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Case Report

The Role of Fetal MRI and US in the Prenatal Diagnosis of Mixed Adrenal Neuroblastoma: A Case Report and Brief Literature Review

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Abstract

Neuroblastoma is a malignant tumor which develops from the neural crest cells of the sympathetic nervous system. Neuroblastoma represents 8-10% of all childhood neoplasms making it the most common extracranial solid tumor in childhood. It is sometimes occasionally detected prenatally during second and third trimester screening. Cross-sectional imaging is needed not only for assessment of the origin, extent of the tumor, local invasion, vascularity and calcifications, but also for staging of neuroblastoma, presence of lymphadenopathy and metastatic spread. Prenatal ultrasound (US) is first-line modality in evaluating neuroblastomas followed by fetal magnetic resonance imaging (MRI) for precise determination the extend of the tumor within the abdominal cavity or the retroperitoneal space. This report aims to present the role of fetal MRI as a valuable adjunct to US in a case of prenatally detected neuroblastoma, which is an important issue for prognostic counseling. Furthermore, beside this case study we would like to contribute a brief review of the literature.

Keywords: Neuroblastoma; antenatal ultrasound; fetal MRI

Introduction

Neuroblastoma is a medullary adrenal neoplasm as well as ganglioneuroblastoma and ganglioneuroma both of them arising from the primitive neural crest cells [1]. It is the most common extracranial solid tumor of childhood, accounting for 8-10% of all cancers in children [2-4]. Two thirds of neuroblastomas develop in the abdomen and of those, from two thirds up to 90% originates from the adrenals [1,5]. Despite of the median age of neuroblastoma diagnosis of 19 months, sometimes they are incidentally detected prenatally during routine ultrasound screening [6]. Neuroblastomas are usually asymptomatic and treatment is not necessary in the neonatal period, however they have the potential for a metastatic spread in utero, which may lead to further surgical resection [7]. Fetal Ultrasound (US) and Magnetic Resonance Imaging (MRI) presents a variety of imaging characteristics and appearances. Like many other neoplasms cross-sectional imaging is necessary for staging and evaluation of the organ of origin, extend of the mass lesion, local invasion of surrounding organs and structures, vascular involvement or displacement of vessels, calcifications, lymph nodes and metastases. Antenatal US is usually diagnostic and is performs as initial examination, however MRI is preferred method for detection of metastases [8]. In our case we present the different US and MRI features of a fetal neuroblastoma as an incidental finding in the 28-th week of gestation. We discuss the advantages of each imaging method in regard to patient counseling, management and postpartum follow-up. Informed consent was obtained from the patient to publish this case presentation.

Case Presentation

A 30-year-old female at 28-th gestational week was referred to our department for fetal MR examination following diagnostic ultrasonographic examination which revealed a fetal tumor close to the left kidney. The demand for MRI evaluation was addressed in order to differentiate between adrenal hemorrhage and suprarenal mass. The US examination revealed an oval lesion with solid and cystic character, located between the stomach and the left kidney, measuring 17/13mm at the parasagittal plane, without any signs of neovascularization. A rounded cystic portion is evident within the

lesion, without intracystic septations. The MR examination was performed on 3T Philips Ingenia MR unit using Single-Shot Turbo Spin Echo (SS-TSE) sequences in coronal, axial and sagittal plane, FOV 266x188mm, slice thickness 3mm and 4mm and matrix 512x256 (Figures 1,2). Fetal MR exam confirmed the extrarenal localization of this soft tissue mass measuring 17/11mm with a central cystic compound of 12/9mm at the level of left adrenal gland and confirms the diagnosis of neuroblastoma based on the macromorphology and the imaging features of the lesion. The Signal Intensity (SI) of the lesion was heterogenous and exclusively hyperintense on T2 - weighted images for the cystic component with lack of diffusion restriction on Diffusion-Weighted Images (DWI). The placenta was intact and located dorsally, no other defects or associated lesions were found in the fetal body and no evidence of metastases was suspected. The patient was informed about the adrenal origin of the lesion and was counseled to perform an US follow-up, which has been done in the 30-th week of pregnancy. No progression of size or change of US appearance was noted on follow-up, there was no signs of metastatic dissemination. Thus, makes easier the final parental decision to continue the pregnancy course and to elaborate a postnatal treatment plan depending on the further imaging results. Surgical resection was advised in case of tumor size progression.

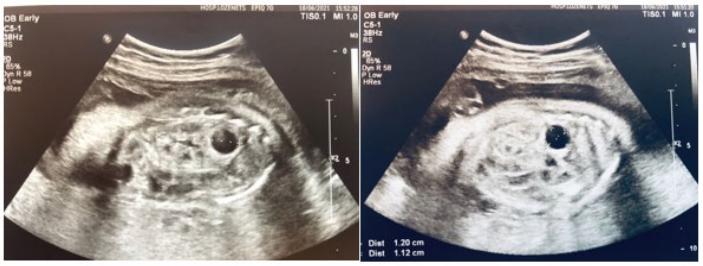

Figure 1: Ultrasonographic appearance of mixte neuroblastoma at 28 weeks of gestation (Figure 1a.b). No internal septations are present within the rounded cystic component of the lesion.

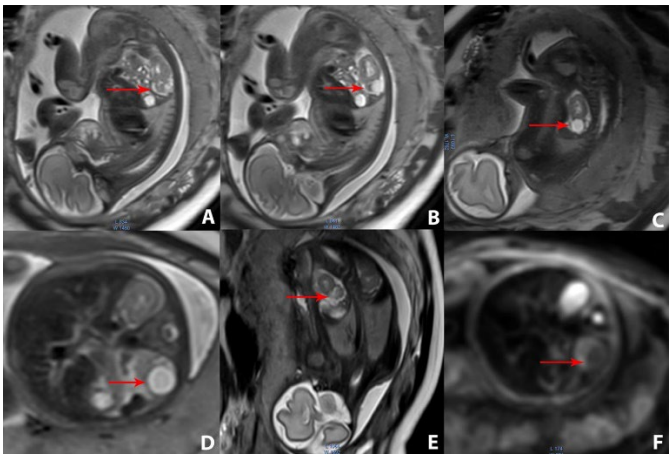

Figure 2: Sagittal T2 SS-TSE images show (Figure 2a,bc), Axial (Figure 2d), Coronal (Figure 2e) demonstrate well-defined solid (Figure 2a,e) and cystic (Figure 2b,c,d) nodular lesion in the left adrenal gland localization consistent with MR appearance of fetal neuroblastoma at 28 weeks of gestation. No diffusion restriction is evident on axial DWI images (Figure 2f).

Discussion

With average age of diagnosis at age of 2 years and nearly all cases diagnosed by age of 10 years, the diagnosis of fetal neuroblastoma could be a challenging task prenatally as these tumors are rarely observed in this period [9]. However, improvements in fetal imaging and widespread use of fetal ultrasonography leaded to higher rate of prenatal diagnosis of fetal neuroblastoma. These malignant tumors composed of immature neuroblasts occurs within the adrenal medulla and along the sympathetic chain ganglia [10]. Imaging findings of neuroblastomas shows a large, often ill-defined abdominal or retroperitoneal mass with or without compression and displacement of the kidney. When large enough it may cross the midline, displace and invade adjacent organs and structures and encase large vessels. Locoregional and

distant lymphadenopathy could be observed frequently. Metastatic dissemination in children varies between 50% and 70% at the time of discovery [2,11].

Imaging findings include a heterogenous infiltrative predominantly soft tissue mass which may contain echogenic calcifications, may encase rather than invade great vessels and may give metastases to liver and bones, rarely cutaneous metastases. High resolution prenatal ultrasound is is a safe, noninvasive imaging procedure and is usually used as initial imaging as it was performed in our case presentation. The features of neuroblastoma on the antenatal ultrasound are variable and range from cystic, mixed solid and cystic, and completely solid with or without calcification. The US findings consists of non-homogeneous mass with anechogenic parts depending on the cystic composition and presence of echogenic calcifications. In regards of the lesion size anterior displacement of the aorta could be observed as well as encasement of the mesenteric vessels. Doppler ultrasound is suitable to assess patency despite of the rare cases of vessel invasion [12]. MRI is a valuable complimentary technique and benefits with superior tissue resolution, which facilitates the correct depiction of the organ of origin and provide a better evaluation of potential metastatic dissemination and bone marrow infiltration [1,12]. In our case MRI was confirmatory for the diagnosis and excluded adrenal hemorrhage by terms of signal intensity characteristics. The centrally located cystic area presented hyperintensity and no sign of bleeding or hemosiderin deposition were detected on T1sequences. Although conventional MRI is most commonly used in fetal MRI, advanced MRI techniques such as Diffusion-Weighted Imaging (DWI) and Magnetic Resonance Spectroscopy (MRS) have been applied to fetal MRI [13-15]. Diffusion imaging of the fetal brain is useful tool for tumor processes [13] however, its clinical application is currently limited by its long acquisition times [14]. No diffusion restriction was evident on DWI sequences in our case. Future developments and improvements of fetal parallel imaging could be helpful in order to decrease the scan time as well as to increase the Signal to Noise Ratio (SNR) and image resolution [14].

The main differential diagnosis, as in our observation, opposes adrenal neuroblastoma and benign adrenal lesions like adrenal cysts or haemorrhage, the latter been the most common cause of adrenal mass during the perinatal period with a frequency of 1, 9/1000 live births [16]. Other secondary differentials that may be challenging include Wilm's tumor, teratoma, lymphoma and rhabdomyosarcoma and they differ in imaging characteristics. Usually patient management refers to "do not touch" lesion and pregnancy course are not affected, especially if no metastases are found as it was in our case with good prognosis and further follow-up which was uneventful. This wait and see strategy in performed in recent years in low stage neuroblastomas [16]. As other tumors, the prognosis of neuroblastoma is related to the age of the newborn, infant, the stage at presentation and the tumor site [10]. Neuroblastomas are graded in accordance to the International Neuroblastoma Staging System (INSS), however newer staging systems based on imaging instead of surgical have been proposed in the last decade [17]. Favorable prognosis occurs in newborns and infants less than 1 year old with low-stage of the disease at presentation and extra-abdominal tumor site. Some authors like Acharya et al. observe spontaneous regression of fetal neuroblastomas [18]. Similar results has been documented in mass-screening studies in infants by Brodeur [19].

Poor prognostic factors such as intratumoral necrosis in case of lactic acidosis and coagulopathy or diffuse metastases are described by Kume et al. [20]. Another study from George et al. describes a relationship between neuroblastoma and congenital heart defects and provides recommendation of echocardiography for congenital cardiovascular malformation screening in patients with newly diagnosed neuroblastoma [21]. Isaacs at al. found patients with cystic neuroblastoma to have a better outcome than noncystic tumors [22]. Similar results are described by Kozakewich et al. which thought cystic neuroblastoma to be a form of neuroblastoma in situ that is associated with highest survival rate of all forms of neuroblastoma [23]. The genetics of adrenal neuroblastomas is another present-day hot topic a part of the advances of modern imaging techniques. By observing higher rates of MYCN amplification, chromosome 1q gain, and chromosome 11q deletion among adrenal tumors Oldridge et al. conclude that adrenal neuroblastomas are more likely to harbor structural DNA aberrations including MYCN amplification compare to thoracic localization [24].

Conclusion

Reported prevalence of neuroblastoma cases in prenatally detected suprarenal masses ranges between 81% and 85 % according to the literature [25,26]. Antenatal ultrasound is first method of choice with variable features of neuroblastoma ranging from cystic, mixed solid and cystic, and completely solid with or without calcification. According our experience fetal MRI is a valuable complimentary technique in cases of US difficulties of determination of the site of origin, metastatic complication in the prenatal period and to rule out coexisting anomalies. The advantageous role of fetal MRI is powerful in cases of neuroblastoma mimickers such as adrenal hemorrhage, adrenal nodular hyperplasia, adrenal cyst and rarely adrenal carcinoma due to the better anatomical visualization and tissue resolution providing multiplanar imaging in a large field of view. In recent years the strategy of surgical exploration of adrenal masses has been replaced by "do not touch" strategy and follow-up in low stage tumors. In advanced grades with poor prognostic factors a combination of surgery, chemotherapy and radiation therapy is usually considered depending the stage at presentation.

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