Appearance of Systemic Amyloidosis in a Chronic Hemodialysis Patient

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Dear Sir,

Since 1977, amyloid deposits in the transverse carpal ligament of patients on regular dialysis treatment (RTD) were reported. Two unanswered questions persist: (1) Are they localized deposits of a systemic amyloidosis? (2) What is the type of the amyloid? We report a case of systemic non-immunoglobulin-induced (AA) amyloidosis in a patient on RTD.

In 1951, Mme C. was 30 years old when a proteinuria was found during her third pregnancy. In 1967, renal failure was diagnosed during a preoperative workup: high blood pressure, urea nitrogen 12mmol/l, proteinuria 1 g/24 h, Addis count: 300 H/min, 2,000 L/min. IVP showed two regular atrophied kidneys. In 1968, subtotal hysterectomy was performed for endometriosis. Regular hemodialysis was started in 1972. From 1972 to 1981 the principal events had been hepatitis and hyperparathyroidism. In 1981, removal of a sacrococcygeal cyst led to the discovery of subcutaneous deposits with the histochemical behavior of amyloid: positive Congo red staining and green birefringence. There were no clinical signs of carpal tunnel syndrome (CTS), nor systemic amyloidosis. Gamma-pathy exploration was negative. In 1982, bone biopsy and parathyroidectomy were performed: no amyloid deposits were found in the tissues. In 1983, during a routine examination a stage I cancer of the cervix uteri was found and treated by irradiation.

In 1983, spontaneous hemarthrosis of the knees, ankles, wrists and shoulders occurred. In the synovial biopsy specimen, amyloid deposits disrupting the synovial membrane but respecting the underlying vessels were found. In 1984, a massive digestive hemorrhage necessitated a laparotomy: there were no metastases, the bleeding originated in the terminal ileum which was resected, a liver biopsy was performed. Large amyloid deposits were found in the level of ileal submucosa and submucosa vessels, smaller deposits in the hepatic arterioles and sinusoid capillaries. Using the Wright's technique, potassium permanganate treatment eliminated Congo red stainability. The patient died postoperatively. No autopsy was performed.

The successive discovery of the amyloid deposits over a period of 4 years led us to relate them to the duration on RTD.

The absence of CTS in a patient on RTD does not eliminate multiple joint involvements. In addition to the latent subcutaneous amyloid deposits [1], which should be investigated by systematic biopsy, there are latent digestive tract and liver deposits. Spontaneous hemarthrosis and bleeding from the ileum must be considered as being two clinical manifestations of amyloidosis in patients on RTD.

Diagnostic investigations for systemic amyloidosis in patient on RTD with CTS are rarely described [2, 3, 5]. Kachel et al. [3] reported in 1 patient the presence of subcutaneous, synovial and rectal deposits. Biopsies of rectal mucosa or liver are not without risk in the patients on RTD, therefore only the systematic examination for amyloidosis of all specimens from biopsies obtained during the life of the patients will permit a better elucidation of this pathology.

The appearance and the extent of the deposits are either related to the time on RTD or to the duration of terminal renal failure [3]. The number of patients on RTD for more than 10 years are increasing, therefore the first question should be answered in the near future.

The Wright's technique gives, here, arguments in favor of an AA amyloidosis, contrary to other previous report [4]. This type of amyloid should agree with an experimental model [3], considering however that all the experimental amyloidosis are AA: leukocyte pyrogene produced during hemodialysis should induce serum amyloid A protein synthesis. Amyloid deposition in long-term hemodialysis patients could originate from this chronic synthesis.

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