Dear Sir,

Carpal tunnel syndrome in patients with long-term hemodialysis is due to β2-microglobulin-derived amyloidosis [1]. Furthermore, dialysis osteoarthropathy [2] and kidney stones of uremic patients [3] were shown to be derived from β2-microglobulin-related amyloidosis. We examined the amyloid deposition in the kidneys of 8 patients with the acquired cystic disease of the kidney on long-term hemodialysis [4–6].

Method and Case Report

Method

Specimens were obtained from 3 autopsy and 5 surgical exploration cases. Surgical resections were performed because of acquired cystic disease of the kidney complicated with renal cell carcinoma. The specimens were fixed with formaldehyde and stained with hematoxylin and eosin. Congo-red staining was performed with and without potassium permanganate treatment. β2-Microglobulin and amyloid AA protein were stained by the avidin-biotin peroxidase complex (ABC) method. Rabbit antihuman β2-microglobulin and AA protein were obtained from Dako Co., USA, ABC kit Vectastain® was obtained from Vector Laboratories, USA.

Case Report

A 64-year-old male who had been on hemodialysis for 13 years was admitted for a reconstruction of an arteriovenous fistula. His original disease was a suspected chronic glomerulonephritis. He had no history of rheumatoid arthritis, tuberculosis or myeloma. He was screened for renal cell carcinoma using CT scan because he had been treated by hemodialysis for a long time and a renal cell carcinoma with acquired cystic disease was found. The right-side nephrectomy (kidney volume: 189 ml) was performed on June 17, 1985, and a 4.5-cm granular cell carcinoma with multiple cystadeno-mas and solid tumors consisting of clear or granular cells was found. Amyloid was positive around the four cyst walls with single-layered epithelium, in a projection of the cyst wall and around a cyst wall with multilayered epithelium. The pericystic area of single-layered cyst wall was positive for Congo red (fig. 1a) and showed apple-
green birefringence (fig. 1b); β2-microglobulin was stained at the same place (fig. 1c). These Congo-red stains were sensitive to potassium permanganate, but amyloid AA protein staining was negative. The patient did not have any matrix stones and tubular casts were not positive for β2-microglobulin-derived amyloid. Focal amyloid deposits were noted in the renal arteries. The patient did not have carpal tunnel syndrome and was not examined for amyloid deposits in the rectal mucosa.

This case prompted us to examine another 7 patients with acquired cystic disease complicated with renal cell carcinoma except 1. None of them showed any amyloid deposition around the cyst. One case with acquired and atypical cysts exhibited amyloid deposits in the vessel media, tubular casts and matrix stones. Another case had amyloid deposits in the tubular casts and matrix stones. Four patients undergoing hemodialysis for more than 10 years tended to show more frequent β2-microglobulin-derived amyloid deposits in acquired cystic disease of the kidney than the 4 patients receiving hemodialysis for less than 10 years.

Comments

Our long-term hemodialysis patient is a typical case of acquired cystic disease of the kidney complicated with renal cell carcinoma [4, 5]. The original disease was suspected to be glomerulonephritis. The acquired cystic disease associated with glomerulonephritis, not with amyloid nephropathy, was potassium-permanganate-sensitive but the amyloid deposits were negative for amyloid AA. This was not a case of renal cell carcinoma complicated with systemic amyloidosis with AA protein [7, 8] which is also sensitive to potassium permanganate. Amyloid deposits around the cysts contained β2-microglobulin, as has been reported for amyloid deposits in other organs of dialysis patients [1, 2].

The relationship between amyloid deposits and the genesis of acquired cysts and adenocarcinoma is unknown. While the amyloid deposits in the dialysis kidney seem to be associated with epithelial hyperplasia, this is different from amyloid nephropathy with AA or AL deposits of amyloid are involved in the genesis of acquired cysts. Our study does not exclude the possibility of tubular obstruction due to cast or matrix stones composed of β2-microglobulin [3] in the genesis of acquired cysts.

In summary, amyloid deposition can occur in the kidney of some patients with acquired cystic disease of the kidney on long-term hemodialysis. These amyloid deposits were potassium-permanganate-sensitive and negative for amyloid AA protein. The deposits contained β2-microglobulin. This β2-microglobulin-derived amyloid deposition differs from renal cancer-related amyloidosis [7, 8].

Fig. 1. a Amyloid deposits around the cyst wall stained with Congo red under ordinary light. × 200. b Same section photographed under polarized light. Amyloid deposits are birefringent. c Rabbit antihuman β2-microglobulin is markedly stained by the ABC method.

References


