A Contribution to Nephrotic Syndrome and Focal Glomerulosclerosis in a Patient with Adult Polycystic Kidney Disease

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Dear Sir,

Montoyo et al. [1] have recently described a case of nephrotic syndrome (NS) and focal glomerulosclerosis (FGS) in a young 35-year-old man affected by adult polycystic kidney disease (APKD). According to Montoyo et al. and to our experience moderate proteinuria is observed in many patients with APKD [2], but the development of NS with massive proteinuria has rarely been described; this finding is considered to be a very rare complication of this disease. The aim of this letter is to report a case that resembles that of Montoyo et al. in several clinical and histopathological features.

A 58-year-old man was hospitalized in our Department of Nephrology for polycystic kidney disease and NS with proteinuria ranging from 6 to 8 g/24 h. He did not have an APKD family history; however, in about 25% of patients a family history consistent with APKD is not reported [3] and the spontaneous mutation rate reaches about 10% [4]. Renal function was slightly impaired; urinary sediment showed microhematuria and cy-lindruria. Physical examination revealed large and palpable kidney, urography and abdominal echography diagnosed APKD. Because of his large cysts the patient underwent surgical biopsy. The renal parenchyma was altered by numerous cysts of various sizes filled with hematic or serous liquid.

Histologic examination showed an average of 30 glomeruli/section; more than 50% of these revealed FGS (fig. 1); a marked tubulo-interstitial fibrosis and flogosis with some cysts were also extensively observed (fig. 2).

As usually reported in the literature about FGS, in our patient NS did not respond to steroid treatment at conventional doses; therefore, the drug was withdrawn. Metoprolol, angiotensin-converting enzyme inhibitor and a lower-protein dietary intake were ad-

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Fig. 1. Glomerulus showing segmental sclerosis. × 400.

Fig. 2. Renal parenchyma showing some cysts, atrophic changes in tubules, interstitial flogosis and fibrosis. $\times 100$.

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ministered. Despite this treatment proteinuria did not decrease, renal function worsened progressively and dialysis was started 6 years later.

In our patient proteinuria and histologic glomerular lesions seem to precede the loss of renal function due to kidney cysts, probably owing to a superimposed glomerulonephritis (idiopathic FGS), rather than to an adaptive hemodynamic response to the progressive ne-phron loss.

However, we agree with the authors that both hypotheses must be considered.

References

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