

Congenital Mesoblastic Nephroma in a Newborn: Case Report

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Abstract: Background: Congenital mesoblastic nephroma (CMN) is a rare mesenchymal renal tumor of the newborn. The objective of this study was to review the clinical particularities, paraclinical and therapeutic characteristics of this tumor. It is about a case report of pediatric patient admitted to a Pediatric Neonatal Reanimation, A. Harouchi Hospital, Ibn Rochd university Hospital centre, Casablanca. Results: We report the observation of a 4-day premature newborn who was hospitalized for exploration of an abdominal mass occupying the right flank since birth. Abdominal ultrasonography revealed a predominantly fleshy right solidocystic renal mass, a site of calcifications laminating the cortex, measuring 57x33x58, with minimal pyelocaliceal dilatation and diffuse infiltration of perrenal fat. The CT scan revealed a kidney increased in size, seat of a voluminous mass rather well limited, of tissue density, heterogeneous, seat of necrosis, realizing a sign of the spur, measuring 40x47x58mm. The diagnosis was confirmed by a kidney biopsy. Conclusion: Mesoblastic nephroma is considered a benign tumor. The diagnosis is discussed on the basis of clinical data, ultrasound and CT scan. Surgery is the primary treatment for NMC and is based on extended nephrectomy. Most patients with NMC have a favorable outcome, the overall prognosis is very good.

Keywords: Renal Tumor, Congenital Mesoblastic Nephroma, Neonatology

1. Introduction

Congenital mesoblastic nephroma (CMN) is a mesenchymal tumor of the kidney, the most frequently occurring solid tumor in newborns and young children. It is a rare pediatric renal tumor with a peak incidence in the first three postnatal months, representing approximately 3% of all renal tumors in children, and is essentially benign. [1].

2. Case Report

A newborn female infant aged 4 days received with an abdominal mass that was found by the pediatrician on delivery at the postnatal visit.

She was delivered prematurely at 31 weeks to a 19 year old, Gravida 3 Para 2. An antenatal ultrasound reported polyhydramnios, she got premature rupture of membranes >18 hours (PPROM) and gave birth to a live female child who weighed 1000g by spontaneous delivery (SVD) and the child was reported to have cried right after birth.

The neonate had a heart rate of 141 beats per minute, tachypneic with a respiratory rate at 72 breaths per minute, and SPO₂ of 98% in room air, she was in moderate respiratory distress rated at 3/10 according to silverman's score, with a high blood pressure at 75/48 mmhg, without hematuria, without gastro-intestinal symptoms or signs of dehydration. She was afebrile at 36.6°C and the clinical malformative assessment was negative.

His abdomen was undistended with a mass palpable bimanually in right quadrant, flank and right iliac pit, measuring 6 × 3 cm. It was non-tender, fixe, solid, had regular edges and the skin covering it was mobile and normally colored.

The newborn benefited from an epicutaneous-cave catheter indicated in view of his low weight and the difficulty of taking a venous line, antibiotic therapy, a serum glucose infusion and enteral feeding by continuous gavage.

Blood investigations reported a haemoglobin of 16.7g/dl, white cell count of 6420/μL, with PNN at 2450/μL, platelets of 172000/μL. C - reactive protein was négatif at 0.3 mg/L.

The creatinine was 6.5mg/L, urea of 5/L and calémiá was at 113mg/L.

A chest X-ray was performed, showing a clear decrease in lung transparency with correct compliance.

Abdominal ultrasonography revealed a predominantly fleshy right solidocystic renal mass, a site of calcifications laminating the cortex, measuring 57x33x58mm, with minimal pyelocalicial dilatation and diffuse infiltration of perrenal fat.

The CT scan revealed a kidney increased in size, seat of a voluminous mass rather well limited, of tissue density, heterogeneous, seat of necrosis, realizing a sign of the spur, measuring 40x47x58mm.

At the top it represses the right liver and gallbladder without separation border with suspicion of dilatation of the intrahepatic bile ducts.

At the bottom it exerts a mass effect on the rest of the renal parenchyma responsible for minimal dilatation of the excretory tract associated with significant infiltration of perrenal fat, which may be related to tumor rupture.



Figure 1. CT scan revealed a kidney increased in size, seat of a voluminous mass rather well limited, of tissue density, heterogeneous, seat of necrosis, measuring 40x47x58mm.

A puncture biopsy was performed confirming congenital mesoblastic nephroma.

The histological examination concerns four biopsy cores measuring between 7 and 3 mm long. it corresponds to a renal parenchyma infiltrated by a tumor proliferation composed of intertwined bundles of fibroblastic cells with elongated nuclei with fine chromatin, an eosinophilic

cytoplasm. the tumor includes islets of renal parenchyma.

An immunohistochemical study was performed, AML took focal AML, WT1 took on most tumor cells. CD56 is negative.

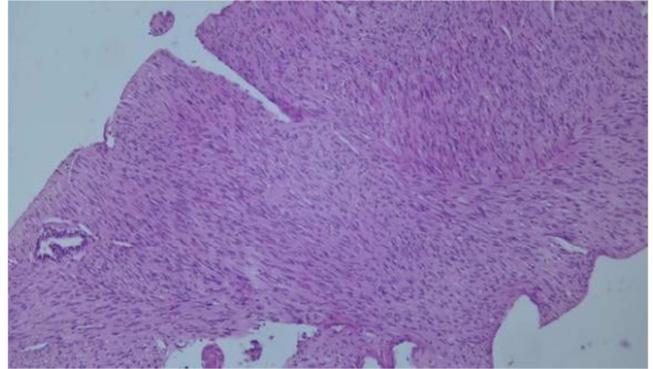


Figure 2. The tumor with hemateine eosin staining at different magnifications 20 times.

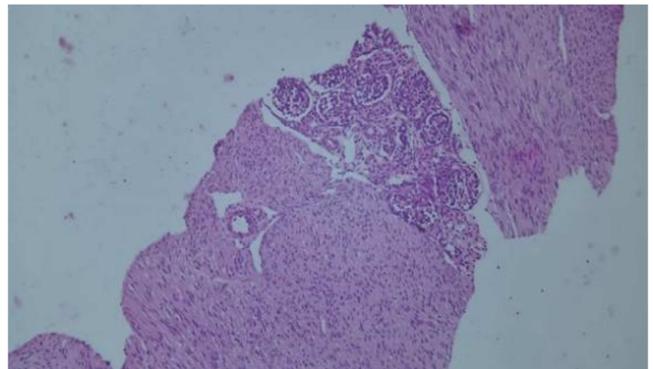


Figure 3. The tumor with hemateine eosin staining at different magnifications 40 times.

The patient's file was discussed with the multidisciplinary staff whose decision was to postpone the surgical treatment due to the benign nature of the tumor and the risk that the surgery may cause due to the small weight.

As no surgical specimen was available, since our patient died of hemorrhagic shock due to the tumor rupture.

3. Discussion

Congenital mesoblastic nephroma is the commonly occurring renal tumor in newborns. It is diagnosed in 90% of cases during the first year of life, usually between 3 and 6 months. [2, 3].

CMN generally manifests as an asymptomatic abdominal mass fortuitously discovered in the newly born infant in the nursery or subsequently by the parents. A less common presentation around or after birth is intratumoral hemorrhage or ruptured tumor with hemoperitoneum and shock, which requires immediate emergency surgery. Inconsistently, high blood pressure, polyuria, and occasionally hematuria, jaundice, and vomiting may be encountered.

Such as our patient, the mesoblastic nephroma was an incidental finding of an initially asymptomatic abdominal

mass, which later developed into hypertension.

Vido L. et al. had reported that tumor cells may generate excessive levels of prostaglandin E and induce hypercalcemia, which may lead to polyuria in the fetus. This is most likely the origin of polyhydramnios, which can induce prematurity and premature delivery such as our patient. [4].

Another study by Malone PS et al. reported that hyperreninism is the principal explanation for the hypertensive mechanism, which is due to increased renin production by afferent arterioles supplying trapped normal tumor glomeruli. [4].

The diagnosis of a renal tumor depends on the exclusion of a mass originating in the liver or adrenal gland. An abdominal ultrasound is the most appropriate initial examination to determine the location and nature of the mass.

CMN is usually discovered antenatally by ultrasound, and often presents as a well-delineated, homogeneous, hypoechoic unilateral mass. For the determination of the origin and morphology of the tumor, magnetic resonance imaging (MRI) is the appropriate paraclinical examination for identification.

In most cases, cross-sectional imaging will also be needed to determine the location of the tumor, local extension, and damage to the surrounding structures. MRI will be the investigation of choice because of its multiplanar properties, high sensitivity to hemorrhage and cystic lesions, and no radiation exposure. In cases of suspected neuroblastoma, MRI can be helpful in assessing tumor extension into the neural foramen or spinal canal. It also has the advantage of clearly differentiating the border of the tumor from normal kidney and suprarenal gland tissue. The major limitation of MRI examination at this age is often the necessity of sedation or general anesthesia. [5, 6].

Our patient could not have MRI because of the difficulty of performing it in the hospital and its high cost.

CT, especially with multi-slice volumetric acquisition, can be an alternative to MRI. The fast examination time allows to avoid the use of sedation [6].

CT scan usually reveals a large, homogeneous, solid intrarenal mass with smooth borders that enhances less than the surrounding normal kidney parenchyma after the administration of intravenous contrast product. [7, 8].

Pathologically, three variants of CMN exist: the classical one is present in 24% of cases, while the cellular variant is observed in 66% of cases and the mixed variant is the least common subtype. [9].

The congenital mesoblastic nephroma always contains in its mesoblastic substance foci of cystic, dysplastic or immature renal tubules.

Sometimes foci of immature and disorganized nephroblastic epithelium can be observed. It is this mixed character that may suggest an origin in the embryonal nephric blastema, somewhat like Wilm's tumor.

The classic type is characterized by a solid, firm, whorled, yellowish/whitish appearance similar to that of a leiomyoma. [3].

Histologically it resembles infantile fibromatosis characterized by spindle cells arranged in fascicles. Abnormal metaplastic changes are seen in the tubules or glomeruli trapped by the lesion, with reduced mitotic activity and absence of necrosis. [10].

In contrast, the cellular variant presents macroscopically with a multiseptated appearance with hemorrhage, necrosis, and cystic areas, while histologically it presents with high cellularity, mitosis, necrosis, hemorrhage, and consists of solid ovoid or spindle-shaped cells with reduced cytoplasm. [10].

In our case, it was not possible to specify the type of mesoblastic nephroma since the surgical specimen could not be obtained due to the death of the patient just after puncture biopsy by hemorrhagic shock.

CMN is generally considered as a benign tumor but recurrence locally and even metastasis remotely are described in 5–10% mainly affecting the lung, liver, bone or brain.

This aggressive tendency is, however limited to cellular and mixed subtypes of CMN and, therefore, adjuvant chemotherapy is recommended.

Surgical treatment is mandatory and is the first-line treatment for all patients with CMN.

In order to achieve complete resection, it is mandatory to remove the kidney, including the entire adipose capsule.

For that reason, partial nephrectomy is not indicated.

A positive tumor margin microscopically/macrosopically is not an evidence to start chemotherapy, which should be reserved for relapsed unresectable neoplasms. Nevertheless, a second surgery excision needs to be considered as the mainstay of treatment, even in the relapsed situation.

The decision on chemotherapy is a difficult one given the relatively Radiation therapy can be considered, but its short-, medium-, and long-term effects must be carefully evaluated on an individual basis. [11].

Most patients with CNM have a favorable outcome, the overall prognosis is very good. [12].

4. Conclusion

CMN is a rare benign tumour to be evoked in front of any solid kidney tumors in the first few months of life and to differentiate from nephroblastoma. Its character non-aggressive leads to a treatment exclusively surgical procedure that leads to healing.

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