Stage III Atypical Cellular Mesoblastic Nephroma

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A six-week-old female infant presented with acute progression of abdominal distension and sudden pallor. Abdominal CT scan revealed large renal mass. The patient underwent total nephrectomy. The pathologic features and immune staining confirmed the presence of atypical cellular congenital mesoblastic nephroma. The patient died two months later because of chemotherapy toxicity.

Bahrain Med Bull 2016; 38 (3): 171 - 172

Congenital mesoblastic nephroma (CMN) is a rare tumor. It accounts for less than 5% of renal tumors in children. It arises from the proliferating nephrogenic mesenchyme. The majority of fetuses with CMN would be the product of polyhydramnios and 25% are premature[1-4]. Neonatal hypertension, hematuria and hypercalcemia are clinical features associated with this tumor[5-9]. It has three variants: the classic form, the cellular or atypical form and the mixed kind[1,6-13]. The classic form has no capsule, minimal cellularity and mitosis, no necrosis and no hemorrhage and has very excellent prognosis. The cellular or atypical form has a high mitotic index, significant necrosis and hemorrhage. The third kind is the mixed variant[1-6,8-11]. The causes of the tumor are unknown because it is rare. The cellular form has been reported in two infants who were the product of in-vitro fertilization[7].

The aim of this report is to present a rare case of mesoblastic nephroma.

THE CASE

The patient was 6-weeks old at presentation. She was born at term by cesarean section due to breech presentation; there was no history of polyhydramnios. The patient's mother had always observed abdominal distension of the baby; however, on the day of presentation, the patient had a significant increase in the size of the abdomen and became pale, listless and tachycardic. The blood pressure was high 100/60. Hemoglobin was low at 7.8 g/dl (normal 10-14.5 g/dl). The patient received a blood transfusion and antihypertensive therapy. The abdominal ultrasound revealed large heterogeneous right upper quadrant mass measuring 9.7 cm x 7 cm. Abdominal and thoracic CT studies revealed a large iso to hypodense infrahepatic soft tissue and encapsulated mass arising from the right kidney. The mass was 9.7 x 8.2 x 7.6 cm (Slx Trans x AP) with an evidence of intra-tumor hemorrhage and suspicion of intra-abdominal hemorrhage.

There was no evidence of distant metastasis, see figure 1 (A and B). The patient had laparotomy and total nephrectomy. The tumor had ruptured in the abdominal cavity. Histopathologically, the tumor was highly cellular and composed of spindle cells with bland nuclei that are arranged in fascicles with mitosis (25-30/10hpf) with large areas of necrosis and hemorrhage. The immunostaining was negative to Desmin, SMS, AE1, AE3, positive to vimentin and negative to WT-1. The histologic findings and immunostaining confirmed the diagnosis of atypical or cellular congenital mesoblastic nephroma.

The tumor was staged III. The patient was followed by a pediatric oncologist for chemotherapy.

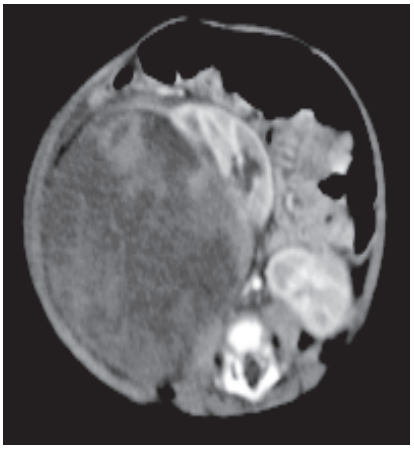

Figure 1 (A): Coronal CT Revealed Large Mass with Moderate Amount of Slightly Dense Fluid in the Abdomen and Pelvis Suggestive of Hemorrhage. No Evidence of Calcification

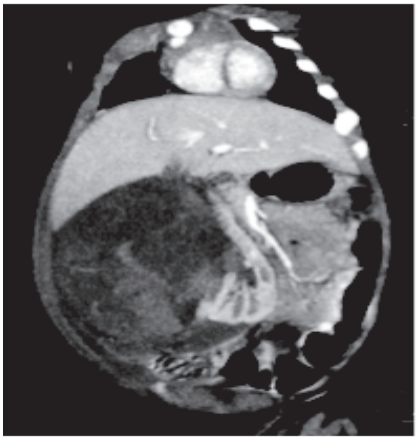

Figure 1 (B): Axial Section of Abdominal CT Scan Shows a Large Encapsulated Iso to Hypodense Right Infrahepatic Soft Tissue Mass Arising from the Medial Aspect of the Right Kidney Just below the Hilum

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DISCUSSION

Congenital mesoblastic nephroma (CMN) is a rare pediatric tumor[1-4,12,13]. It is reported mainly in infants less than six months of age and rarely diagnosed beyond the age of three years. The most common presentation is asymptomatic abdominal mass[1-4]. Hypertension could be an associated finding due to an increase in renin secretion from renal tissue infiltration[1-3,10-13]. This tumor could be suspected in-utero on ultrasonographic examination. Seventy-one percent of gestations are associated with polyhydramnios[1-4]. The atypical or cellular variant could metastasize to adjacent structures and tends to recur[1-4,13]. All three variants of CMN immune react to fibroblastic markers and do not react to epithelial markers. The curative treatment for the classic variant of the congenital mesoblastic nephroma is total nephrectomy, while the atypical needs adjuvant therapy, such as chemotherapy to reduce the possibility of metastasis and recurrence[1-4,13]. The 5-year-survival rate of the cellular variant is 85%, and the overall survival rate is 90%[4].

Our patient had the classic presentation of abdominal distension and also had hypertension. The tumor was a cellular variant, which is an aggressive type compared to the classic variant. The tumor was found ruptured during the resection, which made the administration of chemotherapy essential to reduce the likelihood of recurrence.

This case should add to our limited knowledge about a rare congenital mesoblastic nephroma because the infant had cellular form and was a product of normal conception rather than in-vitro fertilization. Chemotherapy risk, morbidity, and mortality should be considered at an early age.

CONCLUSION

Cellular mesoblastic nephroma is a rare but aggressive congenital renal tumor. The index of suspicion should be high in a patient who presents in the neonatal period with a renal mass, especially in the presence of gestational polyhydramnios.

Author Contribution: All authors share equal effort contribution towards (1) substantial contribution to conception and design, acquisition, analysis and interpretation of data; (2) drafting the article and revising it critically for important intellectual content; and (3) final approval of manuscript version to be published. Yes.

Potential Conflicts of Interest: None.

Competing Interest: None.

Sponsorship: None.

Submission Date: 15 April 2016.

Acceptance Date: 26 June 2016.

Ethical Approval: Approved by the Department of Pediatrics, Salmaniya Medical Complex, Bahrain.

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