DOI: 10.3179/jjmu.JJMU.A.126

◇ CASE REPORT ◇

# シャント術で胎児水腫を改善させ得ず Mirror 症候群の増悪から児を救命できなかった Macrocystic CPAM の 1 例

泉　りりこ　　日高　庸博　　城戸　　咲　　甲斐翔太朗　　中野　嵩大
蜂須賀正紘　　加藤　聖子

**抄　　録**

39 歳の 1 回経産婦．Macrocystic type の胎児先天性肺気道形成異常（CPAM）の診断で妊娠 20 週に当院へ紹介された．超音波断層法で胎児左肺を占拠する 56 × 37 × 26 mm の多房性嚢胞性病変を認め，CPAM volume ratio（CVR）は 1. 61 と算出された．少量の胎児腹水を認め，胎盤は肥厚していた．妊娠 22 週に入院した際には胎児皮下浮腫も認められ，胎児水腫と診断した．数日前から出現した著明な悪心と嘔吐の訴えがあり，母体顔面と手背に浮腫を認めた．血液検査で，軽度の貧血（Hb 10. 2 g/dL）と血小板数低値（10. 0 万/μl），低アルブミン血症を認め，血中 hCG は 178, 077 mIU/ml と高値であった．Mirror 症候群と診断した．妊娠 22 週 5 日に嚢胞羊水腔シャント術を行った．ただし，多房性であったが故に一定サイズの嚢胞が残存することとなり，妊娠 23 週 1 日の CVR は 1. 20 であった．胎児胸壁の皮下浮腫は消失せず，腹水に減少を認めなかった．妊娠 23 週 5 日，一過性に母体の酸素化不良となり，胸部X線で両側胸水貯留と肺うっ血，心拡大を認めた．血液検査で血液希釈が進行しており，尿量も減少した．妊娠継続は困難と判断し，分娩誘発の方針とした．うっ血性心不全に対してアルブミンやフロセミドの投与を行いながら，妊娠 24 週 2 日，経腟分娩に至った．児は 860 g の女児で早期新生児死亡となった．胎盤重量は 725 g で浮腫著明であった．産後母体症状は速やかに改善した．巨大 macrocystic CPAM が多房性である場合の問題点として，シャント術後にも腫瘤が残存し，その結果として胎児水腫の改善にも至らないケースがあることを認識した．

# A case of fetal macrocystic CPAM in which associated hydrops did not resolve despite fetal therapy resulting in worsened mirror syndrome and early neonatal death

Ririko IZUMI, Nobuhiro HIDAKA, FJSUM, Saki KIDO, Shotaro KAI, Takahiro NAKANO, Masahiro HACHISUGA, Kiyoko KATO

**Abstract**

A 39-year-old mother was referred to us at 20 gestational weeks owing to a large fetal left lung multilocular mass（macrocystic CPAM）and minimal ascites. The overall size of the mass lesion corresponded to a CPAM volume ratio（CVR）of 1. 61. Placentomegaly was also noted. On admission at 22 weeks' gestation, the fetus exhibited skin edema, and hydrops fetalis was diagnosed. The patient demonstrated fairly stable vital signs, but complained of emesis and vomiting that had developed over the last several days. Upon physical examination, she exhibited edema of the face and legs. Moreover, the patient's laboratory values showed anemia, low platelet counts, and hypoproteinemia. The serum $\beta$-hCG concentration was 178, 077 mIU/ml. Based on the above results, mirror syndrome was diagnosed. The mother underwent thoracoamniotic shunting at $22^{+5}$ gestational weeks, with successful drainage of the largest cyst. However, a fair-sized mass composed of multiple small cysts persisted, and the CVR was calculated as 1. 20. Fetal pleural skin edema became somewhat less severe, but neither fetal ascites nor placentomegaly improved. At $23^{+5}$ gestational weeks, the mother complained of breathing difficulties. Chest X-ray demonstrated bilateral pleural effusion and cardiomegaly. Maternal hypoproteinemia and edema had gradually worsened, and the patient showed a decrease in urinary volume. We determined that continuation of the pregnancy was not ideal, and induced labor under administration of albumin and diuretics. At $24^{+2}$ gestational weeks, a female infant weighing 860 g was delivered and died immediately after birth. The edematous placenta weighed 725 g. After delivery, the mother's condition improved. Based on this experience, we have re-affirmed that successful drainage of the largest cyst does not necessarily improve the hydropic state of fetuses with multilocular macrocystic CPAM. The fetal and maternal condition should both be treated with caution via physical examination, hematological examination, and chest radiography.

**Keywords**

congenital pulmonary airway malformation, hydrops fetalis, mirror syndrome, shunting operation

九州大学病院産科婦人科
Department of Obstetrics and Gynecology, Kyushu University Hospital, 3−1−1 Maidashi, Higashi, Fukuoka 812−8582, Japan
Received on September 7, 2018; Revision accepted on October 11, 2018　J-STAGE. Advanced published. date: December 3, 2018