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双胞胎肝母细胞瘤3例报告并文献复习

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Hepatoblastoma in Twins: A Report of Three Cases with Literature Review

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摘要 分析肝母细胞瘤的家族遗传倾向, 为临床遗传咨询及早诊断、早治疗提供依据。方法: 收集中国医科大学附属盛京医院2005年6月至2011年11月间收治的3例双胞胎肝母细胞瘤患儿的资料, 结合患儿生化、影像、治疗方案、生存期、家族史及相关文献报道分析遗传相关因素。结果: 本文中3例双胞胎患儿均为单发, 无家族史, 其中1例为早产儿。3例患儿均采用手术治疗, 经病理证实为肝母细胞瘤, 术后1例患儿因失血性休克死亡, 余2例采用不同的化疗方案, 均获得完全缓解。自1969年至今, 已有8例关于兄弟姐妹同患此病的文献报道。8组文献中5组患儿家族中有肿瘤(多数为结肠癌)及家族性结肠息肉病史, 其中3组还有基因异常改变。结论: 肝母细胞瘤是一种罕见的小儿肝脏恶性肿瘤, 具有家族遗传倾向, 与结肠癌及家族性结肠息肉综合征密不可分, 在诊疗过程中, 有必要加强家属的认知, 做好预防保健, 争取早发现早诊断早治疗。

关键词: [肝母细胞瘤](#) [双胞胎](#) [单发](#)

Abstract: To analyze genetic predisposition to hepatoblastoma, to provide clinical genetic counseling, and to encourage early detection, early diagnosis, and early treatment. Methods: A retrospective analysis was performed based on the clinical records of three hepatoblastoma cases among twins from June 2005 to November 2011. The analysis consisted of biochemical data, images, therapeutic schedules, survival, family history, and analysis of the literature related to genetic factors. Results: Only eight articles have been published on hepatoblastoma among siblings since 1969 when Fraumeni first reported hepatoblastoma in infant sisters. Five groups of children in the literature had familial histories of tumors (mostly colon cancer) and familial colon polyps syndrome, including three groups with abnormal gene change. In this article, three cases of hepatoblastoma in twins were reported. The tumor only affected one of the twins that had no family history of the disease. The present report included one case of premature infants and two patients who underwent a surgical operation combined with chemotherapy. They achieved complete remission, but one died shortly after the surgical operation because of hemorrhagic shock. Conclusion: Hepatoblastoma is a rare malignant tumor in children associated with colon cancer and familial colon polyp syndrome. The disease has a genetic predisposition. Therefore, clinical doctors should help family members understanding the disease, facilitate preventive care, and strive for the early detection, early diagnosis, and treatment.

Key words: [Hepatoblastoma](#) [Twins](#) [Solitary](#)

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