Electron Microscopic Studies in a Long-Term Follow-Up of a Case of Congenital Nephrotic Syndrome

J.C.		Davin
P.R.		Mahieu
C.		Dechenne

State University of Liège, Belgium

Key Words

Congenital nephrotic syndrome
Basal membrane
Hearing loss
Electron microscopy

Abstract

A boy presenting with a severe congenital nephrotic syndrome diagnosed by histological analysis at the age of 3 weeks was biopsied again 7 years later. The ultrastructural glomerular basement membrane abnormalities depicted in the first biopsy were no longer present in the second one. The number of completely hyalinized glomeruli was not significantly decreased. The GFR remained normal, but a moderate persistent, non-selective proteinuria (800 mg/24 h) was noted without oedema. The patient however developed a progressive perceptive deficit of hearing.

Dr. J.C. Davin, Hôpital de Bavière, Service de Pédiatrie, 6. 66, boulevard de la Constitution, B-4020 Liège (Belgium)

Case Report

The prognosis of nephrotic syndrome appearing during the first weeks of life is often poor [1–3]. In a preceding issue of this journal [4], we reported a case of a precocious and very severe nephrotic syndrome healing almost completely after 12 months. In this paper, we stress the association in the same patient, 7 years later, of perceptive loss of hearing with congenital nephrotic syndrome and the disappearance of the ultrastructural glomerular basement membrane (GBM) abnormalities concomitantly with the improvement of clinical signs. After 7 years of evolution, the patient presented no oedema, but a moderate, glomerular non-selective, proteinuria (800 mg/24 h). The creatinine clearance was equal to 120 ml/min. A perceptive deficit of hearing developed and needed acoustic assistance.

The optical microscopic analysis of the first renal biopsy, performed at the age of 3 weeks, showed a focal and global hyalinosis of 20% of glomeruli, whereas this percentage decreased to 12,5% 7 years later (table I). The immunofluorescent microscopic studies showed no major glomerular deposits in both biopsies.

The electron microscopic studies of the first biopsy mainly

Table I. Number of completely sclerosed glomeruli

Date of biopsy

Normal glomeruli

Completely

sclerosed

glomeruli*

Completely sclerosed glomeruli, %

13/3/1975 25/2/1982

28

7

7 1

20 12.5

*The difference in the number of sclerosed statistically significant ($p > 0.05$).

glomeruli is not

showed the following abnormalities of all GBM: a decrease of the mean thickness and a focal accumulation of sheets of basement membrane material under the visceral epithelial and endothelial cells (fig. 1). This aspect might result from a lack of fusion between the epithelial and endothelial lamina densa. 7 years later, these abnormalities of the basement membranes disappeared. They looked homogeneous, albeit slightly focally thickened (fig. 2). This thickening might be the consequence of a long-lasting nephrotic syndrome which is known to induce an accelerated ageing of the basement membrane. Indeed, the other ultrastructural findings in the second biopsy are those generally observed in nephrotic syndromes lasting for a long time: swelling of endothelial and epithelial glomerular cells, focal fusion of podocytes and some increase of the mesangial matrix.

In summary, we have reported for the first time, as far as we are aware, the evolution of the electron microscopic aspect of basement membranes in two biopsies performed at birth and 7 years later in a boy presenting with a congenital nephrotic syndrome. The two major following findings in this case are: (1) the strong decrease of proteinuria associated with the disappearance of the ultra-structural lesions observed at birth, and (2) a deficit of perceptive hearing developing during the course of the illness.

References

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Fig. 1. First biopsy. The lamina densa is homogeneous but the lamina rara interna has a patchy aspect with a very inconstant width. The total thickness of the basement membrane is about 150 nm but reaches 400 nm in some rare areas. Subepi-thelial (asterisks) and subendo-thelial (arrow) accumulation of basement membrane is observed. Ur. Pb. $\times 12,500$.

Fig. 2. Second biopsy. Mes-angial overload by mesangial matrix and ballooning of endocapillary cells is observed. Basement membrane width ranges from 325 nm to 650 nm (mean thickness: about 500 nm); their structures are homogeneous. Some of the capillary loops are collapsed. Ur. Pb. $\times 6,500$.

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