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Cystic neuroblastoma in a newborn: A case report

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An abdominal cystic mass with a 2.5x1.8 cm diameter was diagnosed in a 28 week- gestation female fetus using antenatal ultrasonography (US). Postnatal US confirmed a cystic mass in the right adrenal region. The abdominal computed tomography revealed a hypodense, thick walled cyst which pushed the right kidney to the lower side. Intraoperatively, the mass appeared to be within the right adrenal gland, and a right adrenalectomy was performed. Histology confirmed a poorly differentiated neuroblastoma.
Cystic neuroblastoma (CNB) should be considered in the differential diagnosis of suprarenal cystic masses. It can be suggested that any cystic adrenal mass with characteristic US sign (thick wall) in a neonate must be removed since there is a probability of malignancy.

Key Words: Neuroblastoma, Neonate, Cystic, Prenatal

Bir Yenidoğanda Kistik Nöroblastoma: Olgu Sunumu

Yirmisekiz haftalık kız fetüste antenatal ultrasonografide (US) 2.5x1.8 cm çaplı abdominal kistik kitle saptandı. Postnatal yapılan US de kistik kitlenin sağ adrenal bölgede olduğu görüldü. Abdominal kompüterize tomografide hipodens, sağ böbreği aşağı iten kalın duvarlı kist mevcuttu. Ameliyatta kitlenin sağ adrenal glandda olduğu görüldü ve sağ adrenalektomi yapıldı. Histolojik olarak kitle az diferensiye nöroblastoma olarak değerlendirildi.
Suprarenal kistik kitlelerin ayırıcı tanısında kistik nöroblastoma (CNB) düşünülmelidir. US de karakteristik olarak kalın duvarlı kistik kitlesi olan bir yenidoğanda kitlenin malign olabilme ihtimaline karşı çıkarılmasını öneriyoruz.

Anahtar Kelimeler: Nöroblastoma, Yenidoğan, Kist, Prenatal

Neuroblastoma is the most common neonatal solid suprarenal tumor in infants less than 1 year old, however, CNB is exceedingly rare. There are only 50-60 CNB cases in the world literature.[1-4] Many neonatal intra-abdominal cystic lesions are increasingly recognized with the use of antenatal US.[5-8] We report a cystic neuroblastoma with antenatal diagnosis and early treatment.

CASE REPORT

An abdominal cystic mass with a 2.5x1.8 cm diameter was diagnosed in a 28 week- gestation female fetus using antenatal US. Postnatal US confirmed a cystic mass of 3x2.5 cm in diameter in the right adrenal region. The abdominal computed tomography (CT) revealed a hypodense, thick walled cyst which pushed the right kidney to the lower side. There wasn't calcification (Figure 1). Results of hematological and biochemical tests were normal. Vanilmandelic acid (VMA) and Homovanillic acid (HVA) levels in the 24-hour urinary collection were within normal range. Postnatal US performed again on the tenth day, it was seen that the diameter of the cystic mass was not changed. Intraoperatively, the mass appeared to be within the right adrenal gland, and a right adrenalectomy was performed. Histology confirmed a poorly differentiated neuroblastoma, and tumor was completely confined within the adrenal gland (Evans stage 1) (Figure 2). Immunohistochemically, S-100 protein staining was negative, neuron-specific enolase (NSE) and synoptophysin staining were positive. N-myc oncogene expression and DNA flow cytometry couldn't be determined.

Postoperative course was uneventful. The patient was discharged on the postoperative day 4 and she was sent to the pediatric oncology department for further workup.

Fig 1 Computed tomography scan of the abdomen shows a cystic mass in the right adrenal region

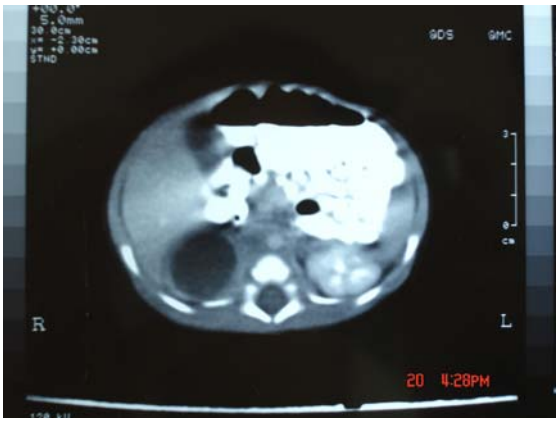
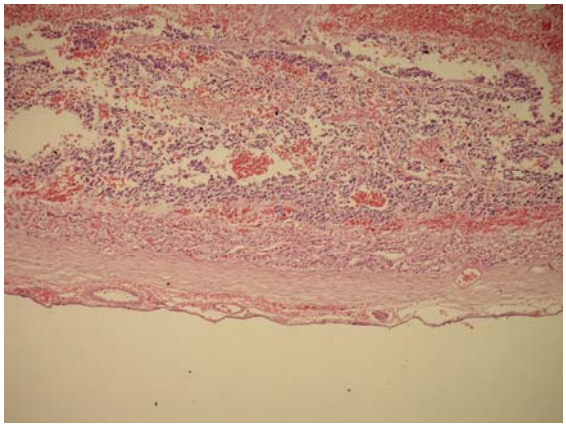


Fig 2 Under the adrenal capsule, and neuroblastoma showing small, undifferentiated cells



DISCUSSION

Cystic neuroblastoma is an uncommon variant of neuroblastoma. Other neonatal suprarenal cystic masses are adrenal hemorrhage, bronchogenic cyst, cystic hygroma or hydronephrosis.[7,9] Most fetal neuroblastomas are incidental findings on obstetrical sonograms; there are occasional reports of maternal and/or fetal symptoms and signs that appear to be either direct or indirect consequences of the presence of a neuroblastic tumor. Maternal symptoms of catecholamine excess are nervousness, sweating, vomiting, flushing, headache, and/or weakness. Frank preeclampsia is associated with widely disseminated fetal neuroblastoma.[8] In our patient, there weren't any maternal symptoms.

Because of the high sensitivity of routine prenatal US and the growing use of US to examine neonates, the discovery of a suprarenal mass before or a few months after birth is increasingly common.[5-8] US shows the size of the mass, the presence of calcification, echogenicity (cystic, solid or heterogeneous), shape (round or triangular) and the characteristics of the connection with the adrenal gland and the kidney in the abdominal masses. The diagnosis of cystic masses with prenatal US is easier than a solid mass. There are masses consisting of mixed solid and cystic components, with the neuroblastoma isolated in the solid component. These lesions also have been described as CNBs. It is postulated that such cysts are formed from degenerative changes within a neuroblastoma.[5] Petit T et al reported that thick wall complex cyst may be neuroblastoma.[1] Similarly, in our patient, the cyst had a thick wall. CNB must be differentiated from adrenal hemorrhage, which is the most common cause of adrenal mass in the newborn, with an estimated incidence of 1.9/1000 live births. Among reliable signs of adrenal hemorrhage are increasingly heterogeneous internal echoes, the late appearance of calcifications and a continuous decrease in size on serial US.[7] Masses identified by prenatal US need careful evaluation as they may represent normal structures, nonsignificant variants, or physiologically significant anomalies. For this reason, postnatal US examination is fundamental for diagnosis. CT and magnetic resonance imaging are highly contributory to the diagnosis of suprarenal masses. On the other hand, the exact diagnosis of a suprarenal mass is impossible without an operation.

Fetal neuroblastoma is normally diagnosed during the third trimester at an average age of 33 weeks, although there are reports of sonographic diagnosis as early as 23 weeks.[10] Adrenal hemorrhage usually occurs at birth or during the first days of life and is favored by obstetric trauma, infection, and hemodynamic or coagulation disorders.[6] However, adrenal hemorrhage may be observed during the second or third trimester of pregnancy.[7] Subdiaphragmatic extralobar pulmonary sequestrations appear during the second trimester as solid, brightly echogenic masses. Color flow Doppler examination can sometimes detect the systemic arterial branch projecting from the thoracic aorta. Several entities included in the differential diagnosis of a cystic mass derive from the kidney, including renal cyst, obstructed upper pole duplication, and cystic Wilm's tumor.[8]

Solid neuroblastoma with elevated VMA, HVA, and NSE are functioning in 70% of cases.[10] Most cystic neuroblastomas are nonfunctioning tumors. Urinary catecolamines values are negative in some prenatally detected cases of neuroblastoma.[2,6] Similarly, the results of 24-hour urine collection for VMA and HVA were negative in our patient. The majority of perinatal neuroblastoma cases are stage 1 or 2 tumors with favorable biological features, which correlates with a 4-year survival of greater than 95 % .[3,8]

Cystic adrenal neuroblastoma is highly unusual. Richards et al. reviewed of the world literature; they identified 31 cases of CNB, including 20 prenatally diagnosed cases.[4] Hamada et al. reviewed of the world literature; they identified 25 cases of prenatally diagnosed CNBs.[1] In Hamada and Richard's reviews, 5 cases are the same patients. Only a few reports of CNBs are found in the literature after 1999.[1,3,4]

Cystic neuroblastoma should be considered in the differential diagnosis of suprarenal cystic masses in a neonate. Prenatal routine US examination is necessary for early determination of any malignancy and diagnosis of other abnormalities. It can be suggested that any cystic adrenal mass with characteristic US signs (thick wall) in a neonate must be removed even though the mass size is not increased. Therefore, probability of the neonatal excision of the cystic neuroblastoma is increased and patient's survival may be improved.

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