Minimal-Change Nephrotic Syndrome Associated with Renal Angiomyolipoma

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mesangial matrix; tubules, interstitium and vessels were normal. Immunofluorescence for IgG, IgA, IgM, fibrinogen and C3 was negative. Electron microscopy showed effacement of epithelial cell foot processes without deposits. Six days after surgery, serum creatinine had raised to 3.9 mg/dl and for this reason prednisone 1 mg/kg/day was begun. Complete remission of proteinuria and the nephrotic state and normal renal function was achieved 6 days after the start of steroid therapy and 12 days after surgical removal of the renal mass. Steroids were discontinued. Now, 12 months after surgery she remains in remission.

Most patients with malignancy-related MCNS have lymphomatous disorders [1].

Dear Sir,

Whereas the association of minimal change nephrotic syndrome (MCNS) with Hodgkin's, non-Hodgkin's lymphoma and other malignant neoplasms is well known [1], its relationship with benign tumors is less well recognized. There is a single case reporting the occurrence of MCNS in renal oncocytoma [2]. We hereby describe the association of MCNS with angiomyolipoma, a benign renal tumour.

A 51-year-old woman presented with a week history of peripheral edema and weight gain. Physical examination revealed: blood pressure 160/95 mm Hg; pulse 86 beats/min; and 3+ bilateral edema extending below the knees and lumbosacral area. Microscopic examination of the urine disclosed 4-5 white blood cells, 1-2 red blood cells; urinary protein excretion was 8.3 g/24 h. The serum creatinine was 76 µmol/l; serum albumin 261 µmol/l; cholesterol 9.8 mmol/l; antinuclear antibody test was negative; C3c and C4 were in the normal range. A renal ultrasound examination demonstrated a normal right kidney; on the left side there was an intrarenal and hyperechogenic mass approximately 2.5 cm in diameter. The computerized tomography (CT) suggested that the mass contained fat. Intrarenal mass excision and renal biopsy was performed. The renal mass measured 3 cm by 2 cm by 2 cm, was well demarcated and not encapsulated. At microscopic examination, the tumor consisted of mature adipose tissue, smooth muscle and thick-walled blood vessels, sporadically including crystalloids and PAS+, diastase-resistant granules (3), typical of renal hamartoma or angiomyolipoma (fig. 1). On light microscopy, renal biopsy demonstrated normal glomeruli with only slight increase in the

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Fig. 1. Microscopic examination of the renal mass shows mature adipose tissue, smooth muscle and thick-walled blood vessels, typical of angiomyolipoma. HE. $\times 108$.

Evidence suggests that the tumor may be involved directly in the pathogenesis of the MCNS, perhaps as a result of a T-cell function disorder [4]. MCNS has also been associated with oncocytoma, a benign renal tumor [2]. An etiologic association between angiomyolipoma and MCNS has not been reported previously. In our case, the diagnosis of both MCNS and renal angiomyolipoma was clear-cut. The relationship is suggested by the coincidence in time and by the remission of MCNS after tumor removal. Certainly steroids could have contributed to the remission of MCNS. However, the few days that elapsed between surgery and remission suggest that elimination of the mass also played a role and supports the hypothesis of

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an etiologic association between renal angio-myolipoma and MCNS rather than that of an incidental finding. The nature of this relationship is unknown, but perhaps renal angio-myolipoma is capable of secreting tumoral factors like cytokines that could alter glomerular permselectivity and induce MCNS, but we are not aware of this having been proven. Regardless of the mechanism, clinicians should consider renal angiomyolipoma in the differential diagnosis of patients presenting with MCNS.

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