

# 初诊颅内静脉窦血栓形成的硬脑膜动静脉瘘一例

王雪\*, 温蒲圆, 刘宏#

青岛大学附属烟台毓璜顶医院, 山东 烟台  
Email: 1312983786@qq.com, #mmemm1968@sina.com

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## 摘要

目的: 探讨硬脑膜动静脉瘘患者临床初始误诊为颅内静脉窦血栓形成一例的诊疗经过。方法: 分析1例初步诊断为颅内静脉窦血栓形成的硬脑膜动静脉瘘患者的临床资料。结果: 本例患者为中年男性, 临床表现为意识障碍、肢体活动不灵等症状, 早期颅脑影像学检查提示深静脉梗死、符合静脉栓塞表现, 经积极抗凝及对症治疗后临床病情进行性加重, 为寻找病因行全脑血管造影术提示为硬脑膜动静脉瘘。结论: 获得性硬脑膜动静脉瘘的早期诊断虽较为困难, 但及时行脑血管造影检查是确诊硬脑膜动静脉瘘的金标准, 结合影像学检查做出正确诊断, 可减少疾病的漏诊误诊。当遇到早期影像学结果支持颅内静脉窦血栓形成诊断且治疗后效果不佳的患者时, 应考虑硬脑膜动静脉瘘的可能性并积极行脑血管造影检查明确诊断, 避免错误治疗。

## 关键词

硬脑膜动静脉瘘, 颅内动静脉血栓形成, 全脑血管造影术

# Diagnosis and Treatment of Dural Arteriovenous Fistula with Intracranial Venous Sinus Thrombosis: A Case Report

Xue Wang\*, Puyuan Wen, Hong Liu#

Yantai Yuhuangding Hospital of Qingdao University, Yantai Shandong  
Email: 1312983786@qq.com, #mmemm1968@sina.com

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\*第一作者。

#通讯作者。

## Abstract

**Objective:** To investigate the diagnosis and treatment of a patient with dural arteriovenous fistula initially misdiagnosed as intracranial venous sinus thrombosis. **Method:** The clinical data of 1 case with dural arteriovenous fistula (DAVF) initially diagnosed as intracranial venous sinus thrombosis were analyzed. **Results:** The patient was a middle-aged male with symptoms of disturbance of consciousness and poor limb movement. Early brain imaging examination showed deep vein infarction, which was consistent with the manifestation of venous embolism. After active anticoagulation and symptomatic treatment, the clinical condition was gradually aggravated. In order to find the cause, whole brain angiography showed dural arteriovenous fistula. **Conclusion:** Early diagnosis of acquired dural arteriovenous fistula is difficult, but timely cerebral angiography is the gold standard for the diagnosis of dural arteriovenous fistula. Combined with imaging examination, correct diagnosis can reduce missed diagnosis and misdiagnosis of the disease. When the early imaging results support the diagnosis of intracranial venous sinus thrombosis with poor treatment results, the possibility of dural arteriovenous fistula should be considered and cerebral angiography should be performed actively to confirm the diagnosis to make a definite diagnosis and avoid wrong treatment.

## Keywords

Dural Arteriovenous Fistula, Intracranial Arteriovenous Thrombosis, Total Cerebral Angiography

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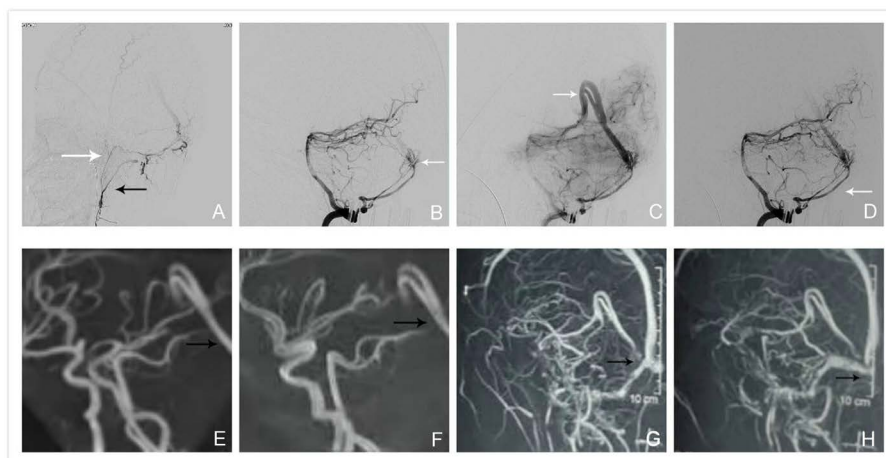
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## 1. 引言

硬脑膜动静脉瘘(dural arteriovenous fistula, DAVF)是指硬脑膜动脉与硬脑膜静脉、脑静脉窦、皮质静脉形成异常血管吻合,是一种少见的脑血管畸形,发病率不足 15% [1]。DAVF 的病因及发病机制目前尚存争议,其临床表现复杂多样,早期影像学常不典型,不易与颅内静脉窦血栓形成(intracranial venous sinus thrombosis, IVST)相鉴别,极易出现漏诊和误诊[2]。全脑血管造影术(Digital subtraction angiography, DSA)目前仍然是诊断和分类 DAVF 的金标准方法,但最新研究认为无创成像检查具有更高的实用性及安全性。本文通过分享一例初诊 IVST 的 DAVF 典型病例的诊疗经过,以期提高对该疾病的早期诊断及临床治疗。

## 2. 临床资料

男性患者,36岁,因“嗜睡、左侧肢体无力、言语不清4d”于2021年2月2日入院,发病前未服用药物。患者4天前出现睡眠增多、懒动,问话仅可回答单音节词;伴左侧肢体无力,左上肢无自主活动,左下肢搀扶下可行走,伴小便失禁。曾于当地医院就诊,查颅脑MRI+MRV(图1)显示:双侧丘脑异常信号,考虑深静脉梗死;直窦及右侧横窦、乙状窦、颈内静脉异常信号,符合静脉栓塞表现。给予抗凝及对症治疗,患者肢体活动较前略好转,左上肢可于床面平移,话语较前增多,时有自言自语,内容不被理解。患者病后精神萎靡不振、食欲较差,睡眠增多,大便正常,有小便失禁,体重无明显减轻。既往史和家族史无特殊,无手术和外伤史,否认家族中有类似患者。结合患者症状及病例资料初步诊断为颅内静脉窦血栓形成。



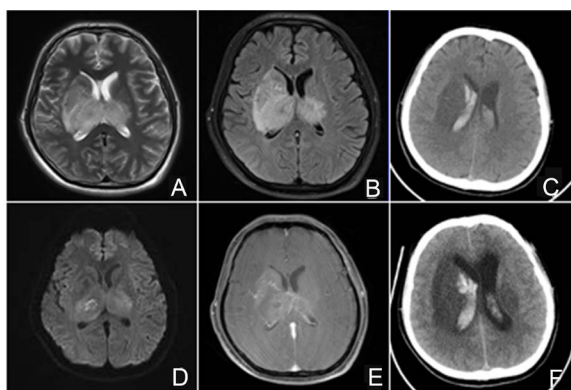
注: A: DSA 示左侧颈外动脉分支(白色箭头咽升动脉、黑色箭头枕动脉)供血; B、C: DSA 示左侧脑膜后动脉向窦引流经直窦反流; D: DSA 示左侧脑膜后动脉供血; E、F: 颅脑 MRA; G、H: 颅脑 MRV。

**Figure 1.** DSA, MRA, MRV

**图 1.** DSA、MRA、MRV

入院查体: T 36.8°C, P 84 次/分, R 16 次/分, BP 146/87 mmHg (1 mmHg = 0.133 kPa)。心肺腹查体无异常。神经系统查体: 嗜睡, 反应迟钝, 高级智能检查欠配合。脑膜刺激征阴性。双侧瞳孔等大正圆, 对光反射灵敏。构音不清。左上肢肌张力减低, 左上肢粗测肌力 3 级, 左下肢粗测肌力 2 级, 右侧肢体肌力 5 级。共济运动检查不配合。感觉系统正常对称, 四肢腱反射存在, 双侧病理征阴性。

实验室及影像学检查: 入院后腰椎穿刺示脑初压 180 mm H<sub>2</sub>O (80~180 mm H<sub>2</sub>O), 无色透明; 脑脊液常规: 白细胞  $13 \times 10^6 \cdot L^{-1}$  ( $0 \sim 8 \times 10^6 \cdot L^{-1}$ ); 脑脊液生化: 蛋白  $1412.7 \text{ mg} \cdot L^{-1}$  ( $150 \sim 400 \text{ mg} \cdot L^{-1}$ ); 脑脊液细菌培养无阳性结果; D-二聚体  $0.37 \text{ mg} \cdot L^{-1}$  ( $0 \sim 0.8 \text{ mg} \cdot L^{-1}$ ); 血常规: 白细胞  $9.21 \times 10^9 \cdot L^{-1}$  ( $3.5 \sim 9.5 \times 10^9 \cdot L^{-1}$ )、中性粒细胞百分率 77.2% (40%~75%); 自身抗体相关检验、肿瘤标记物正常。查颅脑增强 MRI (图 2) 示: 双侧丘脑、大脑脚及右侧基底节区异常信号, 符合深静脉梗死伴少量出血。2021 年 2 月 9 日患者出现反复剧烈呕吐, 考虑颅内高压、怀疑出血, 行颅脑 CT (图 2) 见: 双侧丘脑、基底节区、右侧放射冠区、双侧大脑脚改变, 符合深静脉血栓所致梗死伴脑室内出血表现。复行腰椎穿刺示脑内压 330 mm H<sub>2</sub>O, 脑脊液性状红色浑浊。2021 年 2 月 13 日颅脑 CT (图 2): 右侧侧脑室旁血肿较前稍增大, 左侧脑室内积血较前吸收, 第三四脑室积血较前稍增多, 脑水肿较前进展。



A: 颅脑 MRI T2 相; B: 颅脑 MRI Flair 相; C: 颅脑 CT; D: 颅脑 MRI DWI 相; E: 颅脑 MRI 增强相; F: 复查颅脑 CT

**Figure 2.** Brain imaging examination

**图 2.** 颅脑影像学检查

诊疗经过：患者入住科室后立即给予甘露醇(山东齐都药业股份有限公司, H37020780) 125 ml q6h 联合甘油果糖(辰欣药业股份有限公司, H20057114) 250 ml q12h 脱水减轻颅内水肿、低分子肝素钠(齐鲁制药有限公司, H20030429)5000IU q12h 抗凝、注射用纤溶酶(北京赛升药业股份有限公司, H11022110) 100 IU · d<sup>-1</sup> 溶栓及对症等治疗。住院期间患者出现颅高压症状, 查头部影像学检查考虑出血量不大, 停止抗凝、溶栓用药, 行“侧脑室引流术”降低颅内压力, 余继续予以艾地苯醌 30 mg 3 次/日、银杏叶片 80mg 3 次/日营养神经、依达拉奉 30 mg 2 次/日清除自由基、雷贝拉唑 10 mg · d<sup>-1</sup> 抑酸护胃及补液等药物保守治疗。术后患者病情进行性加重, 且 2021 年 2 月 13 日头部影像学提示颅内出血增多、脑室铸型, 为明确临床诊断, 进一步行 DSA (图 1), 术中发现直窦、右侧横窦未显影; 左侧颈外动脉经左侧枕动脉、左侧咽升动脉、左侧颞浅动脉分支直接供血至窦汇; 左侧椎动脉无明显异常; 右侧椎动脉由右侧脑膜后动脉供血, 引流至窦汇。术中并行“硬脑膜动静脉瘘栓塞术 + 颅内动静脉畸形切除术 + 颅内血肿清除 + 去骨瓣减压术”。术后患者病情继续进展加重, 且出现多脏器功能衰竭, 向家属充分说明患者病情及预后, 家属放弃治疗, 并签字出院。

### 3. 讨论

本例患者为中年男性, 虽然缺乏 IVST 常见的病因和危险因素, 如: 全身衰竭、颅脑外伤、血液疾病、自身免疫性疾病、妊娠等[3] [4], 但颅脑 MRI + MRV 呈现静脉栓塞表现, 因此得出颅内静脉窦血栓形成的初步诊断。依照 IVST 急性期的治疗原则, 对该患者给予低分子肝素联合尿激酶溶栓, 治疗效果不满意。因患者以双侧丘脑对称性病变为主要表现, 治疗后疗效不佳遂考虑除 IVST 外其他疾病成立的可能性: 1) 硬脑膜动静脉瘘: DAVF 好发于横窦、乙状窦和海绵窦区[5], 发生于直窦附近的 DAVF 可造成双侧丘脑病变, 由于累及双侧丘脑上行激活网状系统与大脑皮质间的往返投射联系[6] [7], 患者不易从临床表现上与 IVST 导致的丘脑梗死相区别。2) 乙型脑炎: 乙脑病毒感染引起, 流行于夏秋季。起病急, 体温可急剧上升至 39℃ 以上, 部分患者有嗜睡, 可表现为癫痫发作、意识不清。颅脑 MRI 可见双侧丘脑、中脑等部位对称性病变。本患者无发热, 脑脊液异常, 有待排除诊断。3) 渗透性脱髓鞘疾病: 多有颅内渗透压快速改变病史, 与慢性酒精中毒、快速补钠、肝移植等相关, 出现皮质脊髓束和皮质脑干束或基底节区受损的表现。颅脑 MRI 表现为双侧丘脑、基底节区对称性病变, 本患者病史不支持。4) 中枢神经系统淋巴瘤: 病程多在 6 个月以内, 病理性占位效应或弥散性脑水肿引发症状。本患者表现为双侧丘脑对称性病变, 但病变周围水肿占位效应不明显, 肿瘤标记物无异常, 暂不考虑。通过多角度的思考仍不能明确诊断, 遂行脑血管造影。

静脉窦血栓 MRI 直接表现为静脉窦内血栓信号, 间接表现为脑缺血或白质内水肿、实质出血、皮质水肿等信号; MRV 可见静脉窦走行方向上的充盈缺损及血栓形成。但是早期 DAVF 的影像学表现不典型。虽然 MRI 可以显示动静脉瘘口位置及供血动脉, 但 DAVF 由颈外动脉供血病例检查常表现为阴性结果, 从而出现漏诊[8] [9]。因此, 疾病早期单纯行 MRI + MRV 并不能为鉴别 DAVF 和 IVST 提供有效依据。由于本例患者早期 MRI + MRV 符合深静脉梗死表现, 临床症状也可以用深静脉梗死解释, 因此在初期得出颅内静脉窦血栓形成的诊断。患者颅内出血进行性增多, 结合影像结果考虑静脉性梗死后脑疝形成, 脑室引流术后效果不佳。回顾患者影像结果, MRA (图 1)上可见早期显影的静脉血管, 且 MRI 增强相(图 1)出现流空征, 不支持静脉窦血栓的诊断。DSA 是确诊 DAVF 的金标准, 可显示相关血管情况, 降低出血风险[10]。为明确诊断, 后期对患者行 DSA 发现直窦、右侧横窦不显影, 左侧动脉供血窦汇形成动静脉瘘, 故而最终确诊为硬脑膜动静脉瘘。

硬脑膜动静脉瘘具体病因及发病机制有待进一步深入研究, 有观点认为成人 DAVF 后天获得性因素大于先天[11], 可能与创伤、炎症、雌激素水平、缺血相关[12], 其中, IVST 被认为是导致 DAVF 的独



立因素,且IVST和DAVF二者互为因果[13][14][15]。目前受到大多数学者认可的观点是,静脉窦高压对DAVF起到关键作用[16][17][18][19]:①动静脉吻合开放学说:硬脑膜动静脉间存在生理性通道,相关区域静脉窦内高压后出现静脉回流受阻,血液瘀滞,动静脉短路开放和血管改造,动静脉间交通支病理性开放,演变成为DAVF[20]。②静脉窦高压诱导的血管化生学说:静脉窦高压和脑组织缺血缺氧时,硬膜中血管内皮生长因子(vascular endothelial growth factor, VEGF)大量产生,VEGF协同缺氧诱导因子 $1\alpha$ 、碱性成纤维生长因子、转移生长因子 $\alpha$ 、基质金属蛋白酶(MMP)-2-1306 C/T,引起血管重塑,最后形成DAVF[21][22]。③静脉窦血栓激化学说:颅内静脉窦血栓形成后可引起静脉窦高压,形成DAVF后血液湍流损伤血管壁又促进血栓形成,进一步增加静脉窦压力[23]。概而言之,静脉窦高压是DAVF形成的核心,而静脉窦血栓形成则是静脉窦高压的重要危险因素。

#### 4. 结论

对于确诊的IVST患者抗凝治疗被视为急性期首选治疗措施[24],但是DAVF后正常静脉窦因废用最终狭窄及闭塞,长期增高的皮层静脉压力会造成血栓形成、出血或梗死。DAVF患者若误诊IVST给予长期抗凝,其出血风险会增高[25][26]。获得性DAVF的早期诊断虽较为困难,但及时行DSA是确诊DAVF的金标准,结合影像学检查做出正确诊断,可减少疾病的漏诊误诊。在本次病例中,早期由于未关注MRV检查的局限性而导致误诊脑静脉血栓,表明一部分不典型动静脉瘘案例可能会因为检查手段的局限性而误诊。且神经科介入医师在本病例中行静脉窦内选择性造影时未行测压以判断静脉内血栓形成。因此,当遇到早期影像学结果支持IVST诊断且治疗后效果不佳的患者时,应考虑DAVF的可能性并积极行DSA明确诊断。近期多项临床研究显示,血管内治疗具有创伤小、住院时间短的双重优势,同时积极施行通畅静脉引流、减轻颅内高压治疗,将有效改善患者预后[27][28]。

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