Renal Amyloidosis in a Patient with Epidermal Inclusion Cysts

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Dear Sir,

Chronic antigenic stimulation by chronic infections, including cutaneous ones, may be responsible for the induction of amyloid deposition [1-3].

Epidermal inclusion cysts (EIC) are round subcutaneous tumors, slow growing, usually located on the back [4]. Their wall is epidermal, and atrophic in older cysts. EIC are usually single, sometimes numerous. In this paper we present a case with numerous EIC who developed nephrotic syndrome due to renal amyloidosis.

Case Report

A 27-year-old man noted the appearance of skin nodules in 1978. The skin of his face, neck, back and arms was affected. He was admitted to our department 10 years later (in May 1988) because of edema. He had no manifestations of systemic disease. Urinalysis showed the presence of proteinuria (13 g/l) with a normal sediment. Serum creatinine was 80 $\mu\text{mol/l}$, urea 3,6 mmol/l, plasma proteins 40 g/l, and albumins 18 g/l. Serum complement and immunoglobulins were normal. Some of the cysts were fistulous and Staphylo-coccus aureus grew in culture medium. Bence Jones proteinuria was negative. Chest radiography was normal, as were cardiac and abdominal echocardiographic studies. Kidney size was 11,5 cm, parenchyma hyperreflective. A renal biopsy was performed and 40% of the glomeruli presented amyloid deposits on optical microscopy. The walls of two arterioles were also affected.

There was no noted improvement of the nephrotic syndrome and proteinuria during

Fig. 1. Skin EIC of the face and neck.

follow-up, and slow deterioration of renal function. Increase of serum urea (to 12,8 mmol/l) and creatinine (to 181 $\mu\text{mol/l}$) was noted in 1991, and the patient is still without dialysis treatment. Our patient had EIC for 10 years. The presence of other known disorders associated with reactive amyloidosis was ruled out, and there was no evidence of serum or urine monoclonal immunoglobulins [1]. In our case, the most severe consequence of systemic amyloidosis was renal involvement, charac-

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Fig. 2. EIC on the back.

terized by nephrotic syndrome with slow deterioration of renal function.

Chronic antigenic stimulation from cutaneous infections may be responsible for the induction of amyloid deposition in this kind of cutaneous lesion. Other chronic dermatoses such as acne conglobata, dystrophic epidermolysis bullosa, hidradenitis suppurativa and secondary infected burns have been reported with systemic amyloidosis [2, 3].

Besides the esthetic problems, EIC can induce serious systemic disease – amyloidosis.

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Fig. 3. Wall of a cyst removed by surgical treatment. H&E. × 200.

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Fig. 5. Amyloid deposition in the wall of an intrarenal blood vessel. Periodic acid-Schiff. × 200.

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Fig. 4. Intraglomerular amyloid deposition Periodic acid-Schiff. ×200.

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