·综 述 General review·

硬脊膜动静脉瘘诊断、治疗和预后进展

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【摘要】 硬脊膜动静脉瘘(SDAVF)是一种临床表现多样、误诊率高的罕见疾病。随着无创血管成像技术发展,SDAVF 诊断有了一定提高,但脊髓血管造影仍是诊断 SDAVF 金标准。SDAVF 是一种非自限性疾病,一旦发病均需外科手术或血管内栓塞治疗。随着血管内栓塞材料和技术进步,血管内治疗成功率逐渐增高,但外科手术治疗成功率仍数倍于血管内栓塞治疗。尤其是近年外科手术中微创技术发展,使得 SDAVF 患者获益更大。治疗成功后大部分患者症状可稳定或改善,而未能获早期诊断和治疗常可导致不可逆性神经功能损伤。该文就 SDAVF 诊断、治疗和预后研究进展作一综述。

【关键词】 硬脊膜动静脉瘘;诊断;治疗;预后
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Recent advances in the diagnosis, treatment and prognosis of spinal dural arteriovenous fistula *XU Tianming, CAI Dongyang, XUE Jiangyu, HE Yingkun, LI Tianxiao. Section of Cerebrovascular Disease, Intervention Centre, Henan Provincial People's Hospital, Zhengzhou, Henan Province 450003, China*

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【**Abstract**】 Clinically, spinal dural arteriovenous fistula(SDAVF) is a rare disease with various clinical manifestations and high misdiagnosis rate. With the technological development of non-invasive angiography, the diagnostic ability for SDAVF has been certainly improved, although digital subtraction spinal angiography is still the gold standard for the diagnosis of SDAVF. SDAVF is a non-self-limited disease, it requires surgical or endovascular embolization treatment once the disease comes on. With the advances in materials and techniques for endovascular embolization, the success rate of endovascular therapy has gradually increased, but the success rate of surgical treatment is still several times higher than that of endovascular embolization treatment. In recent years, the development of minimally-invasive surgical techniques has greatly benefited patients with SDAVF. In most patients who has achieved a successful treatment the clinical symptoms become stabilized and improved, while in the patients who has not got early diagnosis and treatment the irreversible neurological impairment often occurs. This article aims to make a detailed review about the recent advances in the diagnosis, treatment and prognosis of SDAVF. (J Intervent Radiol, 2021, 30: 1300-1304)

【**Key words**】 spinal dural arteriovenous fistula; diagnosis; treatment; prognosis

硬脊膜动静脉瘘(spinal dural arteriovenous fistula, SDAVF)是一种罕见的发生在椎间孔处硬脊膜表面根硬膜动脉与髓周静脉间获得性、低流量动静脉短路性疾病,年发病率为 5～10/100 万[1],但又是最常见的脊髓血管畸形,约占 70%[2-3]。好发于中老年人,55～60 岁多见,罕见于儿童和青少年[4];男性好发,男女比 5:1[5]。由于罕见且临床表现不具特异性,早期诊断非常困难、误诊率高,发病至确诊平均需要 15 个月[6-7]。文献报道表明发病至确诊时间与患者预后相关[5,8],因此早期认识 SDAVF 至关重要。现就本病诊断、治疗及预后研究进展作一综述。

1 SDAVF 血管解剖和病理生理学

SDAVF 可发生于任何脊髓水平,单瘘口多见,2 个或 2 个以上瘘口发病率低于或接近 2%[9]。SDAVF 好发于中下胸段,其次是腰段和上胸段[10];骶部约

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占 4%,颅颈交界区约占 2%,发生于 C2 以下和 T1 以上下颈段非常罕见[11];高度发达的颈髓静脉网络可阻止 SDAVF 所致静脉压升高,因此一些无症状 SDAVF 好发于颈部[12],下颈段 SDAVF 发病率可能被低估。

节段动脉(肋间动脉、腰动脉、髂内动脉、椎动脉分支、颈深动脉、颈升动脉)根脊膜支参与硬脊膜供血,因炎性反应、外伤或引流静脉血栓形成等原因位于神经根袖套或附近的硬脊膜内动静脉间潜在通道被迫开放而形成 SDAVF[13-14],并通过根髓静脉引流至髓周静脉系统,与正常脊髓静脉合并形成逆流。压力较高的动脉经瘘口流入根髓静脉,并引流入脊髓冠状静脉丛,而脊髓静脉引流只通过少量根髓静脉流入硬膜外静脉丛,且脊髓静脉系统无静脉瓣,引起脊髓静脉淤血、静脉高压,髓内静脉压可达平均动脉压 74%[15],导致脊髓组织灌注减少、水肿、缺氧和血脊髓屏障破坏。慢而广泛的静脉引流是 SDAVF 特征,静脉引流向上可至颈髓和后颅窝,向下可至脊髓圆锥[16]。硬脊膜支供血不同于根髓动脉,后者供应脊髓前、后动脉并供应脊髓,与脊髓动静脉畸形不同,SDAVF 瘘口由不需供应脊髓的硬脊膜支供应,故不易发生脊髓盗血症状。

2 临床表现与诊断

SDAVF 好发于中老年男性,临床表现主要为慢性进展性脊髓病继发的神经功能缺陷,包括步态障碍、下肢无力、感觉障碍、括约肌功能障碍等,以下肢无力和感觉障碍为首发症状患者超过 70%,其次为括约肌障碍,临床上常遇主诉单侧下肢进行性乏力发展到双下肢,有脚踩棉花异常感觉;上肢不常受累,当上肢受累时提示病变可能在颈部,神经根痛和腰痛也可见[17]。颅颈交界区 SDAVF 患者蛛网膜下腔出血发生率较高[18],也可表现为脑干功能障碍[19]。腰部 SDAVF 也有出血报道[20],但病变低流量引流特性使得胸腰区出血极为罕见。性功能障碍,直肠、膀胱功能障碍通常是 SDAVF 疾病进展先兆,伴发症状和症状恶化是疾病进展特征。部分 SDAVF 患者在运动期间症状会出现加重[21],可能是由于运动期间平均动脉压升高引起脊髓静脉压升高所致。部分患者在给予激素治疗后原有症状会加重[22-24],可能是因为激素应用引起脊髓血流灌注发生改变,使得脊髓静脉压升高,导致脊髓进一步损伤。部分患者在脊髓血管造影术后会出现症状加重[25]。临床特征与其他诊断(如退行性椎间盘疾病、脊髓炎、前列腺肥大等)常常重叠,感觉障碍平面多位于腰骶部,易误诊[8,22]。发病至确诊时间较长,病程进展通常呈渐进性,约 5%患者为急性发病[26]。神经内科多以视神经脊髓炎、脊髓炎、多发性硬化等进行治疗,部分患者以腰椎间盘突出、椎管狭窄在骨科手术治疗[27]。由于患者多为中老年人,部分患者因括约肌功能障碍被误诊为前列腺增生在泌尿外科治疗[28]。有文献报道患者病程越短预后越好[5],因此早期诊断对 SDAVF 患者至关重要。

MRI 检查能清楚显示脊髓及其周围软组织,可用作 SDAVF 初步筛查。脊髓水肿在 T2 加权成像上高信号对 SDAVF 诊断非常敏感[27]。但这些表现也可见于创伤、脱髓鞘病变及椎间盘突出等,不具有特异性。髓周静脉在病情发展过程中逐渐迂曲、扩张,在 T2 加权成像上表现为血管流空影,这对 SDAVF 诊断有特异性[29]。髓内 T2 高信号和髓周静脉迂曲扩张是 SDAVF 常见影像学改变,可见于 70%以上患者[10],然而在约 50%病例中这些影像学改变往往与瘘口位置无关[27]。

高分辨率和对比增强 MRA 在显示 SDAVF 迂曲扩张的引流静脉方面具有很高准确性,对瘘口识别有一定帮助。CTA 不仅能检出迂曲、扩张髓周静脉,且对瘘口识别有较高准确率[30]。CTA 扫描速度快、范围广,但由于 SDAVF 瘘口可能位于脊髓任何水平,对瘘口位置不明确者需扫描整个脊髓,会使患者所受辐射剂量增大。虽然 MRA 和 CTA 并不总能精确定位瘘口,但可为脊髓血管造影提供瘘口最有可能位置预判,避免所有节段动脉不必要造影。

脊髓血管造影是确诊 SDAVF 金标准[10],既能准确定位瘘口位置,又能评估 SDAVF 引流静脉和供血动脉血流动力学。脊髓血管造影可在中度镇静和局部麻醉下进行,也可在全身麻醉下进行,后者可保持静止和应用间歇呼吸暂停,以消除呼吸伪影,获得更好的图像质量。术前通过仔细阅读脊髓 MRA 和 CTA 评估瘘口可能位置,先行此节段动脉造影。如未发现病变,因病变好发于胸腰椎,一般选择自 T12 肋间动脉开始向上、向下逐个节段动脉造影。造影中应对 Adamkiewicz 动脉进行定位(典型起源为 T9～T12[26])并评估其静脉引流,严重静脉高压和脊髓病常导致静脉引流延迟或缺失。典型病例血管造影正位相上,这些纵向的硬膜供血动脉和引流静脉呈水平 T 型[31],自硬脊膜支至根静脉血流可能是缓慢的,因此在行脊髓血管造影寻找瘘口时应以低帧率并坚持至少 4 s 以上,从而排除延迟的根静

脉逆行充盈,减少假阴性发生。

3 治疗

SDAVF 治疗方式有血管内栓塞和外科手术。外科手术主要是切断硬膜内引流静脉近端[32],血管内治疗主要应用液体栓塞剂,在超选至供血硬脊膜支后进行栓塞,栓塞剂必须经瘘口闭塞引流静脉近端[33-34]。动脉阻塞会导致症状短暂改善,但因硬膜良好的吻合网,瘘口易在术后几个月内复发[35]。与单纯血管内栓塞相比,外科手术阻断 SDAVF 具有明显优越性。单纯血管内栓塞与外科手术相比初始失败发生率、晚期复发率显著增高。随着血管内治疗技术和材料进步,血管内治疗成功率逐渐提高,达到 80%,但外科手术治疗成功率仍是单纯栓塞的数倍[2,36]。

考虑到外科手术创伤性,具有创伤小、定位准确、并发症少等优点的血管内治疗仍在临床决策中占有很重要位置,尤其是针对具有合适血管解剖结构患者[2]。乙烯-乙烯醇共聚物(Onyx)栓塞与 α-氰基丙烯酸正丁酯(NBCA)相比,治疗失败和晚期复发概率显著增加[2],这可能由于 Onyx 胶到达不了引流静脉,而引流静脉阻断对于最终治疗至关重要。

外科手术中微创技术发展使得手术阻断更有效、更可行,切口更小,住院时间更短[37]。手术关键是瘘口准确定位。打开硬脊膜通常可见扩张迂曲的静脉呈动脉化改变,瘘口往往在神经根附近硬脊膜上,临时动脉夹阻断引流静脉近硬膜处通常可见动脉化静脉塌陷、颜色由鲜红变暗。术中应用吲哚青绿(ICG)血管造影,可很好地识别瘘口、供血动脉及引流静脉。有文献报道 ICG 应用与良好预后相关[38]。随着复合手术发展,对于复杂 SDAVF,术中可重复造影确定瘘口位置,调整夹闭硬膜内引流静脉近端,确保安全、准确阻断瘘口[39]。常规有效的抗凝治疗,可能不会显著降低术后急性恶化风险,也不会影响远期疗效,但能逆转术后急性恶化[40]。

4 预后

SDAVF 成功治疗后,约 90%患者症状稳定或改善,运动功能障碍得到改善概率与感觉和膀胱功能障碍相比更高,性功能障碍很少改善,疼痛也可能持续存在[41-42]。综合文献分析,影响患者预后的因素主要有术前症状严重程度、发病至确诊时间、脊髓水肿程度、髓周静脉迂曲长度等。下面就这些影响因素进行分析。

通常认为,SDAVF 患者术前症状严重程度决定了预后改善程度[10,42]。但也有研究表明两者无相关性[43]。SDAVF 患者术前症状严重程度是否与术后改善情况相关,目前尚无统一定论。但 SDAVF 治疗对包括截瘫在内的严重神经症状患者是有益的[44],因此即使是晚期神经功能恶化也不能排除改善可能。

SDAVF 患者发病至确诊时间是预测预后的另一重要因素。Jablawi 等[5]报道将病程分为 6 个月、7～18 个月、18 个月以上,通过术后平均 52 个月随访发现病程越短,预后越好。然而 Cenzato 等[45]在术后 3 年随访中发现,患者症状缓解程度与病程长短并无明确相关关系。由于 SDAVF 发病率低,且上述随访结果均为回顾性研究,对于病程对预后的影响仍需大样本前瞻性研究证实。

脊髓水肿程度和髓周静脉迂曲长度也是判断预后的重要因素。术前 T2 加权成像所示脊髓水肿范围与患者严重程度有关,但术后脊髓水肿消退与症状缓解并无明确相关性[33]。术前脊髓水肿范围与预后无相关性[5,10]。Hetts 等[46]报道在 31 例患者随访中发现,患者预后与诊断时髓周静脉扩张程度呈正相关。然而 Jablawi 等[5]发现,髓周静脉扩张程度与预后无相关性。但脊髓静脉系统慢性病理性动脉化,可引起脊髓静脉引流不可逆改变,即使在瘘口充分阻断情况下,也可能显著影响功能恢复[47-48]。

5 结语

上述 SDAVF 血管解剖、病理生理学、临床表现、诊断、治疗和预后等方面阐述提示,SDAVF 患者早期诊断、及时治疗很重要,因为该病变自然史是一临床恶化过程,有可能造成不可逆性脊髓损伤。为了获得最佳临床结果,神经内科、神经外科及脊柱外科医师应充分认识该疾病,早识别、早诊断、早治疗、早康复。

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