Renal Amyloidosis and Rheumatic Heart Disease

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Dear Sir,

Although amyloidosis secondary to collagen vascular diseases, in particular to Beh-çet's disease [1], is frequently reported, rheumatic heart disease with amyloidosis is rare. Herein, we report a patient with mitral valve stenosis due to rheumatic heart disease and renal amyloidosis.

A 40-year-old woman with a history of mitral stenosis and mitral valve commisuro-tomy performed 10 years before admission was referred to our hospital because of uremia. On physical examination, blood pressure was 110/60 mm Hg and heart rate 80 beats/min. On auscultation, a 2°/6 systolic murmur was determined along the left sternal border and lung fields were clear. The liver and spleen were nonpalpable. Hematocrit was 20% and WBC was 15,000/mm³, while BUN was found to be 50 mg/dl and serum creatinine level was 4.5 mg/dl. Erythrocyte sedimentation rate was 120 mm/h. Blood chemistry was otherwise normal. Creatinine clearance was 9 ml/min and proteinuria was 7 g/day. ECG showed sinus rhythm with a negative terminal deflection of P wave in lead V₁, and nonspecific ST-T wave changes in the D₂, D₃ and AVF leads. Left-ventricular hypertrophy voltage criteria were present. Chest roentgenogram showed cardiac enlargement. Abdominal ultrasonographic examination demonstrated a reduction in kidney size and an increase in kidney echo pattern. Echocar-diography revealed an increase in left-ventricular wall thickness and mitral stenosis with dilatation of the left atrium. Renal biopsy demonstrated renal amyloidosis (AA). Diet, vitamin D, CaCO₃ and colchicine were initiated. The renal functions of the patient deteriorated rapidly, but she refused hemodi-alysis. She died because of septicemia at the end of the 1-year follow-up period.

In our patient, the renal amyloidosis may have developed secondary to the present rheumatic heart disease or the cardiac pathology may be related to primary cardiac amyloid infiltration as a part of systemic amyloidosis.

Amyloid infiltration of the heart may show several echocardiographic features as in our patients [2], but it is well-known that the diagnosis of cardiac amyloidosis cannot be assumed from the findings of amyloid in other organs and it may not rule out that diagnosis despite the absence of left-ventricular hypokinesia and low-voltage ECG [3].

In addition, most of the amyloid lesions are limited to the atria and there may be involvement of valves and ventricles in only about one-third of the cases [4].

Furthermore, it may be concluded that a long-standing inflammatory process such as rheumatic heart disease can lead to secondary amyloidosis [5].

Other interesting features of our patient were normal serum albumin levels despite massive proteinuria (7 g/day) and bilateral reduced sizes of the kidneys although amyloid deposition was confirmed histopathologically.

References

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