

# 糖尿病非酮症偏侧舞蹈症1例及文献复习

张婵娟<sup>1</sup>, 高明康<sup>1</sup>, 温丽民<sup>1</sup>, 史亚茹<sup>1</sup>, 褚旭<sup>2\*</sup>

<sup>1</sup>济宁医学院临床医学院, 山东 济宁

<sup>2</sup>济宁医学院附属医院神经内科, 山东 济宁

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## 摘要

目的: 通过探讨非酮症高血糖性偏侧舞蹈症患者的临床表现、诊断和治疗、发病机制, 提高对本病的认识, 做到早期识别、早期治疗, 避免误诊、漏诊。方法: 分析1例济宁医学院附属医院的糖尿病非酮症偏侧舞蹈症患者的临床症状、体征、影像学资料、治疗方法, 并复习相关文献。结果: 本例糖尿病非酮症偏侧舞蹈症患者, 舞蹈样动作较明显, 既往患者血糖控制不规律。患者入院随机血糖: 14 mmol/L, 糖化血红蛋白: 11%, 血酮体: 3.6 mg/dl; 颅脑CT示左侧基底节区高密度影; 颅脑MRI T1WI可见左侧基底节区高信号。入院后经积极控制血糖水平, 1周后舞蹈样症状完全消失。结论: 对于糖尿病非酮症偏身舞蹈症的患者做到早期识别、早期干预, 积极控制血糖水平, 科学合理地加用抗精神病药物, 控制患者舞蹈样动作, 其预后良好, 并提高了患者的生活质量。

## 关键词

糖尿病, 非酮症, 舞蹈症

# Hemichorea Associated with Non-Ketotic Hyperglycemia: 1 Case Report and a Review of the Literature

Chanjuan Zhang<sup>1</sup>, Mingkang Gao<sup>1</sup>, Limin Wen<sup>1</sup>, Yaru Shi<sup>1</sup>, Xu Chu<sup>2\*</sup>

<sup>1</sup>Jining Medical University, Jining Shandong

<sup>2</sup>The Affiliated Hospital of Jining Medical University, Jining Shandong

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\*通讯作者。

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## Abstract

**Objective:** To improve the understanding of this disease by discussing the clinical manifestations, diagnosis, treatment and pathogenesis of patients with non-ketotic hyperglycemic hemichorea, to achieve early recognition and early treatment, and to avoid misdiagnosis and missed diagnosis. **Methods:** The clinical symptoms, signs, imaging data and treatment of a patient with non-ketotic hyperglycemic hemichorea from the Affiliated Hospital of Jining Medical College were analyzed, and the relevant literature was reviewed. **Results:** This case of hemichorea associated with non-ketotic hyperglycemia had more pronounced dance-like movements and irregular glycaemic control in previous patients. The patient was admitted with random blood glucose: 14 mmol/L, glycosylated hemoglobin: 11%, blood ketone bodies: 3.6 mg/dl; cranial CT showed high-density shadow in the left basal ganglia region; cranial MRI T1WI showed high signal in the left basal ganglia region. After admission, the chorea-like symptoms completely disappeared after 1 week after active control of blood glucose level. **Conclusion:** For patients with non-ketotic hyperglycemic hemichorea to achieve early identification, early intervention, active control of blood glucose level, scientific and reasonable addition of antipsychotic drugs to control the patient's chorea-like movements, their prognosis is good and the quality of life of patients is improved.

## Keywords

Diabetes Mellitus, Non-Ketotic, Chorea

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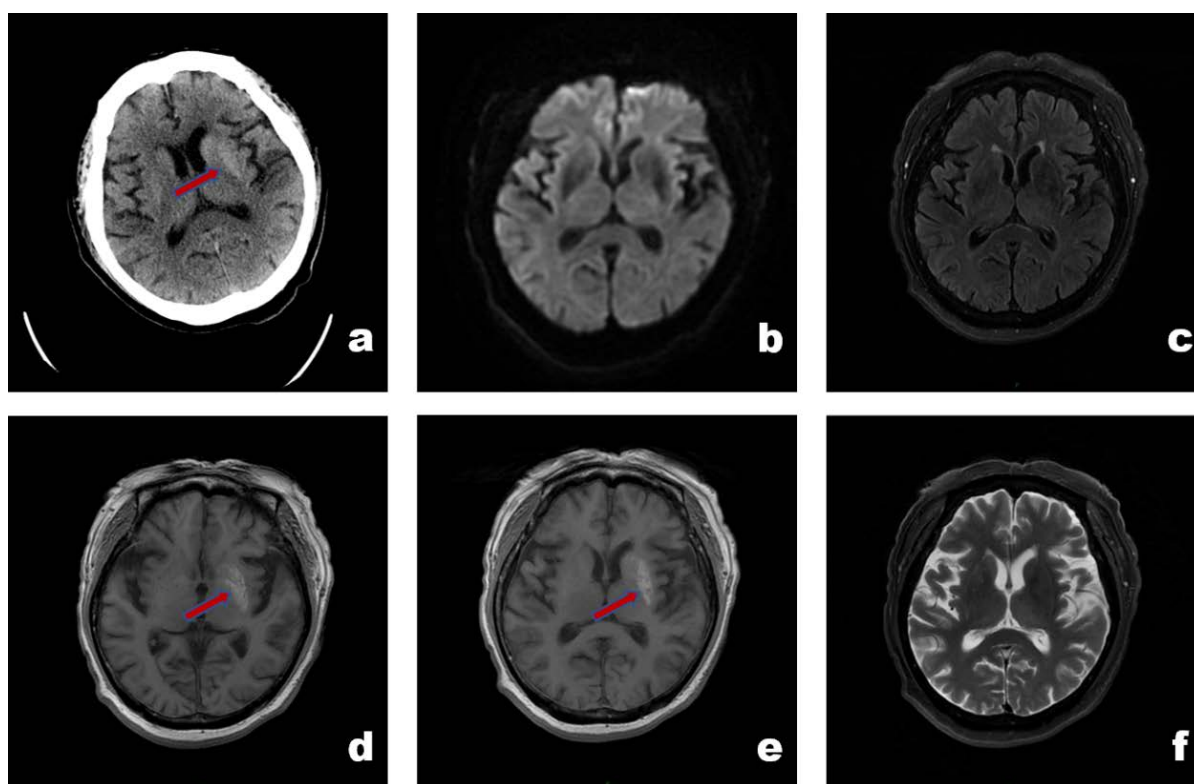
## 1. 引言

糖尿病是一种以高血糖为特征的代谢性疾病,若患者的血糖水平长期控制欠佳,会进一步造成眼底、肾脏、心脏、血管、神经等组织的慢性损害、功能障碍,而中国作为全世界糖尿病患者数量最多的国家,由其所导致的人体各系统并发症问题也日趋严峻。非酮症高血糖(Non-Ketotic Hyperglycemia, NKH)可产生各种神经系统症状,包括精神改变、癫痫发作和自主运动,如舞蹈和颤抖[1],舞蹈症(chorea)是一种不规则、不协调、自主的运动障碍,主要累及单侧肢体(偶可为双侧肢体同时受累),脑血管功能不全、神经退行性疾病、肿瘤性疾病、免疫性疾病、感染性疾病和代谢性疾病是这种罕见疾病的继发性原因[2]。糖尿病非酮症偏侧舞蹈症多见于长期血糖控制不良的老年患者,主要的临床表现为急性起病的偏身舞蹈样症状,其辅助检查的特点是非酮症性的高血糖,影像学特征性表现为舞蹈动作肢体对侧的基底节区 CT 高密度影或 MRIT1 加权像上高信号灶[3]。糖尿病与中枢神经系统晚期并发症的风险增加有关[4],虽然高血糖是舞蹈症最常见的代谢原因,但其致病机制现仍不非常明确。本文通过对我院 1 例糖尿病非酮症偏侧舞蹈症进行研究分析,并结合复习国内外文献以提高对该病的认识。

## 2. 病例资料

患者,女性,81 岁。因“右侧肢体不自主活动 4 h”于 2020 年 10 月 30 日收住济宁医学院附属医院。患者 4 小时前无明显诱因出现右侧肢体不自主活动,站立时行走不稳,偶有头晕,无头痛、言语不清、饮水呛咳等。既往“2 型糖尿病”病史 20 年,未系统监测及诊治,“高血压病”病史 10 年,未系统诊

治,“冠心病、房颤”病史5年,患者入院前均未规律口服相关治疗药物。入院查体:T 36.5℃,P 105次/min,R 16次/min,BP 100/59 mmHg,神志清楚,精神欠佳,言语流利,双侧瞳孔等大等圆,直径约3 mm,对光反射敏感,眼球活动自如,无眼震。颈软,双侧鼻唇沟对称,伸舌居中,四肢肌力5级,肌张力正常,右上肢舞蹈样动作。双侧巴氏征未引出,克氏征、布氏征均为阴性,感觉、共济无异常。实验室检查:血糖 14 mmol/L,糖化血红蛋白 11%,血酮体 3.6 mg/dl,氯 96 mmol/L,碳酸氢盐 30.8 mmol/L,B型钠尿肽 339 pg/ml,同型半胱氨酸 26.3 umol/L,余血细胞分析、肝功、肾功、血脂、心肌酶谱、电解质、凝血常规、D-二聚体、甲功三项等化验结果未见明显异常。神经影像学检查(见图1):颅脑CT示左侧基底节区高密度影(见图1(a));颅脑MRT1加权像示左侧基底节区高信号灶(见图1(d)、图1(e)),MR DWI、Flair像、T2加权像等均未见明显异常(见图1(b)、图1(c)、图1(f))。患者晚间入院后临时给予胰岛素注射一次,次日监测患者血糖6次:10.4、17.6、19.4、17.5、20.1、19.9,给予格列齐特缓释片、阿卡波糖片控制患者血糖水平,并加强血糖监测,经系统治疗后,患者血糖控制在6.1~11.7 mmol/L之间,5天后患者右侧肢体不自主运动症状明显减轻,1周后舞蹈样动作完全消失。经治疗好转后出院。出院3个月电话随访,患者血糖控制可,右侧肢体不自主活动未再出现。



**Figure 1.** Neuroimaging tests: (a) cranial CT scan; (b) cranial magnetic resonance DWI; (c) cranial magnetic resonance T2 Flair; (d), (e) cranial magnetic resonance T1; (f) cranial magnetic resonance T2

**图1.** 神经影像学检查: (a) 颅脑CT; (b) 颅脑MRDWI; (c) 颅脑MR T2 Flair; (d)、(e) 颅脑MRT1; (f) 颅脑MRT2

### 3. 讨论

糖尿病非酮症偏侧舞蹈症(hemichorea associated with nonketotichyperglycemia, HCNH)平均发病年龄为71岁[5],主要见于患有2型糖尿病的亚洲老年妇女[6],但也有相当数量的男性,虽然患者的糖尿病病程各不相同,但都处于严重的高血糖状态,大多数患者的HbA1c水平超过10% [7]。

作为主要假说的代谢紊乱理论认为,在高血糖这一危险因素的影响下,三羧酸(Krebs)循环变得失活,导致大脑转向无氧代谢[8],大脑将 GABA ( $\gamma$ -氨基丁酸)代谢成琥珀酸,从而提供了另外一种能量来源。然而,这种 GABA 分流只满足基底神经节所需能量的 10%~40%左右,进而导致代谢性酸中毒的发生。与可以重新合成 GABA 的酮症高血糖不同,非酮症高血糖时 GABA 和醋酸盐会迅速被耗竭,而醋酸盐的耗竭又会减少乙酰胆碱的合成。因此,基底节内 GABA 和乙酰胆碱的水平降低,再加上代谢性酸中毒和能量产生不足,可能会导致基底节功能障碍以及偏侧舞蹈症的发生[9]。此外,多巴胺受体上调、雌激素相关的[10]多巴胺能超敏反应以及血脑屏障的破坏也被认为与发病有关。然而,还没有一种单一的机制可以解释所有的症状特征[11]。

影像学检查对其早期诊断非常重要,典型的影像学表现是 CT 上基底节单侧高密度区或 MRIT1 加权像上信号增强[12], DWI 弥散受限,这也与本病例影像学表现相一致。相关研究显示,基本上所有病例均累及壳核,多数病例累及尾状核头部,少数病例累及苍白球[13],但为什么是基底节,特别是壳核易受影响,目前具体机制还不清楚[14]。并且 MR 检查较 CT 检查对病灶的发现具有更强的敏感性和准确性[15],部分患者 CT 检查可无明显异常表现,要注意对此类患者漏诊的可能性。

据报道, NKH 舞蹈病的预后良好,罕见例外[8],治疗的主要手段是积极控制血糖,同时根据患者所出现的临床症状进行针对性的治疗,例如对于舞蹈动作明显的患者,科学合理地加用氟哌啶醇、丁苯那嗪、苯二氮卓类等抗精神病药物在控制舞蹈动作方面很有疗效[14] [15]。

非酮症性高血糖是偏侧舞蹈症的罕见原因之一[16],虽然偏侧舞蹈症少见,但也可以作为糖尿病的首发症状出现[17] [18],在临床工作中,对于不明原因突发肢体舞蹈样动作为主要临床表现就诊的患者,要考虑合并该疾病的可能。在纠正此类患者的高血糖状态之后,通常会使其舞蹈样症状和体征完全消失[19],并且早期治疗可使患者的神经影像学异常表现部分甚至是完全消退[4] [11],因此对糖尿病非酮症偏侧舞蹈症患者做到早期识别、早期干预是非常必要的。

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