

. 综 述 .

颅颈交界区硬脊膜动静脉瘘的诊治进展

徐田明 蔡栋阳 薛绛宇 贺迎坤 综述 李天晓 审校

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颅颈交界区硬脊膜动静脉瘘(cranio cervical junction spinal dural arteriovenous fistulas, CCJ-SDAVFs)是一种发生在枕骨大孔至第2颈椎之间的罕见的血管畸形^[1],约占SDAVFs的2%^[2,3],好发于中老年人,男性多发^[4]。由于CCJ-SDAVFs的发病率低、解剖结构复杂且临床表现广泛,极易造成漏诊、误诊^[5-8],从而造成严重的临床后果。本文就该病的血管构筑、发病机制、临床表现、诊断及治疗的研究进展进行综述。

1 CCJ-SDAVFs的血管构筑和发病机制

CCJ的神经、血管解剖结构较胸腰椎更为复杂^[9],神经组织包括脑干、上颈髓,第1、2脊神经,副神经的脊神经支等^[5];血管结构包括椎动脉及其第1、2脊膜支,根动脉,脊髓前动脉,脊髓后外侧动脉,根静脉和椎旁静脉丛等^[5,10]。与颅后窝和脊髓相比,CCJ的硬脑膜有发达的血管网络^[11]。

CCJ-SDAVFs的供血动脉主要为椎动脉的硬脊膜支^[12],右侧椎动脉多见^[1];也可见咽升动脉和枕动脉的脑膜支参与供血^[4,13]。CCJ硬膜供血动脉常存在吻合^[14],所以,CCJ-SDAVFs常合并有脊髓软膜动脉的参与供血^[12,15]。Adrianto等^[16]报道显示14%的CCJ-SDAVFs有脊髓前动脉或脊髓后外侧动脉的参与供血。

CCJ-SDAVFs通常引流至髓静脉、冠状静脉丛、颅内静脉系统,少数直接汇入硬膜外静脉和椎旁静脉丛^[4]。根静脉从瘘口流出,并将动脉化的静脉血经无静脉瓣的冠状静脉及放射状静脉引流至脊髓,引

起,最终导致静脉高压综合征^[17,18]。正常情况下,脊髓的静脉引流是下行的,而脑干静脉和颈髓静脉之间存在丰富的吻合^[1,3]。当正常的引流静脉受阻时,可向上引流至颅内静脉系统,如海绵窦、岩下窦、窦汇或者皮层静脉^[4],颅内引流通常静脉流速较快因血流动力学原因可至静脉曲张或静脉瘤的形成,易发生静脉性蛛网膜下腔出血^[14]。

CCJ-SDAVFs的发病机制尚不清楚,常合并脑动脉狭窄及脑动脉瘤等疾病,表明CCJ-SDAVFs可能为一种血管退行性疾病^[1]。高血压所致的血流动力学改变对CCJ-SDAVFs的起源和发展有重要意义^[19]。年龄、创伤、感染等后天因素可引起静脉纤维化和血栓形成,引起节段性静脉引流障碍和静脉高压,而动物模型研究显示静脉高压可诱导动静脉瘘的形成^[1]。

2 CCJ-SDAVFs的临床表现及诊断

CCJ-SDAVFs是一种罕见的血管畸形,具有广泛的临床表现,常表现为蛛网膜下腔出血、脊髓病、脑干功能障碍等^[14]。有些病人还可表现为神经根病变^[20]、颅神经麻痹^[21]。罕见的临床表现包括小脑出血^[6]、桥脑出血^[7]、癫痫^[22]等。CCJ-SDAVFs导致的蛛网膜下腔出血好发于颅后窝和脑干前部,部分病人仅表现为第四脑室少量出血^[1]。与颅内动脉瘤性蛛网膜下腔出血相比,CCJ-SDAVFs病人临床表现相对较轻,95%的病人Hunt-Hess分级为1、2级^[23]。

以蛛网膜下腔出血为首发症状的病人,行CT检查;对于出血量较少、CT显影不清者,可行腰椎穿刺术检查。表现为脊髓病或脑干功能障碍而怀疑CCJ-SDAVFs的病人,可行头颈部MRI检查,多可见脑干或脊髓水肿,颈髓腹侧或背侧面的虫蚀样血管流空影^[24]。由于CTA无法反应血流动力学因素且受颅后窝解剖结构的影响,对CCJ-SDAVFs的确诊比较困难,但是可为解剖入路提供参考。Fujimoto

等^[5]报道应用计算机图形学将动脉、静脉、脊髓和硬脑膜等融合,了解其解剖结构,有助于提高复杂 CCJ-SDAVFs 显微手术的效果。目前,DSA 仍然是诊断 CCJ-SDAVFs 的金标准^[1,4,5],但存在一定概率的假阴性^[25]。假阴性多见于无静脉曲张的低流量瘘或合并急性脑积水的 CCJ-SDAVFs 病人^[1];对于无静脉曲张的低流量瘘所致的假阴性,在造影寻找瘘口时,应用低帧率并坚持至少 4 s 以上以排除延迟的根静脉逆行充盈^[24];对于蛛网膜下腔出血后血肿压迫或脑积水继发颅内压增高引起瘘口暂时性闭塞的病人,急性期后再次行 DSA 明确诊断。此外,超过一半的 CCJ-SDAVFs 的供血动脉来自右侧椎动脉的硬脊膜支^[1,25],而左侧椎动脉通常为血流优势侧,当有足够的血流进入右侧椎动脉使右侧小脑后下动脉显影时,一般只行左侧椎动脉造影^[25],这也会造成假阴性。

3 CCJ-SDAVFs 的治疗及预后

CCJ-SDAVFs 治疗方式有手术治疗和血管内栓塞治疗^[13]。手术治疗主要是切断硬膜内引流静脉的近端^[26];血管内栓塞治疗主要应用液体栓塞剂,在超选至供血的硬脊膜支后栓塞,栓塞剂必须通过瘘口闭塞引流静脉的近端^[27],以防复发。

血管内治疗具有创伤小、定位准确等优点。随着血管内治疗技术及材料的进步,血管内栓塞治疗的成功率逐渐增加^[28]。但 SDAVFs 的血管内治疗的可行性在很大程度上取决于病变的血管结构,而 CCJ-SDAVFs 供血动脉通常以直角从椎动脉发出,且供血动脉小且弯曲,微导管难以到达瘘口位置^[1,18];且椎动脉、枕动脉和咽升动脉的脑膜支之间存在复杂的吻合,使 CCJ-SDAVFs 的栓塞复杂化^[14]。然而,如果病变位于高风险部位或来自高风险病人群体,血管内治疗的优点将大大超过上述限制。

手术治疗是 CCJ-SDAVFs 最佳的治疗方案^[15,18,20],手术的关键是术中准确定位瘘口。术中根髓动脉可能被误认为 CCJ-SDAVFs 的引流静脉,且 CCJ-SDAVFs 硬膜内动脉常与根脊膜支存在吻合^[5],因此术中可以一枚动脉夹临时夹闭可疑血管,行术中血管造影或吲哚菁绿造影加以辨别^[5,18]。手术治疗 CCJ-SDAVFs 成功率高、复发率低^[28],但存在术后感染、脑脊液瘘等并发症,术中应严密缝合硬膜,若硬膜无法严密缝合,应去枕骨肌上取一块筋膜严密缝合。

对于以蛛网膜下腔出血为首发表现的 CCJ-

SDAVFs,即使出血量少,因有再次出血的风险^[14],治疗仍然是必须的。存在蛛网膜下腔出血的 CCJ-SDAVFs 病人手术治疗预后良好^[1]。

对于表现为脊髓病的 CCJ-SDAVFs,通常认为术前症状的严重程度与其预后直接相关^[29,30],但也有研究表明两者无相关性^[31]。从发病至确诊的时间也是评估预后的因素,通常认为病程越短,预后越好^[31]。也有文献表明病人的预后与病程的长短无明确相关性^[32]。所以,表现为脊髓病的 CCJ-SDAVFs 的预后,仍需进一步研究。

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