kappa-light chains.[1] When isolated proteinuria is first detected in an asymptomatic pregnant woman, it is not common for physicians to consider primary kidney disease and kidney disease secondary to systemic disorders in the differential diagnosis.[8,9] Therefore, the diagnosis may be delayed.

In our case, the patient was pregnant, and she had no history of proteinuria in previous pregnancies. The clinical features were consistent with the preliminary diagnosis of preeclampsia; however, kidney biopsy confirmed LCDD of lambda type. Therefore, we suggest that kidney biopsy should be conducted to rule out primary or secondary kidney diseases if the presenting features are atypical for preeclampsia, or response to standard therapy is delayed, and we should recognize the possibility of plasma cell dyscrasia through the analysis of urine proteins in pregnant women with proteinuria.

CONFLICT OF INTEREST

None declared.

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