Advances in Clinical Medicine 临床医学进展, 2024, 14(2), 2732-2737
Published Online February 2024 in Hans. https://www.hanspub.org/journal/acm
https://doi.org/10.12677/acm.2024.142385

特发性颅内高压并静脉窦狭窄致视乳头水肿1例合并文献复习

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收稿日期：2024年1月7日；录用日期：2024年2月1日；发布日期：2024年2月8日

摘 要

目的：报道1例特发性颅内高压并静脉窦狭窄致视乳头水肿的临床特征和诊治要点，分析该病的发生机制及治疗预后。方法：回顾该病例的诊疗过程，并结合相关文献分析该病表现及治疗预后。结果：根据患者病史、症状、体征及腰椎穿刺，全身辅助检查，眼底照相，颅内CTV结果，诊断为“特发性颅内高压并静脉窦狭窄”，行颅内静脉窦支架置入术，随访2个月，病情稳定。结论：特发性颅内高压是相对罕见的不明原因的颅内高压，导致的视乳头水肿对患者视功能及生活质量造成严重损害，早期诊断对于挽救患者的视功能尤为重要。

关键词

特发性颅内高压，视乳头水肿，静脉窦狭窄

A Case of Idiopathic Intracranial Hypertension with Venous Sinus Stenosis Causing Papilledema Combined with Literature Review

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Received: Jan. 7th, 2024; accepted: Feb. 1st, 2024; published: Feb. 8th, 2024

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文章引用: 李晓晶, 李元彬. 特发性颅内高压并静脉窦狭窄致视乳头水肿 1 例合并文献复习[J]. 临床医学进展, 2024, 14(2): 2732-2737. DOI: 10.12677/acm.2024.142385

Abstract

Objective: To report the clinical features and diagnostic and therapeutic points of a case of idiopathic intracranial hypertension with venous sinus stenosis resulting in optic papillae edema, and to analyze the mechanism of the disease and the prognosis of the treatment. Methods: The diagnostic and treatment process of this case was reviewed, and the manifestations and treatment prognosis of this disease were analyzed with relevant literature. Results: Based on the patient's medical history, symptoms, signs and symptoms, lumbar puncture, systemic auxiliary examination, fundus photography, and intracranial CTV results, the patient was diagnosed as "idiopathic intracranial hypertension with venous sinus stenosis", and underwent intracranial venous sinus stenting, and her condition was stabilized after 2 months of follow-up. Conclusion: Idiopathic intracranial hypertension is a relatively rare and unexplained form of intracranial hypertension, resulting in optic papillae edema, which causes serious damage to the patient's visual function and quality of life, and early diagnosis is particularly important to save the patient's visual function.

Keywords

Idiopathic Intracranial Hypertension, Papilledema, Sinus Stenosis

1. 背景

特发性颅内高压是一种在常规检查中无法找到任何病理发现的颅内压增高而引起的综合征，以头痛、视乳头水肿等颅内压增高的相关症状和体征为主要临床表现，特发性颅内高压(idiopathic intracranial hypertension, IIH)是视盘水肿最常见的原因，尤其是在 50 岁以下的患者中[1]；大多数 IIH 患者呈良性自限病程，但仍有 10%的 IIH 患者因慢性视盘水肿最终发展为双眼盲[2]，其病程和预后可能是多变的，需要多学科的诊断和治疗方法[3]。

2. 临床资料

患者男性，35 岁，因“双眼阵发性黑矇 1 月，左眼著。”于 2023 年 9 月 14 日来我院就诊，患者 1 月前无明显诱因出现双眼阵发性黑矇，伴有头痛，无色觉异常、眼球转动痛，否认药物接触史、否认眼外伤史，既往史及个人史无特殊。眼科查体：矫正视力：右眼 1.0 左眼 0.6，双眼眼压 20 mmHg。裂隙灯检查：双眼眼前节正常，相对性传入性瞳孔障碍检查阴性，双眼视盘高度充血水肿，静脉轻度迂曲，盘周视网膜出血(见图 1)，视网膜平伏。视野检查示右眼仅在注视中央附近存在极小部分视野，左眼盲？P-VEP 示双眼 P-100 波峰时延迟，波幅降低。入院后完善血常规、血生化、血沉、C 反应蛋白、免疫及感染标志物，颅脑 CT 未见明显异常。进一步行腰椎穿刺，测脑脊液(Cerebrospinal Fluid, CSF)压力大于 350 mmH_2O，脑脊液检查未见明显异常。神经内科会诊意见：查体未见神经系统阳性体征，初步诊断为颅内高压，给予甘油果糖降颅压治疗，建议完善头颅 MRI、MRV、MRA；头颅 MRV 示：左侧横窦略细，局部显影浅淡，头颅 MRI、MRA 未见明显异常。神经外科会诊意见，进一步明确横窦情况，考虑择期行静脉窦支架手术。给予金纳多，甲钴胺，维生素 B1、七叶皂苷钠营养神经，改善微循环，甘油果糖氯化

钠降颅压治疗，视盘水肿稍好转(见图 2)出院。患者于 2023-10-08 至北京某医院完善头颅 CTV 示：左侧横窦－乙状窦较对侧纤细，局部充盈欠佳，见充盈缺损(见图 3)。行左侧横窦支架植入术，术后 2 月，头痛症状明显好转，眼科查体：矫正视力：右眼 1.0 左眼 1.0，双眼视盘水肿消退，盘周出血消失(见图 4)，视网膜平伏。视野检查示双眼视野缺损范围较前明显缩小。

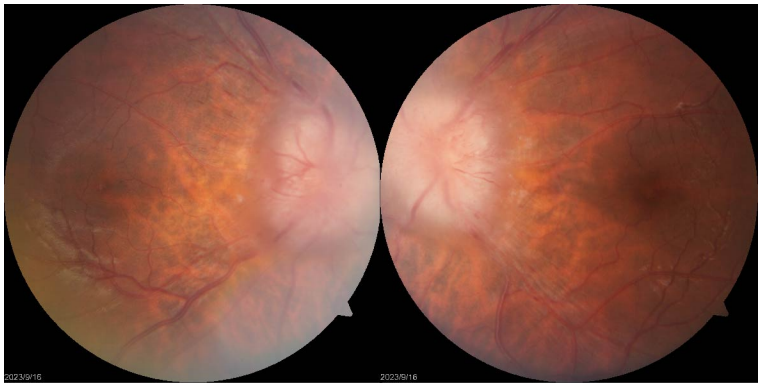

Figure 1. Fundus image of the IIH patient at the first diagnosis
图 1. IIH 患者首诊时眼底图像

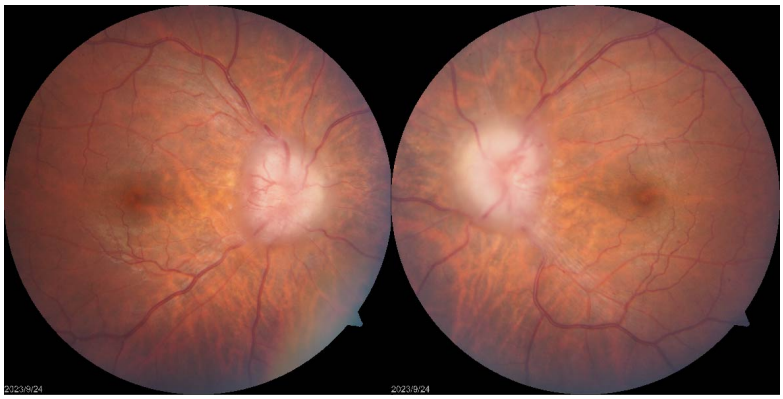

Figure 2. Fundus image of IIH patient after cranial pressure lowering treatment in our hospital
图 2. IIH 患者于我院降颅压治疗后眼底图像

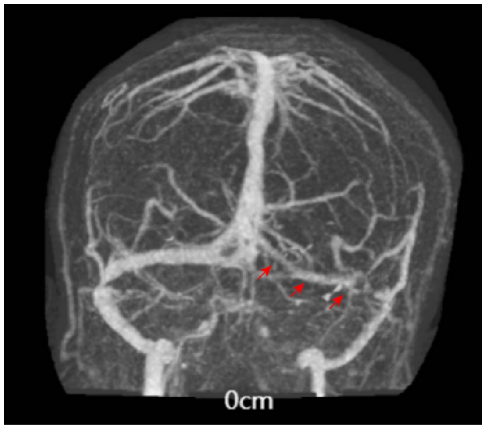

Figure 3. The left transverse-sigmoid sinus is more slender than the contralateral side and is poorly filled locally; filling defects are seen
图 3. IIH 左侧横窦－乙状窦较对侧纤细，局部充盈欠佳，见充盈缺损

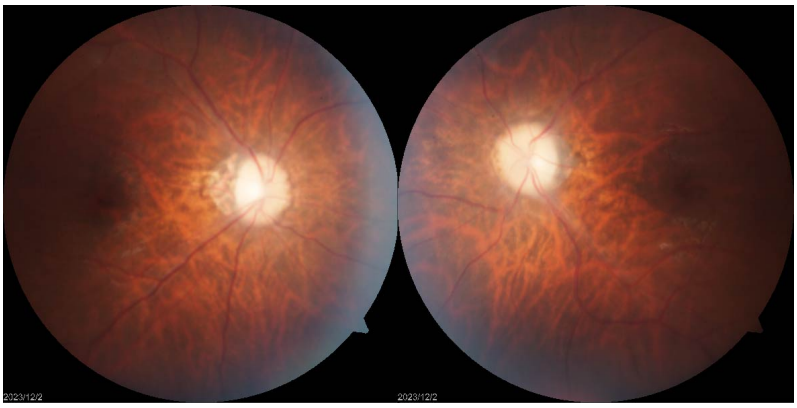

Figure 4. Fundus image of a patient with IIH 2 months after venous sinus stent placement
图 4. IIH 患者静脉窦支架置入术后 2 月眼底图像

3. 讨论

特发性颅内高压(IIH)是一种病因不明、以颅内压升高为特征的疾病。典型的病人是超重的育龄妇女。最常见的症状包括头痛、短暂性视觉障碍(Transient visual obscurations, TVOs)、搏动性耳鸣、背痛和复视[4]。TVOs 是指一只或两只眼睛的视力丧失或混浊，持续一分钟或更短时间。其原因被认为是视神经内压力升高导致视神经乳头头短暂缺血[5]。特发性颅内高压所致的急性期视乳头水肿的特征性表现是双侧视神经肿胀，伴视功能异常或接近正常。这些特征与其他引起视乳头水肿的疾病如视神经炎和前部缺血性视神经肿大区别开来[6]，并且这类患者 RAPD 检查常常是阴性，本例患者双眼视乳头水肿，但该患者右眼视力正常，并且 RAPD (−)，这些特征都与其他引起视乳头水肿的疾病不一致。特发性颅内高压的诊断标准由 Wall M.和 Corbett J.J.于 2014 年修订的特发性颅内高压的诊断标准如下：“诊断假性脑肿瘤综合征的必要条件：A) 视乳头水肿；B) 除脑神经异常外，其他神经检查正常；C) 典型患者(女性和肥胖)的 MRI (加或不加钆)显示脑实质正常，无脑积水、肿块或结构性病变，脑膜未见异常强化；D) 脑脊液成分正常；E) 在正确操作的腰椎穿刺中，CSF ≥ 250 mmH_2O [7]”。本例患者双眼阵发性黑矇，左眼著，伴有头痛症状符合 IIH 的临床表现，眼科查体发现患者双眼视盘高度充血水肿，静脉轻度迂曲，盘周视网膜出血，符合标准 A，腰椎穿刺测定 CSF 大于 350 mmH_2O 符合标准 E，神经系统查体、常规检查、检验、风湿免疫指标均未见明显异常，颅内 CT、MRI 未见任何能引起颅内压增高的病理发现符合标准 B、C、D，由此可得出该患者特发性颅内高压的诊断明确，推测患者阵发性黑矇的症状可是由于特发性颅内高压所致的 TVOs。

特发性颅内高压的发生机制尚未清楚，目前研究表明它的发生发展可能与脑脊液动力学失调、肥胖[8]、荷尔蒙失调[9]等因素密切相关，其中脑脊液动力学失调包括 CSF 分泌过多、蛛网膜颗粒重吸收减少和静脉窦压力梯度异常等[3] [10]。许多研究表明，颅内静脉窦压力升高，进一步导致颅内静脉窦压力梯度异常，颅内脑脊液压力相应升高，此类患者常出现头痛、视乳头水肿伴视功能障碍等症状[11] [12]。静脉窦狭窄所导致的静脉窦压力梯度异常是 IIH 中颅内压升高的一个重要因素[13]。30%~90%的 IIH 患者均存在静脉窦流出梗阻[14] [15]。静脉窦狭窄的正反馈循环假说[16]表明：静脉窦的狭窄会导致上游静脉的压力升高，上游静脉的压力升高导致脑脊液的回收受阻，脑脊液压力随之升高，而升高的脑脊液压力又可进一步压迫静脉窦，导致静脉窦的进一步狭窄，形成恶性循环。升高的 ICP 传递到视神经周围的蛛网膜下腔，阻碍了神经节细胞轴突内的轴浆运输，导致视乳头水肿的发生[1]，且长期的视乳头水肿会导致神经节细胞层的变薄[17]。本例患者 CTV 显示左侧横窦－乙状窦较对侧纤细，局部充盈欠佳，见充盈缺损，提示我们该患者可能合并静脉窦的狭窄，推测其发生视乳头水肿的机制可能为静脉窦狭窄的正反馈

循环假说。

静脉窦支架置入术正在兴起，并已被接受为治疗具有病理性压力梯度的静脉窦狭窄所致 IIH 患者的主要手段[16] [18]，一篇纳入 418 名患者接受静脉窦支架植入术的 IIH 患者的 Meta 分析报告表明[19]，通过静脉窦支架置入术的治疗方式，近 80%的患者头痛得到改善，90%的患者耳鸣得到改善，近 94%的患者乳头水肿得到改善。本例患者行静脉窦支架置入术后，头痛、阵发性黑矇症状消失，双眼视乳头水肿消退，视野缺损范围较前显著缩小，视功能得到明显改善，又一次印证了静脉窦支架置入术对于 IIH 合并静脉窦狭窄的患者疗效显著。近年来，随着 MRV、DSA 检查的普及，静脉窦狭窄在 IIH 患者中的检出率越来越高。30%~93%的 IIH 患者存在局灶性脑静脉窦狭窄[20]。静脉窦狭窄是颅内压升高的结果还是原因仍存在争议，影像学狭窄支架置入术的临床疗效是显著的[21]，2021 年脑静脉狭窄介入诊疗专家共识指出，术前充分完善静脉窦狭窄相关无创影像学评估，明确静脉窦狭窄情况，同时穿刺股动脉及股静脉；股动脉通路用于常规动脉造影和支架释放时的路图定位。股静脉通路，通过合理使用长鞘、导引导管和中间导管有助于通路头端通过乙状窦与颈静脉结合部的迂曲及颈静脉球结构，从而提高静脉窦支架置入过程的效率和成功率[22]。

大多数 IIH 的中国患者，当出现临床症状时，头痛和搏动性耳鸣等症状容易受到忽视，而视乳头水肿所致的视功能障碍给患者带来更直观的不适感[23]，因此 IIH 的患者常常首诊于眼科，眼科医生应该充分掌握 IIH 的临床表现及诊断标准，当接诊双眼视盘水肿的患者时，在排除视神经炎、缺血性视神经病变、颅内占位等常见病因外，还应行腰椎穿刺明确 CSF 压力，进一步完善颅脑 CTA、CTV、MRA、MRV、DSA 等检查明确颅内血管情况，及时发现并诊断，联合多学科的诊断和治疗，避免患者出现不可挽回的视功能丧失。

基金项目

烟台市科技局项目(2021YD025)。

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