Imaging Findings of Neonatal Adrenal Disorders

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In newborn infants, normal adrenal glands are characterized by a relatively thin echogenic center surrounded by a thick, hypoechoic cortical rim as seen on ultrasound (US). Various disorders involving the neonatal adrenal gland include adrenal hemorrhage, hyperplasia, cyst, Wolman’s disease, and congenital neuroblastoma. Adrenal hemorrhage is the most common cause of an adrenal mass in the neonate, though differentiation between adrenal hemorrhage and neuroblastoma is in many cases difficult. We describe characteristic US, CT and MR imaging findings in neonates with various adrenal disorders.

Index words: Infants, newborn, genitourinary system
Adrenal gland, abnormalities

Normal Neonatal Adrenal Glands
The appearance of neonatal adrenal glands varies; they may be lambdoid, V- or Y-shaped, and are relatively large (1). On US, corticomедullary differentiation is evident, with an outer echolucent cortex and a central echogenic line representing the medulla (Fig. 1A and B). The cortex is prominent in normal newborn infants because of the presence of a thick transient fetal cortex, which involutes rapidly during the first three weeks of life and completes the synthesis of maternally produced precursor steroids to make estrogens during the third trimester of pregnancy. After the neonatal period, the hypoechoic peripheral zone becomes smaller on US as the fetal cortex involutes. When ipsilateral renal agenesis or ectopia is present, the shape of adrenal gland is discoid and elongated (Fig. 1C).

Adrenal Hemorrhage
Idiopathic neonatal adrenal hemorrhage is usually asymptomatic and frequently discovered as an incidental finding on US (1). A large baby, difficult delivery, and perinatal hypoxia have been reported as predisposing factors in neonatal adrenal hemorrhage. On US, hemorrhage can be echogenic, echolucent or
mixed in appearance, depending on its age (Fig. 2, 3). The adrenal gland may be discernible in part or completely obliterated. A diagnosis of adrenal hemorrhage is based on sequential US examinations and involves...

Fig. 1. Normal adrenal glands in newborn infants.
A. Abdominal US in a newborn infant shows lambda-shaped right adrenal gland (arrows) with thick echolucent cortex and echogenic central line of the medulla. RK = right kidney.
B. US with prone position in another newborn infant demonstrates large and thick limbs of the adrenal gland (arrows). The corticomedullary differentiation is clearly seen.
C. Longitudinal US through the right renal fossa shows the elongated adrenal gland (arrow). In this case, right kidney (not shown) was seen in lower abdominal cavity and was fused with the lower pole of left kidney (crossed fused ectopy).

Fig. 2. Neonatal adrenal hemorrhage.
A. Longitudinal US shows left adrenal hemorrhage (calipers) with mixed solid and cystic appearance. Note relative preservation of the normal triangular shape. LK = left kidney.
B. Three months later, left adrenal hemorrhage seen on the figure A is completely resolved.
demonstration of liquefaction and regression. Calcification at its periphery is common (Fig. 3C). Due to the paramagnetic effect of methemoglobin, MR imaging (Fig. 3B) clearly shows adrenal hemorrhage of high signal intensity on precontrast T1-weighted images. Adrenal hemorrhage may involve only a part of the adrenal gland even when normal contour is preserved (2). When it primarily involves the adrenal medulla, normal configuration of the gland and the cortex is preserved (Fig. 4).

Fig. 3. Neonatal adrenal hemorrhage with calcification. A. Longitudinal sonogram shows adrenal hemorrhage (H) above right kidney (RK), with a predominantly solid appearance. B. T1-weighted axial MR image shows a large hyperintense hematoma (arrowheads) in the right adrenal gland with central isosignal intensity suggesting resolving hemorrhage. C. Two weeks later, right adrenal hemorrhage has decreased in size with eggshell-like calcification in the periphery of the resolving hematoma (arrowheads). RK = right kidney.

Fig. 4. Adrenal medullary hemorrhage in a newborn. Transverse (A) and longitudinal (B) US scans of the adrenal region show normal hypoechoic cortices of both adrenal glands (arrowheads). However, echogenic adrenal medullae (m) are markedly thickened.
Congenital Adrenal Hyperplasia

Enzymatic defects at any site of adrenal steroid biosynthesis result in cortisol deficiency and secondary overproduction of pituitary corticotropin, which in turn overproduces androgens. Congenital adrenal hyperplasia or adrenogenital syndrome causes ambiguous genitalia, pseudohermaphroditism in females and an enlarged penis and precocious puberty in males. Deficiency of 21-hydroxylase is the most common form of congenital adrenal hyperplasia, and diffuse or nodular enlargement of the adrenal cortex occurs in many but not all babies with this condition (2, 3). US may show diffuse adrenal enlargement with a wrinkled, wavy, cerebriform appearance and loss of normal corticomedullary differentiation (Fig. 5). Occasionally, adrenal enlargement may mimic adrenal mass but bilateral involvement with relative preservation of gland configuration can exclude adrenal tumor (Fig. 5C and D).

Congenital Lipoid Adrenal Hyperplasia

Conversion of cholesterol to pregnenolone is an essential step in the synthesis of all adrenocortical hormones. Infants with 20, 22-desmolase deficiency cannot synthesize any glucocorticoid, mineralocorticoid, or sex steroid. The adrenal glands are enlarged and filled with cholesterol and other lipids, which explains echogenicity on US and hypodensity on CT scan (5) (Fig. 6).

Adrenal Cyst

Adrenal cysts may be parasitic, epithelial, endothelial or pseudocysts; true epithelium-lined adrenal cysts are rare in the neonate. Cystic lesions of the adrenal glands are usually pseudocysts (hemorrhagic cysts) secondary to resolving adrenal hemorrhage.

Fig. 5. Congenital adrenal hyperplasia in a newborn girl with virilization and dark pigmentation. Right (A) and left (B) adrenal glands show diffuse enlargement with cerebriform appearance (arrows). L = liver, S = spleen. C. Transverse US in another female infant with congenital adrenal hyperplasia reveals diffuse enlargement of bilateral adrenal glands (arrowheads). D. T2-weighted coronal MR image in the same patient to C shows diffuse enlargement of the adrenal glands (arrows) with preservation of the normal triangular configuration.
Adrenal cystic masses have been described in association with complete or incomplete forms of Beckwith-Wiedemann syndrome (6, 7) (Fig. 7). Because an adrenal cyst cannot be clearly distinguished from the cystic neuroblastoma occurring in association with Beckwith-Wiedemann syndrome, close observation, as well as US or surgical confirmation, is needed.

**Wolman’s Disease**

Wolman’s disease is a fatal xanthomatosis of autosomal recessive inheritance and is characterized by failure to thrive, hepatosplenomegaly, vomiting, and steatorrhea. Uniform bilateral adrenal enlargement with calcification is pathognomonic (8). Calcification

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**Fig. 6.** Congenital lipoid adrenal hyperplasia.

A. Longitudinal US scan of the left flank shows enlarged adrenal gland (arrows) above the left kidney (LK). The gland appears diffusely echogenic with loss of corticomедullary differentiation.

B. Unenhanced CT scan at the level of adrenal gland shows thickened both adrenal glands with low attenuation, indicating fatty infiltration.

**Fig. 7.** Adrenal cystic mass in a newborn girl with Beckwith-Wiedemann syndrome.

Longitudinal US scan (A) shows a large, well-demarcated cystic mass with homogeneous internal echoes above the right kidney (RK).

T2-weighted (B) and postcontrast T1-weighted (C) coronal images show a large suprarenal cystic mass (M) with homogeneous high signal intensity on T2-weighted image without enhancing solid portion. Right kidney (RK) is displaced and rotated inferiorly and medially. Surgical removal with histologic examination confirmed hemorrhagic cyst of the right adrenal gland.
Fig. 8. Wolman disease in a 45-day-old boy.
A. Plain radiograph shows triangular calcifications in both suprarenal regions (arrowheads).
B. Transverse US scan shows calcifications along both adrenal limbs with acoustic shadowing (arrows).
C. Unenhanced CT scan clearly shows extensive adrenal calcifications with lamellated appearance (arrows). Coronal TI (D) and T2-weighted (E) MR images show bilateral, symmetrical enlargement of the adrenal glands (arrows) with alternating low and isosignal intensities suggesting lamellated calcifications. Note preservation of normal adrenal gland configuration.

Fig. 9. Congenital neuroblastoma.
A. Longitudinal US scan in a newborn girl shows a well-demarcated, homogeneously echogenic mass (M) above the right kidney (RK). Surgery confirmed neuroblastoma originating from the right adrenal gland.
B. Extraadrenal neuroblastoma in a newborn baby. Transverse US scan shows a retrocaval mass (arrows) which is partly cystic and solid. At surgery, it was proved to be a retroperitoneal neuroblastoma with hemorrhage in the vicinity of right adrenal gland. V = inferior vena cava, RK = right kidney.
throughout the glands is clearly demonstrated on plain radiograph, US, and CT (Fig. 8). On MR images, both adrenal glands are diffusely thickened with linear iso or hypointensity, suggesting calcification (Fig. 8 D and E).

**Congenital Neuroblastoma**

Congenital neuroblastoma should be considered in the differential diagnosis of a neonatal suprarenal mass. Cystic neuroblastoma is more common during the neonatal period than at any other age, though it may be difficult or even impossible to differentiate neonatal neuroblastoma from adrenal hemorrhage, especially when the tumor is cystic or hemorrhagic (9). Neuroblastoma tends to remain static over the first two weeks of life as compared to adrenal hemorrhage. US demonstrates a cystic or solid-appearing suprarenal mass originating from the adrenal gland or paravertebral sympathetic chain (Fig. 9).

References


신생아부신질환의영상소견

1. 성균관대학교의과학대학삼성의료원 방사선과

윤혜경·김보경·이민희

신생아의정상부신은초음파에서두터운저에코의피질과암은고에코의수질로보이는것이특징이다.신생아의부신을침범하는질환은다양하며부신출혈,부신과증식,낭종,선천성신경아세포종등이있다.이중부신출혈이가장흔한데,신경아세포종의감별이어려운경우도있다.저자들은신생아의여러부신질환에서특징적인초음파,전산화단순촬영,자기공명영상소견을보고하고자하였다.
### 4th REFRESHER COURSE ON OBSTETRIC SONOGRAPHY

**- FETAL THORAX AND ABDOMEN -**

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<th>Time</th>
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<td>8:30-9:00</td>
<td>登陸</td>
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<td>9:00-9:30</td>
<td>Normal sonographic anatomy of the fetal thorax and abdomen</td>
<td>이영호(삼성제일병원)</td>
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**♠ 수강 신청 안내 ♠

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