当院で管理した胎児脊髄髄膜瘤の診断背景と生後予後

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抄 録

目的: 脊髄髄膜瘤(myelomeningocele:MMC)胎児の神経学的予後改善を目的とした妊娠 26 週未満での胎児治療の早期安全性試験が本邦でも始まった。本研究では当院で管理した MMC 胎児の出生前診断背景と生後予後を検討し、今後の課題を明らかにする。方法: 2014 年 11 月から 2022 年 8 月に当院で妊娠管理した胎児 MMC 症例を対象とし後方視的に検討した。出生前診断時期,診断契機となった超音波所見、生命・運動予後を主要評価項目とした。結果: 対象は 26 例あり、紹介週数の中央値は妊娠 25 週(11 - 34 週)、11 例(42%)は 26 週以降に紹介されていた。5 例は 22 週未満で診断され、うち 3 例は人工妊娠中絶となった。紹介契機は MMC 単独が 11 例、脳室拡大単独が 5 例、脊椎所見および脳室拡大が 6 例、レモンサイン・バナナサインが 1 例、他の合併疾患が 3 例であった。中絶例を除いた 23 例の分娩週数中央値は 38 週(26 - 40 週)で、生後フォローアップ期間の中央値 23 か月(0 - 67 か月)において全例生存していたが、14 例(61%)で運動機能障害を指摘された。生後 18 か月以上を経過した児 15 例中 10 例(67%)が神経因性膀胱と診断、自己導尿を必要とした。結論:胎児診断された MMC 症例は生後に運動機能障害を呈するものも多い。胎児診断の時期が現状では十分早いとは言えず今後診断時期を早める必要性が認識された。

Diagnosis and postnatal prognosis of fetuses with myelomeningocele managed at our institution

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Abstract

Purpose: A safety trial for early fetal treatment before 26 gestational weeks to improve the neurological prognosis of fetuses with myelomeningocele (MMC) was conducted in Japan. The aim of the study was to examine the prenatal diagnostic background and postnatal outcomes of fetuses with MMC at our institution and identify future challenges for fetal treatment. Methods: We retrospectively reviewed the medical records of MMC cases in our institution encountered between November 2014 and August 2022. The timing of prenatal diagnosis, ultrasound findings leading to diagnosis, and the vital/motor prognosis were evaluated. Results: The study included 26 cases, all of which were referred from other institutions. The median gestational age at referral was 25 weeks (range: 11-34 weeks), with 11 cases (42%) being referred after 26 weeks. Five cases were diagnosed before 22 weeks, of which three opted for elective termination. The referral indications were isolated MMC in 11 cases, isolated ventriculomegaly in five cases, combined spine and ventricular abnormalities in six cases, lemon and banana signs in one case, and other associated conditions in three cases. Excluding pregnancy terminations, the median gestational age at delivery was 38 weeks (range: 26-40 weeks). At a median follow-up of 23 months (range: 0 - 67 months), all cases survived; however, 14 cases (61%) exhibited motor function impairment. Ten of 15 children (67%) who were at least 18 months of age were diagnosed with neurogenic bladder or required self-urination. Conclusion: While the vital prognosis of fetuses with MMC is favorable, many exhibit motor function impairment postnatally. The current timing of prenatal diagnosis is not sufficiently early, highlighting the need to expedite the diagnosis.

Keywords

fetal myelomeningocele, fetal surgery, prenatal diagnosis

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