·病例报告·

帕金森综合征合并肌萎缩侧索硬化症一例

胡雅娉 郭宇 胡艺漾 梁战华

【关键词】 帕金森障碍； 肌萎缩侧索硬化； 病例报告
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Parkinsonian syndrome combined with amyotrophic lateral sclerosis: one case report

HU Ya-ping, GUO Yu, HU Yi-ying, LIANG Zhan-hua
Department of Neurology, the First Affiliated Hospital of Dalian Medical University, Dalian 116011, Liaoning, China
Corresponding author: LIANG Zhan-hua (Email: zhanhualiang@163.com)

患者 男性，67岁，主因进行性左侧肢体运动迟缓1年，于2013年7月24日入院。患者1年余前无明显诱因出现左手动作迟缓，如擀饺子皮时左手旋转面皮等精细动作不如以前灵活，后逐渐累及左下肢，表现为行走时左下肢拖拉，为求进一步诊断与治疗，至我院就诊。患者近年来常出现便秘，偶有尿急。既往慢性萎缩性胃炎病史40余年，复发性阿佛他溃疡40余年。余无异常。

体格检查 患者体温36.2 ℃，呼吸16次/min，脉搏72次/min，血压140/80 mm Hg（1 mm Hg = 0.133 kPa）。发育良好，体型消瘦，神志清楚，语言流利、说话声音欠洪亮，精神和智力未见明显异常，面具脸，伸舌偏右，咽反射存在，舌肌束颤、右侧欠饱满。左上肢肌力4级、肌张力1^{+}级，其余肢体肌力5级、肌张力0级。紧张或用力时双上肢出现不自主抖动，静止性震颤不明显，双上肢腱反射亢进，右侧Babinski征阳性，左侧Chaddock征阳性、Hoffman征阳性，无体位性低血压。

诊断与治疗经过 实验室检查：血尿常规、心肌酶谱、肝肾功能试验、甲状腺功能试验、免疫球蛋白三项、补体C3和C4、抗核抗体（ANA）谱、抗双链DNA抗体（dsDNA）、狼疮细胞、红细胞沉降率（ESR）、血清镁均于正常值范围。血清梅毒螺旋体（TP）抗体、人类免疫缺陷病毒（HIV）抗体均呈阴性。血清肿瘤标志物筛查呈阴性。腰椎穿刺脑脊液外观清亮、透明，压力140 mm H_2O（1 mm H_2O = 9.81 × 10^{-3} kPa，80～180 mm H_2O），白细胞计数2 × 10^6/L［（0～8）× 10^6/L］，蛋白定量0.58 g/L（0.12～0.60 g/L）、葡萄糖2.61 mmol/L（2.20～3.90 mmol/L）、氯化物水平132 mmol/L（120～132 mmol/L），IgG 46.40 mg/L（0～34 mg/L），余未见明显异常。神经心理学测验：简易智能状态检查量表（MMSE）评分为25分，无痴呆表现；蒙特利尔认知评价量表（MoCA）评分22分，有认知功能障碍表现。影像学检查：腹部超声显示膀胱残留尿量50 ml。头部MRI显示，双侧半卵圆中心轻度脱髓鞘改变，扩散张量成像（DTI）可见中央前回皮质、皮质下白质和半卵圆中心部分各向异性（FA）值降低（图1）。颈腰椎MRI显示，椎间盘膨出，但未见明显脊髓压迫或椎管狭窄。^{18}F-脱氧葡萄糖（^{18}F-FDG）PET显示，右侧豆状核葡萄糖代谢降低（图2）。肌电图提示四肢肌肉、胸锁乳突肌、胸段脊旁肌、腹直肌广泛神经源性损害可能性大；4个月后复查肌电图仍提示广泛神经源性损害，符合肌萎缩侧索硬化症（ALS）表现，且对应的神经传导速度（NCV）减慢、波幅降低，提示前角细胞损害加重，表明肌萎缩侧索硬化症进展（图3）。临床诊断：帕金森综合征合并肌萎缩侧索硬化症。予普拉克索0.50 mg（3次/d）、多巴丝肼（美多芭）187.50 mg（3次/d，空腹）、巴氯芬10 mg（3次/d）口服改善帕金森病症状。住院16 d，出院时一般情况良好，生命体征平稳，自觉左侧肢体欠灵活较前稍改善，但神经系统查体与入院时大致相同。出院后继续按照上述方案治疗，治疗4个月，患者自觉症状稍改善，神经系统检查：左上肢肌力4级、肌张力

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作者单位：116011 大连医科大学附属第一医院神经内科
通讯作者：梁战华（Email：zhanhualiang@163.com）

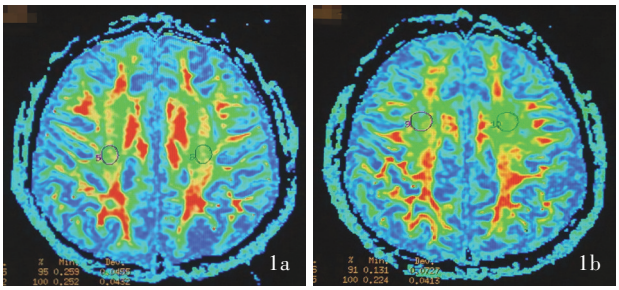

图1 头部DTI检查所见 1a 近侧脑室顶部层面双侧半卵圆中心FA值分别为0.37(左侧)和0.36(右侧) 1b 胼胝体上层面双侧半卵圆中心FA值分别为0.34(左侧)和0.31(右侧)

Figure 1 Head DTI examination findings. Bilateral centrum semiovale on the proximal ventricular top level showed that FA value was 0.37 on the left and 0.36 on the right (Panel 1a). Bilateral centrum semiovale on the corpus callosum level showed that FA value was 0.34 on the left and 0.31 on the right (Panel 1b).

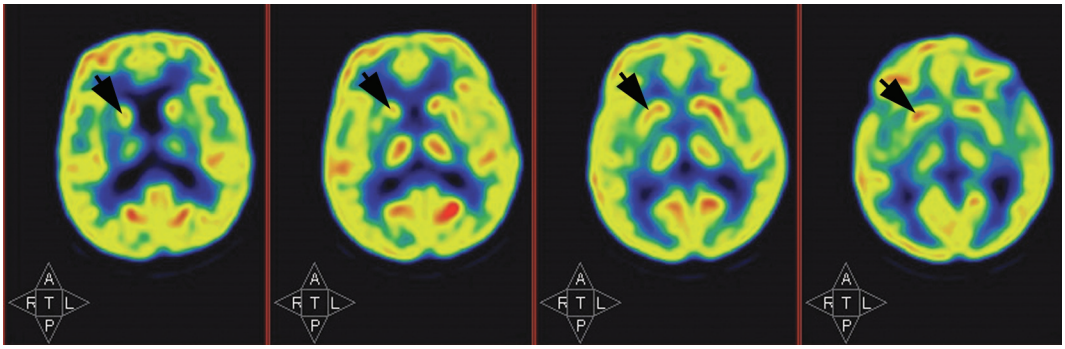

图2 ^{18}F-FDG PET显示,右侧豆状核葡萄糖代谢降低(箭头所示)

Figure 2 ^{18}F-FDG PET showed glucose hypometabolism of right lenticular nucleus (arrows indicate).

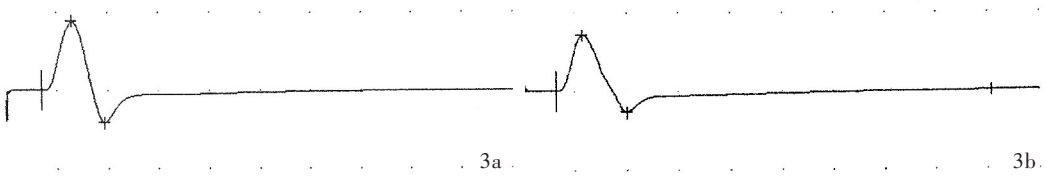

图3 左侧腓总神经传导速度检查所见 3a 首次检查(2013年7月)时波幅为12.90 mV 3b 4个月后复查(2013年11月)时波幅为9.80 mV,较前次明显降低,提示前角细胞损害加重。结合两次肌电图检查结果提示广泛神经源性损害,表明肌萎缩侧索硬化症进展

Figure 3 EMG findings of left common peroneal nerve. First examination (July 2013) showed the amplitude was 12.90 mV (Panel 3a). Four months later (November 2013), second examination showed obviously reduced amplitude (9.80 mV), suggesting aggravated anterior horn cell damage. These two examinations revealed extensive nerve-derived damage, indicating progressive ALS (Panel 3b).

1^{+}级,其余肢体肌力5^{-}级、肌张力1级。但逐渐出现左下肢无力,自觉双下肢近端偶有肌肉跳动感,结合两次肌电图结果,明确合并肌萎缩侧索硬化症,遂加用利鲁唑[1]50 mg(1次/12 h,空腹)口服治疗。

讨 论

该例患者有运动迟缓、肌张力增高等锥体外系症状,根据Otsuka等[2]的研究,^{18}F-FDG PET显示基底节区如豆状核(包括壳核和苍白球)、尾状核等部位葡萄糖代谢降低可排除原发性帕金森病,故帕金森综合征诊断明确。两次肌电图结果均证实下运动神经元疾病,结合腱反射亢进、病理征阳性等上运动神经元疾病体征,符合2000年修订的El Escorial肌萎缩侧索硬化症诊断标准[3];Nelles等[4]的研究显示,肌萎缩侧索硬化症患者DTI序列可见中央前回、半卵圆中心和内囊后肢FA值降低。头部MRI表现亦支持肌萎缩侧索硬化症诊断。虽然该例患者肌萎缩、肌肉跳动等肌萎缩侧索硬化症表现并不典型,但结合影像学和神经电生理学检查可明确诊断为帕金森综合征合并肌萎缩侧索硬化症(亦称Brait-Fahn病[5])。近年来,越来越多的研究显示二者相互重叠并非巧合,可能具有共同的发病机制[6-7]。肌萎缩侧索硬化症临床表现不典型的病例,影像学和神经电生理学检查尤为重要,在临床工作中应积极开展,以免漏诊和误诊[8]。

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