A prenatally detected adrenal cyst treated by adrenal-sparing surgery: case report and review of the literature

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A neonatal case of left adrenal cyst detected *in utero* and successfully treated by adrenal-sparing surgery is presented and discussed with review of the literature. Incidentally discovered prenatal adrenal masses present a diagnostic dilemma. Benign and malignant conditions can present as a fetal suprarenal mass. There is a wide spectrum of management modalities ranging from follow-up by serial sonographic scanning during pregnancy to early primary excision of the mass. We report a neonate with prenatal diagnosis of a cystic mass arising from the left adrenal gland. Postnatal excision of the mass without adrenalectomy was carried out. Frozen sections of the mass and a biopsy of the left adrenal gland confirmed the benign nature of the cyst and normal adrenal tissue. The uniloculated cyst was reported as a pseudocyst. After surgery, the recovery was uneventful, and the patient was discharged 4 days postoperatively in good condition. On the basis of this case and review of the literature, we may conclude that early primary surgical excision is recommended for either diagnosis or treatment if the results of prenatal or postnatal imaging studies are unreliable for the precise diagnosis of suprarenal mass. Adrenal-sparing surgery is recommended if pathological evaluation of frozen sections has confirmed the benign nature of the mass. *Ann Pediatr Surg* 7:152–154 © 2011 Annals of Pediatric Surgery

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Introduction

The increasing use of ultrasound for routine obstetrical evaluation has led to an increase in the incidental detection of fetal suprarenal masses 1. Possible diagnoses of a suprarenal mass identified *in utero* include congenital adrenal cystic neuroblastoma, cystic Wilm's tumor, cortical renal cyst, mesoblastic nephroma, obstructed upper pole duplication anomalies with ectopic ureteral implantation, hepatic cysts, choledochal cysts, ovarian cysts, enteric cysts, and adrenal cysts 2.

Adrenal cysts in infants are usually of the pseudocyst variety and are thought to be secondary to neonatal adrenal hemorrhage [3]. An adrenal hemorrhage incidence of 1.7 per 1000 births has been reported on the basis of a necropsy series. However, the incidence of adrenal hemorrhage *in utero* is yet unknown [4]. Although the etiology of prenatal adrenal hemorrhage is not yet clear, in a study it was explained as a sudden increase in venous pressure transmitted from the inferior vena cava back to the adrenal gland. In the same study, the investigators also suggested that this mechanism could explain why the right adrenal gland with its short vein is involved in 70% of reported cases than the left gland [5].

The aim of this study was to present a case of prenatally diagnosed left-sided adrenal cyst with successful management by adrenal-sparing surgery and to review the literature based on the pathogenesis, the differential diagnosis, and the management of adrenal cysts.

Case study

A 2500 g full-term boy was born by spontaneous vaginal delivery to a gravida 1, para 1 mother. Antenatal history revealed an abdominal mass diagnosed at 24 weeks of gestational age during routine obstetrical evaluation. The mother neither had any history of medication use during pregnancy, nor of trauma, hypertension, diabetes mellitus, toxemia, smoking, or alcohol ingestion. The paternal age was 30 years. The mother and father were not consanguineous and the family history was not contributory. No other data could be obtained of the follow-up sonographic evaluation for the rest of the pregnancy until birth. Postnatal sonography on the day 2 of birth in the referring center demonstrated a 38×29 mm cystic mass localized between the tail of the pancreas and the upper part of the left kidney. The patient was admitted to our clinic at the age of 6 days with a diagnosis of abdominal mass. Physical examination showed an immobile mass in the left lower abdomen of the newborn. The initial blood count revealed: red blood cells of $3.89 \times 106/mm^3$; hemoglobin level of 14.7 g/dl; hematocrit of 41.8%; white blood cells of $6.04/103/mm^3$; platelet count of $282 \times 103/mm^3$; α-fetoprotein of 60 500 ng/ml; β-human chorionic gonadotropin of 2.93 mIU/ml; neuron-specific enolase of 33.13 ng/ml; and ferritin of 305.9 ng/ml. All ranges were normal, except for a slight increase in ferritin level.

Abdominal and Doppler ultrasound examinations showed a cystic mass measuring $30 \times 40 \times 42$ mm in the left lower abdominal area, containing unechogenic material with no vascularity inside or peripheral to the mass. A computed tomography scan study revealed normal-sized kidneys and an ovoid-shaped hypodense lesion between the spleen and the upper pole of the left kidney with an unidentified solid echogenity medially located to the mass (Fig. 1). A laparotomy was performed at the age of 16 days in the

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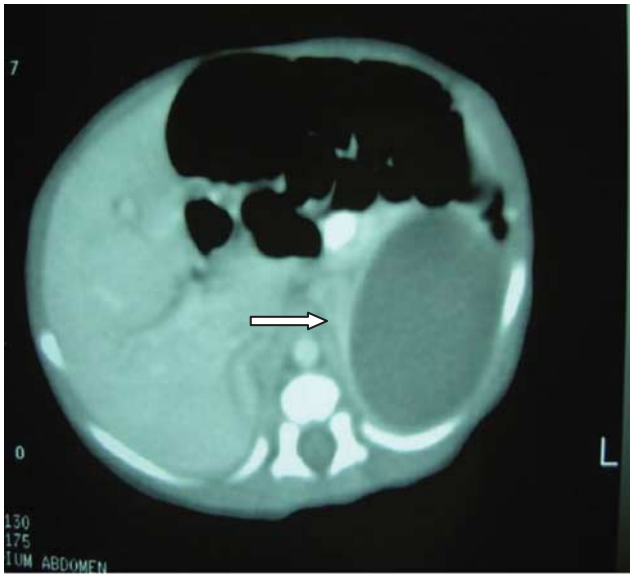
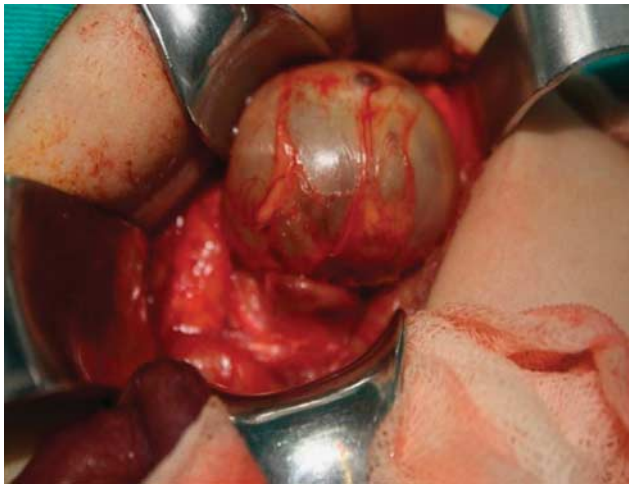
Fig. 1



Computed tomography showing the cystic mass and adjacent unidentified solid echogenity. Solid echogenity (arrow).

Fig. 2



Operative view of the adrenal cyst.

postnatal period for differential diagnosis and treatment. During the operation, a well-encapsulated cystic mass arising from the left adrenal gland, measuring 30 × 35 mm, was excised totally without adrenalectomy (Fig. 2). Frozen sections of the mass and a biopsy of the left adrenal gland confirmed the benign nature of the cyst and normal adrenal tissue. On pathologic examination, the uniloculated cyst, which consisted of hemorrhagic fluid with walls consisted of dense, fibrous connective tissue, devoid of a recognizable endothelial layer, was reported as a pseudocyst. The walls were 1-mm to 2-mm thick, and islands of adrenal cortical tissue were incorporated into the walls.

After surgery, the recovery was uneventful, and the patient was discharged 4 days postoperatively in good condition.

Discussion

Fetal suprarenal masses incidentally detected *in utero* usually present a diagnostic dilemma. Benign and malignant conditions can present as a fetal suprarenal mass, and the sonographic appearance of prenatal adrenal hemorrhage does not always follow the typical course. Moreover, congenital cystic neuroblastoma (CCNB) and adrenal hemorrhage may coexist [6]. Definite diagnosis of fetal adrenal hemorrhage is difficult before birth because the condition presents variable sonographic features ranging from cystic, mixed solid, and cystic, to a solid mass, which are similar to the features seen for CCNB [7]. The patient should be followed by serial sonographic scanning during pregnancy and after birth to observe the gradual resolution and regression of the congenital adrenal mass. However, in our case, although an intra-abdominal mass was detected early at the second trimester of pregnancy, we could not obtain any information about the size or sonographic appearance of the mass that would allow the diagnosis of adrenal hemorrhage and avoid surgery.

The etiologies of neonatal adrenal hemorrhage include birth trauma, perinatal hypoxia, septicemia, shock, thrombocytopenia, congenital syphilis, disseminated intravascular coagulation, and renal vein thrombosis [2]. In this case, there was no prenatal or postnatal insult that could explain the prenatal adrenal hemorrhage. Most earlier studies noted the mass during the third trimester of pregnancy. However, with the progress of obstetric ultrasonography, recent studies have demonstrated that prenatal adrenal hemorrhage can occur as early as the second trimester, as in our case [5].

Adrenal hemorrhage involves the right side three to four times more often than the left side, possibly because of susceptibility of the enlarged gland to compression between the liver and the spine [8]. In the case we presented, adrenal hemorrhage may have occurred on the left side, without any trauma or intrauterine complications, because of the relatively large size and vascularity that make the gland sensitive to compression and changes in venous pressure.

Neuroblastoma is the most common neonatal malignancy, and the adrenal gland is the most common primary site for neuroblastoma [9]. The currently available radiological modalities and urinary catecholamine levels or tumor markers are not reliable enough for differentiating benign conditions from CCNB. Early detection of CCNB is associated with a greater than 90% cure, as metastases have usually not occurred. Another concern about adrenal cysts is the association of incidental malignancy other than CCNB; a 7% incidence of malignancy was reported from a review of over 600 cases of adrenal cysts from the literature [10]. Besides malignancy, spontaneous hemorrhage into an adrenal cyst resulting in shock may occur because of the vascularity of the adrenal gland; the cyst may also be infected. No general agreement exists for therapy, with some investigators advocating surgery and others prescribing a more conservative approach.

Potential interventions include percutaneous needle aspiration or sclerotherapy, surgical resection, or cyst unroofing. For pseudocysts and complex cysts, when detecting malignancy by imaging modalities and tumor markers is difficult, there has been a recent shift toward earlier surgical excision. Adrenal-sparing resections as simple enucleation of the cyst, with preservation of kidney and adrenal gland, are the preferred surgical choice [11]. In our case, there was an ovoid-shaped hypodense multiseptated lesion between the spleen and the upper pole of the left kidney with an unidentified solid echogenity medially located to the mass. Because the diagnosis was uncertain with laboratory and radiological examination, primary surgical excision was attempted. At surgery, we found the cystic mass arising from the left adrenal gland well encapsulated and consisting of hemorrhagic fluid. After confirming the benign nature of the cyst and normal adrenal tissue by frozen sections of the mass and a biopsy of the left adrenal gland, we excised the cyst completely using the adrenal-sparing approach and preserved the left adrenal gland and the kidney.

In their studies, Cohen *et al.* [12] described three types of adrenal hemorrhage: (a) central hematoma formation, in which the hematoma distorts the medulla and attenuates the overlying cortex; (b) total necrosis of either or both sides, in which the gland (or glands) is enlarged but maintains its shape; and (c) segmental lesions in which normal adrenal tissue can be seen adjacent to the lesion. The case we presented was of the third type in which adjacent normal adrenal tissue was demonstrated by a frozen biopsy during surgery.

Adrenal cysts are classified into four histopathological subtypes: endothelial, pseudocyst, epithelial, and parasitic. Pseudocysts are the most common subtype and usually result from hemorrhage within normal adrenal tissue or tumor [13]. Organization of the hemorrhagic adrenal area results in a unilocular cyst. The walls of this pseudocyst consisted of dense fibrous tissue and are not lined with epithelium. Adrenal cortical tissue may be incorporated into the walls of the pseudocyst in up to 19% of the cases [14]. On pathologic examination, the cyst we excised was found to be uniloculated and had hemorrhagic fluid. The walls were 1-mm to 2-mm thick, consisting of dense fibrous connective tissue, and there was no epithelium lining the walls of the cyst. Adrenal tissue was detected in the walls of the cyst. All these features indicated a pseudocyst for histopathological diagnosis.

In summary, benign and malignant conditions can present as a fetal suprarenal mass. Among benign conditions, prenatal adrenal cyst due to adrenal hemorrhage can occur as early as the second trimester of pregnancy without a significant insult. Follow-up obstetric sonography is required for observing the gradual resolution and regression of the congenital adrenal hemorrhage. If the currently available radiological modalities, prenatal or postnatal, are unreliable for differential diagnosis, early primary surgical excision is recommended for either diagnosis or treatment.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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