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CONGENITAL MESOBLASTIC NEPHROMA: CASE REPORT

NEFROMA MESOBLÁSTICO CONGÉNITO: REPORTE DE CASO

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ABSTRACT

Congenital mesoblastic nephroma (CMN) is the most frequent renal tumor in newborns and infants. under 3 months. The clinical case of a patient younger (under) than 3 months, with prenatal diagnosis, referred in a timely manner for evaluation and management of the Institution by the Institution is presented. CMN is a low incidence tumor. Early diagnosis as well as complete excision of the tumor are predictors of good prognosis, as in the case of our patient.

Keywords: Congenital mesoblastic nephroma; Renal tumor; Infants. (Source: MeSH NLM).

RESUMEN

El nefroma mesoblástico congénito (NMC), es el tumor renal más frecuente en recién nacidos e infantes menores de 3 meses. Se presenta el caso clínico de una paciente menor de 3 meses, con diagnóstico prenatal, referido de manera oportuna para evaluación y manejo de la Institución. El NMC es un tumor de baja incidencia. El diagnóstico precoz así como la exéresis completa del tumor son predictores de buen pronóstico, como en el caso de nuestra paciente.

Palabras claves: Nefroma mesoblástico congénito; Tumor renal; Lactantes. (Fuente: DeCS BIREME).

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INTRODUCTION

Congenital mesoblastic nephroma (CMN), also called fetal renal hamartoma, leiomyomatous or mesenchymal hamartoma, is a tumor with a good prognosis, except for the cell line⁽¹⁾. Described for the first time by Bolande in 1967, as a tumor different from nephroblastoma due to its histology, treatment and prognosis⁽²⁾. It represents 3 to 5% of all renal tumors in pediatrics⁽³⁾, more frequently in newborns and infants under 3 months⁽²⁾. Local recurrence and metastasis is 5% in the first year⁽⁴⁾.

The origin is probably due to nephrogenic mesenchymal proliferation⁽⁵⁾. The NMC has a survival of 85% at 5 years. Metastasis is more frequently local, followed by lung, liver, brain, and heart involvement. Survival after recurrence or with metastasis is 57% at 5 years⁽⁶⁾.

CLINICAL CASE

It's the clinical case of a 54-day-old infant, born at term, with no history, from Ancash. Referred to institution with diagnosis of right renal mass on prenatal ultrasound during third trimester. The mother is a young woman with no significant history or harmful habits.

In the physical evaluation of the patient, a mass was palpated on the right flank, neither mobile nor painful. The laboratories did not show alterations, nor did he present any other symptoms. The ultrasound confirmed a heterogeneous tumor with regular borders in the upper 1/3 of the right kidney. Figura 1a

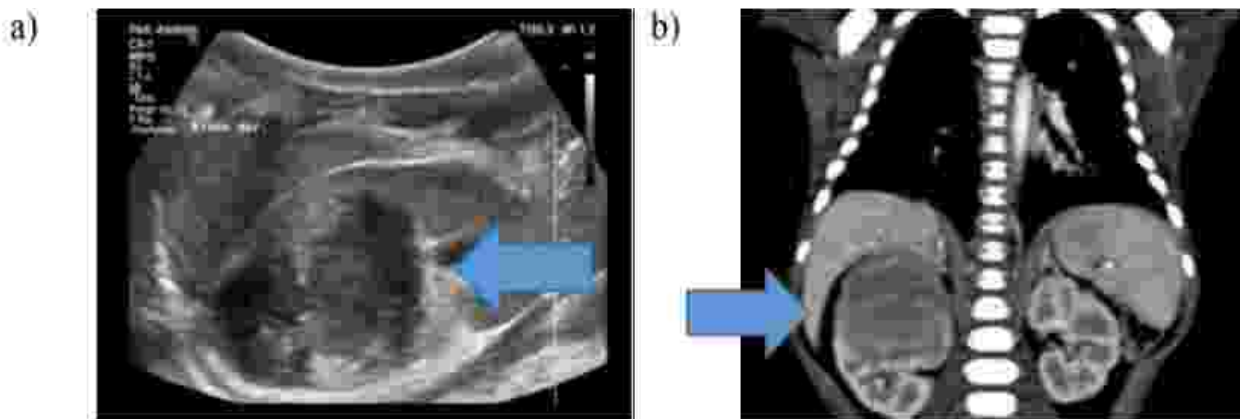
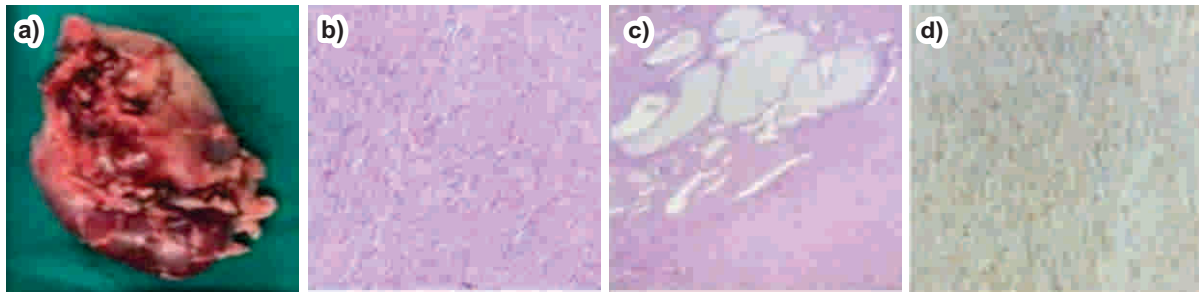


Figure 1. Imaging study: a) Renal ultrasound: heterogeneous tumor with regular borders in the upper 1/3 of the right kidney. b) Abdominal CT with contrast: expansive lesion of slightly heterogeneous density, 41mm x 35mm x 36mm.

A computed tomography scan was performed showing an expansive lesion of slightly heterogeneous density, located in the upper half of the right kidney, measuring 41mm x 35mm x 36mm (DL x DBH x SD). The mass was confined to the renal parenchyma, reaching the renal pelvis with mild pyelic ectasia and scant vascularity without significant contrast uptake or internal calcifications in the tumor. The right kidney presents an anatomical variant with two right renal veins. Figura 1b

She presented herself to the Institution's Oncology Committee and it was decided to perform a right radical nephrectomy, extracting the piece without complications, and she was discharged 9 days after surgery. The right kidney, right suprarenal gland and right ureter are received in the pathological anatomy department. The tumor measures 4.2cm x 3.9cm x 3cm, located in the upper 2/3 of the right kidney, gray-white in color, fibrous and myxoid in appearance, without necrosis or hemorrhage, representing 50% of the right renal volume.



a) Kidney with tumor in the upper pole and perirenal fat. b) Neoplasm with thin spindle cells arranged in intersecting fascicles. c) Presence of islands of cartilaginous tissue, glomeruli and spindle cells. d) Positive immunohistochemistry for smooth muscle actin.

Figure 2. Pathological study of renal tumor

Microscopy revealed CMN, a classic variant, with infiltration of the renal capsule, perirenal fat, renal pelvis, and Gerota's fascia. The renal vein, renal sinus, and surgical border of the ureter were free of neoplasia. Immunohistochemistry reports negative WT-1 and Desmin, positive actin and vimentin, and Ki-67 of 2%. Figure 2

The patient did not receive neoadjuvant or adjuvant chemotherapy and is controlled in an outpatient clinic, without presenting surgical complications or recurrence, with survival to the date of more than 3 years.

DISCUSSION

Approximately 5% of perinatal tumors arise from the kidney. CMN is the second most common tumor after Wilms' tumor during the first year and is the most common in the first 6 months of life⁽²⁾ as was the case of the patient with 2 months of life. Unlike those published by Geramizadeh, with patients aged between 18 months and 11 years⁽⁷⁾.

There are 3 histological types: classic (24%), cellular (66%), and mixed (10%) being the classic one with the best prognosis⁽⁴⁾. Intrauterine manifests with polyhydramnios and hydrops fetalis, associated with premature delivery and hypercalcemia⁽⁵⁾. The clinical picture presents as a palpable mass in the flank (31.8%), hematuria (27.3%), lumbar pain (22.7%), and hypertension due to increased renin⁽⁸⁾, it may present pulmonary hypertension and heart failure⁽⁹⁾.

It is more frequent in females⁽⁸⁾, differing from the results found by Pachl, where males predominated⁽¹⁰⁾. The differential diagnosis is with Wilms tumor, metanephric

stromal tumor, and clear cell sarcoma of the kidney, with a worse prognosis⁽¹⁾. Obstetric ultrasound allows intrauterine diagnosis⁽¹²⁾. While the computerized axial tomography of the abdomen allows to make the diagnosis, as well as to differentiate between the histological types⁽⁵⁾.

The initial treatment is surgical, except for those patients with a high suspicion of cellular histology and older than 3 months, where chemotherapy is indicated from the preoperative period⁽⁴⁾. Also, when the tumors are larger, chemotherapy allows a reduction, with less risk of intraoperative complications⁽²⁾. Adjuvant chemotherapy is indicated in patients with incomplete tumor resection or tumor rupture during excision, as well as in local recurrence and metastasis.

There are 3 histological variants of CMN: classic, cellular, and mixed. The classic type is characterized by being a solid and firm tumor with the presence of a capsule and fibroblastic cells, with low mitotic activity and abundant collagen deposition. The cell type with long hemorrhagic areas with a necrotic component, high mitotic activity, invasion of peripheral fat and connective tissue⁽¹¹⁾. In immunohistochemistry, CMN is positive for vimentin and smooth tissue actin, negative for desmin and Cd34⁽¹²⁾.

The pathological anatomy of the patient-reported classic-type histology with tumor-free surgical margins. Currently, the patient is free of disease, without recurrence 3 years after surgery. Unlike what was published by Jehangir, with a recurrence of up to 71%



and metastasis of 42% in patients with cell-type histology, this being a risk factor⁽¹³⁾.

Reported poor prognostic factors are age over 3 months, positive surgical margins, cellular histological variant, and tumor rupture during excision⁽¹⁴⁾. The patient did not present any of these factors.

CONCLUSIONS

ECMN is a low-incidence tumor, more frequent in

newborns and infants under 3 months. Good prognostic factors allow greater survival. The initial treatment is surgical, except for patients with risk factors.

Comprehensive evaluation, identifying warning signs and early intervention are factors that will favor survival in pediatric patients with solid abdominal tumors, which is why it is important to know this pathology.

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