Antenatal Diagnosis of Wilms Tumor

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Renal tumors present very rarely in utero, with mesoblastic nephroma being more common than Wilms tumor. Antenatal detection of mesoblastic nephroma has been reported previously on 13 occasions.[1] Prenatal diagnosis of nephroblastomatosis, which is believed to be a premalignant precursor of Wilms tumor, also has been reported.[2] We report a case of antenatally detected Wilms tumor.

CASE REPORT

A 23 year old gravida 3 para 1 aborta 1 woman was referred for a second opinion. The previous scan, done to evaluate polyhydramnios, revealed a large abdominal mass. The patient had no risk factors for congenital anomalies. Real-time B-mode ultrasonography showed a single fetus in cephalic presentation at 37 weeks of gestation with all fetal parameters corresponding to gestational age. A large, solid, well-encapsulated mass measuring 8.5 × 5.7 × 6.0 cm was seen in the left renal area. A portion of the left kidney was visualized and appeared normal (Figs. 1, 2). Color Doppler sonography detected flow in the mass and pulsed wave Doppler sonography showed an increased diastolic flow (resistive index = 0.5), indicating a low-resistance circulation (Fig. 3). Although mesoblastic nephroma is more common in the antenatal and neonatal period, the presence of a well-encapsulated tumor prompted us to make a diagnosis of Wilms tumor. The contralateral kidney appeared normal. The small bowel appeared compressed. Bilateral clubfeet and polyhydramnios were present. Biophysical activity of the fetus was normal.

The following day, the patient reported loss of fetal movements. A stillborn male fetus weighing 2.8 kg was delivered by cesarean section. An elective cesarean section was performed in view of suspected abdominal dystocia and a previous cesarean section. External examination revealed abdominal distention, bilateral clubfeet, and a normal head and neck. Autopsy confirmed the presence of a mass in the left renal region. All the other abdominal viscera and the head and neck appeared normal. No enlarged lymph nodes were found. The cause of stillbirth could not be ascertained. Histopathologic examination of the mass revealed biphasic histologic findings with sheets of primitive blastema cells interspersed with foci of glomerular differentiation consistent with Wilms tumor (Fig. 4).

DISCUSSION

Wilms tumor has an annual incidence of 7.8 per 1,000,000 children under the age of 15 years with a random risk of developing the tumor estimated to be 1 in 10,000 live births.[3] Most of the tumors are unilateral. The recurrence risk for siblings after one child is affected is 1% for unilateral and 1 to 2% for

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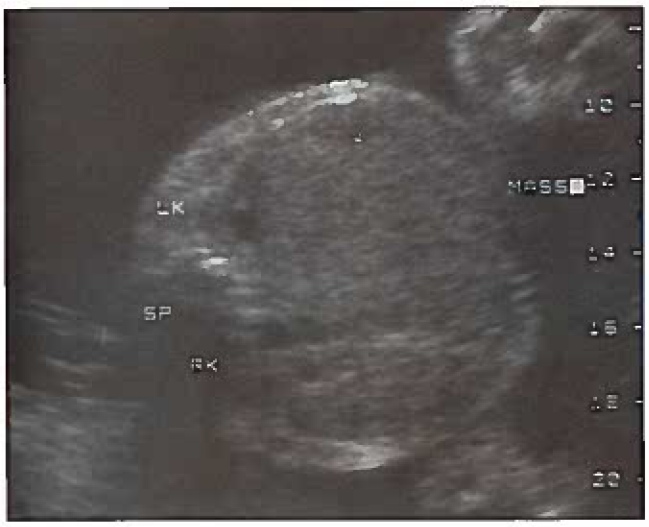

Figure 1 Transverse section of the abdomen shows a well-encapsulated mass in the left kidney. RK, Right kidney; LK, portion of the remaining normal left kidney.

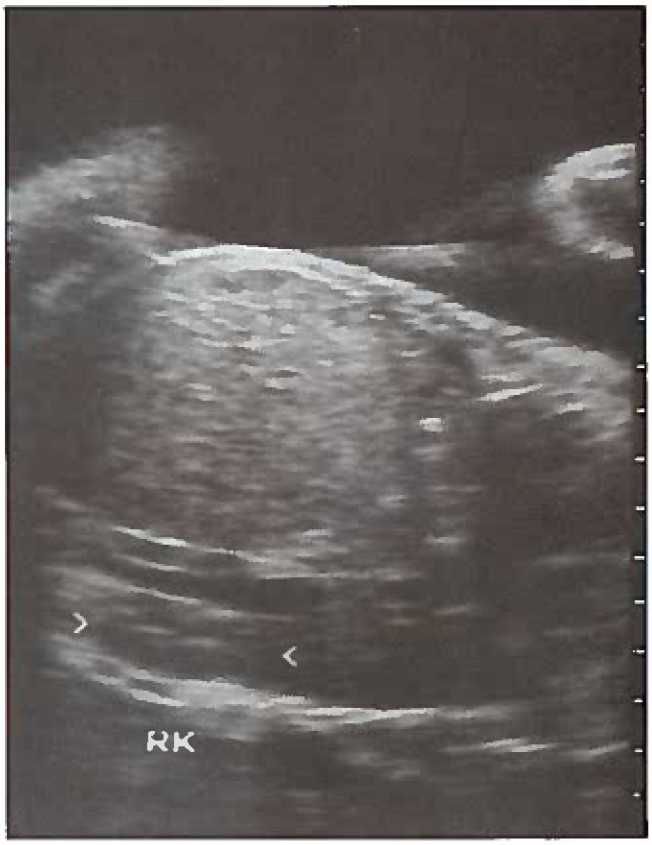

Figure 2 Coronal section shows a well-encapsulated mass in the left renal area. RK, Right kidney.

bilateral tumors. The recurrence risk is 5% for unilateral and 32% for bilateral tumors if the parent is affected.[3]

Mesoblastic nephroma is a solitary hamartoma of the kidney. It appears as a solid infiltrating tumor with medium level echoes on ultrasonography.[4] It is difficult to define the outline of this tumor sonographically, as it is contiguous with the normal nephrons, does not have a well-defined capsule, and blends with the remaining normal kidney.[5–7] In contrast, Wilms tumor appears as a solid echogenic mass that is well encapsulated.[8] The capsule is well visualized and can be seen separating the mass from the remaining portion of the normal kidney. The mass may be interspersed with anechoic spaces representing areas of hemorrhage and necrosis.[9] The well-defined borders of the mass represented a pointer to the diagnosis of Wilms tumor in our patient.

An increased prevalence of congenital anomalies occurs in association with Wilms tumor. Hemihypertrophy (total, segmental, or crossed) of the body, more common with bilateral tumors, is not present at the time of birth and not diagnosable in utero.[3] The antenatal scan should include a careful search for ultrasonographic markers of chromosomal anomalies, as trisomy 18 and Turner syndrome have been reported to occur with Wilms tumor.[10]

Dysplastic kidneys may show changes that are focal or involve the whole kidney. Focal renal dysplasia may produce a mass lesion mimicking a renal tumor.[11] The presence of other malformations of the urinary tract, such as ureteropelvic junction obstruction, ureteral duplication with ectopic ureterocele, posterior urethral valves, ectopic ureter or "prune belly" syndrome, in association with dysplastic kidneys differentiates it from renal tumors.[11]

Polyhydramnios is a consistent feature associated with renal tumors. Decreased gastrointestinal uptake due to compression by the mass and increased blood flow leading to increased urine production are the factors that have been postulated to cause polyhydramnios.[12] Both of these factors could have contributed to the presence of polyhydramnios in our patient.

Fetal kidney biopsy can be performed if the mass is detected at an earlier gestational age. Lin and coworkers have reported that increased urinary hyaluronic acid is a marker for Wilms tumor.[13] Hence, the possibility of detecting increased levels of hyaluronic acid in the amniotic fluid of fetuses with Wilms tumor needs to be explored.

Mesoblastic nephroma usually is a benign tumor and the prognosis is good. It can be treated with nephrectomy alone. Chemotherapy or radiotherapy can be offered in the rare instance when the histologic findings are suggestive of malignancy.[14] The

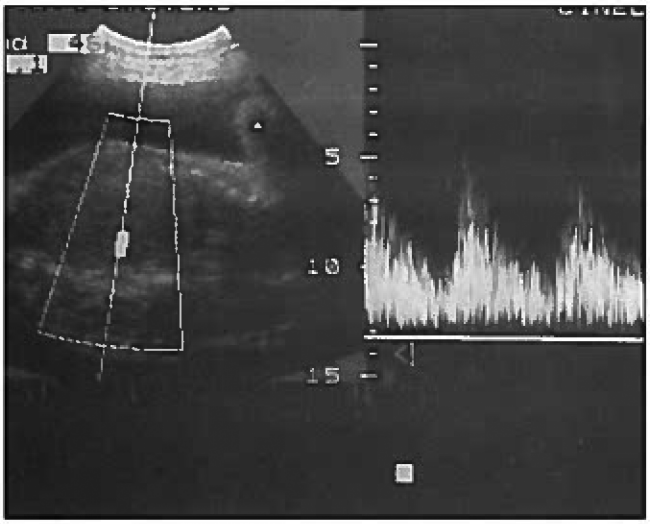

Figure 3 Doppler sonogram of the mass shows increased diastolic flow.

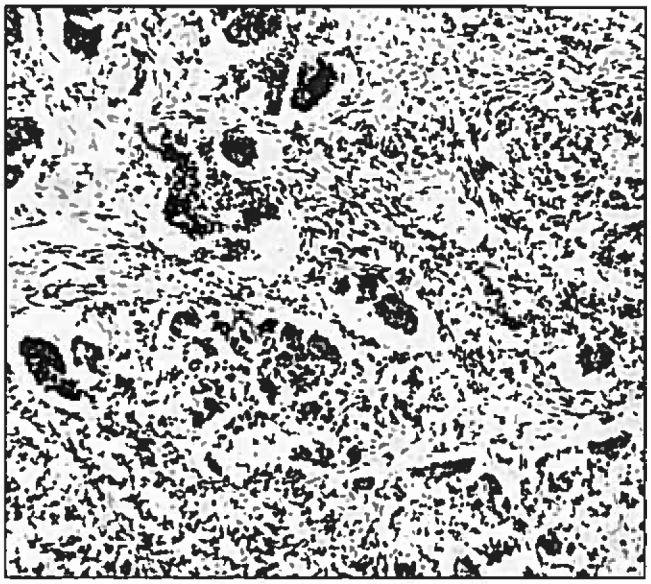

Figure 4 Histologic examination of the mass shows sheets of primitive blastema cells interspersed with foci of glomerular differentiation.

prognosis in a patient with Wilms tumor depends on the age at detection, stage, size, lymph node invasion, and histologic type, with a 90% survival rate for tumors of stages I, II, and III with favorable histologic composition.[15] Nephrectomy followed by radiotherapy or chemotherapy is the treatment of choice.[3] The overall cure rate is 80 to 90% in unilateral tumors.[16] The prognosis is good when the tumor is detected before the age of 2 years. The presence of extensive tubular differentiation, massive amounts of rhabdomyoblastic differentiation, and absence of anaplasia also denote a good prognosis.[16]

In conclusion, Wilms tumor can be detected antenatally by delineating the well-defined margins of the tumor, and the diagnosis can be confirmed by a fetal renal biopsy. Amnioreduction for the polyhydramnios can prevent preterm labor and its associated complications. The parents should be counseled about the prognosis for the child. Prenatal diagnosis can alert the pediatric surgeon and the neonatologist to decide on the time of surgery and course of management after the baby is delivered, as detection and treatment at an earlier age have a better prognosis.

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