

论著

骨髓移植治疗Duchenne型肌营养不良模型鼠的实验研究

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摘要 目的: 观察骨髓移植治疗Duchenne型肌营养不良(DMD)模型鼠(mdx鼠)后,骨骼肌中抗肌萎缩蛋白的表达,以及病理、生理和运动功能的变化情况,从而正确评价骨髓移植治疗DMD的疗效。方法: 以正常C57鼠作为供者,以致死剂量放疗后的mdx鼠作为受者进行骨髓移植,在移植后4个月和6个月分别用荧光免疫组化法观察抗肌萎缩蛋白表达情况,用HE染色观察并计算骨骼肌细胞核中心移位纤维比例(CNF),同时进行mdx鼠腓肠肌电生理检查和运动功能检测。结果: 骨髓移植后的mdx鼠骨骼肌细胞膜上有部分抗肌萎缩蛋白表达,CNF比例和肌肉湿重下降,mdx鼠的被动运动功能增强。结论: 骨髓移植后,骨髓干细胞在mdx鼠体内的骨骼肌和骨髓中定居,并分化成可以表达抗肌萎缩蛋白的骨骼肌细胞,使mdx鼠病理、生理得到部分改善,最终导致了肌肉的运动功能的增强。说明骨髓移植治疗DMD是有一定价值的,为临床应用提供实验基础。

关键词 [骨髓移植](#); [干细胞](#); [肌营养不良症](#)

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Experimental treatment of the model mice of Duchenne muscular dystrophy by bone marrow transplantation

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Abstract

AIM: To detect dystrophin expression in skeletal muscles of mdx mice after bone marrow transplantation (BMT), and to evaluate the effect of BMT on Duchenne muscular dystrophy (DMD). METHODS: Bone marrow cells were cultured for three days, and then transplanted into mdx mice irradiated lethally through tail veins. After 4 and 6 months, dystrophin expression on myocytes membranes in mdx mice was detected by fluorescent immunohistochemical staining. The centrally nucleated fibers (CNF) were calculated by HE staining, and the physiologic parameters measured and the motor function detected by traction test, rotating rods test and rotating wheels test were also observed. RESULTS: Until 4 and 6 months after BMT, dystrophin was expressed partly on myocytes membranes in mdx mice, and the ratio of CNF decreased, physiologic functions improved, the motor ability reinforced in treated group. CONCLUSION: After BMT, marrow stem cells settled in injured skeletal muscles and bone marrow, then differentiated into myocytes with dystrophin expression and caused the improvement of pathology, physiology and motor function in treated group finally. These results give a powerful proof for the treatment of DMD with BMT.

Key words [Bone marrow transplantation](#) [Stem cells](#) [Muscular Dystrophy](#)

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