CASE REPORT

Prenatal diagnosis of an adrenal mature teratoma mimicking a neuroblastoma

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ABSTRACT

Teratomas are defined by the presence of cell types from different germ layers, they typically involve the gonads or the sacrococcygeal region and are rarely retroperitoneal. Prenatally detected adrenal teratomas are extremely uncommon. Aim of this paper is to share our experience with an adrenal antenatal mass initially diagnosed as a left adrenal neuroblastoma that turned out to be a mature teratoma after microscopical examination. We present the case of a male fetus with antenatal diagnosis of a left adrenal cystic image at the 22nd week of amenorrhea. The fetal magnetic resonance imaging showed a non-calcified cystic mass of the left adrenal gland, compatible with a neuroblastoma. At birth an ultrasound confirmed the presence of an anechogenic lesion of the left adrenal gland. The infant was closely monitored during his first year and in the absence of significant regression of the adrenal mass, it was decided to perform a laparoscopic left adrenalectomy. Unexpectedly, the final pathological diagnosis was mature cystic adrenal teratoma. In conclusion, an adrenal mass diagnosed antenatally is generally a hemorrhage or a neuroblastoma. Adrenal teratomas are very rare and those diagnosed antenatally even more. At present, we have no clinical, biological, or radiological evidence to suspect them before surgical removal. There are only two other cases of unexpected adrenal teratoma in infants described in Literature.

KEYWORDS: adrenal mass; mature teratoma; antenatal diagnosis; laparoscopy; children

INTRODUCTION

Teratomas are defined by the presence of cell types from different germ layers and they are classified as mature or immature based on the degree of differentiated tissue. Mature teratomas have above all well-differentiated component, while immature teratomas are composed of at least 10% immature tissue and are at a higher risk for malignant behavior [1].

Adrenal teratomas are rare neoplasms and are reported to account for only 1–4% of surgically excised adrenal nodules [1-3].

Adrenal masses could be diagnosed from 20 weeks of amenorrhea (WA) when adrenal glands turn visible sonographically. It most often consists of hemorrhages or neuroblastomas [4], but there are several other differential diagnoses. [5].

It remains a challenge for clinicians to distinguish the nature of asymptomatic adrenal mass based only on antenatal imaging findings. The accurate diagnosis of neonatal adrenal mass depends mostly on their dynamic observation by imaging exams like enhanced CT and ultrasonography [4].

We report the case of an antenatally detected left adrenal cystic mass that was initially managed like congenital neuroblastoma, which turned out to be a mature teratoma post-adrenalectomy.

Antenatally detected suprarenal masses are likely to be neuroblastomas or adrenal hemorrhages, but may be rare benign lesions (subdiaphragmatic extralobar pulmonary sequestration, bronchogenic cyst, renal dysplasia).

Teratomas typically involve the gonads or the sacrococcygeal region and are rarely retroperitoneal.

Prenatally detected adrenal teratomas are extremely uncommon, we therefore decided to describe our recent experience and to review the literature.

CASE PRESENTATION

A male infant with a history of a prenatally detected left suprarenal mass was addressed to the surgical unit. Antenatal ultrasonography at 22 WA disclosed a cystic left suprarenal mass about 2 cm in diameter. The fetal magnetic resonance imaging (MRI) showed a 20x13 mm non-calcified

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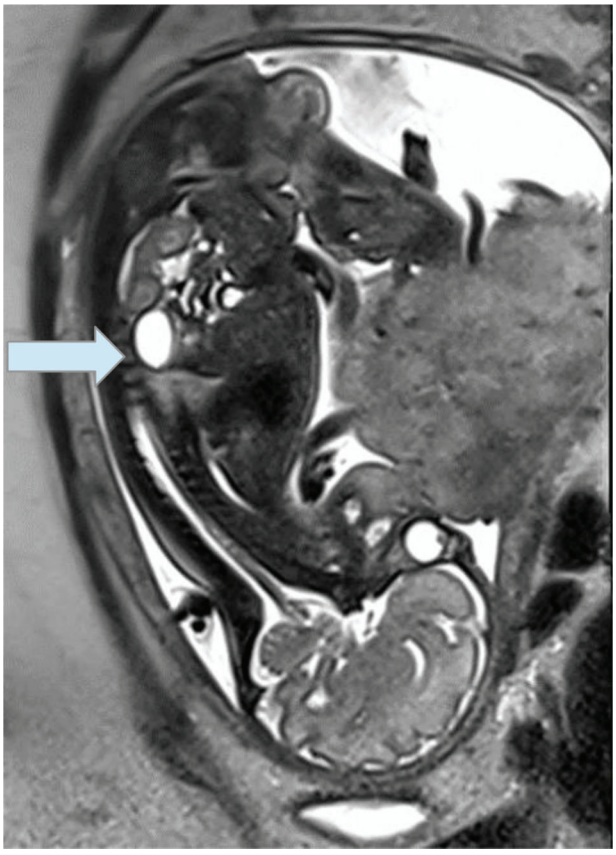

Fig. 1. Fetal MRI image demonstrates a retroperitoneal, suprarenal, 20 x13 mm, homogeneous, high T2 signal intensity, non-calcified, cystic mass (arrow).

cystic mass of the left adrenal gland, compatible with a neuroblastoma (Figure 1). Vaginal delivery was uneventful.

Results of routine laboratory tests including complete blood count, blood biochemistry, and urinalysis were normal, except for a major iron-deficiency anemia. Ultrasonography at 1 months of life showed an anechogenic image of the left adrenal gland of 17x12 mm, with thin walls (Figure 2).

In the 9 months interval between the birth's ultrasound and the last imaging, the mass minimally diminished in size and stabilized at about 2 cm.

The infant was closely monitored during his first year of life and in the absence of symptomatology or significant regression of the mass, we decide to proceed with its removal.

We performed a preoperative CT which showed a partially cystic 20 x 22 mm mass in the left adrenal region, non-enhanced, without calcification (Figure 3). It was therefore decided to surgically remove the tumor. An uneventful laparoscopic adrenalectomy was performed. The patient was monitored for 24 hours, restarted alimentation immediately after surgery, and was discharged in good general conditions at 48 hours. The patient had no symptoms during 5 months of follow-up.

The pathological result described a mass with mixed composition of ciliated columnar epithelium of the respiratory type, well differentiated cartilaginous structures as well as small seromucous glands. The specimen was diagnosed as a mature cystic adrenal teratoma, without any suspicious sign of malignancy, which was completely resected (Figure 4).

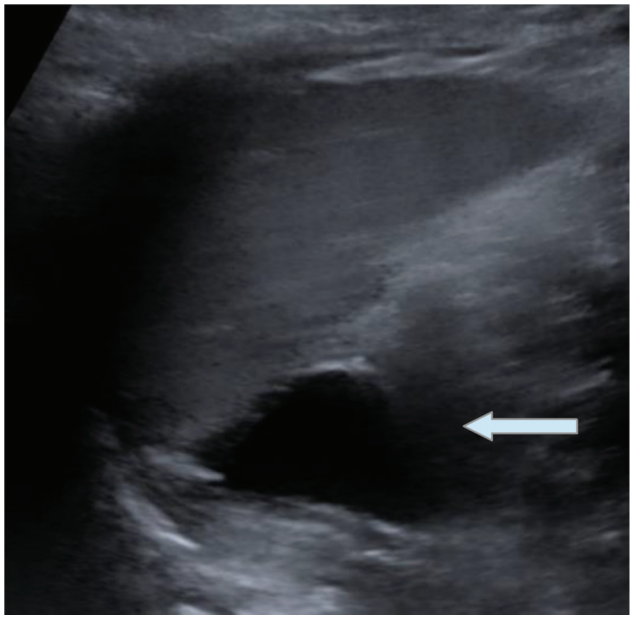

Fig. 2. US of the left adrenal, one month after birth (arrow).

DISCUSSION

Adrenal masses could be diagnosed from 20 WA when adrenal glands turn visible sonographically. It most often consists of hemorrhages or neuroblastomas [4], but other differential diagnoses are represented by adrenal cysts, subdiaphragmatic extralobar pulmonary sequestrations, bronchogenic cyst, hepatic tumors, adrenogenital syndrome (secondary to congenital adrenal hyperplasia) or renal dysplasia [5].

It remains a challenge for clinicians to distinguish the nature of asymptomatic adrenal mass based only on antenatal imaging findings. The accurate diagnosis of neonatal adrenal mass depends mostly on their dynamic observation by exams like enhanced CT and ultrasonography.

Several studies [6] suggest that prenatal diagnosis of suprarenal masses should be preferentially focused on neuroblastoma because usually they prove to be congenital neuroblastomas [4].

For this reason, we initially managed our patient like a congenital neuroblastoma. We based our workup only on ultrasound monitoring, accordingly with several authors [6-8]. As a matter of fact, congenital neuroblastoma generally has a favorable prognosis with regression of lesions in most cases during the first year of life [9,10].

We decided not to perform any specific blood test to support our diagnosis. In fact, catecholamine metabolites often demonstrated to be not a valuable index for the diagnosis of adrenal masses [4]. Also, the role of alpha-fetoprotein (AFP) is controversial: AFP levels are normally very high at birth and fall exponentially to normal adult levels by the age of 1 year.

In our case, was the atypical aspect at the imaging and above all the atypical behavior (lack of regression, stability of lesion) that take made us to perform an adrenalectomy.

We were rather surprised by the result of the pathological report that showed a mixed composition of ciliated columnar epithelium of the respiratory type, well differentiated cartilaginous structures and seromucous glands, diagnosed as a mature cystic adrenal teratoma.

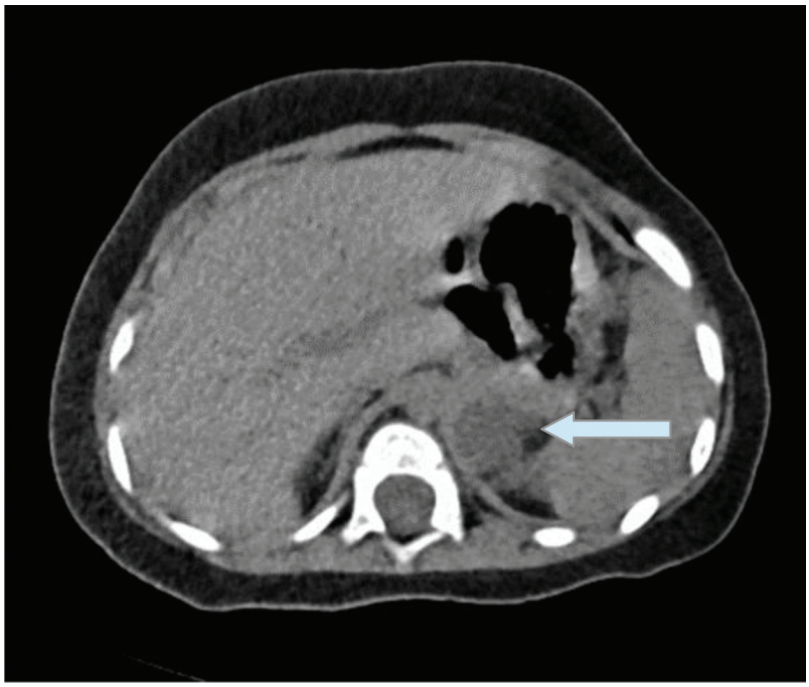

Fig. 3. Preoperative CT showing a partially cystic 20 x 22 mm mass in the left adrenal region without calcification (arrow).

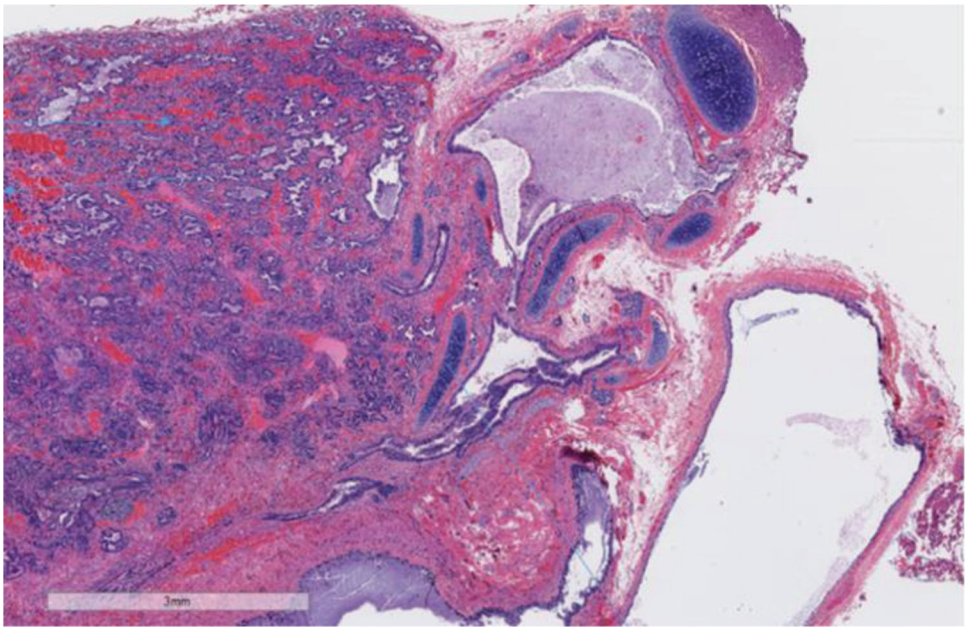

Fig. 4. Microscopic image of the adrenal teratoma showing mixed composition of ciliated columnar epithelium of respiratory type, well differentiated cartilaginous structures and seromucous glands (HE, x50).

Teratomas are rare germ cell neoplasms that originate from totipotent cells that differentiate into tissue components representing derivatives of ectoderm, mesoderm, and endoderm [1]. Commonly they affect the gonads but also may occur at extragonadal sites. The common extragonadal sites include anterior mediastinum, sacrococcygeal region, pineal region and retroperitoneum [1].

The retroperitoneal localization is extremely rare, above all in childhood [11].

To the best of our knowledge, only very few other cases of primary mature adrenal teratomas affecting children are reported in literature. [12,13] Most of these cases were incidentally detected or clinically manifested because of abdominal distension. Is extraordinarily rare that the tumor is diagnosed antenatally. To date, there are only two other cases of adrenal antenatal mass firstly diagnosed as neuroblastoma which turned out to be a mature teratoma reported in literature [12,13].

The most characteristic imaging feature of mature teratomas is a heterogeneous mass containing fluid, fat, and calcification [7]. None of these features are considered diagnostic for teratomas: they are non-specific and it is rather the dynamic observation by CT and ultrasound that can help for differential diagnosis [13].

Mature teratoma's prognosis is generally very favorable, especially if the tumor is completely removed: complete surgical excision provides the best chance of cure. Therefore, the correct diagnosis is essential for optimal postnatal treatment, but it remains a challenge.

CONCLUSION

It remains a challenge for clinicians to distinguish the nature of asymptomatic adrenal mass. Adrenal teratomas are very rare and those of antenatal diagnosis even more. At present, we have no clinical, biological, or radiological evidence to suspect it before surgical removal, but they should be included in the differential diagnosis of a prenatally detected adrenal masses, and resection is advised.

Ethics approval and consent to participate

Informed consent was properly documented by both the patient's parents. Ethical approval by the local hospital ethical committee was given properly.

Availability of data and materials

All data generated or analyzed during this study are included in this article.

Competing interests

The authors have no financial or personal relationships with people or organizations that could inappropriately influence or bias their work.

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