Multiple Tendineal Tumoral Amyloid Masses in a Patient on Long-Term Dialysis

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Dear Sir,

Amyloid deposits are a frequent finding in patients on long-term dialysis during the second decade of treatment [1]. These deposits are believed to be responsible for the carpal tunnel syndrome, chronic arthropathy and bone lacunae. The nature of this particular form of amyloid was elucidated by Gejyo et al. [2] who showed that the β_2 -microglobulin (B2m) is the major constituent of the amyloid fibers. After the first reports it became evident that joints and periarticular tissues are the most common targets where even large masses may be clinically observed.

We report here the case of a uremic patient who developed still undescribed multiple paratendinous B2m amyloid masses, responsible for a progressive and invalidating clinical picture.

Table 1. Time table of amyloid-related complications

Year Complication

1971 start of regular dialysis treatment

1981 bilateral shoulder pain

1983 right CTS (operated in May)

1983 left CTS (operated in November)

cervical spondyloarthropathy with spondylolisthesis of C5

X-ray evidence of bone lacunae at the level of the wrist, femoral heads and hip, bilaterally

1988 macroscopic evidence of multiple large tendineal masses in both forearms and in the distal part of the left leg

CTS = Carpal tunnel syndrome.

Case Report

A 57-year-old woman with chronic uremia was admitted in 1971 to a regular dialytic program using Cuprophane® dialyzers until 1986 and, thereafter, AN 69 hollow-fiber dialyzers. After 10 years of treatment she developed chronic multiple articular stiffness and pain, suggesting synovial involvement related to amyloid deposition. The clinical sequence of these complications is reported in table 1. After 17 years of dialysis treatment the patient developed several masses

which grew in a few months (fig. 1); their consistency was soft and their mean size was $2 \times 3 \times 2$ cm. Congo red staining (fig. 2) revealed amyloid in one of the masses removed and cytochemistry showed a strong presence of B2m. The patient also developed a rupture of the long head of the right biceps humeral muscle, thus further limiting her quality of life.

There are several reports providing evidence of amyloid depositions in hemodialysis patients. The main targets of this complication are synovia, thus explaining the frequent involvement of the carpal tunnel and articular and periarticular subsynovial bone structures. In addition there are anecdotal reports of systemic visceral involvement (heart, lung, liver, spleen, gastrointestinal tract, vessels) whose clinical incidence remains to be defined [3].

Large pseudotumoral periarticular masses of amyloid have already been described [4] and attributed to enlargement of articular synovial bursa filled with amyloid material. The present case provides further proof that the primary targets of amyloid deposition are synovial structures. In fact all nodules were close to tendineal synovia. The tendineal involvement and the large size of the nodules explain the patient's discomfort and joint stiffness. The subsequent biceps tendon rupture added new evidence for the heavy amyloid infiltration of tendineal structures. These findings are showing the strong affinity of B2m amyloid for synovial structures.

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Fig. 1. The physical examination of the patient revealed multiple subcutaneous masses of the right humeral biceps distal tendon (A), of the left carpal ulnar flexor (C) and peroneal (D) tendons; a rupture of the long head of the right biceps humeral muscle was also present (B).

Fig. 2. Microscopic examination of the right humeral biceps paratendinous mass: an apple green birefringence under polarized light is observable. Congo red.

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