Radiology Case Reports xxx (2016) 1–5

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: http://Elsevier.com/locate/radcr

Case Report

Diagnosis of neonatal neuroblastoma with postmortem magnetic resonance imaging

James Davis MD[a,*], Nathan Novotny MD[b], Jacqueline Macknis MD[c], Zeynep Alpay-Savasan MD[d], Luis F. Goncalves MD[a]

[a] Department of Diagnostic Radiology, Beaumont Hospital, Royal Oak, MI
[b] Department of Pediatric Surgery, Beaumont Hospital, Royal Oak, MI
[c] Department of Anatomic Pathology, Beaumont Hospital, Royal Oak, MI
[d] Department of Maternal Fetal Medicine, Beaumont Hospital, Royal Oak, MI

ARTICLE INFO

Article history:
Received 5 May 2016
Received in revised form
24 August 2016
Accepted 29 August 2016
Available online xxx

Keywords:
Fetal imaging
Postmortem imaging
MRI
Metastatic neuroblastoma
Virtual autopsy

ABSTRACT

Postmortem magnetic resonance imaging (MRI) is emerging as a valuable tool to accompany traditional autopsy and has potential for use in cases when traditional autopsy is not possible. This case report will review the use of postmortem MRI with limited tissue sampling to differentiate between metastatic neuroblastoma and hepatoblastoma which could not be clearly differentiated with prenatal ultrasound, prenatal MRI, or emergent postnatal ultrasound. The mother presented to our institution at 27 weeks gestation after an obstetric ultrasound at her obstetrician's office identified a large abdominal mass. Fetal ultrasonography and MRI confirmed the mass but were unable to differentiate between neuroblastoma and multifocal hepatoblastoma. The baby was delivered by cesarean section after nonreassuring heart tones led to an emergent cesarean section. The baby underwent decompressive laparotomy to relieve an abdominal compartment syndrome; however, the family eventually decided to withdraw life support. At this time, we performed a whole body postmortem MRI which further characterized the mass as an adrenal neuroblastoma which was confirmed with limited tissue sampling. Postmortem MRI was especially helpful in this case, as the patient's family declined traditional autopsy.

Introduction

Postmortem magnetic resonance imaging (MRI) is emerging as a valuable tool to accompany traditional autopsy. This case report will review the use of postmortem MRI with limited tissue sampling to differentiate between metastatic neuroblastoma and hepatoblastoma which could not be clearly differentiated with prenatal ultrasound, prenatal MRI, or emergent postnatal ultrasound. Postmortem MRI was especially helpful in this case, as the patient's family declined

Competing Interests: The authors have declared that no competing interests exist.

* *Corresponding author.*
E-mail address: james.davis@beaumont.org (J. Davis).

http://dx.doi.org/10.1016/j.radcr.2016.08.019

traditional autopsy. The clinical scenario, in-utero imaging, and postmortem imaging will be discussed and compared with pathology. Finally, conclusions regarding the utility of postmortem MRI will be drawn.

Case report

A 33-year-old G1 P0 with history of multiple unsuccessful infertility treatments presented to William Beaumont Hospital at 13 weeks and 4 days gestation with heavy vaginal bleeding. She was examined in the emergency center, and pelvic ultrasonography revealed a single-live intrauterine pregnancy with subchorionic hemorrhage. At 19 weeks gestation, a routine anatomy scan revealed no fetal abnormalities. At 27 weeks gestation, she returned to the hospital after an ultrasound in her obstetrician's office identified a fetal abdominal mass.

An obstetrical ultrasound demonstrated a live intrauterine pregnancy with a large mass compressing the liver and kidneys along with bilateral hydronephrosis and ascites. The origin of the mass was indeterminate sonographically; however, retroperitoneal origin was favored based on the relative displacement of the abdominal organs (Figs. 1A and B).

At 28 weeks gestation, a fetal MRI was performed which also revealed an intra-abdominal mass, measuring 9.4 × 6.8 × 8.6 cm, likely arising from the retroperitoneum Fig. 2. The mass was displacing the liver and kidneys and demonstrated restricted diffusion. There was hepatic heterogeneity, likely representing metastatic disease or possibly multifocal hepatoblastoma. In addition, there was significant skin thickening and placentomegaly consistent with hydrops. At this time, a biophysical profile was performed which was scored as 2/8, and the patient was transferred to the high-risk obstetrical unit.

On August 25, 2015, nonreassuring fetal heart tones prompted delivery by low transverse cesarean section. Both the mother and baby survived delivery; however, the baby required intubation for oxygen desaturation. An emergent ultrasound performed at the bedside also failed to determine the origin of the tumor conclusively. There was sliding of the liver over the mass with respiratory movements, suggesting that the mass did not originate from the liver. An emergent bedside decompressive laparotomy was performed to treat the abdominal compartment syndrome. The laparotomy revealed a tumor arising in the right abdomen, extending well into the left abdomen. Liver metastases were noted. As the patient was profoundly coagulopathic, no biopsy or resection was possible, and a makeshift vacuum-assisted device was used to manage his open abdomen (Figs. 3A and B).

Renal failure and difficulty with ventilation led to irreversible acidosis. After a lengthy discussion, the family decided to withdraw support on August 26, 2015. On August 27, 2015, a postmortem full body MRI was performed along with limited tissue sampling. Three dimensional T1-weighted, T2-weighted, and T2*-weighted sequences were obtained on a Philips 3T scanner with voxel size of 0.6 × 0.6 × 0.6 mm (slice thickness), then reconstructed in the sagittal, coronal, and transverse planes.

A 9.3 × 6.2 × 6.7-cm heterogeneous mass was identified. It was predominantly hyperintense on T1 and hypointense on T2, with extensive blooming artifacts on T2*, consistent with internal calcifications. With the improved resolution of postmortem imaging, it was clear that the mass originated from the right adrenal gland and crossed the midline, displacing both kidneys and the liver. The left adrenal gland was normal. The liver contained multiple round lesions which had high T1 signal, low T2 signal, and blooming artifact on T2*, consistent with metastatic disease (Figs. 4A-C).

The parents consented to the procurement of neoplastic tissue for diagnostic purposes only. On gross pathologic evaluation, the abdomen was bulging, measuring 32 cm in circumference. The abdomen was opened, revealing a protruding liver with tan-white nodules and areas of necrosis and hemorrhage. A biopsy specimen was procured from the liver demonstrating a small round blue cell tumor with positivity for neuroendocrine markers (chromogranin and synaptophysin). Morphology was consistent with a stroma-poor, poorly differentiated neuroblastoma with favorable histology (Fig. 5). It was also noted that there were multiple cross sections of the placenta that demonstrated metastasizing small round blue tumor cells within the fetal vessels.

Discussion

Neuroblastoma is the most common solid extracranial tumor of infants and children. Up to 50% of neuroblastomas are diagnosed before the age of 2 but can be diagnosed later in childhood as well. The earliest documentation of prenatal diagnosis of neuroblastoma was in 1983 by Fenart et al when a 1.4-cm hyperechoic adrenal mass was identified, and then removed following birth [1]. Since then, many case reports of fetal neuroblastoma have been published; however, relatively few demonstrate metastatic disease [2]. Improvements in prenatal imaging and widespread use of fetal ultrasonography have led to an increased rate of prenatal diagnosis of fetal neuroblastoma. 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. Patients with cystic neuroblastoma have a better outcome than those noncystic tumors [3]. Despite metastatic disease and large tumor burden, the prognosis is often considered favorable. Most notably, in patients with stage 4S neuroblastoma, which includes metastases to the liver, skin, or bone marrow, patients may have complete spontaneous regression of the tumor [4]. Ninety percent of fetal neuroblastomas arise in the adrenal glands, but can arise anywhere along the sympathetic chain. When a suprarenal mass is identified in utero, primary differential considerations often include neuroblastoma, hepatoblastoma, rhabdomyosarcoma, extralobar bronchopulmonary sequestration, and possibly adrenal hemorrhage. Ultrasound can often narrow the differential, and in cases where the diagnosis is not clear, fetal MRI can be of diagnostic aid [2,5].

The typical gross pathologic, histologic, and MRI characteristics of neuroblastomas are discussed in the following paragraphs.