[27] Papageorgiou AC, Croft PR, Ferry S, et al. Estimating the prevalence of low back pain in the general population. evidence from the South Manchester back pain survey[J]. Spine(Phila Pa 1976),1995,20(17):1889—1894.

[28] Nolan L, Wit A, Dudziñski K, et al. Adjustments in gait symmetry with walking speed in trans-femoral and trans-tibial amputees[J]. Gait Posture,2003,17(2):142—151.

[29] Kulkarni J, Adams J, Thomas E, et al. Association between amputation, arthritis and osteopenia in British male war veterans with major lower limb amputations[J]. Clin Rehabil,1998,12(4):348—353.

[30] Liu K, Tao T, Wang A, et al. Surgical revision for stump problems after traumatic above-ankle amputations of the lower extremity[J]. BMC Musculoskelet Disord, 2015, 16:48.

[31] 卢中道,王义生.创伤性截肢术后并发症及处理[J].医学与哲学(临床决策论坛版),2008,29(6):26—28.

[32] Le JT, Scott-Wyard PR. Pediatric limb differences and amputations[J]. Phys Med Rehabil Clin N Am, 2015, 26(1): 95—108.

[33] Yin Y, Zhang L, Xiao H, et al. The pre-amputation pain and the postoperative deafferentation are the risk factors of phantom limb pain: a clinical survey in a sample of Chinese population[J]. BMC Anesthesiol,2017,17(1):69.

[34] Burger H, Marincek C. Upper limb prosthetic use in Slovenia[J]. Prosthet Orthot Int,1994,18(1):25—33.

[35] Almassi F, Emadi N, Mousavi B, et al. Dermatosis contributing factors in bilateral lower limb war-amputees[J]. Pak J Biol Sci,2010,13(2):78—82.

[36] Wollina U, Konrad H, Graefe T, et al. Botulinum toxin A for focal hyperhidrosis in leg amputees: a case report [J]. Acta Derm Venereol,2000,80(3):226—227.

[37] Potter BK, Burns TC, Lacap AP, et al. Heterotopic Heterotopic ossification following traumatic and combat-related amputations. prevalence, risk factors, and preliminary results of excision [J]. J Bone Joint Surg Am, 2007, 89(3): 476—486.

[38] Gonda K, Nakaoka T, Yoshimura K, et al. Heterotopic ossification of degenerating rat skeletal muscle induced by adenovirus-mediated transfer of bone morphogenetic protein-2 gene [J]. J Bone Miner Res,2010,15(6):1056—1065.

[39] Friberg O. Biomechanical significance of the correct length of lower limb prostheses: a clinical and radiological study [J]. Prosthet Orthot Int,1984,8(3):124—129.

[40] Braaksma R, Dijkstra PU, Geertzen JHB. Syme amputation: a systematic review[J]. Foot Ankle Int, 2018, 39(3): 284—291.

[41] Patzkowski JC, Blanck RV, Owens JG, et al. Comparative effect of orthosis design on functional performance[J]. J Bone Joint Surg Am,2012,94(6):507—515.

·病例报告·

Pisa综合征的康复:1例报告

刘 颖[1] 舒 璇[1] 王 含[2,3]

Pisa综合征(Pisa syndrome,PS)是一种姿势性疾病,见于包括帕金森病(Parkinson's disease, PD)在内的多种神经退行性疾病[1],是PD,尤其是晚期PD中一种较常见的致残性并发症,但相关临床报道并不多见。本文报道1例PS患者的康复诊疗情况。

患者郭某某,男性,73岁,于2019年7月来北京协和医院康复科就诊。主诉:右手不自主抖动4年半,步行不利1年,加重伴躯干右侧倾斜3个月。现病史:2015年起出现右手不自主抖动,静止时出现,运动时减轻,睡眠时消失。当地医院诊为"PD",给予美多芭半片口服,每日3次,右侧手抖仍缓慢加重,并逐渐出现左手笨拙。近1年渐出现步行不利,表现为小碎步,走路前冲,伴行动迟缓,翻身及起立困难,自觉起步及转弯尚可。2019年3月入住北京协和医院神经科,诊为帕金森叠加综合征。加用米多吡5mg口服,每日1次,治疗

DOI:10.3969/j.issn.1001-1242.2021.04.016

1 中国医学科学院北京协和医学院北京协和医院物理医学康复科,北京100730; 2 中国医学科学院北京协和医学院北京协和医院神经科; 3 通讯作者

第一作者简介:刘颖,女,主任医师; 收稿日期:2020-04-20

后手抖明显减轻。出院后渐出现坐立位及步行时躯干向右侧倾斜，步行时间延长和距离增加时明显，伴间断腰痛，卧床休息后躯干倾斜和腰痛可减轻。起步及转身时偶有冻结现象，近3个月跌倒2次。睡眠时偶有喊叫和拳打脚踢。自诉嗅觉减退多年。无饮水呛咳。无体位相关性头晕。否认幻觉、记忆力减退及其他认知障碍。既往高血压病史40年，近几年未服药，血压正常。

查体：神志清晰，构音可，音调略低。面部表情少。双眼垂直注视欠灵活，动作缓慢，幅度尚可，下视疑似明显。右侧上、下肢肌张力高，左侧无明显异常，四肢肌力5级。右侧轮替动作、叩指、双足拍地动作缓慢。后拉试验(-)。姿势评估：坐位及立位下脊柱右侧弯，仰卧及俯卧位侧弯明显减轻。步态检查：偏斜步态，步幅小，未见明显冻结及前冲。步行时右上肢伴随动作少，躯干向右侧倾斜渐加重。

辅助检查：左旋多巴负荷试验阴性。帕金森综合征专项MRI检查：中脑短轴/桥脑短轴=0.56，MRPI=(桥脑面积/中脑面积)×(小脑中脚/小脑上脚)=12.07。立位全脊柱X线正位相示：脊柱左侧凸，Cobb 角18°；胸、腰椎骨质疏松，骨质增生。俯卧位胸、腰椎正位相示：胸椎无明显侧凸；腰椎左侧凸较立位略减轻，Cobb角9°。步态分析示：摆动相左侧髋、膝关节屈曲不充分，右踝背屈不足。Berg平衡功能评分35分。跌倒风险筛查：起立行走试验(timed up and go，TUG)：16"95；起立测试(chair rising test，CRT)：20"72；走直线步态测试(tandem gait test，TGT)：1/8，提示跌倒高风险。

经北京协和医院疑难病会诊中心多学科会诊，诊断为帕金森叠加综合征、PS、骨质疏松、脊柱关节炎；平衡功能障碍、步行障碍、跌倒高风险；ADL轻度依赖；社会参与局限。因患者不便长住北京，给予居家康复指导，包括核心肌群及下肢肌力训练、躯干和四肢屈曲肌群牵伸、脊柱伸展及胸廓扩张运动、一字步站立、单足站立及负重平衡训练、姿势镜前保持脊柱中立位的姿势控制及加强右侧椎旁肌力量的脊柱侧凸矫形训练、左手置于左髋诱导脊柱回复中立位的感觉技巧手势引导、起坐训练、步态训练(原地高抬腿踏步，喊口令迈步，大步行走，双臂大幅摆动，在地面上划标志线，在视觉引导下大步行走，跨越障碍物行走)、面部表情肌训练、眼球旋转及移动训练、励协夫曼言语训练等。

1个月后复诊评估示：立位下脊柱轻度右侧弯；步行时躯干轻度右偏，左侧髋、膝屈曲及右踝背屈角度增加。Berg评分46分。TUG：12"24；CRT：13"06；TGT：6/8，跌倒风险降低。患者站立位脊柱右侧弯，俯卧位、仰卧位脊柱侧弯减轻。见图1—3。

PS被定义为可以通过被动活动或仰卧位减少的躯干侧弯[2]，以冠状面上的姿势畸形为主要表现，也可能合并一定程度的躯干前屈和旋转[3]。PS侧弯的特征是在坐、站和行走时脊柱偏向一侧。PS患者也可能对其垂直位置的感知受损，不能通过头部代偿行为来纠正视觉输入的校准，同时可能合并的腰痛和不稳也会导致跌倒发生[4]。PS可在亚临床发作和进行性恶化的情况下以慢性方式发展，也可在数天或数周内以快速进展的亚急性方式出现[5]。Doherty等[2]建议PS的诊断标准为躯干侧弯至少10°。根据躯干侧弯角度可将PS分为轻度(<20°)或重度(≥20°)[1]。

PS最早见于抗精神病药物治疗的患者[2]。除了抗精神病药、多巴胺受体拮抗剂等药物以外，该病现已逐渐归因于神经退行性疾病，如PD、Alzheimer病、多系统萎缩[6—12]。在

图1　立位全脊柱正位X线检查示脊柱左侧凸，Cobb角18°

图2　仰卧位胸椎正位X线检查示胸椎无明显侧凸

图3　仰卧位腰椎正位X线检查示腰椎左侧凸较立位略减轻，Cobb角9°

PD患者中,PS的发病率为8.8%—21%[1,13]。PS患者年龄较大,病程较长,Hoehn-Yahr分期较高,体重指数较低,生活质量较差。PS患者跌倒更为频繁,呈现“偏斜步态”。骨质疏松和关节炎是PS最常见的相关疾病。多变量logistic回归模型证实PS与以下变量相关:Hoehn-Yahr分期,左旋多巴和多巴胺激动剂持续联合治疗,相关疾病,偏斜步态。有PS的PD患者的病理生理学特征尚不明确,可能由中枢或外周机制造成[2—3,14—16]。有PS的PD患者比没有PS的PD患者表现出更大的运动不对称性[17],但尚不清楚脊柱侧凸是倾向于受累较重的一侧还是较轻的一侧[1,14]。躯干肌张力障碍可能在PS发病中起关键作用[18]。肌电图研究发现与健康受试者相比,PS患者双侧、侧弯同侧或对侧脊旁肌和/或非脊旁肌的激活程度更高[17,19—20]。PS的躯干肌张力障碍通常可通过感觉技巧来改善,这种现象在PD患者中很少见。感觉运动整合障碍也被视为PS发病的一个关键因素。PD患者不仅在体位定向和体位稳定性方面受损[21—22],而且这两个系统之间相互作用方面也受损。与PD相比,PS只会增加静态条件下的姿势不稳定性,似乎不影响步态,这可能反映了主要依靠视觉输入的代偿系统的存在。有研究发现,PS患者在闭眼行走时会表现出偏斜步态[15]。但是,PS患者有时不知道自己姿势异常,甚至不能通过倾斜头部来校正视野水平轴。所以,康复训练应借助姿势镜来帮助患者进行主动调节。外周机制如肌病、脊柱退行性变和软组织改变,都可能导致肌肉失衡,无力和代偿性姿势[2]。研究表明,肌肉废用或萎缩主要发生在与躯干倾斜侧的同侧,而脂肪退化可发生在双侧[17,19]。轴性骨骼畸形发生在合并长期、中重度PS的PD患者,并且PS患者脊柱侧凸很大程度与塌陷或姿势张力受损有关,而不是骨改变[23]。背痛是PS患者的常见主诉,发生率70.6%[1],患者为暂时缓解疼痛而采取的代偿姿势可能会对本体感觉和前庭系统产生长期负面影响,从而导致身体结构异常。诊断PS需通过站立位和仰卧位x线检查以排除结构性骨改变和脊柱侧凸[24—25],但有时脊柱侧凸可能与PS合并存在。为明确诊断,我们请放射科为本例患者进行卧立位全脊柱X线检查,但由于卧位不便进行全脊柱摄像,故分别进行了胸腰段检查,结果发现患者在仰卧位脊柱侧凸角度较站立位时减小,但并未完全消失,结合之前住院期间腰椎MRI检查发现腰椎侧弯,考虑该患者是PS合并脊柱侧凸,腰痛可能与合并的骨质疏松及脊柱关节炎有关。

PS是PD早期死亡的危险因素之一[13],通过适当治疗,PS是可逆或至少部分可逆的[15—16,26]。因此,PS的早期识别、药物调整及康复治疗非常重要。PS可能在任何类型的药物改变后亚急性发展[5,7]。因此,治疗第一步是恢复与PS发病相关的任何药物改变。当没有与PS相关的药物改变时,可以考虑增加或减少左旋多巴,因为PS可能是多巴胺太少的结果或者多巴胺能刺激过多[27]。另外,多巴胺的缓慢释放可能是另一种选择[2]。虽然药物诱导性PS的药物治疗尚未建立,但据报道,抗胆碱能药物对40%药物诱导性PS发作有效,其余患者对停药或减少每日抗精神病药物剂量有反应[7]。基于肉毒杆菌毒素(botulinum toxin,BoNT)对过度肌肉活动的疗效,已有学者提出采用BoNT治疗PS[28]。迄今为止,有限的临床研究表明BoNT能够增强康复治疗的效果,可能是PS的治疗选择之一[12,29—31]。已有数项研究表明运动康复对合并PS的PD有效,是一项非常有希望的治疗策略[29,31—34]。尽管各研究所采用的康复计划不同,包括有氧热身、姿势控制、个体化的本体感觉和触觉刺激、牵伸训练、力量训练、平衡训练、步态训练、放松训练、灵活性训练、桥式运动、普拉提等,但训练强度均较大,通常每周训练3—5天,持续2—4周,疗效维持时间3—6月。另有研究表明,合并PS的PD患者较同龄健康人及单纯PD患者身体摆动速度更大,更难获得良好的重力力线,因此,PS合并PD患者的康复计划,除了针对PD的常规康复训练外,还应包括脊柱力线和动态姿势训练[15]。本例患者康复指导是在康复评估基础上,结合患者个体化需求进行,以改善姿势和平衡障碍、预防跌倒、缓解腰痛、预防继发功能障碍为主,疗效显著。未来需要进一步研究明确PS康复的最佳方案和持续时间。

参考文献

[1] Tinazzi M, Fasano A, Geroin C, et al. Pisa syndrome in Parkinson disease: An observational multicenter Italian study [J]. Neurology, 2015, 85:1769—1779.

[2] Doherty KM, Vande Warrenburg BP, Peralta MC, et al. Postural deformities in Parkinson's disease [J]. Lancet Neurol, 2011, 10: 538—549.

[3] Tinazzi M, Geroin C, Gandolfi M, et al. Pisa syndrome in Parkinson's disease: an integrated approach from pathophysiology to management [J]. Mov Disord, 2016, 31: 1785—1795.

[4] Geroin C, Gandolfi M, Bruno V, et al.Integrated approach for pain management in Parkinson disease [J]. Curr Neurol Neurosci Rep, 2016,16:28.

[5] Yokochi F. Lateral flexion in Parkinson's disease and Pisa syndrome [J]. J Neurol, 2006, 253(suppl 7): Ⅶ17—Ⅶ20.

[6] Kwak YT, Han IW, Baik J,et al. Relation between cholinesterase inhibitor and Pisa syndrome [J]. Lancet, 2000, 355:2222.

[7] Suzuki T, Matsuzaka H. Drug-induced Pisa syndrome (pleurothotonus): epidemiology and management[J]. CNS Drugs, 2002,16:165—174.

[8] Patel S, Tariot PN, Hamill RW. Pisa syndrome without neuroleptic exposure in a patient with dementia of the Alzheim-

er type [J]. J Geriatr Psychiatry Neurol, 1991,4:48—51.

[9] Colosimo C. Pisa syndrome in a patient with multiple system atrophy [J]. Mov Disord, 1998,13:607—609.

[10] Davidson M, Powchik P, Davis KL. Pisa syndrome in Alzheimer's disease [J].Biol Psychiatry, 1988, 23:213.

[11] Huvent Grelle D, Boulanger E, Puisieux F. Relationship between Pisa syndrome and cholinesterase inhibitor use for elderly adults with Alzheimer's disease [J]. J Am Geriatr Soc, 2014, 62:2450—2453.

[12] Bonanni L, Thomas A, Varanese S, et al. Botulinum toxin in treatment of lateral axial dystonia in Parkinsonism[J]. Mov Disord, 2007, 22: 2097—2103.

[13] Laudisio A, Lo Monaco MR, Vetrano DL, et al. Association of Pisa syndrome with mortality in patients with Parkinson's disease [J]. J Am Med Dir Assoc, 2019, 20: 1037—1041.

[14] Castrioto A, Piscicelli C, Perennou D, et al. The pathogenesis of Pisa syndrome in Parkinson's disease [J].Mov Disord, 2014, 29:1100—1107.

[15] Geroin C, Smania N, Schena F, et al. Does the Pisa syndrome affect postural control, balance, and gait in patients with Parkinson's disease? an observational cross- sectional study [J]. Parkinsonism Relat Disord, 2015, 21:736—741.

[16] Vitale C, Falco F, Trojano L, et al. Neuropsychological correlates of Pisa Syndrome in patients with Parkinson's disease[J]. Acta Neurol Scand, 2016, 134:101—107.

[17] Tassorelli C, Furnari A, Buscone S, et al. Pisa syndrome in Parkinson's disease: clinical, electromyographic, and radiological characterization [J]. Mov Disord, 2012, 27: 227—235.

[18] Albanese A, Bhatia K, Bressman SB, et al. Phenomenology and classification of dystonia: a consensus update[J]. Mov Disord, 2013, 28:863—873.

[19] Tinazzi M, Juergenson I, Squintani G, et al. Pisa syndrome in Parkinson's disease: an electrophysiological and imaging study [J]. J Neurol, 2013, 260:2138—2148.

[20] Matteo AD, Fasano A, Squintani G, et al. Lateral trunk flexion in Parkinson's disease: EMG features disclose two different underlying pathophysiological mechanisms [J]. J Neurol,2011, 258:740—745.

[21] Vaugoyeau M, Viel S, Assaiante C, et al. Impaired vertical postural control and proprioceptive integration deficits in Parkinson's disease [J]. Neuroscience, 2007, 146: 852—863.

[22] Schoneburg B, Mancini M, Horak F, et al. Framework for understanding balance dysfunction in Parkinson's disease [J].Mov Disord, 2013, 28:1474—1482.

[23] Doherty KM, Davagnanam I, Molloy S, et al. Pisa syndrome in Parkinson's disease: a mobile or fixed deformity? [J]. J Neurol Neurosurg Psychiatry, 2013, 84:1400—1403.

[24] Vrtovec T, Pernus F, Likar B. A review of methods for quantitative evaluation of spinal curvature[J].Eur Spine J, 2009,18: 593—607.

[25] Vrtovec T, Pernus F, Likar B. A review of methods for quantitative evaluation of axial vertebral rotation [J].Eur Spine J, 2009,18: 1079—1090.

[26] Barone P, Santangelo G, Amboni M, et al. Pisa syndrome in Parkinson's disease and parkinsonism: clinical features, pathophysiology, and treatment [J]. Lancet Neurol, 2016,15:1063—1074.

[27] Cannas A, Solla P, Floris G, et al. Reversible Pisa syndrome in patients with Parkinson's disease on dopaminergic therapy [J]. J Neurol, 2009, 256:390—395.

[28] Jankovic J. Botulinum toxin in clinical practice [J]. J Neurol Neurosurg Psychiatry, 2004,75:951—957.

[29] Tassorelli C, De Icco R, Alfonsi E, et al. Botulinum toxin type A potentiates the effect of neuromotor rehabilitation of Pisa syndrome in Parkinson disease: a placebo controlled study [J]. Parkinsonism Relat Disord, 2014, 20: 1140—1144.

[30] Dupeyron A, Viollet E, Coroian F, et al. Botulinum toxin-A for treatment of Pisa syndrome: a new target muscle [J]. Parkinsonism Relat Disord, 2015, 21:669—670.

[31] Santamato A, Ranieri M, Panza F, et al. Botulinum toxin type A and a rehabilitation program in the treatment of Pisa syndrome in Parkinson's disease [J]. J Neurol, 2010, 257:139—141.

[32] Capecci M, Serpicelli C, Fiorentini L, et al. Postural rehabilitation and Kinesio taping for axial postural disorders in Parkinson's disease [J]. Arch Phys Med Rehabil, 2014, 95: 1067—1075.

[33] Bartolo M, Serrao M, Tassorelli C, et al. Four- week trunk- specific rehabilitation treatment improves lateral trunk flexion in Parkinson's disease [J]. Mov Disord, 2010, 25:325—331.

[34] Kataoka H, Ikeda M, Horikawa H, et al. Reversible lateral trunk flexion treated with a rehabilitation program in a patient with Parkinson's disease [J]. Parkinsonism Relat Disord, 2013,19: 494—497.