Neonatal Adrenal Hemorrhage Presenting as a Multiloculated Cystic Mass

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Neonatal adrenal hemorrhage presenting as an abdominal mass in the newborn is not uncommon. However, judging the nature of a suprarenal mass is sometimes difficult, especially when the structure is more complex with unusual clinical course. We report a male newborn with neonatal adrenal hemorrhage presenting as a multiloculated cystic mass. The margins between the mass and left kidney were indistinct. All laboratory data including complete blood cell count, serum electrolytes, liver function, renal function, blood sugar, α-fetoprotein, β-human chorionic gonadotropin, urinalysis, and 24-hour urine vanillylmandelic acid were within normal limits. Serial sonographic follow-up revealed failure to decrease in size without change in echogenicity. Surgical exploration was performed to exclude the possibility of malignancy. This case highlights the diagnostic problems that arise when a space-occupying lesion is found near or at the adrenal gland in the neonate. We suggest that early surgical intervention for the suprarenal mass without sufficient evidence of malignancy would not be prudent. [*J Chin Med Assoc* 2008;71(9):481–484]

Key Words: adrenal hemorrhage, neonate, suprarenal mass

Introduction

Neonatal adrenal hemorrhage (NAH) is not uncommon. The reported incidence ranges from 1.9 to 5.5 per 1,000 newborns detected by ultrasonographic screening.[1,2] The relatively large size and increased vascularity of the adrenal glands in the newborn may result in vulnerability to mechanical compression and sensitivity to changes in venous pressure during delivery. Also, any factor leading to hypoxia may result in redistribution of blood toward the central nervous system, heart, and adrenal glands. The increased pressure associated with congestion and the damaged endothelial cells associated with hypoxia may cause adrenal hemorrhage.[3] The clinical presentation is variable; infants may be asymptomatic, with the diagnosis made incidentally. The most common clinical symptoms are poor feeding, vomiting, persistent jaundice, anemia, and abdominal mass. Judging the nature of a suprarenal mass is sometimes difficult, especially when the structure is more complex with unusual clinical course. We report a case of NAH presenting as a multiloculated cystic mass. Surgical exploration was performed to exclude the possibility of malignancy.

Case Report

A male infant was born after 39 weeks' gestation to a gravida 2 para 2 woman with an uneventful pregnancy. He was delivered vaginally without instrumental assistance at a private clinic; his birth body weight was 3,450 g, and Apgar scores were 9 and 10 at 1 and 5 minutes, respectively. An abdominal mass was palpated in the left flank, and bile-stained vomiting was noted. Indirect hyperbilirubinemia developed gradually, with a peak bilirubin level of 17.1 mg/dL on the 3rd postnatal day, which was treated with phototherapy.

He was transferred to our hospital on the 8th postnatal day with the following vital signs: blood pressure 80/52 mmHg, temperature 36.7°C, pulse 140 beats/minute, respiratory rate 35/minute. Laboratory investigations showed: white blood cell count 19,400/mm^3; hemoglobin 16.6 g/dL; platelets 541,000/mm^3; Na^+ 142 mmol/L; K^+ 5.1 mmol/L; Cl^- 107 mmol/L; alanine aminotransferase 28 U/L; aspartate aminotransferase 46 U/L; blood urea nitrogen 7 mg/dL; creatinine 0.4 mg/dL; total bilirubin 12.2 mg/dL; direct bilirubin 0.8 mg/dL; blood sugar 70 mg/dL.

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Also, α-fetoprotein, β-human chorionic gonadotropin, urinalysis, and 24-hour urine vanillylmandelic acid were within normal limits. The infant was normotensive and did not exhibit signs of adrenal insufficiency or bleeding tendency.

Abdominal ultrasound showed a multiloculated cystic mass, 4.5 × 3.3 cm in size, at the upper pole of the left kidney. The margins between the mass and the left kidney were indistinct (Figure 1). Abdominal computed tomography (CT) revealed a well-defined cystic lesion measuring about 5.0 × 4.3 × 5.0 cm with internal septa over the left suprarenal region; this mass caused anterior displacement of the pancreatic tail and body, spleen, and stomach (Figure 2).

Serial sonographic follow-up revealed failure to decrease in size without change in echogenicity. Surgical exploration was performed to exclude the possibility of malignancy on the 18th postnatal day. Histopathologic examination revealed adrenal cortical tissue surrounding a hematoma; calcification and fibrosis were also noted. This was compatible with adrenal hemorrhage. After 6 months of follow-up, the infant was healthy, with normal growth and developmental milestones.

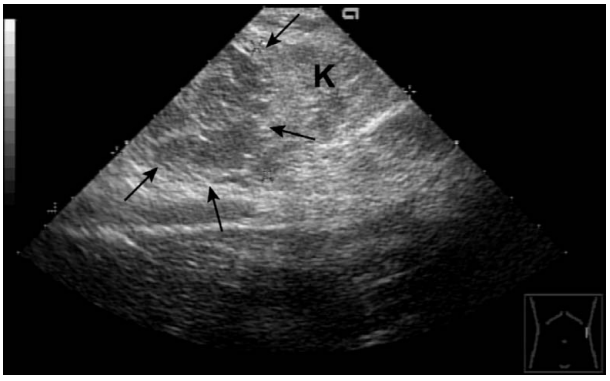

Figure 1. Abdominal ultrasound shows a multiloculated cystic mass, 4.5 × 3.3 cm in size, at the upper pole of the left kidney. The margins between the mass and left kidney are indistinct. K = kidney.

Discussion

NAH is a well-known clinical entity. Predisposing factors include birth trauma, prolonged labor, hypoxia, asphyxia, shock, septicemia, and hemorrhagic disorders. However, spontaneous occurrence is also recognized in otherwise normal neonates. The right adrenal gland is involved 3–4 times more than the left due to its greater likelihood of compression between the liver and spine. Since the right adrenal vein usually drains directly into the inferior vena cava, compression is likely to induce venous pressure changes.[4] The clinical presentation is variable; infants may be asymptomatic, with the diagnosis made incidentally. The most common clinical symptoms are poor feeding, vomiting, persistent jaundice, anemia, and abdominal mass. NAH is usually self-limited with resolution.

Differential diagnoses of cystic lesions near or at the adrenal gland include adrenal hemorrhage, adrenal cyst, adrenal abscess, neuroblastoma, pulmonary sequestration, bronchogenic cyst, enteric cyst, splenic cyst, and cystic lymphangioma. Some cysts arising from the upper pole of the kidney may be similar in the image investigation, including duplication of the renal collecting system, hydronephrosis, multicystic dysplastic kidney, Wilms' tumor, and cystic nephroma.[5] In neonates, ultrasound is the preferred modality for both the initial screening and the follow-up evaluation because

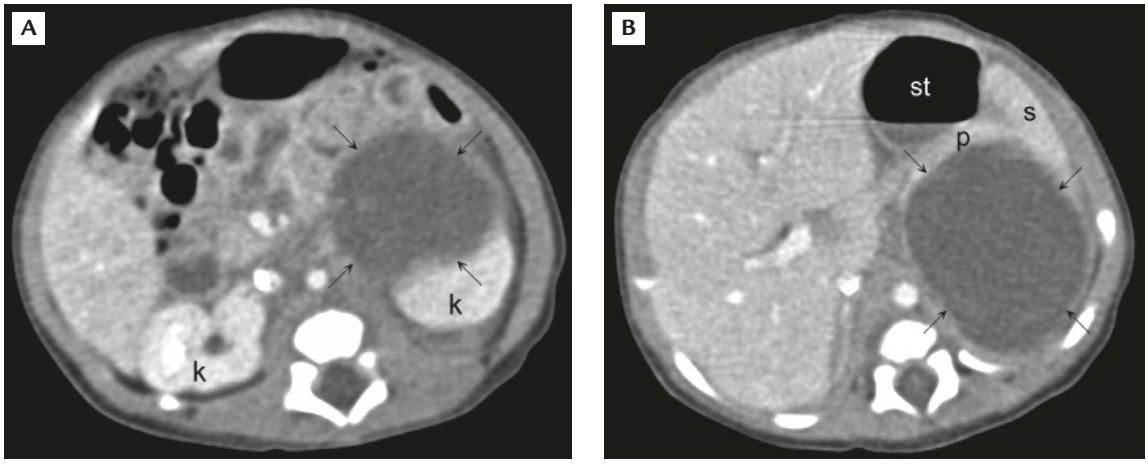

Figure 2. (A) Abdominal computed tomography shows a well-defined cystic lesion (arrows), measuring about 5.0 × 4.3 × 5.0 cm in size, with internal septa over the left suprarenal region. (B) This mass (arrows) caused anterior displacement of the pancreatic tail and body, spleen, and stomach. k = kidney; p = pancreas; s = spleen; st = stomach.

it is portable, rapid, sensitive, noninvasive, and free from ionizing radiation.

The ultrasound images are different at different stages. In the early stage, adrenal hemorrhage appears solid and echogenic. As liquefaction occurs, the mass demonstrates mixed echogenicity with a central hypoechoic region, and gradually, cystic change occurs. Calcifications may be seen as early as 1–2 weeks after onset. Sometimes, a complex picture may be found due to the coexistence of necrotic tissue, blood clots, calcifications, and cysts. Eventually, it resolves completely and becomes anechoic about 2 months later.[6]

Since neuroblastoma is a relatively frequent neonatal malignancy, it is certainly important to distinguish between NAH and neuroblastoma. Imaging study is not always conclusive and does not allow an unquestionable differential diagnosis. Neuroblastoma can present as a solid, cystic, or mixed mass. The cystic formation associated with a neuroblastoma may be related to hemorrhage or necrosis of the tumor. That means neuroblastoma and adrenal hemorrhage may coexist.[7] Color Doppler sonography may be useful in some cases of NAH that are characterized by diminished or absent blood flow in contrast to neuroblastoma.[8] CT and magnetic resonance imaging (MRI) are useful in confirming the presence of hemorrhage and progression of hemoglobin breakdown by typical signals, but they usually do not provide additional information.[9] Further, nuclear scintigraphy and urinary catecholamine metabolites assay do not offer much help due to their limited sensitivity during the neonatal stage.[5] If the diagnosis remains uncertain, serial sonographic follow-up is the first choice. Since some lesions including NAH, pulmonary sequestration, and neuroblastoma can spontaneously regress in infancy, unnecessary surgery should be avoided.

Table 1 summarizes cases of NAH receiving surgical exploration for various reasons associated with suspicion of malignancy, including this case.[10–13] All had common characteristics: no significant predisposing factors; left-side hemorrhage; large size more than 2 cm; complex picture; underwent operation at a relatively early age compared with the average time of NAH resolution, except 1 case whose operation time was not mentioned. In retrospect, the operations occurred too early when there was insufficient evidence of malignancy, and were probably unnecessary.

Our case highlights the diagnostic problems that arise when a space-occupying lesion is found near or at the adrenal gland in the neonate. An accurate diagnosis of a suprarenal mass requires meticulous investigation and close follow-up. Surgical exploration should only be considered under the following conditions: positive

Table 1. Summary of 5 cases of neonatal adrenal hemorrhage who received surgical exploration

Authors	BBW (g)	GA (wk)	Sex	Mode of delivery	Clinical presentation	Lesion side	Lesion size at birth (cm)	Imaging	Other laboratory data	Age at operation (d)	Indication for operation
Burbige[10] (1993)	3,700	38	Female	CS	Palpable mass	Left	7.0×9.0	US, VCUG, DTPA	Normal 24-hr urine VMA & HVA	5	Complex structure with increasing echogenicity
Brame et al[11] (1999)	3,400	39	NM	Induced vaginal delivery	Palpable kidney	Left	2.0	US, MRI, DTPA	Normal 24-hr urine VMA & HVA	NM	Expanding nature of mass
Patankar et al[12] (2002)	3,000	Term	Male	NSD, uncomplicated	Palpable mass	Left	3.9×3.2	US, CT, DTPA	NM	7	Non-homogeneous cystic mass
Hsieh et al[13] (2005)	3,458	40	Male	NSD, uncomplicated	Palpable mass	Left	3.5×4.2×3.0	US, CT	NM	8	Persistent echogenic appearance
This case (2009)	3,450	39	Male	NSD, uncomplicated	Palpable mass, vomiting, jaundice	Left	4.5×3.3	US, CT	Normal AFP, β-hCG & 24-hr urine VMA	18	Failure to decrease in size without echogenicity change

BBW = birth body weight; GA = gestational age; NM = not mentioned; CS = Cesarean section; NSD = normal spontaneous delivery; US = ultrasound; VCUG = voiding cystourethrography; DTPA = diethylenetriaminepentaacetic acid scintigraphy; MRI = magnetic resonance imaging; CT = computed tomography; VMA = vanillylmandelic acid; HVA = homovanillic acid; AFP = α-fetoprotein; β-hCG = β-human chorionic gonadotropin.

tumor markers, elevated urinary catecholamine metabolites, high uptake by the suprarenal mass on nuclear scintigraphy, evidence of metastasis, persistent echogenic appearance without resolution, unusual clinical course under conservative treatment, and progression in size. Early operation of the suprarenal mass without sufficient evidence of malignancy would not be prudent.

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