Fetal Adrenal Haemorrhage – Two-Dimensional and Three-Dimensional Imaging

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Key Words

Fetal adrenal mass • Adrenal haemorrhage • Prenatal ultrasonography • Three-dimensional sonography • Fetal neuroblastoma • Adrenal gland

Abstract

A case of prenatal adrenal haemorrhage first detected by 2-dimensional and 3-dimensional sonography at 27 weeks' gestation is reported. Ultrasound examination showed a large cystic mass (32 × 27 × 27 mm) in the right suprarenal region of the fetus. Two weeks later, the mass had slightly increased in size demonstrating hyperechoic areas within the cyst. Further serial ultrasound examinations revealed a progressive organisation of the cystic mass associated with a moderate reduction in size. The diagnosis of adrenal haemorrhage was confirmed by postnatal follow-up sonograms as the mass decreased in size from 28 × 21 × 21 mm on day 1 to 23 × 18 × 17 mm on day 42. Course and sonographic signs were typical for adrenal haemorrhage and the neonate was therefore managed without surgical exploration. The child is developing normally at 6 months of age.

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Introduction

Routine use of ultrasonography as a screening test during pregnancy has led to the discovery of an increasing number of fetal suprarenal masses. These masses usually represent benign lesions albeit some are malignant. Clinicians are faced with a diagnostic and therapeutic dilemma as the differentiation between benign and malignant adrenal masses may prove difficult, even with high-resolution ultrasound technology.

Case Report

A 41-year-old woman, gravida 2 para 1, was referred to our Fetal Medicine Unit for further assessment of an adrenal mass at 27 weeks' gestation. Her medical history had been unremarkable. On detailed sonography, a solitary cystic mass located superior to the right kidney measuring 32 × 27 × 27 mm was found (fig. 1, 2). Two-dimensional and 3-dimensional ultrasound clearly demonstrated that the tumour was arising from the adrenal gland. Biophysical profile, fetal biometry and routine Doppler examinations of the umbilical cord and the middle cerebral artery were normal. The size of the mass initially increased to 39 × 29 × 29 mm at 29 weeks' gestation and eventually decreased to 28 × 21 × 21 mm at birth.

At 38 weeks' gestation, the mother was delivered of a healthy male neonate weighing 3,400 g. No anaemia or palpable abdomi-

Fig. 1. Prenatal tomographic ultrasound imaging at 27 weeks showing an anechoic suprarenal cystic mass.

Fig. 2. Power Doppler examination of the suprarenal region at 27 weeks showing the renal arteries. No flow can be seen within the mass.

Fig. 3. Postnatal power Doppler sonography at day 4 of life showing only a few vessels within the capsule of the mass and absence of perfusion within the tumour. Arrows indicate the outline of the adrenal mass.

Fig. 4. Postnatal sonography at day 42 of life showing a solid echogenic tumour with a central hypoechoic region.

nal mass were found in the neonate and sonography showed a solid mass with heterogenous echotexture in the position of the right adrenal gland. Neither with colour-coded Doppler sonography nor with power Doppler sonography was perfusion detected inside the mass (fig. 3). The size of the mass decreased after birth to $23 \times 18 \times 17$ mm at 42 days of life (fig. 4). Tumour markers, including urinary vanillylmandelic acid, urinary homovanillic acid and neuron specific enolase, were within normal ranges. As the findings were suspicious of an adrenal haemorrhage, invasive examinations as well as CT and ^{131}I -labelled meta-iodobenzylguanidine scans were deemed unnecessary. Ultrasonography at 6 months showed progressive resolution of the mass.

Discussion

The majority of large abdominal masses detected prenatally are of renal origin (mesoblastic nephroma, nephroblastoma, i.e. Wilms' tumour, multicystic dysplasia, hydronephrosis), or duplications of the urinary or intestinal tract, followed by teratoma and especially adrenal tumours. Prenatal ultrasound should be able to differentiate abdominal masses from suprarenal masses in most cases. The main differential diagnoses of suprarenal masses are adrenal haemorrhage and neuroblastoma [1]. Suprarenal masses, however, may also be due to extralobular pulmonary sequestration, adrenal abscess, adrenal nodular hyperplasia, adrenal cyst, bronchogenic cyst [2] or rarely adrenal carcinoma [3].

The incidence of adrenal haemorrhage in newborns ranges from 2 out of 1,000 (detection at birth) to 3 out of 1,000 (postnatal ultrasound screening). The detection

Fig. 5. Three-dimensional reconstruction and volume measurement using virtual organ computer-aided analysis.

rate in utero is much lower. Adrenal haemorrhage affects the right side 2–3 times more frequently than the left and is bilateral in 5–15% of cases [4, 5].

The aetiology of adrenal haemorrhage and the reasons for predilection of the right side have not been fully elucidated. One possible explanation is the compression of the adrenal gland between the liver and spine and direct drainage of the right adrenal vein into the inferior vena cava inducing higher venous pressure changes compared to the left side where the adrenal vein connects to the renal vein [6]. The adrenal glands are particularly vulnerable to haemorrhage because of their size (about 20 times the relative size in the adult) and high vascularity with supply from the inferior phrenic artery, abdominal aorta and renal artery [7].

Haemorrhage is typically contained within the capsule of the adrenal gland. If rupture of the capsule occurs, blood may spread into the retroperitoneal space or peritoneal cavity.

Several cases of scrotal haematoma or hydrocele due to neonatal adrenal haemorrhage are described in boys with patent processus vaginalis. Therefore, scrotal swelling should always lead to scrotal and abdominal postnatal ultrasound examinations to rule out adrenal or renal masses [8]. Adrenal insufficiency is found very rarely even in cases with severe bilateral adrenal haemorrhage. Residually functioning adrenal tissue in the subcapsular region is left in most cases and adrenal insufficiency only manifests clinically when more than 90% of each gland are destroyed [5, 9].

The normal adrenal glands can be clearly visualised by ultrasound in neonates and consist of a hypoechoic cortex and a thin echogenic medulla. The sonographic pattern of adrenal haemorrhage depends on its age. Active adrenal bleeding appears sonolucent. Later on, a solid clot with diffuse echogenicity is found, retracting continuously. Finally, as liquefaction occurs, the mass demonstrates mixed echogenicity, often with a central hypoechoic region, and eventually becomes completely anechoic [10, 11]. While 3-dimensional sonography is not necessary to differentiate between adrenal haemorrhage and neuroblastoma, it can be helpful in determining the exact size of the mass. In particular, 3-dimensional reconstruction and measurement of the volume of the mass with a rotational method using virtual organ computer-aided analysis is a valuable tool for repeated evaluation during follow-up (fig. 5).

Congenital neuroblastoma is the most frequent suprarenal malignancy, with an incidence of approximately 6 out of 1,000 liveborns [12] and a diverse sonographic appearance. In contrast to the peripheral rim-like calcifica-

tions and rim-like Doppler pattern of adrenal haemorrhage, neuroblastoma often shows a network of microscopic vessels with characteristic high-velocity Doppler shifts inside the tumour and stippled calcifications [7]. It has been suggested that many of the neonatal cases of neuroblastoma can regress spontaneously. Yamamoto et al. [13] found tumour regression in 11 of 12 cases of early-stage neuroblastoma detected by mass screening. Therefore, recommendations for treatment of suprarenal masses, discovered incidentally during fetal life, have changed in recent years. Surgical exploration in every case gave way to a wait-and-see attitude based on close pre- and postnatal ultrasound follow-up.

Suspect masses that progress or remain unchanged during the first few months of life raise suspicion of malignancy and should be explored surgically. Appropriate prenatal assessment and close sonographic monitoring may avoid surgery in cases of benign masses like adrenal haemorrhage or spontaneously regressing neuroblastomas.

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