·论　著·

成人脊髓纵裂畸形导致的脊髓拴系综合征的治疗

谢京城　陈晓东　杨　军

【摘要】**目的** 总结成年脊髓纵裂畸形(SCM)导致的脊髓拴系综合征(TCS)的临床特点及手术治疗经验。**方法** 回顾性分析2007年9月至2019年3月手术治疗的21例成年SCM导致的TCS病人的临床资料。采用后路椎板切除、骨性或软骨性及纤维中隔分离切除、脊髓拴系松解、脊膜囊重建术。**结果** 21例中,男6例,女15例;年龄18~65岁,平均28.7岁;Ⅰ型7例,Ⅱ型14例。21例显微手术将中隔切除,脊髓拴系达Ⅰ级松解,无手术并发症。术后疼痛缓解或消失,双下肢无力及大小便功能障碍逐渐恢复。术后随访9个月至12年,平均5.8年;神经功能稳定及好转;随访期间未见拴系复发。**结论** 成人由SCM引发的TCS少见,以慢性腰腿痛为主要临床表现,手术在显微镜下行中隔分离切除、脊髓拴系松解及硬膜重建,疗效满意。

【关键词】脊髓拴系综合征;脊髓纵裂;成人;脊柱裂;显微手术

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Surgical treatment for adult patients with tethered cord syndrome due to split cord malformation

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【**Abstract**】**Objective** To summarize the clinical manifestation, imaging characteristic and experience of surgical treatment of tethered cord syndrome (TCS) due to split cord malformation (SCM) in adults. **Methods** The clincal data of 21 adult patients with TCS due to SCM who were surgically treated under microscope from September 2007 to March 2019 were retrospectively analyzed. **Results** There were 6 males and 15 females with an average age of 28.7 years (range, 18~65 years). Clinically, all the patients presented with chronic back and legs pain and lower extremities weakness or bowel and bladder dysfunction. Preoperative CT and MRI imaging showed spina bifida, characteristic bony, cartilaginous, or fibrous septum, and associated anomalies. According to CT and MRI features, 7 patients were categorized into Type Ⅰ and 14 into type Ⅱ. All the patients had low-lying conus medullaris and thickened filum terminale and 3 patients had meningeal cyst of filum terminal at sacral spine. 21 operations had been performed including resection of the median septum, de-tethering the spinal cord, and reconstruction of the dura sac. No operative complication occurred. The local pain was relieved, and the lower limbs weakness or bowel and bladder dysfunction were gradually relieved after the operation. The period of follow-up ranged from 9 months to 12 years, with an average of 5.8 years. The neurological function was improved or stable in all the patients. **Conclusions** TCS due to SCM in adults is rare, which is characterized with insidious onset and chronic progressive procedure. The surgical treatment including resection of the median septum, untethering of spinal cord, reconstruction of dura sac under microscope have good outcomes.

【**Key words**】Tethered cord syndrome; Split cord malformation; Spina bifida; Microsurgery; Adult patient

脊髓纵裂畸形(split cord malformation,SCM)是少见的神经管缺陷疾病,分为两型,其中Ⅰ型特征为脊髓分为两个半脊髓,每个半脊髓有自己的硬膜囊,两者之间为骨性/软骨性中隔;Ⅱ型特征为两个半脊髓在同一个硬膜囊内,两者之间为纤维性分隔[1,2]。由于中隔组织将脊髓分为两半,导致脊髓受牵拉,发生脊髓拴系综合(tethered cord syndrome,TCS),往往在儿童期发病。成人脊柱脊髓相对长度比较稳定,因此,成人SCM少见[3,4]。成人SCM导致的TCS起病隐匿,容易误诊。2007年9月至2019年3月收治成人SCM导致的TCS共21例,现报道如下。

1 资料与方法

1.1 一般资料　21例中,男6例,女15例;年龄18~65岁,平均28.7岁;病程8个月~10年。

1.2 临床表现　腰背痛17例,其中14例伴有放射性下肢痛;下肢无力11例,其中10例步态异常,8例足部或小腿肌萎缩导致高弓足或足内翻;大小便功能障碍9例;腰骶部异常皮肤红斑、凹陷或肿物5例。术前神经功能按Hoffman分级[5]:0级4例,1级7例,2

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级10例。

1.3 影像学检查 21例术前腰骶椎X线及CT检查均显示脊柱裂，位于L_{2-3}水平5例、L_{3-4}水平10例、L_{4-5}水平5例、S_{1-2}水平1例。术前MRI显示脊髓被中隔组织一分为二，脊髓圆锥位于L_2以下（图1）。近端见脊髓空洞12例，合并骶管终丝囊肿3例、肠源性囊肿2例、畸胎瘤1例。Ⅰ型7例，Ⅱ型14例。

1.4 手术方法 手术在静脉-吸入复合麻醉下进行。取俯卧位，术中采用神经电生理监测。逐层切开皮肤、皮下组织至筋膜层，切除沿途异常皮肤及皮下组织。行骨膜下分离椎旁肌显露病变部位异常棘突及椎板，行畸形棘突椎板切除术。①对Ⅰ型SCM，显露骨嵴插入双侧硬膜囊间位置，围绕双脊膜囊间骨嵴自背侧向腹侧剥离切除，注意腹侧异常血管的止血，自头、尾端正常硬膜中线剪开，然后于双侧硬膜囊背侧纵行剪开硬膜，向两侧悬吊，将双侧硬膜囊内侧残留硬膜与骨质切除，将残留硬膜平铺于椎管内腹侧。②对Ⅱ型SCM，沿正常硬膜囊头、尾端向异常处剪开，悬吊，显露硬膜下腔，自背侧向腹侧，分离切除纤维中隔。③利用同一个切口或另外设计切口，行终丝探查切断术。④对合并的先天性肿瘤，同期切除。⑤松解沿途蛛网膜粘连，将脊髓还纳于同一硬膜囊，见脊髓及马尾神经松弛于椎管内腹侧，搏动好，顺行探查脊髓、马尾神经结构完好，脑脊液流动通畅。⑥将蛛网膜自硬膜粘连处分离下，覆盖在脊髓及神经表面，用生理盐水冲洗清亮，取5-0无损伤线连续缝合硬膜层。⑦术后俯卧7~9 d，切口区域沙袋压迫。

1.5 术后临床效果评价 ①以Kirollos分级[6]评定手术拴系松解程度。②以视觉模拟量表（visual analogue scale，VAS）评分评价疼痛改变。③采用关键肌肉力量0~5级评价下肢运动功能。④用日本骨科学会协会（Japanese Orthopaedic Association，JOA）评分评估括约肌功能[7]。⑤以Hoffman神经功能分级[5]评价神经功能。⑥末次随访时，复查腰椎MRI及动力位X线，结合神经功能评估再栓系及脊柱生物力学。

2 结 果

2.1 手术结果 手术用时1.9~3.5 h，平均2.5 h；术中出血量100~350 ml，平均200 ml。对7例Ⅰ型SCM，手术将骨性软骨性中隔全切除（图2）；对14例Ⅱ型SCM，将纤维中隔组织全切除。终丝切断1.5~2 cm，TCS得到Kirollos分级Ⅰ级松解，合并的终丝囊肿3例及先天性肿瘤3例一期切除。无手术并发症。

2.2 近期临床疗效 ①术前疼痛17例，术后疼痛缓解或消失，术后VAS评分[(4.09±1.20)分]较术前[(6.87±2.80)分]显著降低（$P<0.05$）。②11例运动障碍术后肌力提高1~2级。③9例括约肌功能障碍，术后JOA评分[(2.10±1.19)分]与术前[(2.05±1.02)分]无明显变化（$P>0.05$）。

2.3 随访情况 术后随访9个月~12年，平均5.8年。脊髓功能状态按Hoffman分级：0级7例，1级9例，2级5例；无恶化病例。末次随访MRI检查证实脊髓及马尾神经松弛，未发现再拴系征象。

3 讨 论

TCS是指由于先天及后天因素导致无弹性结构系住脊髓末端，限制其垂直移动所致的神经障碍症候群，于1976年由Hoffman等[8]提出；1981年，Yamada等[9]将TCS概念加以扩展。TCS按病因可以分为：终丝牵张、脂肪脊髓脊膜膨出、SCM、脊髓脊膜膨出、皮窦道等。

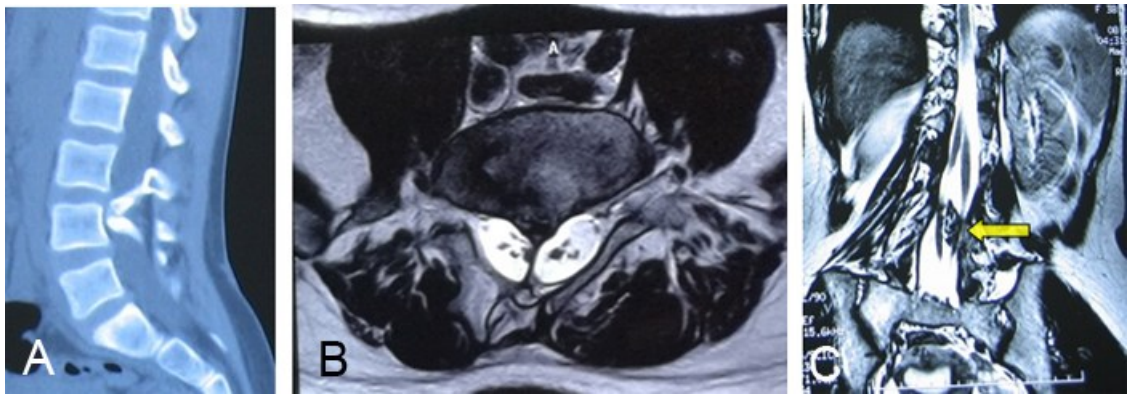

图1 典型L3~4节段Ⅰ型脊髓纵裂畸形影像学表现

A. 术前腰椎CT矢状位，显示骨性中隔走行及伴随的脊柱裂；B. 术前MRI T_2加权像轴位显示骨性中隔、双硬膜囊，双侧的半脊髓及神经根；C. 术前MRI T_2加权像冠状位显示脊髓分叉、骨性分隔（↑示）

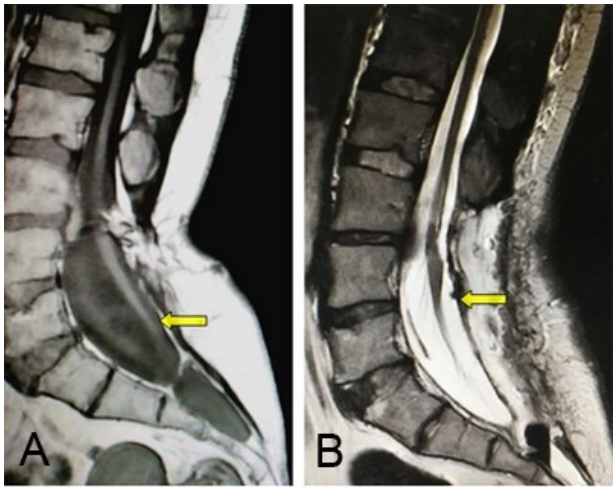

图2 L3节段Ⅰ型脊髓纵裂畸形合并骶管囊肿手术前后MRI
A. 术前MRI T_1加权像矢状位显示L3水平中隔、脊髓圆锥低位于S1水平(↑示),合并骶管内脊膜囊肿;B. 术后MRI复查T_2加权像,显示骨性中隔消失、脊髓松弛于椎管内,脊髓圆锥上升至L_{4-5}水平(↑示)、骶管囊肿消失

SCM是脊髓在某个节段被纵向分为两个半脊髓,目前普遍接受的是Pang[1,2]的SCM分型及胚胎学发病机制学说。其胚胎学机制是源于神经外胚层形成期,外胚层与内胚层黏附分离,卵黄囊与羊膜间形成一被中胚层所包围的副神经肠管,横贯脊索并导致脊索背侧神经板一分为二,形成两个半神经板,管化为两个半脊髓结构,随后的中胚层间充质基发育,形成后期两个半脊髓位于双脊膜囊或单一脊膜囊内,中隔以骨性软骨性分隔或纤维性分隔,随着胎儿期脊柱脊髓相对位置变化,脊髓受牵拉,导致缺血,出现TCS症候群。

成人SCM脊柱与脊髓处于动态平衡状态,一旦此平衡被打破,即产生TCS相应临床表现,因此成人SCM导致的TCS,起病隐匿、进展缓慢,容易漏诊或误诊。本文总结发现,成人SCM导致的TCS,常见症状包括慢性腰背部疼痛、下肢无力及感觉障碍,严重者发生大小便功能障碍,体检可发现皮肤红斑、足部畸形、腿部肌萎缩、步态异常、脊柱侧弯等,MRI可以确诊SCM导致的TCS,结合腰椎CT可以进行分型,明确脊髓纵裂三维解剖结构,并可以显示其头端的脊髓空洞及尾端脊髓圆锥及内终丝,MRI还可以发现合并的脊髓空洞、确定圆锥及内终丝牵张位置,是否合并先天性肿瘤等[10,11]。

对儿童SCM导致的TCS,手术可以去除病灶,防止神经功能进一步损伤,普遍持积极态度[12,13]。而对于成人SCM导致的TCS,一旦出现神经功能障碍及症状进展,应尽早手术,实践证实手术对疼痛缓解及下肢无力改善明显[3,4,14,15]。

SCM导致的TCS手术关键在于术前充分了解其解剖学三维关系,追溯其相应的胚胎发生学基础,合理制定手术方案。术中神经电生理监测非常关键[16,17]。手术原则是逐层解剖,通过皮下-筋膜层、脊柱裂层、硬膜外、硬膜下显微操作,达到充分松解,解除中隔组织压迫及牵拉,然后,逐层恢复正常解剖结构。手术要点:①设计切口时,围绕中隔部位,兼顾异常皮肤及皮下组织切除范围。②切开皮肤与皮下组织后达筋膜,平筋膜层切除异常皮肤及皮下组织瘢痕。③显露脊柱裂时,要遵循骨膜下剥离原则,显露脊柱裂后,逐步咬除或磨除异常骨质以显露脊柱异常部位。④对Ⅰ型SCM,沿背侧向腹侧,切开骨性分隔物两侧硬脊膜,逐渐深入,结合磨除骨性分隔,直到椎管腹侧,注意中隔伴随的血管及纤维组织的分离切除。对Ⅱ型SCM,一般可在脊髓纵裂下端相对应节段发现有棘突裂或椎板裂,在硬膜表面找到纤维束带。沿头、尾端正常硬膜中线处剪开显露纤维中隔,向双侧悬吊硬膜,分离切除纤维中隔。⑤内终丝分离及切断是松解TCS必要的步骤[18],利用同一切口或附加切口,行内终丝分离切断术,切断长度1.5 cm以上,对合并的骶管囊肿及先天性肿瘤同期切除。至于脊髓空洞,与牵张因果关系,解除牵张后,空洞缩小甚至消失[19]。⑥将两个半脊髓还纳于同一个硬膜囊内,松解沿途蛛网膜粘连,直到脊髓及马尾神经松弛于椎管内腹侧,搏动好,此时顺行探查脊髓、马尾神经结构完好,脑脊液流动通畅。⑦重建完整的硬膜囊,将蛛网膜自硬膜粘连处分离下,覆盖在脊髓及神经表面,用生理盐水冲洗清亮,取5-0无损伤线连续缝合硬膜层。所谓重构硬膜囊,关键还是要将蛛网膜自硬膜内壁剥离下,覆盖于脊髓及马尾神经表面,依靠蛛网膜的内皮细胞属性,形成蛛网膜下腔,然后是成型缝合硬膜层。⑧术后俯卧7~9 d,以保证松解后的脊髓及马尾神经始终松弛于椎管内腹侧,避免脑脊液漏发生。

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