Dialysis-Related Amyloidosis and Acquired Renal Cystic Disease with Renal Cell Carcinoma in Uremia

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Dear Sir,

β 2-Microglobulin (β 2-M)-related amyloidosis is a common complication in patients with uremia under long-term dialysis. Clinical manifestations include carpal tunnel syndrome, destructive arthropathy, and lytic bone lesions. Acquired renal cystic disease (ARCD) is also a complication of uremia which may evolve renal cell carcinoma (RCC). Although separately these two illnesses are not rare in patients with chronic renal failure undergoing hemodialysis, their simultaneous occurrence in a patient has only rarely been encountered. One such case is presented here.

A 55-year-old female presented with headache, nausea, vomiting and asthenia in 1975. The diagnosis of chronic glomerulonephritis was established in 1976 based on findings including proteinuria, azotemia, hypertension and contracted kidneys. In 1978, she developed oliguria, weakness, anorexia, and pallor. Additional laboratory examinations revealed hyponatremia, metabolic acidosis, and anemia. The creatinine clearance rate was 1.6 ml/min. She received peritoneal dialysis and oral lavage sequentially followed by regular hemodialysis (3 times a week using a cuprophane membrane). In 1988, a routine laboratory examination disclosed a high serum β 2-M level (64,111 ng/ml). Three months later she noticed numbness of both hands. Radiographs revealed erosion and loss of bony mass in carpal bones. Bilateral carpal tunnel syndrome was shown by nerve conduction velocity/electromyography. The leftside lesion turned to thenar muscle atrophy in 1990. She received surgery to release the tightened median nerve in 1991. Histologic observations of the resected specimens disclosed amyloidosis. Also in 1988, she was found to have bilateral renal cysts during a sonographic checkup of the abdomen. However, there was no evidence of malignancy at that time. In 1991, computerized tomographic scan of the abdomen demonstrated a 3-cm mass over the lower pole of the left kidney, for which she underwent radical nephrectomy. Pathologic evaluations of the resected tissue confirmed RCC. Follow-up abdominal sonographies did

not detect evidence of metastasis or recurrence of the tumor. In May 1992, she suddenly developed lower abdominal discomfort with passage of a large amount of dark red stool. Colon fiberoscopy demonstrated a cauliflower and infiltrative tumor in the region of cecum and ileoceccal valve. The patient underwent right hemicolectomy. During the operation diffuse ulcerations and pseudopolyps over the ileoceccal region were noted. The pathologic assessment of the resected tissues disclosed ischemic bowel disease of the cecum and ascending colon with amyloid deposition along the vascular walls. She is now under regular high-flux hemodialysis using a polysulfone membrane. The serum β_2 -M concentration in December 1992 was 20,959 ng/ml.

The incidence of gastrointestinal manifestations in dialysis-related amyloidosis is low. The presentations include abdominal pain, fullness, diarrhea and melena [1]. This case is interesting in that, in addition to causing carpal tunnel syndrome and lytic bone lesions, the amyloid infiltration on the walls of the colon submucosal vessels resulted in bleeding and ischemic bowel. The deposition even led to huge polypoid mass formation that mimicked a malignant tumor. Elevated serum β_2 -M levels reflect an increased turnover of lymphatic cells. The kidney is the sole organ that eliminates this peptide. Accumulating evidence supports the pathogenetic role of

β_2 -M in dialysis-related amyloidosis. Although the composition of this patient's amyloid deposit was not investigated, the detection of high serum β_2 -M before the onset of carpal tunnel syndrome was a clue to the role of β_2 -M. The predisposing factors for developing β_2 -M amyloidosis might include the cuprophane dialyzer. This patient's β_2 -M decreased after the usage of a highly bio-compatible membrane. The pathogenesis of ARCD and RCC in uremia is still an enigma. Whether β_2 -M accumulation predisposes or causes ARCD and RCC is not known. Bommer et al. [2] reported that β_2 -M may be related to cystic degeneration of the kidneys in uremia. Canaud et al. [3] suggested that unusually high values of β_2 -M in uremic patients may indicate the presence of occult cancers. Since elevation of β_2 -M reflects an increased turnover of the lymphatic cells with compromise of the immune surveillance, it is possible that β_2 -M accumulation may increase the chance of developing RCC in uremic patients with ARCD.

References

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