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Function within mesoblastic nephroma: imaging - pathologic correlation

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Abstract. Congenital mesoblastic nephroma is the only primary renal neoplasm with radiographically demonstrable function. Renal function within two typical congenital mesoblastic nephromas was demonstrated by computed tomography in one case and renal scintigraphy in the other. Excretion of contrast medium and radionuclide was due to functioning nephrons trapped within the tumor.

Renal function was demonstrated by computed tomography and renal scintigraphy in two patients with typical congenital mesoblastic nephroma. This function was due to excretion by functioning nephrons trapped within the stroma of the tumor.

Case reports

Case 1

M. H., a four-day-old female, presented with a palpable right abdominal mass. The patient was a 2836 g, 37 week gestational age infant born to a 22-year-old female by vaginal delivery. Prenatal ultrasound had not been performed.

The infant had a distended abdomen with a firm, right-sided flank mass which did not cross the midline. The physical examination was otherwise normal. Complete blood count, electrolytes, BUN, and creatinine were normal.

Sonography demonstrated a solid intrarenal mass in the lower pole of the right kidney (Fig. 1 A). Renal scintigraphy, using 99m-Tc-DTPA, showed normal function of the left kidney as well as function within the lower pole mass of the right kidney (Fig. 1 B).

The patient underwent a radical right nephroureterectomy. The right renal tumor measured $4.0 \times 3.5 \times 3.25$ cm and had a yellowish-tan, glistening appearance on gross cut section (Fig. 1 C). Microscopically, there were sheets of uniform spindle cells which surrounded normal appearing glomeruli and tubules (Fig. 1 D). The pathologic diagnosis was mesoblastic nephroma.

Case 2

N. A., a two-day-old male, was transferred to Children's Hospital Medical Center with a palpable left abdominal mass. The patient was a 2700 g, 36 week gestational age infant born to a 20-year-old female by low forceps delivery. Prenatal ultrasound at 32 weeks gestation had demonstrated a solid left upper pole renal mass.

The infant had a protuberant abdomen with a firm left-sided flank mass which crossed the midline. The physical examination was otherwise normal. Complete blood count, electrolytes, BUN, and creatinine were normal.

Sonography demonstrated a solid intrarenal mass in the upper pole of the left kidney. CT confirmed the $6.25 \times 4.80 \times 5.5$ cm mass and demonstrated excretion of contrast medium in the periphery of the tumor mass as well as compressed normal renal parenchyma (Fig. 2 A). Delayed CT sections at the same anatomic level showed excretion of contrast into dilated upper pole calyces as well as continued opacification of compressed renal parenchyma (Fig. 2 B).

The patient underwent a radical left nephroureterectomy and adrenalectomy. The tumor had a yellowish glistening appearance on cut section (Fig. 2 C). Grossly, the renal capsule was intact and the mass blended imperceptably with normal renal parenchyma (Fig. 2 C). Microscopically, the neoplasm consisted of sheets and whorls of uniform spindle cells with occasional mitotic figures. Tumor cells surrounded and engulfed normal renal structures (Fig. 2 D). The pathologic diagnosis was mesoblastic nephroma.

Discussion

Congenital mesoblastic nephroma (fetal renal hamartoma) is the most common renal neoplasm occurring in the first few months of life; the prognosis is usually excellent [1]. A recent review [2] and case report [3] have confirmed the pathologic spectrum of congenital mesoblastic nephroma as proposed by Beckwith [4], which ranges from benign renal tumors, through atypical lesions of more aggressive potential, to highly malignant spindle cell tumors akin to clear cell sarcoma of kidney. Typical mesoblastic nephroma tends to be firm, "whorled", and pale yellow to whitish-gray in color without grossly

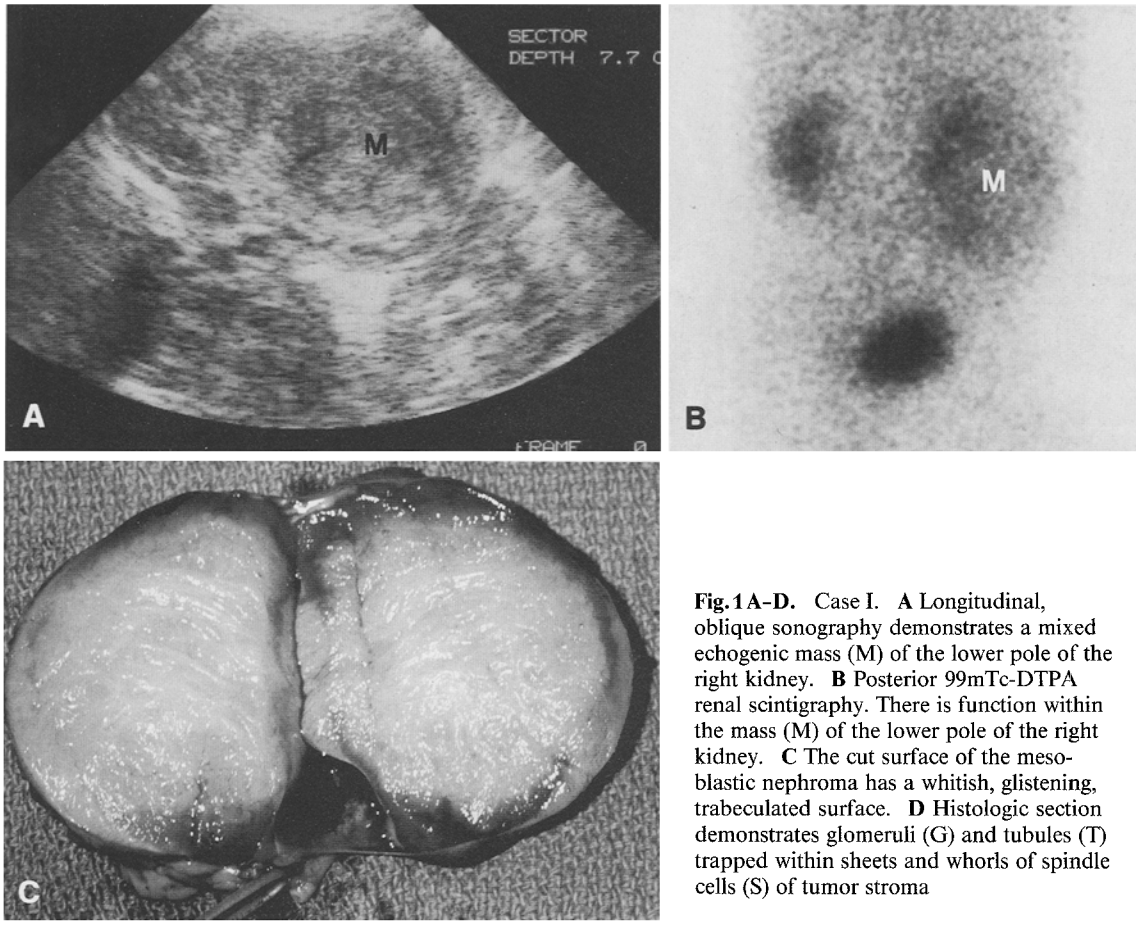
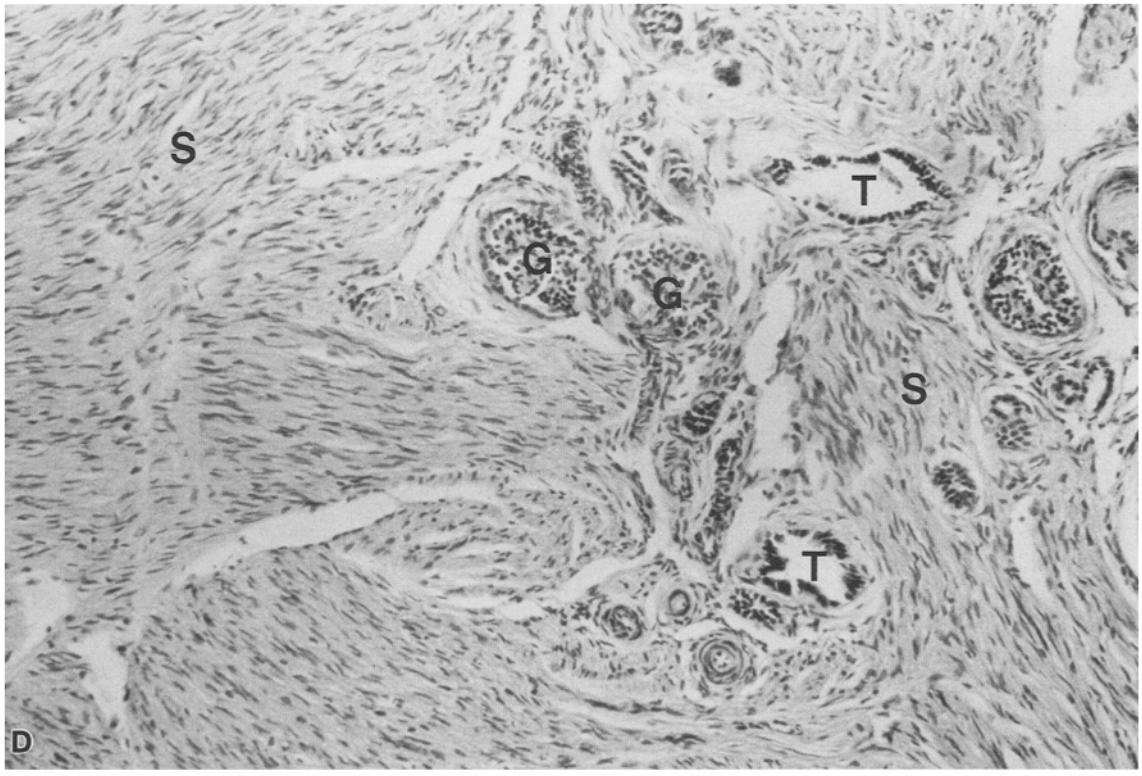


Fig. 1 A–D. Case I. **A** Longitudinal, oblique sonography demonstrates a mixed echogenic mass (M) of the lower pole of the right kidney. **B** Posterior 99mTc-DTPA renal scintigraphy. There is function within the mass (M) of the lower pole of the right kidney. **C** The cut surface of the mesoblastic nephroma has a whitish, glistening, trabeculated surface. **D** Histologic section demonstrates glomeruli (G) and tubules (T) trapped within sheets and whorls of spindle cells (S) of tumor stroma



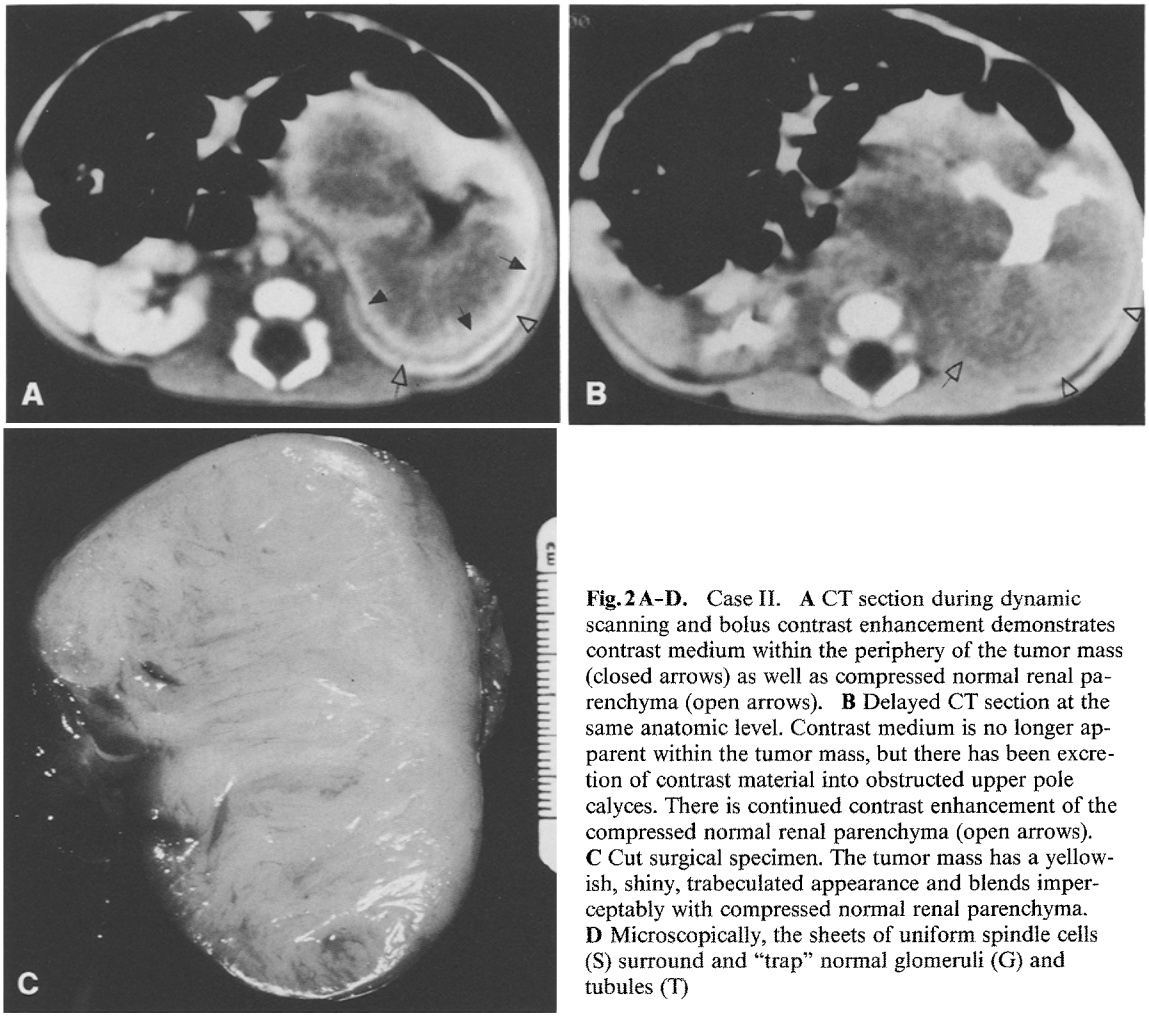


Fig. 2 A–D. Case II. **A** CT section during dynamic scanning and bolus contrast enhancement demonstrates contrast medium within the periphery of the tumor mass (closed arrows) as well as compressed normal renal parenchyma (open arrows). **B** Delayed CT section at the same anatomic level. Contrast medium is no longer apparent within the tumor mass, but there has been excretion of contrast material into obstructed upper pole calyces. There is continued contrast enhancement of the compressed normal renal parenchyma (open arrows). **C** Cut surgical specimen. The tumor mass has a yellowish, shiny, trabeculated appearance and blends imperceptably with compressed normal renal parenchyma. **D** Microscopically, the sheets of uniform spindle cells (S) surround and "trap" normal glomeruli (G) and tubules (T)

evident necrosis. Atypical and more aggressive mesoblastic nephromas tend to be soft, fleshy tumors with areas of gross hemorrhage and necrosis [2]. Microscopically, typical mesoblastic nephroma is composed of uniform, spindle-shaped cells which are arranged in bundles and surround nephrons. Conversely, atypical mesoblastic nephroma is more cellular without recognizable normal glomeruli or tubules [2].

The mean age of patients presenting with typical mesoblastic nephroma is 5 days with a slight predominance of females [2]. The newborn usually presents with a large, non-tender abdominal mass.

Plain films of the abdomen show a large, soft-tissue abdominal mass that is rarely calcified. Sonography most commonly demonstrates a mixed echogenic intrarenal mass indistinguishable from Wilms tumor in the older child. The sonographic findings are similar to those of a non-calcified uterine leiomyoma: a solid renal mass with low-level internal echos [5].

Neovascularity is extremely common within mesoblastic nephroma [6]. Moreover, small accumulations of contrast medium without obvious communication with the collecting system may be seen within the tumor by excretory urography [5]. Sty and Oechler reported function within mesoblastic nephroma demonstrated by 99m-Tc-glucoheptonate renal scintigraphy [7]. They noted that the fibrous and mesenchymal stroma of the tumor "isolated and surrounded islands of normal glomeruli and tubules, but with preservation of complete nephrons" [7]. They postulated that the tracer was filtered by these functioning nephrons and subsequently attached to the normal tubules trapped within the tumor. The function demonstrated in our two patients by 99m-Tc-DTPA renal scintigraphy (Case I) and enhanced CT (Case II) was due purely to glomerular filtration by functioning nephrons since there is no attachment of this radionuclide tracer or contrast medium to tubules.

Function within typical congenital mesoblastic nephroma may be demonstrated by renal scintigraphy or computed tomography. Continued excretion on delayed images verifies that this is function and not merely total body opacification effect within the hypervascular tumor. Our cases support the claim that mesoblastic nephroma is the only primary renal neoplasm with demonstrable function [5]. Such function is due to trapping of normal glomeruli and tubules within the benign stroma of the tumor. Tumor function has only been reported and demonstrated in patients with the typical form of mesoblastic nephroma. Nephrectomy of this benign form of the tumor is curative without the need for supplementary radiation or adjuvant chemotherapy [2, 6].

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