Congenital Mesoblastic Nephroma Associated with Focal and Segmental Sclerosis

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Dear Sir,

Since the classic study of Bolande et al. [1], congenital mesoblastic nephroma (CMN) has been considered to be an entity separate from Wilms' tumor. It is a rather unusual neoplasm of infancy and early childhood [1-8]. Complete histological investigation is crucial for accurate categorization of CMN and diagnosis of simultaneous underlying kidney pathology. A scan view of the contralateral kidney and a long follow-up definitely contribute to a better understanding of the clinical course of these patients. Though CMN usually occurs in the absence of recognized clinical syndromes, there are some reports showing association of this tumor with hemihypertrophy, polyhydramnios and hypercalcemia [2, 3].

We describe here an unusual and interesting case of CMN which to our knowledge represents the first documentation of the association of focal segmental sclerosis (FSS) and progressive renal failure. A 7-month-old white normotense male infant, who had a cryptic right testicle, presented a mass in the right flank which was displacing the ureter and vena cava.

Heterogeneous areas and microcalcifications were observed in the right kidney by computed tomography and ultra-sonography. Urinalysis showed maximum documented proteinuria of 3.4 mg/24 h and no microscopic hematuria. The initial BUN/creatinine ratio was 10/0.9 mg/100 ml. At the time of nephrectomy the imaging of the left kidney was normal.

The patient underwent nephrectomy. The excised kidney weighed 700 g, and measured 16 × 8 cm. The external surface was

smooth and pale tan. On the cut surface there was a well-limited mass measuring 12 × 8 × 4 cm. This mass was completely enveloped by a smooth capsule, with a whorled appearance and of fibroelastic consistency (fig. 1). The remaining renal parenchyma surrounding the lesion was normal. Microscopic examination of the tumor mass revealed the pattern of typical CMN, confined to the capsule limits (fig. 2). Histological examination of the remaining kidney parenchyma disclosed areas of focal glomerulo-sclerosis (FGS).

One year after surgery, arterial hypertension was detected and poorly controlled. Urinalysis showed maximum documented proteinuria 3.4 mg/24 h, intermittent microscopic hematuria and a serum BUN/creatinine ratio of 27/10 mg/100 ml. In November 1993 the patient was 2 years and 8 months old and presented periorbicular edema, a blood pressure of 190/110, and a serum BUN/creatinine ratio of 197/27 mg/100 ml. At that time the left renal ultrasound

showed increased echogenicity suggestive of underlying chronic renal disease. A left renal biopsy was performed showing mesangio-proliferative glomerulopathy leading to global sclerosis (fig. 3). The patient also underwent right orchidopexy. The patient was then submitted to peritoneal dialysis. Two months later he developed peritonitis and died. CMN is a rather unusual neoplasm of infancy and early childhood, generally showing good prognosis after nephrectomy. We report a rare and interesting case of a CMN arising in a kidney with FSS which was characterized and further clarified by long-term follow-up and by a renal biopsy of the contralateral kidney. To our knowledge, this case represents the first documentation of this combination. The simultaneous occurrence of CMN and FSS in our patient indicates a causal relationship but the association between these two conditions remains unproved.

FGS is a common, nonspecific pattern of glomerular response to injury that occurs in a wide variety of glomerular and nonglomerular diseases. The occurrence of CMN and FGS is provocative but unique in the literature. The pathogenesis of FGS lesions is not known in this or in other clinical settings in which it occurs, such as idiopathic nephrotic syndrome. A possible unifying hypothesis for these observations is that FGS can result from glomerulus 'overload' or hyperfiltration. This may explain the occurrence of FGS in a wide variety of chronic renal diseases that severely reduce the number of functional nephrons and, possibly, in the kidneys of our patient. The marked deterioration of renal function following nephrectomy in our patient may result from a worsening of the hyperfiltration injury in the remaining kidney parenchyma. Experimental studies have shown that subtotal (5/6) nephrectomy in young rats results in hyperfiltration in the remaining nephrons and the development of progressive segmental glomerulosclerosis [3, 5]. The critical overload period is at younger ages.

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Fig. 1. Sectioned surface of kidney, showing a tumor with a whorled appearance and indistinct margins separating it from the adjacent renal parenchyma (arrow).

Fig. 2. Histologic aspect of mesoblastic nephroma with woven bundles of closely packed cells with scanty cytoplasm. HE. $\times 400$.

Fig. 3. Histologic aspect of mesangioproliferative glomerulopathy with sclerosis. Hexamine methenamine silver. $\times 400$.

Prompted by the significant occurrence of FGS in patients with congenital solitary kidney (unilateral renal agenesis) or acquired solitary kidney due to nephrectomy, we investigated the possibility that FGS might develop in other patients with reduced renal mass. Although rare, the combined occurrence of CMN and FSS in an individual patient may not only present a diagnostic dilemma but may also raise questions regarding appropriate management. Treatment strategies for both entities depend on accurate clinical follow-up and should be integrated with the pathologic aspects to provide optimal results. A prolonged follow-up of CMN cases would be interesting to determine whether this association is casual or related and to detect alterations developing in the same or in the contralateral kidney.

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