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LARGE HEPATIC HEMANGIOMA COMPLICATED WITH SEVERE POLYHYDRAMNIOS AND KASABACH-MERRITT SYNDROME

VELIKI JETRENI HEMANGIOM KOMPLICIRAN TEŠKIM POLIHIDRAMNIONOM I KASABACH-MERRITTOVIM SINDROMOM

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Case report

Key words: hepatic hemangioma, polyhydramnios, Kasabach-Merritt syndrome

Summary. Infantile hepatic hemangioma is a benign hepatic tumor that can be associated with life-threatening perinatal complications such as output cardiac failure or hydrops. We report a case of large hepatic hemangioma prenatally suspected by two-dimensional ultrasound at 35 weeks of gestation in 24-year-old secundipara. Hemangioma turned to be complicated with severe polyhydramnios and Kasabach-Merritt syndrome. The baby was operated on the second day after birth and the diagnosis was confirmed by histopathology. This is a first case of hepatic hemangioma to be suspected by antenatal sonographic examination and subsequently reported in Qatar.

Prikaz slučaja

Ključne riječi: jetreni hemangiom, polihidramnij, Kasabach-Merrittov sindrom

Sažetak. Novorođenački jetreni hemangiom je dobroćudni tumor jetre koji može biti združen s perinatalnim komplikacijama opasnim po život, kao što su zatajenje srca ili fetalni hidrops. Opisujemo slučaj velikog jetrenog hemangioma na koji se prenatalno postavila sumnja prilikom dvodimenzionalnog ultrazvučnog pregleda u 35 tjednu trudnoće kod 24-godišnje drugorotkinje. Hemangiom je bio kompliciran ekstenzivnim polihidramnijem i Kasabach-Merrittovim sindromom. Dijete je operirano drugog dana života i dijagnoza hemangioma potvrđena je histopatološkim pregledom. Ovo je prvi opisani slučaj jetrenog hemangioma na koji se posumnjalo antenatalnim ultrazvučnim pregledom u Kataru.

Introduction

Hepatic hemangioma (hemangioendothelioma) is the most frequently observed hepatic tumor of early infancy.[1] Large fetal liver hemangiomas may cause severe perinatal complications, particularly high-output cardiac failure and/or Kasabach-Merritt syndrome.[2] It is a rare disease, usually of infants, in which a vascular tumor leads to decreased platelet counts and sometimes other bleeding problems, which can be life-threatening. It is also known as hemangioma thrombocytopenia syndrome.[2] Polyhydramnios (in absence of other fetal, maternal and placental causes) is reported to be a rare complication of these large tumors.[3] In contrast, isolated small hepatic hemangiomas are not associated with any of these fetal and postnatal sequels. In this report, we present a case of large hepatic hemangioma complicated with severe polyhydramnios and Kasabach-Merritt syndrome.

Case report

A 24-year-old secundipara was referred to Department of Obstetrics and Gynecology, in Women's Hospital in Doha at 35 weeks of gestation with ultrasound scan suspecting the tumor of unknown origin in fetal hemi-abdomen. Sonographic examination demonstrated a singleton fetus with normal cardiac activity and fetal movements. The fetal biometry corresponded to the 50-th percentile of standardized biometry for 35 weeks of gestation. The placenta and amniotic fluid were normal with amniotic fluid index (AFI) of 7 cm. Detailed sonographic examination revealed a large hyperechoic abdominal mass, closely adjacent to the left

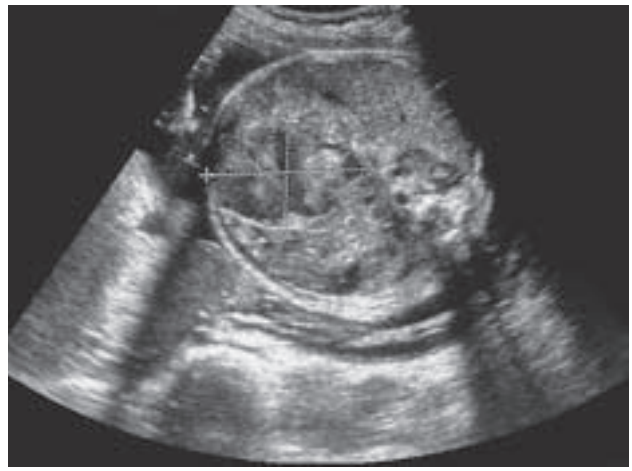

Figure 1. Two-dimensional ultrasonographic picture of fetal abdomen with lines showing hyperechoic abdominal mass (6.7×4.2 cm), closely adjacent to the left kidney and left liver lobe

Slika 1. Dvodimenzionalna ultrazvučna slika fetalnog trbuha s crticama koje pokazuju hiperehogenu tvorbu veličine 6,2×4,2 cm, tijesno uz lijevi bubreg i lijevi jetreni režanj

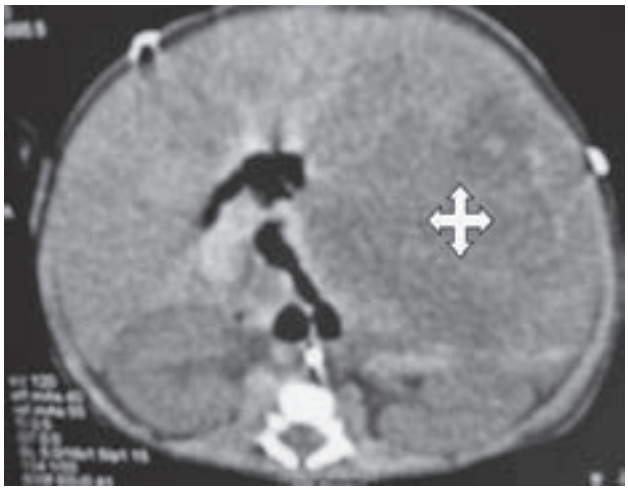

Figure 2. Computed tomography scan of infant's abdomen with arrow showing heterogeneous mass sized 8.5×8.0×6.5cm anterior to left kidney, clearly separable with minimal areas of hyper densities, indicating a hepatic origin of the mass (left lobe)

Slika 2. Kompjuterizirana tomografska slika dječjeg trbuha s križem koji pokazuje heterogenu tvorbu veličine 8,5×8,0×6,5 cm, ispred lijevog bubrega, jasno ograničenu, s minimalnim područjima pojačane gustoće, što upućuje na jetreno porijeklo tvorbe

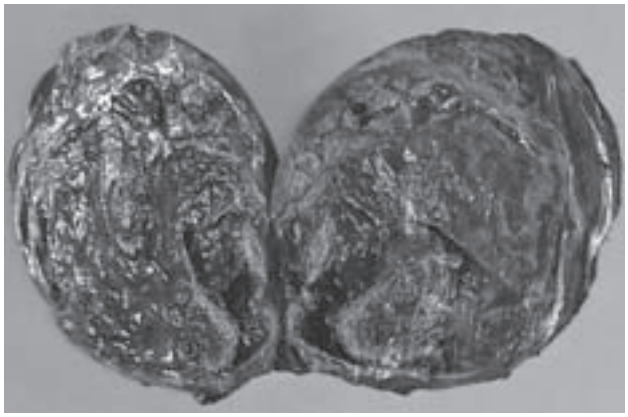

Figure 3. Macroscopic appearance of dissected liver hemangioma: solid structure of tumor can be seen

Slika 3. Makroskopski izgled reseciranog jetrenog hemangioma: može se vidjeti čvrsta struktura tumora

kidney and left liver lobe, measuring 6.7×4.2 cm *(Figure 1)*. Color Doppler demonstrated only a vascular tumor capsule. There was no evidence of fetal hydrops and the rest of the fetal anatomy appeared normal. Differential diagnosis included either benign tumor of adrenal gland or liver. Since the fetus was male ovarian etiology was ruled out. Three days later the patient was urgently admitted to the Department because of symptomatic polyhydramnios (AFI 25.8 cm) and non reassuring cardiotocography (CTG). Decision was taken to augment her labor on 36 weeks with Syntocinon infusion and controlled amniotomy. She delivered vaginally a boy weighting 3340 grams with Apgar scores 6 and 8. At birth there was no evidence of cutaneous hemangiomas. The baby was postnatally karyotyped and the findings were normal. 4 hours after the birth the baby was found to have low thrombocytopenic platelets count (76×10^9) and soon developed cholestatic jaundice (direct bilirubinemia of 212 mmol); the child was diagnosed to have Kasabach-Merritt syndrome.

On a second postnatal day urgent ultrasound showed a mixed echogenic lesion anteroposterior to the left kidney without significant vascularity. Computed tomography (CT) scan showed large heterogeneous mass sized 8.5×8.0×6.5 cm anterior to left kidney, clearly separable with minimal areas of hyperdensities, indicating a hepatic origin of the mass (left lobe) *(Figure 2)*. The probable diagnosis was hepatic hemangioma. Baby was operated on the second day after birth. A complete resection of a tumor measuring around 8×7cm was performed *(Figure 3)*. Histopathology report confirmed the diagnosis of infantile hepatic hemangioendothelioma.

Discussion

Hepatic hemangioma (HH) is the most common tumor of the liver during early infancy. It occurs in up to 10 percent of Caucasian infants, and is generally noticed within the first few days to months of life. HH is of mesenchymal origin and usually solitary, but the actual etiology remains unknown.[4] Mostly, they are composed of masses of arterial and venous connections within the liver. As a result of low resistance flow, high output cardiac failure and hydrops often occur. In one series of 16 infants presenting with HHs within four months of birth, 58 percent were diagnosed with high output cardiac failure.[4] Kasabach-Merritt syndrome is another life-threatening perinatal complication (severe thrombocytopenia, consumptive coagulopathy, and hemolytic anemia), which results from platelets and clotting factors trapping within a large HHs. Polyhydramnios (in absence of other fetal, maternal and placental causes) is rare complication of these large tumors, described only once in the reviewed literature.[3] Etiology of polydramnios in these cases is unknown; probable cause could be gut compression. In contrast, isolated small tumors do not appear to be associated with any of these fetal and postnatal sequels. HHs occurring during the antenatal period is difficult to diagnose and the severe forms discovered by sonography have a poor prognosis.[3,5] Once a baby with a suspected HH is born, attention should focus on establishing a definitive diagnose. In the cases of HHs, more than 50% will have associated cutaneous hemangiomas. The infant's platelet count, fibrinogen, and fibrin split products should be checked to exclude disseminated intravascular coagulation and platelet trapping. An initial bedside ultrasonogram may be helpful in establishing the diagnosis, but CT or magnetic resonance imaging (MRI) scans are usually indicated to more fully define these lesions.[6] Postnatal treatment for these patients can be either surgical (hepatic artery embolization or ligation, liver resection, liver transplantation) or non-surgical (corticosteroids, interferon-α, radiotherapy). However, if HH is diagnosed during pregnancy, compromised preterm fetus can be treated in-utero by maternal corticosteroid administration.[7] In conclusion, if a large hyperechogenic mass of

the suspected liver origin is detected prenatally and complicated with (severe) polyhydramnios, diagnosis of hepatic hemangioma should be considered. In order to prevent serious cardiovascular complication, continual clinical surveillance is indicated.

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