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· 专题研究 ·

肝内胆管结石合并肝叶萎缩-增生综合征行保留尾状叶次全肝切除术1例并文献复习

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摘要

背景与目的: 肝内胆管结石长期反复炎症可导致肝叶萎缩-增生综合征, 部分患者可表现为双侧肝叶明显萎缩而尾状叶代偿性增大, 此类患者手术切除范围及剩余肝体积的精准评估具有较大挑战。本文报告1例肝内胆管结石合并肝叶萎缩-增生综合征患者行保留尾状叶次全肝切除术的诊治过程, 并结合相关文献探讨该术式的适应证、关键技术及临床应用价值。

方法: 回顾性分析2025年3月中国人民解放军总医院第五医学中心收治的1例肝内胆管结石合并肝叶萎缩-增生综合征患者的临床资料。术前采用肝脏三维可视化技术评估肝脏形态、尾状叶体积及其血管和胆道的解剖关系, 结合肝功能储备进行手术规划; 术中联合胆道镜及超声引导, 行保留尾状叶次全肝切除术、胆总管探查取石术及T管引流术。结合国内外相关病例进行文献复习。

结果: 患者为66岁女性, 影像学检查显示左、右半肝明显萎缩, 尾状叶代偿性增大, 肝内胆管多发结石伴扩张, 尾状叶血供、静脉回流及胆道引流通畅。三维重建显示尾状叶体积948 mL, 术后剩余肝体积约占标准肝体积的84%。患者顺利完成保留尾状叶次全肝切除术联合胆总管探查取石及T管引流术, 手术时间420 min, 术中出血量约800 mL, 未输血。术后肝功能恢复良好, 未发生严重并发症, 术后第12天出院。术后病理提示肝内胆管癌, 大胆管型, 切缘阴性。术后2个月复查未见明确结石残留及肿瘤复发; 术后4个月胆道镜检查未见结石残留, 肝功能正常, 拔除T管; 随访8个月, 患者一般状况良好, 影像学检查未见明确结石及肿瘤复发。

结论: 对于肝内胆管结石合并肝叶萎缩-增生综合征的严格筛选患者, 在充分评估肝功能储备、剩余肝体积以及尾状叶血供、静脉回流和胆道引流条件的基础上, 联合三维可视化、胆道镜及术中超声进行精准术前规划和术中操作, 保留尾状叶次全肝切除术具有可行性。但该术式技术要求高, 应严格掌握适应证, 并充分警惕肝内胆管癌等远期并发症。

关键词

胆石症; 胆管, 肝内; 肝叶萎缩-增生综合征; 肝切除术

中图分类号: R657.4

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Caudate lobe-sparing subtotal hepatectomy for hepatolithiasis complicated by liver lobe atrophy-hypertrophy syndrome: a case report and literature review

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Abstract

Background and Aims: Long-standing hepatolithiasis with recurrent biliary inflammation may lead to liver lobe atrophy-hypertrophy syndrome. In some patients, marked atrophy of both hepatic lobes is accompanied by compensatory enlargement of the caudate lobe, making the extent of hepatectomy and accurate assessment of the future liver remnant challenging. We report a case of hepatolithiasis complicated by liver lobe atrophy-hypertrophy syndrome treated with caudate lobe-sparing subtotal hepatectomy and review the relevant literature to evaluate the indications, key technical considerations, and clinical applicability of this procedure.

Methods: The clinical data of a 66-year-old patient with hepatolithiasis complicated by liver lobe atrophy-hypertrophy syndrome who was admitted to the Fifth Medical Center of Chinese PLA General Hospital in March 2025 were retrospectively analyzed. Three-dimensional visualization was used preoperatively to assess hepatic morphology, caudate lobe volume, and the anatomical relationships of its vascular and biliary structures. Surgical planning was performed based on these findings and hepatic functional reserve. Caudate lobe-sparing subtotal hepatectomy combined with common bile duct exploration, stone extraction, and T-tube drainage was subsequently performed with the assistance of choledochoscopy and intraoperative ultrasonography. Relevant cases reported in the literature were also reviewed.

Results: Imaging revealed marked atrophy of the left and right hepatic lobes, compensatory enlargement of the caudate lobe, and multiple intrahepatic biliary stones with intrahepatic bile duct dilatation. The caudate lobe had preserved blood supply, venous drainage, and biliary drainage. Three-dimensional reconstruction showed a caudate lobe volume of 948 mL, corresponding to approximately 84% of the standard liver volume after resection. Caudate lobe-sparing subtotal hepatectomy combined with common bile duct exploration, stone extraction, and T-tube drainage was successfully performed. The operative time was 420 min, with estimated blood loss of approximately 800 mL and no blood transfusion. Liver function recovered well postoperatively, with no severe complications, and the patient was discharged on postoperative day 12. Histopathological examination unexpectedly revealed intrahepatic cholangiocarcinoma, large-duct type, with negative resection margins. At 2 months postoperatively, no definite residual stones or tumor recurrence was detected. Choledochoscopy at 4 months showed no residual biliary stones, and the T-tube was removed. During the 8-month follow-up, the patient remained in good general condition, with no definite evidence of recurrent stones or tumor on imaging.

Conclusion: In carefully selected patients with hepatolithiasis complicated by liver lobe atrophy-hypertrophy syndrome, caudate lobe-sparing subtotal hepatectomy is feasible after comprehensive assessment of hepatic functional reserve, future liver remnant volume, and the vascular, venous, and biliary drainage of the caudate lobe. Three-dimensional visualization, choledochoscopy, and

intraoperative ultrasonography may facilitate precise preoperative planning and intraoperative identification and preservation of critical structures. However, this technically demanding procedure requires strict patient selection, and long-term complications such as intrahepatic cholangiocarcinoma should be carefully considered.

Key words Cholelithiasis; Bile Ducts, Intrahepatic; Liver Lobe Atrophy-Hypertrophy Syndrome; Hepatectomy
CLC number: R657.4

肝内胆管结石是指左右肝管汇合部以上的肝内各分支胆管内发生的结石，具有发病原因复杂、结石部位多样、术中难以除尽、术后易复发、造成并发症严重等特点，长期的结石造成的胆管炎症可致肝叶萎缩-增生综合征^[1]，其中尤为特殊的是广泛的肝内胆管结石致左右半肝萎缩，仅尾状叶代偿性增大，这样的复杂情况为实际的临床治疗带来了一定的挑战。本文报道 1 例肝内胆管结石合并肝叶萎缩-增生综合征患者在三维可视化及术中超声等技术辅助下行保留尾状叶次全肝切除术的诊治经过，并结合相关文献进行复习。

1 病例资料

1.1 一般资料

患者 女，66 岁。主因“间断上腹部疼痛不适 4 月余”于 2025 年 3 月收入中国人民解放军总医院第五医学中心肝病外科。患者入院前 4 个月余间断出现上腹部疼痛不适，较剧烈，伴恶心、呕吐，无寒战、发热。既往无高血压、糖尿病及心脑血管疾病史，无胆道及其他手术史。入院查体：皮肤、巩膜无黄染，腹软，右上腹压痛（+），无反跳痛及肌紧张，墨菲征（-），移动性浊音（-），

肝区叩击痛（-），肠鸣音 3 次/min。血常规：白细胞（WBC） $5.89 \times 10^9/L$ ，血红蛋白 128.00 g/L，血小板 $121.00 \times 10^9/L$ ；生化全项：白蛋白 38 g/L，直接胆红素（DBIL） $7.1 \mu\text{mol/L}$ ，总胆红素（TBIL） $19.1 \mu\text{mol/L}$ ，丙氨酸氨基转移酶（ALT）28 U/L，天冬氨酸氨基转移酶（AST）41 U/L，碱性磷酸酶 175 U/L， γ -谷氨酰基转移酶 96 U/L，钠 139 mmol/L，钾 4.2 mmol/L，二氧化碳结合力 23.0 mmol/L，血清糖类抗原 19-9（CA19-9）313.00 U/mL，凝血酶原活动度（PTA）98.5%。肝脏 MRI 及磁共振胰胆管成像（MRCP）提示，肝脏形态不规整，左、右肝萎缩、尾状叶增大，肝内胆管多发结石伴肝内胆管扩张、局部积气，胆总管下段结石，尾状叶门脉血流通畅（图 1）。肝脏三维重建可见肝尾状叶增生明显、左右半肝萎缩，尾状叶的动脉、门脉供血、静脉回流及胆道通畅（图 2）。评估患者肝功能 Child-Pugh 分级 A 级（评分 5 分）。肝储备功能：吲哚菁绿 15 min 滞留率（indocyanine green retention rate at 15 minutes，ICG-R15）3.5%。行三维重建规划，患者体表面积为 1.59 m^2 ，标准肝体积约 1 125 mL，全肝体积 1 226 mL，尾状叶肝体积 948 mL，剩余肝体积占标准肝体积的 84%，占全肝体积的 77%。

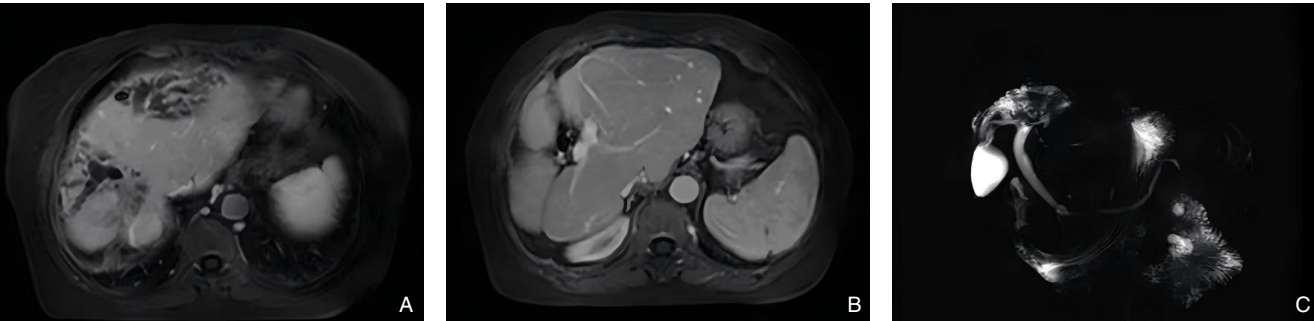


图 1 影像学检查可见肝内胆管多发结石伴扩张，左、右肝叶萎缩，尾状叶代偿性增大 A-B：肝脏 MRI；C：MRCP
Figure 1 Imaging findings showing multiple intrahepatic biliary stones and bile duct dilatation, atrophy of the left and right hepatic lobes, and compensatory enlargement of the caudate lobe A-B: Liver MRI; C: MRCP

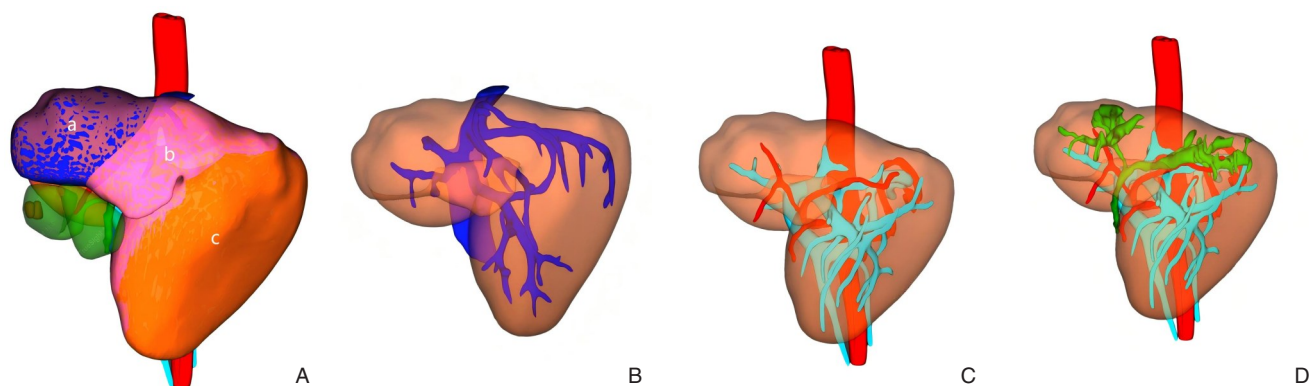


图2 肝脏三维重建图像 A: 显示萎缩的肝右叶 (a)、萎缩的肝左叶 (b)、增生的肝尾状叶 (c); B: 显示肝静脉走行; C: 显示门静脉及肝动脉走行; D: 显示肝内扩张的胆道走行

Figure 2 Three-dimensional reconstruction of the liver A: Atrophy of the right hepatic lobe (a), atrophy of the left hepatic lobe (b), and hypertrophy of the caudate lobe (c); B: Course of the hepatic veins; C: Courses of the portal vein and hepatic artery; D: Dilated intrahepatic bile ducts

经术前评估,患者术前化验提示CA19-9升高,结合病史不排除长期胆管结石导致恶变可能,但术前影像学检查未提示癌变,其余相关化验未见明确异常。术前三维重建评估患者左、右半肝均已萎缩,尾状叶代偿性增大,结合肝功能Child-Pugh评分、ICG检查及尾状叶肝体积占标准肝体积百分比,评估该患者可耐受保留尾状叶次全肝切除术,无绝对手术禁忌证,遂决定行手术治疗。

1.2 手术方法

右上腹反“L”形切口,长约25 cm。逐层进腹,探查见肝左右叶质地硬,明显萎缩,肝尾状叶质地软,胆囊大小正常,囊壁水肿,内可触及质硬结石,肝外胆管明显扩张,胃、肠、胰等其他腹部脏器未探及明显异常。分离胆囊三角,确认胆囊管与胆总管、肝总管关系后,夹闭并剪断胆囊管及胆囊动脉,经胆囊床顺行剥离胆囊。暴露胆总管,于胆总管中段纵行切开约1 cm,置入胆道镜,胆总管远端可见黑色结石1枚,用取石网篮取出,向上观察见肝内胆管黏膜粗糙,伴絮状物,并可见左右肝内多发结石,同时胆道镜探查尾状叶的胆管,可见左尾状叶及右尾状叶胆管直接开口于胆总管且未见明确结石。超声刀分离镰状韧带至第二肝门,解剖第一肝门,放置肝门阻断带备用。离断肝肾、肝结肠、左右三角韧带及左右冠状韧带,充分游离肝脏左右叶,鞘外法分

离出左侧肝蒂,注意保护左尾状叶肝蒂(患者因肝左叶萎缩,将肝左叶掀起后可见其与左尾状叶存在天然界限,同时利用术中超声明确左尾状叶的肝蒂走行,避免损伤),离断后用3-0血管线缝合左侧肝蒂断端。将肝左叶掀起后游离至第二肝门,确认肝左静脉根部与Spiegel叶之间的Arantius管(术中可见其自左侧肝蒂延伸至肝左静脉根部),夹闭后离断,分离出肝左静脉,用一次性切割闭合器离断。沿萎缩肝左叶、Arantius管与质软肝尾状叶之间潜在平面,使用超声刀沿肝尾状叶离断肝实质,分离至肝中静脉,夹闭后离断,继续沿尾状突分离肝实质至右侧第一肝门,离断右侧肝蒂(同理,利用萎缩肝右叶与尾状叶潜在平面,结合术中超声,避免损伤右尾状叶肝蒂),用3-0血管线缝合断端,沿尾状叶腔静脉旁部由足侧向头侧离断肝实质至第二肝门,用一次性切割闭合器离断肝右静脉,完整切除肝左右叶,观察尾状叶血供及回流情况无异常,术中Pringle法阻断肝门,共阻断6次,单次最长阻断时间15 min,间隔5 min阻断1次,累计阻断时间84 min。肝断面以氩气刀喷射电凝及缝扎止血,创面无渗血及胆汁漏等。于胆管前壁放置22号T管,用3-0可吸收线间断缝合并固定,确认无胆汁外漏后,创面放置止血纱布辅助止血。于肝断面放置硅胶引流管,T管及引流管经右侧穿出腹壁,逐层关腹,无菌敷料覆盖切口。术中相关图片见图3。

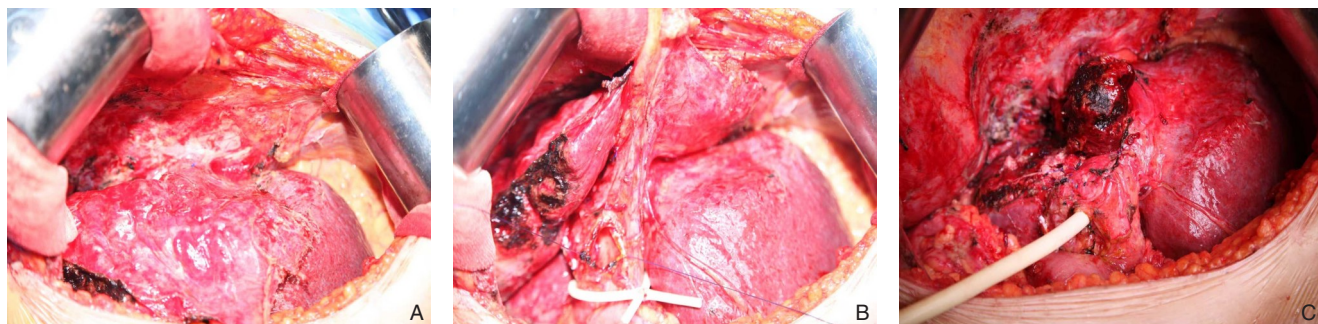


图3 术中相关图片 A: 萎缩的肝叶; B: 抬起萎缩的左、右肝叶后可见代偿性增大的尾状叶; C: 切除术后仅保留的尾状叶

Figure 3 Intraoperative findings A: Atrophic hepatic lobes; B: compensatory enlargement of the caudate lobe after elevation of the atrophic left and right hepatic lobes; C: the caudate lobe remaining after subtotal hepatectomy

1.3 手术与术后情况

手术时间 420 min, 术中出血量约 800 mL, 未输注红细胞及血浆。患者术后予以保肝、抗感染、补液及营养支持等治疗, 并密切监测肝功能及相关实验室指标。术后患者肝功能总体恢复良

好, 主要实验室指标动态变化见图 4。根据 Clavien-Dindo 并发症分级标准, 患者术后出现 II 级并发症, 未发生肝衰竭、术后大出血及胆汁漏等严重并发症。术后第 9 天拔除腹腔引流管, 第 12 天病情稳定后出院。

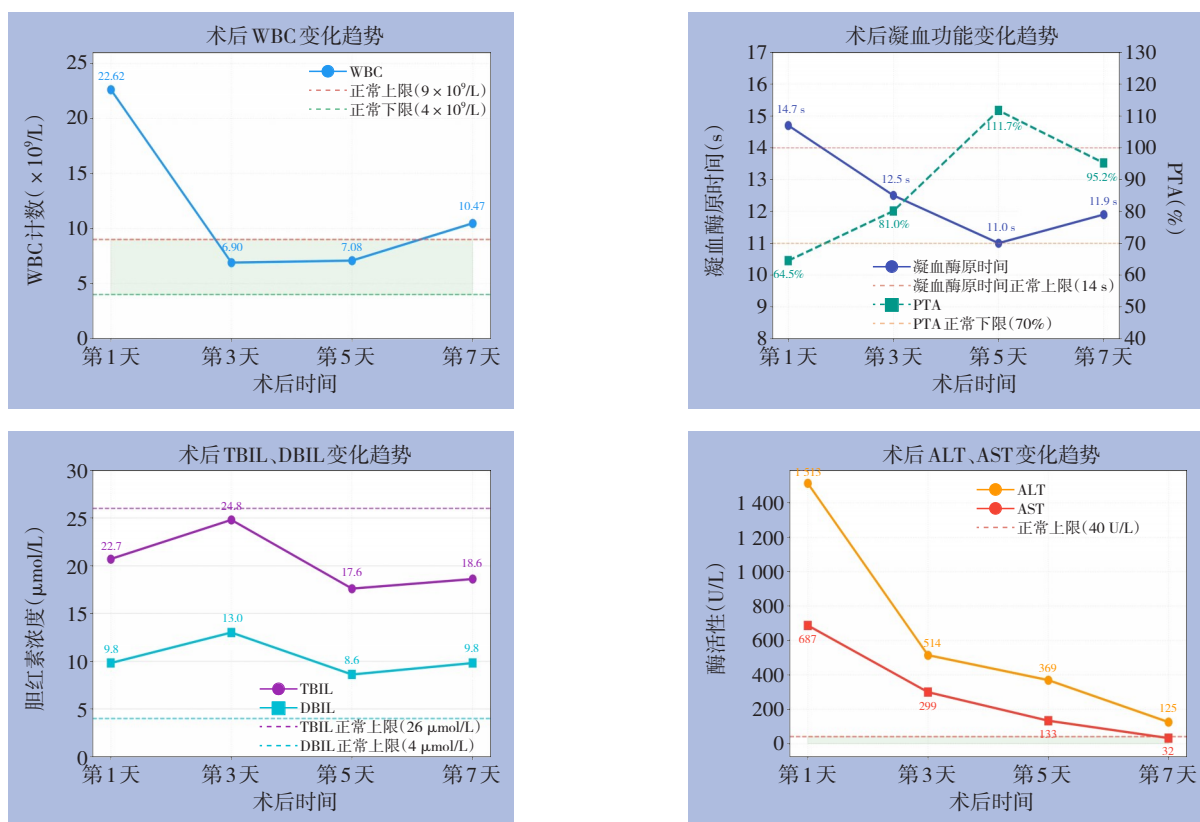


图4 术后主要化验指标变化

Figure 4 Changes in major laboratory parameters after surgery

术后病理检查显示, 切除的左右半肝组织中可见胆管癌, 大胆管型, 肿瘤大小约 3.5 cm × 2.0 cm × 2.0 cm; 脉管内未见明确癌栓, 神经可见肿瘤累及, 切缘未见肿瘤, 病理分期为 pT1aNXMO (AJCC 第 8 版)。背景肝组织为肝内胆管结石 (图 5)。

肿瘤组织免疫组织化学结果: CK19 (+)、GPC-3 (-)、Ki-67 (+30%)、EMA (+)、CEA (+)、CDX-2 (-)、P53 (-)、CK7 (+)、CK20 (-)、Syn (-)、EGFR (-)、CD10 (-)、MUC-1 (+)、MUC-2 (-)、MUC-5AC (+)、MUC-6 (散+)、

HER2 (-)、MLH1 (+)、MSH2 (+)、MSH6 (+)、PMS2 (+)。

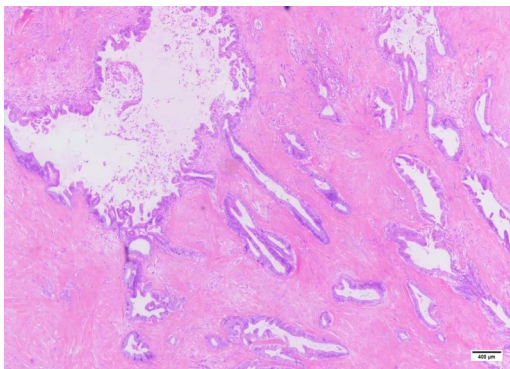


图5 术后病理切片 (HE×40)
Figure 5 Histopathological findings of the resected specimen (HE×40)

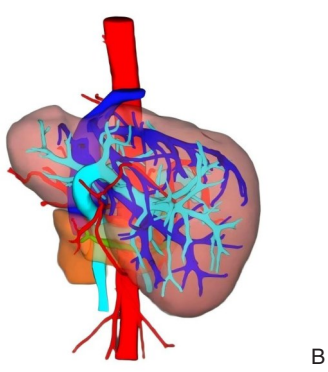
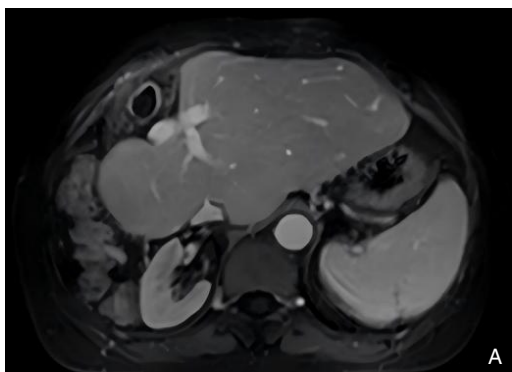


图6 术后2个月复查显示肝脏尾状叶门脉血流通畅,肝内未见明确结石及肿瘤复发迹象 A: 肝脏MRI; B: 三维重建
Figure 6 Imaging findings at 2 months after surgery showing the portal venous flow of the caudate lobe was patent, with no definite residual intrahepatic biliary stones or tumor recurrence A: Liver MRI; B: three-dimensional reconstruction

2 讨论与文献复习

长期的肝内胆管结石造成的胆管炎症可致肝叶萎缩-增生综合征、肝硬化甚至胆管癌变^[2-7]。黄志强院士曾提出“取尽结石、去除病灶、解除梗阻、通畅引流”的16字方针;在此基础上,中华医学会进一步增加“防止复发”,构成20字治疗方针^[8]。手术治疗是肝内胆管结石最有效的治疗方式,保留尾状叶次全肝切除术几乎是肝内胆管结石病的特有术式。尾状叶具有多重血供、多流出道的特点,其具有强大的再生能力,可以看作一个后备肝脏^[9],尾状叶可分为左侧的Spiegel叶、右侧的尾状突和中间的下腔静脉旁部^[10],进出尾状叶的管道相对独立,其门静脉、动脉通常自第一肝门发出,静脉可直接回流入下腔静脉,而胆管可直接汇入左右肝管甚至肝总管^[11-12],故而左、右半肝广泛肝内胆管结石致肝脏萎缩时,尾状叶

考虑患者术前CA19-9升高,结合术后病理结果,提示长期肝内胆管结石及慢性胆管炎症可能与胆管癌发生相关。患者切缘阴性,达到R₀切除。术后2个月复查,患者一般状况良好,肝脏MRI及三维重建显示尾状叶门静脉血流通畅,未见明确肝内胆管结石残留及肿瘤复发征象(图6),相关实验室指标未见明显异常。随后予以信迪利单抗、仑伐替尼联合吉西他滨+奥沙利铂方案进行抗肿瘤治疗。术后4个月行胆道镜检查,未见胆道内结石残留,肝功能正常,遂拔除T管。术后随访8个月,患者无特殊不适,复查相关影像学检查未见明确结石残留或肿瘤复发。

可逐渐增生代偿肝脏功能^[13],这为该术式的成功实施提供了理论基础,该术式目前国内外报道相对较少,我国肝胆外科专家Dong等^[14]首次在国际上报道了采用该术式成功治疗多例肝内胆管结石合并肝叶萎缩-增生综合征的患者,该研究纳入了自2003年3月—2009年12月接受仅保留尾状叶次全肝切除术的患者共12例,所有患者均顺利完成手术并康复出院,平均随访51个月,其中有1例患者因尾状叶胆管反复出现结石,在接受手术后的第17个月因急性化脓性胆管炎离世,其余11例患者均未出现肝内胆管结石复发,生存良好。之后间断有零散的个案报道证实该术式的可行性及有效性(表1)^[15-19]。这些报道中行该术式的患者均为肝内胆管结石合并左右肝萎缩、尾状叶代偿性增大,其中文浩权等^[17]报道采用腹腔镜技术行手术治疗。张浩鹏等^[15]报道的病例因尾状叶胆管汇入肝总管,且患者患有先天性胆管扩张症,故

切除肝外扩张胆管，行尾状叶胆管+空肠吻合术。蒋燦等^[16]在 2020 年报告的病例中，患者左尾状叶胆管汇入左肝管，遂行左尾状叶胆管+空肠吻合术。柴乃俊等^[18]报道的病例中患者术后出现胆管-支气管瘘，这可能与术后持续胆汁漏、炎症感染有关。上述病例报道中均未采用术中超声明确尾

状叶流通管道走行，除蒋燦等^[16]报道的病例外，均未利用三维可视化技术建立术前肝脏的三维重建模型，故术者仅依靠 CT、MRI/MRCP 等影像行术前规划，这不可避免地增加了术中损伤重要管道结构的风险。

表 1 保留尾状叶次全肝切除术治疗肝内胆管结石相关疾病的既往病例报道

Table 1 Summary of previously reported cases treated with caudate lobe-sparing subtotal hepatectomy for hepatolithiasis-related diseases

作者	时间	病例数	适应证	术式	随访结果
张浩鹏, 等 ^[15]	2024 年	1	先天性胆管扩张合并肝内胆管结石	保留尾状叶次全肝切除联合肝外扩张胆管切除,胆肠吻合术(尾状叶胆管+空肠)	未随访
蒋燦, 等 ^[16]	2020 年	1	肝内胆管结石合并肝叶萎缩-增生综合征	保留尾状叶次全肝切除术+(左尾状叶胆管+空肠)胆肠吻合术+胆总管探查取石术+T管引流术	未随访
文浩权, 等 ^[17]	2023 年	1	肝内胆管结石合并肝叶萎缩-增生综合征	腹腔镜下单独保留肥大肝尾状叶的次全肝切除+胆道探查+T管引流术	随访 5 个月, 患者未见结石残留及复发
柴乃俊, 等 ^[18]	2018 年	1	肝内胆管结石合并肝叶萎缩-增生综合征	保留尾状叶次全肝切除术	随访 1 年, 其间患者出现胆管-支气管瘘, 经治疗后病情稳定
彭创, 等 ^[19]	2016 年	1	肝内胆管结石合并肝叶萎缩-增生综合征	保留尾状叶次全肝切除术+胆总管探查取石术+T管引流术	随访 1 年, 患者未见结石残留及复发

该术式主要难点在于如何保留足够的功能性肝体积，避免术后肝衰竭的发生^[20]，准确地评估测算剩余肝体积是否足够成为减少肝衰竭发生的关键^[21]。此外，整个手术过程中如何精准识别并保护进出尾状叶的血管及胆道结构也是预防术后肝衰竭的重要一环。三维可视化技术不仅能够帮助外科医生精准地对剩余肝体积进行测算，同时也能够帮助外科医生清晰地了解肝动脉、门静脉及胆道的立体关系。特别是明确进出尾状叶的通道，从而在术前模拟、规划最佳切除方式^[22-24]，本文报道的病例中，术前利用三维可视化技术重建了肝脏立体三维模型，较为精准地计算出尾状叶的体积，经评估切除病肝后，剩余肝体积占标准肝体积 84%，足以耐受次全肝切除。同时，利用重建后的肝脏三维模型，术者能够清楚且直观地看到尾状叶的血供及静脉回流，能够帮助术者在术前规划手术方案，术中注意保护进出尾状叶的管道，避免医源性损伤，也为患者的术后顺利康复提供了坚实的保障。结合既往的病例报道，在行肝切除术之前，有必要行胆道镜探查，一则可以明确尾状叶胆管内有无结石，二则可明确尾状叶胆管开口部位，可在一定程度上提示术者避免损伤胆道，导致术后胆汁漏、胆管狭窄等并发症

发生，且术后放置 T 管引流，如短期内患者结石残留或复发亦可通过 T 管窦道取石。在行肝实质离断过程中，术者要精准把握肝脏解剖平面，可减少术中出血并快速识别出重要管道结构^[25-27]，如萎缩的左半肝与 Spiegel 叶之间的潜在平面、肝左静脉根部与 Spiegel 叶之间的 Arantius 管平面（沿该平面离断肝实质即可分离出肝中静脉）、萎缩的右半肝与右尾状叶间的潜在平面，同时结合术中超声可有效避免尾状叶血管的损伤^[28-29]。肝内胆管结石通常伴有炎症及血管的代偿性增生^[30]，离断后的肝脏切面，做好确切的缝扎处理，可有效减少术后出血及胆汁漏的发生概率。

综上，对于经严格筛选的肝内胆管结石合并肝叶萎缩-增生综合征且双侧肝叶功能明显受损、尾状叶代偿性增大的患者，在充分评估肝功能储备、剩余肝体积以及尾状叶血供、静脉回流和胆道引流条件后，保留尾状叶次全肝切除术可作为一种个体化治疗选择。三维可视化联合胆道镜及术中超声有助于提高手术规划及重要管道结构保护的准确性。但目前相关证据主要来自病例系列及个案报道，长期疗效和适用范围仍需进一步研究验证。对于长期肝内胆管结石患者，还应加强胆管癌的风险评估及长期随访。

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