·病例报告·

椎基底动脉延长扩张症伴基底动脉夹层动脉瘤致蛛网膜下隙出血一例

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【关键词】 椎动脉； 基底动脉； 扩张,病理性； 动脉瘤,夹层； 蛛网膜下腔出血； 病例报告

【Key words】 Vertebral artery; Basilar artery; Dilatation, pathologic; Aneurysm, dissecting; Subarachnoid hemorrhage; Case reports

Subarachnoid hemorrhage caused by vertebrobasilar dolichoectasia complicated with basilar artery dissecting aneurysm: one case report

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患者 男性,59岁,主因突发头晕伴行走不稳3 d,于2015年9月20日入院。患者3 d前跑步时无明显诱因突发头晕伴行走不稳,休息后症状加重并出现右眼视物模糊和右侧听力下降,当地医院行头部CT和MRI检查提示基底动脉延长扩张症伴动脉瘤,右侧小脑新发梗死(图1a,1b)。为求进一步手术治疗,遂至我院就诊。患者既往有高血压(高血压3级,极高危)20余年,血压最高达180/110 mm Hg(1 mm Hg=0.133 kPa),规律服用硝苯地平20 mg/d,血压控制尚可;无明确的颅内出血和脑梗死病史;个人史及家族史无特殊。入院后体格检查:血压150/95 mm Hg,一般状况良好,粗测双眼视力正常,右侧听力下降,右侧指鼻试验欠稳准,Romberg征阳性,余未见明显异常。影像学检查:头部高分辨力磁共振黑血血栓成像(MRBTI)显示,椎基底动脉延长扩张症伴基底动脉近段夹层动脉瘤(图1c,1d)。数字减影血管造影术(DSA)亦提示椎基底动脉延长扩张症伴基底动脉近段夹层动脉瘤(图1e~1h)。结合临床病史考虑夹层动脉瘤进展使右侧小脑后下动脉开口闭塞,从而导致右侧小脑后下动脉供血区梗死。为防止动脉瘤夹层扩大或破裂,考虑行双支架辅助弹簧圈栓塞术,以达到修复血管夹层之目的。术前3 d常规口服阿司匹林100 mg/d和氯吡格雷(波立维)75 mg/d抗血小板治疗,但计划手术当日清晨患者突发剧烈头痛,进而出现进行性意识障碍。体格检查:浅昏迷,双侧瞳孔等大、等圆,直径约2.50 mm,颈项强直,Hunt-Hess分级Ⅳ级,气管插管后行床旁头部CT检查提示蛛网膜下隙出血(图1i),考虑基底动脉夹层动脉瘤破裂所致,遂急诊行血管内栓塞治疗。患者仰卧位,气管插管全身麻醉,双侧股动脉穿刺置入6F动脉鞘并予常规全身肝素化(首剂量为2~3 mg/kg,1 h后追加前次剂量的1/2),在DSA路径图引导下将2根5F导引导管分别置入双侧椎动脉V2段远端(在不过度干扰后循环血流的情况下分别置入支架微导管和弹簧圈微导管)。首先,采用支架半释放技术经右侧椎动脉经Vasco微导管(法国Balt公司)将4.50 mm×50.00 mm的Leo支架(法国Balt公司)植入基底动脉中下段并释放至其覆盖的动脉瘤瘤颈远端,此时经左侧椎动脉经Traxcess 14微导丝(美国MicroVention公司)将Echelon 10微导管(美国EV3公司)头端送至动脉瘤瘤腔内。然后向动脉瘤瘤腔内致密填塞弹簧圈5枚(包括7 mm×30 mm弹簧圈成篮、5 mm×15 mm弹簧圈1枚、4 mm×12 mm弹簧圈3枚,美国EV3公司),再将Leo支架完全释放,使其尾端覆盖右侧椎动脉V4段病变处;经右侧椎动脉将另1枚4.50 mm×

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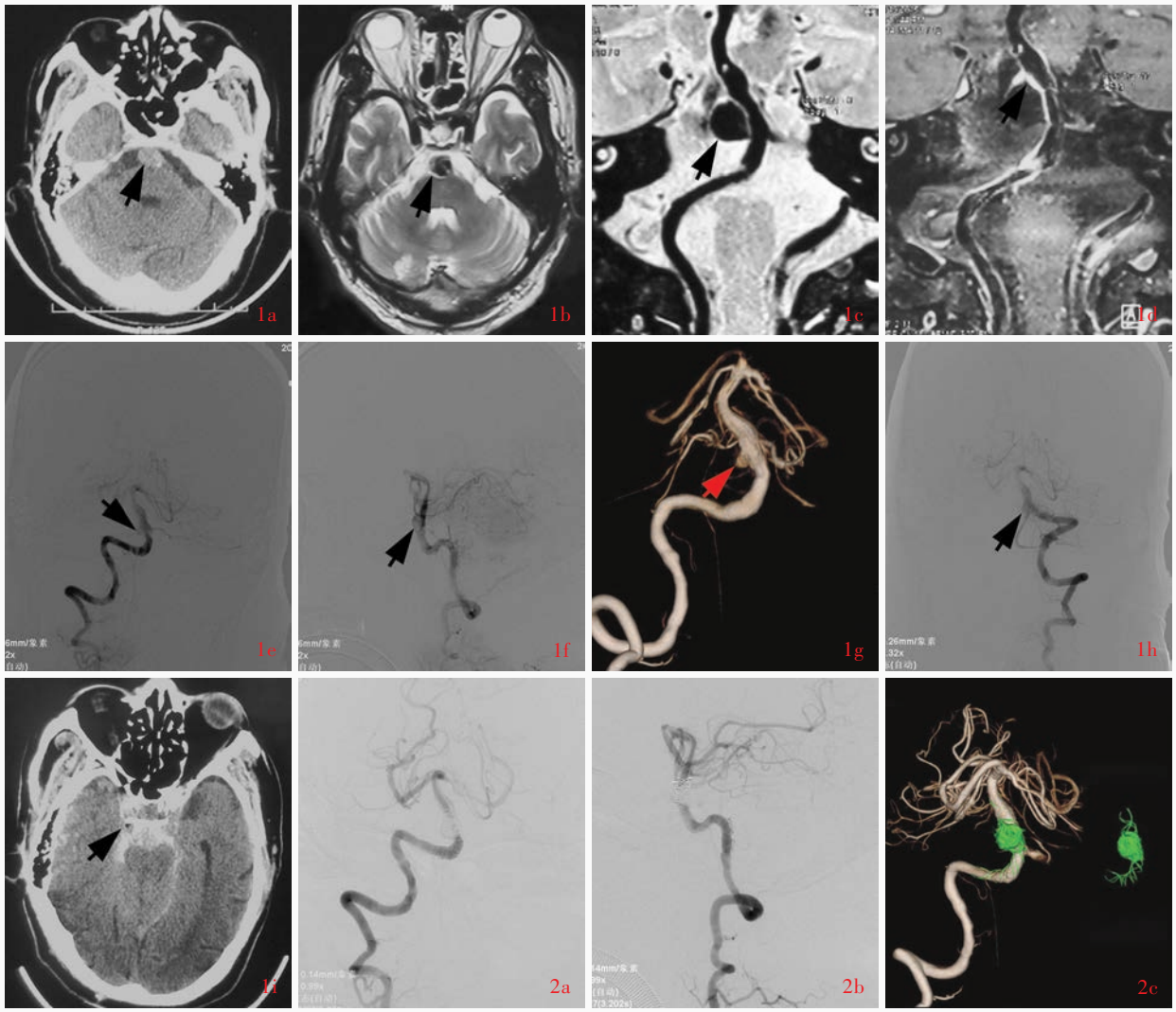

图1 术前头部影像学检查所见 1a 横断面CT显示桥前池高密度影，系增宽的基底动脉（箭头所示） 1b 横断面T_2WI显示，基底动脉近段夹层动脉瘤，可见“双腔征”改变（箭头所示） 1c 冠状位高分辨力MRBTI显示，双侧椎动脉汇合处远端和基底动脉近段夹层动脉瘤（箭头所示），椎-基底动脉走行偏向侧方 1d 冠状位高分辨力增强MRBTI显示，动脉瘤瘤壁明显强化（箭头所示），提示夹层动脉瘤结构不稳定 1e～1g 正位，侧位和3D-DSA显示，右侧椎-基底动脉走行迂曲、扩张，基底动脉近段夹层动脉瘤（箭头所示） 1h 正位DSA显示，夹层动脉瘤累及双侧椎动脉汇合处和右侧椎动脉末端（箭头所示） 1i 横断面CT显示，桥前池、环池、鞍上池和蛛网膜下隙出血，血肿主要位于动脉瘤体周围（箭头所示） **图2** 术后即刻脑血管造影所见 2a，2b 正位和侧位DSA显示，原动脉瘤未显影，载瘤动脉血流通畅 2c 3D-DSA显示，动脉瘤瘤腔填塞较致密，支架贴壁良好

Figure 1 Preoperative head imaging findings Axial CT showed high-density shadow of prepontile cistern, which was widened basilar artery (arrow indicates, Panel 1a). Axial T_2WI showed the dissecting aneurysm was located in the proximal part of basilar artery and there was "double lumen" change (arrow indicates, Panel 1b). Coronal high-resolution MRBTI showed the dissecting aneurysm was located in the distal end of the junction of bilateral vertebral arteries and proximal part of basilar artery (arrow indicates), and the vertebrobasilar artery extended to one side (Panel 1c). Coronal high-resolution enhanced MRBTI showed obvious enhancement of the wall of aneurysm (arrow indicates), indicating the instability of dissecting aneurysm (Panel 1d). Frontal, lateral and 3D view of DSA showed the circuity and expansion of right vertebrobasilar artery, and dissecting aneurysm located in the proximal part of basilar artery (arrows indicate, Panel 1e–1g). Frontal view of DSA showed the dissecting aneurysm involved the junction of bilateral vertebral arteries and the distal end of right vertebral artery (arrow indicates, Panel 1h). Axial CT showed prepontile cistern, ambient cistern, suprasellar cistern and subarachnoid hemorrhage and the hematoma was mainly located around the aneurysm (arrow indicates, Panel 1i). **Figure 2** Immediate postoperative DSA findings Frontal and lateral view of DSA showed the aneurysm was embolized completely and the blood flow of parent artery was well (Panel 2a, 2b). 3D-DSA showed the cavity of aneurysm was embolized densely and there was a good apposition of the stent (Panel 2c).

40.00 mm的Leo支架于第1枚Leo支架内套叠释放。术后即刻复查DSA显示载瘤动脉和左侧小脑后下动脉血流通畅，原动脉瘤未见显影（图2）。术后予低分子量肝素（速碧林）6×10^3 U/12 h皮下注射3 d，阿司匹林100 mg/d和氯吡格雷75 mg/d口服抗血小板治疗3个月。患者共住院18 d，出院时神志

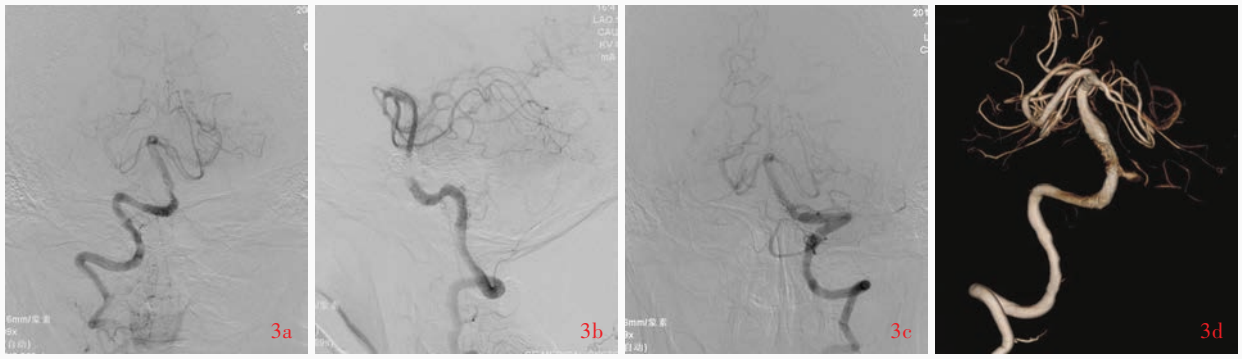

图3 术后3个月复查脑血管造影所见 3a,3b 正位和侧位DSA显示,原动脉瘤未显影,载瘤动脉血流通畅 3c 正位DSA显示,原动脉瘤未显影,左侧椎动脉血流通畅,左侧小脑后下动脉显影良好 3d 3D-DSA显示,动脉瘤瘤腔闭塞完全,支架贴壁良好

Figure 3 DSA findings 3 months after operation Frontal and lateral view of DSA showed the aneurysm was embolized completely and the blood flow of parent artery was well (Panel 3a, 3b). Frontal view of DSA indicated complete embolization of the aneurysm and the blood flow of left vertebral artery was well. The left posterior inferior cerebellar artery developed well (Panel 3c). 3D-DSA showed the aneurysm was embolized totally and there was a good apposition of the stent (Panel 3d).

清楚,精神可,遵嘱肢体活动良好。术后随访10个月,未出现脑出血、脑缺血和脑干压迫症状,体格检查一般状况良好,右侧听力下降基本恢复,右侧指鼻试验稳准,Romberg征阴性。术后3个月复查DSA提示动脉瘤栓塞完全,椎-基底动脉管腔修复良好(图3)。

讨 论

椎基底动脉延长扩张症(VBD)系指椎动脉和基底动脉异常增粗、迂曲并产生神经系统症状的动脉性疾病,属临床少见疾病,占所有颅内动脉延长扩张症(发病率0.06%~5.80%)的78%,长期随访的病死率高达39.7%[1-5]。椎基底动脉延长扩张症最早由Smoker等[6]于1986年提出,其发病机制目前尚不清楚。部分学者认为,全身性动脉高压、动脉内膜和内弹力层损害、动脉粥样硬化等多种因素共同参与该病的发生与进展[5-8],疾病进展至一定程度可以引起多种神经系统症状,如动脉搏动性压迫导致小脑功能障碍、脑积水、睡眠呼吸暂停综合征(SAHS)、短暂性或永久性运动障碍、三叉神经痛及其他脑干和脑神经压迫症状[9-12]。

通常情况下,椎基底动脉延长扩张症的自然史进展缓慢,然而一旦疾病进展,其发生出血性或缺血性卒中的风险明显升高,病死率也明显高于单纯压迫症状或无症状性椎基底动脉延长扩张症[3,13]。有研究显示,未干预的椎基底动脉延长扩张症患者较干预患者具有更高的病死风险,其影像学进展可作为预测预后不良的重要因素[13-16]。Passero等[15]纳入的156例椎基底动脉延长扩张症患者中脑卒中(缺血性卒中占84.8%、出血性卒中占15.2%)、颅内占位效应和无症状患者分别占42.3%、35.9%和21.8%,仅就出血性卒中而言,椎基底动脉延长扩张症患者发生颅内出血的概率为11/(1000人·年),其中发生蛛网膜下隙出血的概率为2.2/(1000人·年),所有患者进行平均为期11.70年的随访,其5、10和15年生存率分别为54.1%、39.5%和23.5%,年总体病死率约6%[13,15]。该例患者以突然发生的小脑后下动脉闭塞导致小脑梗死症状起病,伴脑干受压症状,且高分辨力MRBTI显示动脉瘤瘤壁明显强化,故应予干预治疗,以阻止动脉瘤夹层扩大和降低动脉瘤破裂风险。该例患者术前抗血小板治疗准备阶段发生动脉瘤破裂出血,考虑与其夹层动脉瘤结构不稳定有关,一方面,高分辨力MRBTI显示的动脉瘤瘤壁强化提示夹层动脉瘤结构不稳定,另一方面,右侧小脑后下动脉闭塞导致小脑梗死亦提示该夹层动脉瘤近期存在进展趋势。

椎基底动脉延长扩张症最常见的症状是后循环缺血,但目前预防脑卒中的措施在此类患者中并无明显作用,部分学者认为,抗血小板药有可能增加颅内出血风险[3,13-14]。结合本文患者,笔者的临床经验是,对于椎基底动脉延长扩张症伴基底动脉夹层动脉瘤患者应谨慎应用抗血小板药,其缺血事件的发生有可能由动脉夹层致穿支动脉闭塞所致,故此类患者可考虑积极手术治疗[16-18]。自1962年Mount和Taveras[19]提出闭塞椎动脉以治疗椎基底动脉延长扩张症理论以来,众多学者在显微外科手

术和血管内介入治疗方面进行探索和尝试，但仍有部分基底动脉近端闭塞的患者可出现病情进展，因此，他们认为，血管壁结构和血流动力学共同参与椎基底动脉延长扩张症的病程，单纯降低血流流速可能无法完全改善患者预后[19-20]。此外，有学者采用微血管减压术（MVD）、人工血管移植术、血管修剪吻合术、基底动脉管径缩小术等治疗椎基底动脉延长扩张症，但术后后循环缺血事件发生率并无明显减少[21-22]。

近年来，随着神经介入材料和技术的发展，有学者基于加强或修复管腔的理论采用支架辅助弹簧圈栓塞技术、多支架套叠释放技术、血流导向装置等手段治疗椎基底动脉延长扩张症，但目前仍处于探索阶段，其远期疗效尚不确切[18,23-26]。一方面，支架内衬于椎-基底动脉可部分改变其迂曲程度，通过血流动力学改变而减轻血流对血管壁的冲击，并改善分支血管的血供，降低缺血事件的发生[17,27]；另一方面，支架（联合弹簧圈）的使用可以通过增加动脉瘤瘤颈处的金属覆盖率以促进血管内皮修复，降低出血风险[23,27-28]。Leo支架作为一种自膨式闭环型编织支架，具有较好的径向支撑力和贴壁性，并具有较高的金属覆盖率，目前多应用于椎-基底动脉夹层动脉瘤[29]。该例患者应用2枚Leo支架套叠释放并联合弹簧圈，达到改善椎-基底动脉血流动力学的目的，促进血管自身修复，术后复查DSA显示动脉瘤瘤颈处修复良好，较好地体现出多支架套叠释放技术的优点。

值得注意的是，椎基底动脉延长扩张症的诊断与治疗目前尚无成熟方案，鉴于其高复发率、高病残率、高病死率和预后较差等疾病特点，首都医科大学宣武医院对此类疾病的诊断与治疗主要遵循以下原则：对于血管均匀扩张、无局限性动脉瘤样膨大的患者，应首选保守治疗（严格控制危险因素、抗血小板治疗、调脂治疗等）并定期复查影像学（CTA、MRA、DSA），病情进展者再考虑血管内介入治疗；对于血管在扩张基础上有局限性动脉瘤样膨大的患者，特别是病灶未累及基底动脉末段，应首选多支架套叠释放联合弹簧圈栓塞治疗；对于血管长节段延长扩张，特别是病灶已累及基底动脉顶端而多支架套叠释放难以完成的患者，建议控制危险因素，以药物治疗为主。而对于特定患者，手术或介入闭塞椎动脉联合枕动脉（颞前动脉）-大脑后动脉搭桥术不失为一种治疗选择。

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·小词典·

中英文对照名词词汇（三）

KPS评分 Karnofsky Performance Status（KPS）
破碎红纤维 ragged red fiber（RRF）
腔隙性梗死 lacunar infarct（LACI）
丘脑底核 subthalamic nucleus（STN）
三碘甲状腺原氨酸 tri-iodothyronine（T_3）
三维时间飞跃 three-dimensional time-of-flight（3D-TOF）
三维稳态构成干扰 three-dimensional constructive interference in steady state（3D-CISS）
少突胶质细胞转录因子2 oligodendrocytes transcription factor-2（Olig-2）
视觉模拟评分 Visual Analogue Scale（VAS）
视野 field of view（FOV）
数字减影血管造影术 digital subtraction angiography（DSA）
水通道蛋白4 aquaporin 4（AQP4）
苏格兰颅内血管畸形研究 Scottish Audit Intracranial Vascular Malformations（SAIVMs）
特发性震颤 essential tremor（ET）
梯度回波序列 gradient echo sequence（GRE）
同型半胱氨酸 homocysteine（Hcy）
突触素 synaptophysin（Syn）
微血管减压术 microvascular decompression（MVD）
稳态构成干扰 constructive interference in steady state（CISS）
系统性红斑狼疮 systemic lupus erythematosus（SLE）
细胞色素B cytochrome B（Cyt B）
纤维母细胞生长因子受体1 fibroblast growth factor receptor 1（FGFR1）
线粒体DNA聚合酶γ mitochondrial DNA polymerase γ（POLG）
线粒体脑肌病 mitochondrial encephalomyopathy（ME）
胸苷激酶 thymidine kinase（TK）
选择性5-羟色胺再摄取抑制剂 selective serotonin reuptake inhibitor（SSRI）
血氧水平依赖 blood oxygenation level-dependent（BOLD）
血氧水平依赖性功能磁共振成像 blood oxygenation level-dependent functional magnetic resonance imaging（BOLD-fMRI）
药物治疗未破裂脑动-静脉畸形优于手术试验 Medical Management with or without Interventional Therapy for Unruptured Brain Arteriovenous Malformations（ARUBA）trial
N-乙酰天冬氨酸 N-acetyl-aspartate（NAA）
异柠檬酸脱氢酶1 isocitrate dehydrogenase 1（IDH1）
荧光原位杂交 fluorescence in situ hybridization（FISH）
硬脑膜动-静脉瘘 dural arteriovenous fistula（DAVF）
丝裂原激活蛋白激酶 mitogen-activated protein kinase（MAPK）
Glasgow预后分级 Glasgow Outcome Scale（GOS）
运动诱发电位 motor-evoked potential（MEP）
真性红细胞增多症 polycythemia vera（PV）
中型多棘神经元 medium spiny neuron（MSN）
椎基底动脉延长扩张症 vertebrobasilar dolichoectasia（VBD）
Kearns-Sayre综合征 Kearns-Sayre syndrome（KSS）
左旋多巴诱导异动症 levodopa-induced dyskinesia（LID）