CLINICAL CASE

Classic mesoblastic nephroma


Abstract  Mesoblastic nephroma is usually detected before 12 months of age. It often presents as an intra-abdominal mass and is occasionally related to polyhydramnios during pregnancy, high blood pressure, hematuria, and premature birth. It corresponds to 3% of all renal tumors in pediatric patients and 90% of the cases are diagnosed during the first year of life.

Presented herein is the case and its management of a neonate with an intra-abdominal mass corresponding to a mesoblastoma.

A 30-day-old infant presented with an abdominal tumor detected at birth. He was the product of a second pregnancy and was delivered by cesarean section at 40 weeks of gestation. The pregnancy was unremarkable and the periodic check-ups were registered as normal. Abdominal asymmetry was observed at birth and a mobile, non-painful 5 x 5 cm mass was palpated in the right hemiabdomen. Abdominal ultrasound, Doppler ultrasound, and plain and contrast-enhanced abdominal tomography scans were performed. The infant underwent a right nephrectomy, producing a 75 g right kidney measuring 7 x 5 x 4.5 cm. Its upper pole had a 5 x 5 x 5 cm well-defined white fibrous neoformation. The microscopic study revealed mesoblastic nephroma.
Introduction

Described for the first time by Bolande in 1967,1 mesoblastic nephroma is a congenital kidney neoplasia that is usually detected before 12 months of age. The mean age is 2 months. Nephroma usually presents as an intra-abdominal mass and occasionally is related to polyhydramnios during pregnancy, high blood pressure, hematuria, and premature birth.2 It corresponds to 3% of all kidney neoplasia in pediatric age patients and 90% of the cases are diagnosed during the first year of life.3

Case presentation

A 30-day-old infant boy was referred to our pediatric urology unit to be evaluated for an abdominal tumor detected at birth. He was the product of a second pregnancy and delivered at 40 weeks of gestation by cesarean section. The pregnancy was unremarkable. Abdominal asymmetry was observed at birth and a 5 x 5 cm mobile, nonpainful mass was palpated in the right hemiabdomen. Wilms’ tumor was suspected and an abdominal ultrasound reported a retroperitoneal lesion in the upper pole of the right kidney. Its edges showed well-defined echogenicity, it was predominantly solid and heterogeneous, with a hypoechoic center, and displaced the intestinal segments anteriorly and medially (fig. 1). A color Doppler ultrasound revealed significant flow and a lesion measuring 69 x 53 x 48 mm at its largest diameters with no evidence of lymph node growth (fig. 2). Laboratory tests reported hemoglobin 17.6 g/dL, hematocrit 49.6%, platelets 229,000, leukocytes 26,900, creatinine 0.8, and urea 22.4. Plain and contrasted abdominal computed axial tomography (CAT) scans showed a well-delineated retroperitoneal abdominal mass dependent on the upper pole of the right kidney, with contrast-enhanced edges, measuring 46.7 x 52.3 mm. Its content was heterogeneous with a hypodense central portion (figs. 3 and 4). The patient was clinically stable with no data of acute abdomen, his control laboratory tests were within normal parameters and he had conserved kidney function. The infant underwent right nephrectomy that revealed a moderately vascularized, solid renal tumor in the upper pole of approximately 8 x 6 x 5 cm, with no disease in veins or lymph nodes. The postoperative period was adequate, tending towards improvement, and the patient was released on the 7th day.

The pathology study of the surgical specimen reported a right kidney weighing 75 g and measuring 7 x 5 x 4.5 cm in the upper pole with a white fibrous well-defined neoformation of 5 x 5 x 5 cm. The inferior portion of the specimen measured 3 x 1.8 x 1.8 and in this area the cortex/medulla relation was conserved (fig. 5). Microscopic study revealed myofibroblasts that infiltrated the parenchyma, impeding the development of tubules and glomeruli. They were characterized by a predominance of matrices, without atypia. The histopathologic definitive diagnosis was mesoblastic nephroma (5 x 5 x 5 cm) with infiltration of the renal capsule and classic perirenal fatty tissue. The
operating field was at least 1 mm from the infiltration of the perirenal fatty tissue. The vessels studied from the hilar region were negative for neoplastic permeation.

The patient has continued to have periodic follow-up and has shown adequate growth and development for his age. Control ultrasound study showed no tumor activity, the blood chemistry reported creatinine of 0.9 and urea of 20.4, and the remaining tests were within normal parameters.

Discussion

There are many causes of abdominal and pelvic tumors in neonates, and those of renal origin represent 55% of the cases. The male/female presentation rate of mesoblastic nephroma is 2:1, respectively. It has 2 presentations: classic mesoblastic nephroma and cellular nephroma. Our patient presented with the classic type, typically leiomyomatous. In general, it behaves like a benign renal tumor. Lesions of this type are usually cured through surgical resection, with no need for radio or chemotherapy.

The first-line diagnostic complement is abdominal ultrasound that frequently shows a unilateral, hypoechogenic, and homogeneous renal mass. The presence of hypoechogenic zones and hyperechogenic rings aids in the diagnosis. Color Doppler ultrasound denotes hypervascularization and changes of pattern in the adjacent vessels. The CAT scan showed an intrarenal mass with a nonspecific attenuation pattern that could be homogeneous or heterogeneous.

There were no histologic criteria that consistently correlated with outcome and it appears that age (under 3 months) and adequate complete surgical resection are the best outcome indicators.

Figure 2  Doppler ultrasound image showing a right renal mass with increased vascular flow.

Figure 3  Tomography scan showing a 4.9 x 5.4 cm mass in the upper pole of the right kidney.

Figure 4  Nuclear magnetic resonance image showing the right renal mass.

Figure 5  A solid, well-defined, fibrous neoformation in the upper pole of the kidney.
The classic variant of mesoblastic nephroma that our patient presented with is described as a large firm mass with infiltrating edges and a similar appearance to a uterine leiomyoma. The cellular variant is softer, with cystic areas and irregular contours. As a microscopic finding the main characteristic is the proliferation of fusiform cells with varying degrees of cellularity and characteristics of secondary mesenchyme. The cells in proliferation have myofibroblast, fibroblast, and smooth muscle characteristics.

Recently, the (t12;15) (p13;q25) translocation resulting in the ETV6-NTRK3 fusion has been found in these tumors; the same alteration detected in infantile fibrosarcomas. This suggests that they could be the same neoplasia. Both variants are reactive to fibroblastic markers, such as vimentin, actin, and desmin; they are negative to epithelial markers.

The treatment for both subtypes continues to be radical nephrectomy. During the first year, the patient should undergo periodic control ultrasonography to detect early signs of recurrence. Recurrence factors are positive surgical margins, rupture of the tumor during resection, the age of the patient, and the disease subtype. Patients older than 3 months, with the cellular variant, are candidates for adjuvant chemotherapy. The most frequent chemotherapies are vincristine, cyclophosphamide, and doxorubicin (VCD) or vincristine, doxorubicin, and actinomycin (VDA) and ifosfamide, carboplatin, and etoposide (ICE).

**Conflict of interest**

The authors declare that there is no conflict of interest.

**Financial disclosure**

No financial support was received in relation to this article.

**References**


**Errata sheet**

In the *Revista Mexicana de Urología* Vol. 73, September - October 2013, number 5 and in supplement 1 Vol. 73, November 2013, the following information was omitted in the copyright notice and disclaimer:

*REVISTA MEXICANA DE UROLOGIA*, year 2013, number 5, September-October.

*REVISTA MEXICANA DE UROLOGIA*, year 2013, supplement 1, November.