J Korean Radiol Soc 1999;40:325-328

Prenatally Diagnosed Cystic Neuroblastoma : A Case Report[1]

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Cystic masses of the adrenal gland are unusual in the fetus and most are secondary to hemorrhage. Cystic neuroblastoma is extremely rare, and in contrast to solid neuroblastoma, follows a benign course, is diagnosed earlier, and rarely presents with metastatic lesions(1-4). We report one case of cystic neuroblastoma diagnosed prenatally by ultrasound (US) and magnetic resonance (MR) imaging, and include a review of the literature.

Index words : Adrenal gland, neoplasms
Adrenal gland, MR
Adrenal gland, US
Neuroblastoma

Obstetrical US can diagnose many types of congenital malformations and some tumors before birth. Neuroblastoma is an embryonic tumor of sympathetic origin and one of the most common malignant solid tumors to occur during infancy. Cystic neuroblastoma is extremely rare; only 29 cases have been reported in the medical literature. It is difficult to differentiate from adrenal hemorrhage, adrenal cyst, renal cyst, cystic Wilm's tumor, obstructed upper pole duplication anomalies with ectopic ureteral implantation, hepatic cysts, choledochal cysts, and ovarian or enteric cysts (1, 5, 6).

Case Report

A 30-year-old woman (gravida 1, para 0) was referred for sonographic evaluation at 37 weeks gestation because of a fetal intra-abdominal mass.

US examination(Fig. 1) demonstrated a 5.5×5cm-sized, anechoic mass with internal thin septa superior to the right kidney, which the mass had displaced inferiorly. No solid component in the mass was demonstrated. In addition, a purely cystic mass in the right suprarenal area was detected by prenatal MR imaging (Fig. 2A, B), with low signal intensity on T1-weighted images and high signal intensity on T2-weighted images. Hematoma was excluded. A 3500-gm female baby was delivered by Cesarean section at 39 weeks gestation; labor and delivery proceeded without complication. At birth, physical examination revealed a palpable abdominal mass, and US examination on the first postnatal day showed little interval change. Hepatic metastasis was not demonstrated. A 24-hour urinalysis

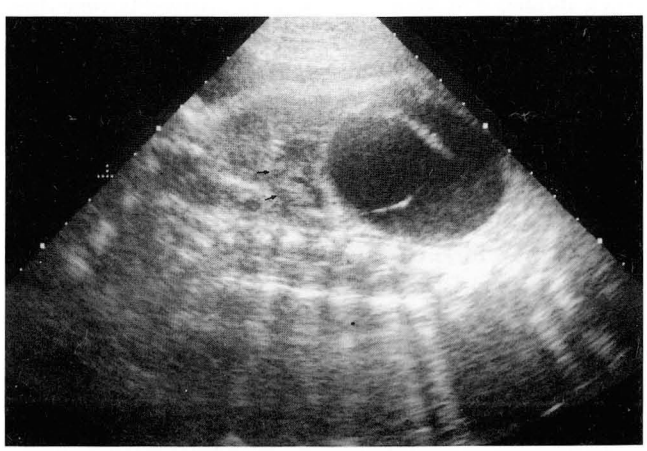

Fig. 1. Longitudinal fetal sonogram at 37 weeks gestation shows well-defined anechoic mass with internal septa superior to the right kidney(arrows).

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Received September 7, 1998; Accepted November 16, 1998
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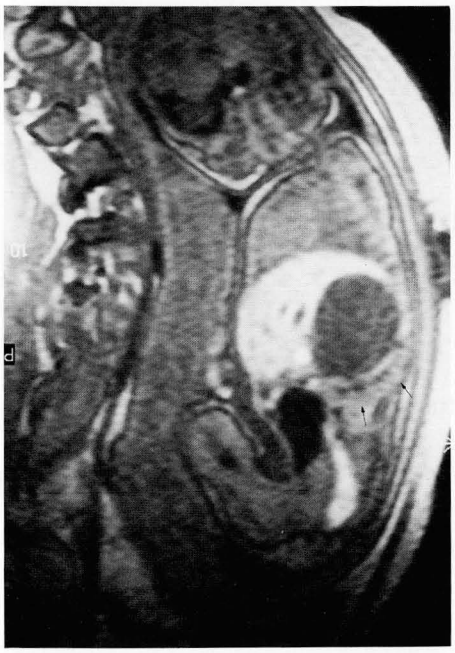
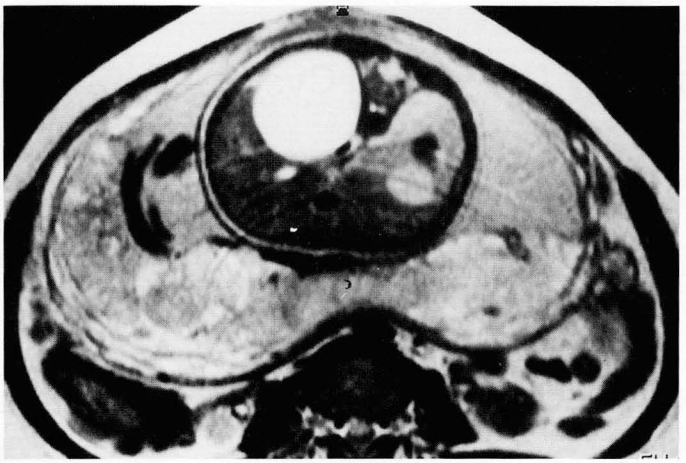


A



B

Fig. 2. Prenatal MR images at same gestational age.
Sagittal T1(**A**)-and axial T2(**B**)-weighted images show purely cystic mass. Right kidney (arrows) is inferiorly displaced.

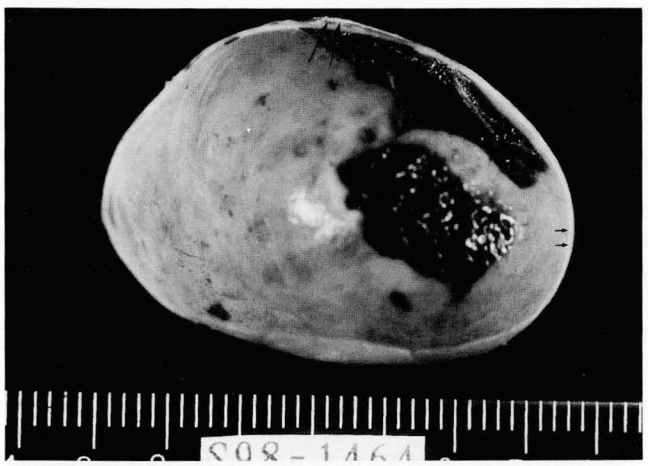


Fig. 3. Gross specimen of the mass.
Cystic mass shows focally shallow blood clots and smooth inner surface. The wall of cyst (small arrows) is very thin. Adrenal remnants (long arrows) is stretched around the cyst.

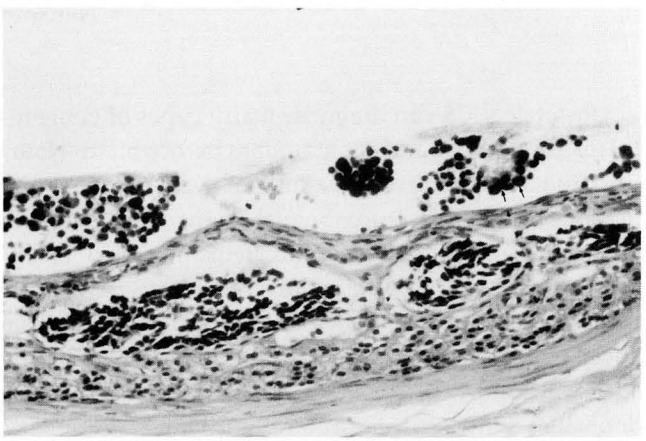


Fig. 4. Microscopic examination(H and E stain, × 200).
The cyst wall shows narrow rim of neuroblastoma at the top and residual cortex below it. A Homer-Wright rosette (arrows) is noted.

for vanillylmandelic acid (VMA) was within normal limits. A repeat US examination 20 days later revealed no significant change in the size of the mass. The infant underwent a total excision of the mass at age 3 weeks, and surgical exploration revealed a 7.0×5.0×4.5 cm cystic adrenal mass. On sectioning(Fig. 3), it was found to be cystic and contained yellowish serous fluid. The cyst wall was fibrotic, less than 1mm thick, and covered with focally shallow blood clots. On pathologic examination(Fig. 4), a cystic neuroblastoma was found to be present. It was considered to be at stage I, the patient underwent no further therapy. At the most recent follow-up at age 4 months, she was doing well, with no evidence of disease.

Discussion

Neuroblastoma is a tumor of the postganglionic sympathetic neurons and is the most common extracranial solid tumor found in children. In half of childhood neuroblastomas, the tumor arises in the adrenal gland. Hemorrhage and necrosis are often seen in neuroblastoma, but cyst formation is uncommon. Purely cystic lesions have been reported in fetal neuroblastoma, but

are rare during infancy(1, 2, 5). In contrast to solid neuroblastoma, the cystic variety follows a benign course, is located almost exclusively in the adrenal gland, is diagnosed earlier, and rarely presents with metastatic lesions. Calcification is rare. Most cystic neuroblastomas are nonfunctioning, and only 9.5% have shown documented elevation of VMA or homovanillic acid (HVA) levels. A majority of patients have favorable biologic features with hyperdiploid cellular DNA content and no amplication of N-myc oncongene. In contrast, solid neuroblastomas with elevated VMA, HVA and neuron-specific enolase occur in 70% of cases.

Because of their characteristics of low stage disease and favorable biologic features(2, 5, 7), purely cystic neuroblastoma may indicate a favorable postnatal prognosis. Cystic neuroblastoma is often diagnosed incidentally through prenatal sonographic studies(2). On prenatal US examination, cystic neuroblastoma may appear as a mass which is either complex and echogenic, or anechoic. The liver is the most common site of metastasis, though metastatic lesions are rare. Skeletal involvement is very rare(8). When US findings are negative or equivocal, computed tomography and MR imaging may be better in defining liver metastases, and MR imaging is the method of choice for distinguishing adrenal hemorrhage(4). Chen CP et al.(5) reported that on initial imaging, 15 of 21 cases of congenital adrenal cystic neuroblastoma were purely cystic while six were mixed. Cyst formation in association with neuroblastoma may be related to hemorrhage and necrosis of a tumor(1, 2, 5). Cyst content has been described as serous, gelatinous, or hemorrhagic, and in our case prenatal US and MR imaging showed a purely cystic mass.

Most reported cases have been diagnosed after 32 weeks of gestation. The differential diagnosis of suprarenal cystic masses in fetuses includes adrenal hemorrhage, adrenal cyst, obstructed upper renal moiety of a duplex kidney, cystic Wilms tumor, renal cysts, mesoblastic nephroma and multilocular cystic nephroma(3, 4, 7). Adrenal hematoma is the most common cause of a suprarenal fetal mass. Because of their relatively large size and vascularity in utero, the fetal adrenal glands are susceptible to hemorrhage. Adrenal hematoma during its natural history may be sonographically entirely echogenic, mixed echogenic or anechoic when first imaged, but its texture will gradually evolve and become more cystic and echolucent on follow-up US examinations(1, 5). Fetal adrenal cysts are extremely rare. Although cystic neuroblastoma is a rare form of fetal neuroblastoma, it should be considered in the differential diagnosis of a cystic adrenal mass(1).

Cystic neuroblastoma shows a high rate of spontaneous regression, and prior to excision, a period of close observation is thus reasonable; surgery can sometimes be avoided(1, 4, 5). Surgical excision may be indicated for tumors that do not become smaller on follow up examination, that become more complex after resolution of the cyst, that have poor biologic features, and that demonstrate sonographic evidence of hepatic metastasis on diagnosis or later(1, 4, 5). For children with prenatally diagnosed neuroblastoma, Acharya et al.(5) recommended observation for several months, and close follow-up with monthly sonograms, especially for tumors that are small(<3 cm) and cystic.

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대한방사선의학회지 1999;40:325-328

산전에 진단된 낭성 신경모세포종 : 1예 보고[1]

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태아기에 부신에서 기원하는 낭성종양은 흔하지 않다. 가장 흔한 것은 부신의 출혈에 동반된 이차적인 낭성변화이다. 낭성 신경모세포종은 신경모세포종의 드문 형태이다. 고형의 신경모세포종과는 달리 낭성 신경모세포종은 거의 대부분 부신에서 기원하고 보다 양성의 경과를 취한다. 또한, 고형의 신경모세포종보다 일찍 진단되어지고 다른 장기로의 전이는 드물다(1-4). 저자들은 산전초음파와 산전 자기공명영상에 의해 진단되어진 낭성 신경모세포종을 1예 경험하였기에 문헌고찰과 함께 보고하고자 한다.