总之,电影磁共振成像能清楚显示DCM的主要病理和病理生理变化,并可准确测量患者的心功能状况,重复性好。有助于DCM的全面诊断和疗效的随访观察。

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Ⅱ型糖原累积病、多发性肌炎患者骨骼肌的超微结构研究

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病例1,女,23岁,因心慌、气短八年余,全身乏力五年余而入院。查体:显示全身躯干肌及四肢肌萎缩,以四肢远端为明显,肌张力降低,入院后按重症肌无力治疗未见好转。

病例2,女,45岁,因心慌、气短二年余,低热、全身乏力,肌痛,出现下蹲起立与上楼困难一年余入院,入院后按心肌炎治疗,病情不见好转。二位患者肌电图示肌原性疾病改变。

肌肉活检标本分别取自三角肌和腓肠肌,按常规方法制备电镜标本,PHILIPS-CM$_{10}$透射电镜观察。

镜下显示:Ⅱ型糖原累积病,肌细胞的肌浆内有大量的糖原颗粒聚集,特别是肌膜下明显,肌原纤维之间和肌丝之间可见糖原聚集,糖原在肌细胞内游离存在。在肌纤维萎缩时,可见原纤维消失处有较多的糖原颗粒充填,亦可见到玫瑰花型糖原颗粒。细胞器移位、受压及被糖原颗粒代替,线粒体增多、肿胀,常见空泡化。少数肌纤维坏死。间质纤维结缔组织及脂肪组织增生。

电镜下显示:多发性肌炎,肌细胞的肌原纤维排列紊乱,肌纤维溶解,肌丝断裂,肌原纤维Z带增粗和变形,Z带物质增多。有的Z带破坏,线粒体明显肿胀和空泡化。内皮细胞及基底膜增厚,肌浆内常可见到淋巴细胞和浆细胞侵润,有的肌细胞胞浆内和胞核内可见与病毒相似的颗粒。

讨论 Ⅱ型糖原累积性疾病和多发性肌炎初起临床症状相混淆,不易区别,而通过肌肉活检,电镜下超微结构形态特点即可获得肯定诊断。Ⅱ型糖原累积病属常染色体隐性遗传代谢障碍,病变主要侵犯骨骼肌,系酸性麦芽糖酶缺乏所致,由于此酶的缺乏,糖原不能被分解葡萄糖,便在肌细胞内大量的聚集。而多发性肌炎系弥漫性疾病。目前文献倾向于免疫性疾病。文献报道在多发性肌炎病人的肌肉中见到似病毒颗粒、细小病毒(picornavirus)包含体,作者在肌细胞浆内和细胞核内可见病毒相似颗粒,因此,病毒感染可能是多发性肌炎的病因,超微结构显示其各自的形态学特征,在病理诊断和鉴别诊断具有一定应用价值。

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