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个案与短篇

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Kleine-Levin 综合征 1 例报道

杨西爱[1],李光勤[2]
（1. 陕西省安康市中心医院神经内科,安康 725000;2. 重庆医科大学附属第一医院神经内科,重庆 410016）

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Kleine-Levin 综合征（Kleine-levin syndrome,KLS）是一种临床上较为少见的疾病,国内报道病例较少。现将临床收治的 1 例女性病例报道如下。

1 临床资料

患者,女,32 岁。因“突发呼之不应 7 h”于 2008 年 5 月 30 日入院。入院 7 h 前患者于夜间睡眠中跌落床下,仍为熟睡状,面色正常,呼吸均匀,呼之不应,无头部及身体其他部位外伤,无恶心、呕吐及二便失禁,无口角歪斜、口吐白沫、双眼凝视等。家属将其扶到床上仍处于睡眠状,次日 11:00 后开始逐渐苏醒,醒后意识清楚,精神差,懒言少动,易激惹,对发作及跌落床下无记忆。追问病史,6 年多来平均每月发作 1 次,发作时睡眠持续时间最长 36 h,中间可有上厕所及饮食等动作,完毕后继续睡觉。自诉对以前发作过程中上厕所和进食断续有记忆,并且在发作后 3 d 左右进食量较平时多两倍。既往史:每年春季有皮肤过敏史,余无特殊。出生史及家族史无特殊。入院查体:内科查体及神经系统查体未见明显异常。辅助检查:三大常规检查无异常。肝肾功、血脂、血糖、电解质均正常。脑电图、头颅 CT、头颅 MRI、头颅磁共振波谱分析均未见异常。诊断为:Kleine-Levin 综合征。入院后给予改善循环及都可喜治疗,治疗 2 周后好转出院。随访 6 月,仍有发作,症状类似。

2 讨 论

KLS 是一种比较少见的疾病, 二十世纪二三十年代由 Kleine 和 Levin 先后描述,由此得名,国内又名周期性嗜睡贪食综合征,主要临床表现为周期性过度睡眠、食欲亢进、性欲增强等。常在青少年时期发病,男性多于女性,患者体型多偏胖。KLS 患者在睡眠发生前有困乏先兆,睡眠发生后持续时间长,常常在 1 d 至 1 周左右。其间可以起床吃饭及上厕所,与人交流少或无,发作过后 3 d 左右常有进食量增加,夜间

（下转第 62 页）

作者介绍:杨西爱(1980-),女,硕士,
研究方向:脑血管病。

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睡眠差。多导睡眠检测可见慢波睡眠相有部分损害。对全球散在的 186 例病例的回顾性研究发现[1],KLS 患者中周期性过度睡眠发生率为 100%、认知改变为 96%、进食紊乱为 80%、性欲增强为 43%、强迫症为 29%、情绪低落为 40%。KLS 的具体病因尚不明确。可能与下列因素有关:①下丘脑功能紊乱,苯基二氢喹唑啉(Orexin)含量下降:适量的苯基二氢喹唑啉可以帮助人们保持苏醒状态。有的病例行 SPECT 检查可发现丘脑灌注不足[2,3]、磁共振波谱分析发现脑内代谢降低,影响了神经传递[4];②反复感染:KLS 发病前可有困倦、流感样症状,有报道 KLS 患者存在下丘脑的炎性反应,提示反复病毒感染可能与本病有关[5];③遗传易感性:有报道该病呈现家族聚集性,犹太人中发病率高,推测可能与遗传易感性因素有关[6]。本病需与以下疾病鉴别:①发作性睡病,本病以不可抵抗的睡眠发作、猝倒、睡眠麻痹和睡眠幻觉四联征为特点,发作持续时间短,一般十几分钟。②癫痫非惊厥性持续状态:该病临床表现为以意识障碍基础上伴或不伴自动症症状,一般持续时间较短,脑电图有癫样放电可鉴别。③器质性复发性嗜睡症:可见于第三脑室肿瘤、脑炎或脑外伤等,神经系统检查和影像学检查可鉴别。KLS 的治疗以支持对症为主,有研究显示中枢兴奋药或锂对部分病例有效[3,4],本病预后较好,发病 10 年左右后有自愈倾向。

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