

· 病例报道 ·

成人先天性支气管胆管瘘1例

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【中图分类号】R734.2 DOI: 10.3779/j.issn.1009-3419.2010.01.18

A Case Report of Congenital Bronchobiliary Fistula in Adults

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1 临床资料

患者，女性，51岁。咳苦胆味淡黄色泡沫样痰51年，加重2年。患者生后即出现咳嗽咯痰，夜间睡眠时加重，每日痰量约40 mL，淡黄色，味苦，最多每日达500 mL。曾于多家医院反复诊断为“肺炎”。既往无白陶土样便，无肝胆疾病史，饮食正常。查体：右侧中下胸部触觉语颤略有增强，叩诊右肺中下部可及轻度浊音，听诊右肺中下部可闻及明显湿啰音，左侧正常。支气管镜检查示：右支气管开口隆突旁可见一漏斗样狭窄分支，管腔内不断漏出胆汁样分泌物。ERCP检示：窦道经左肝管经食管裂孔越过膈肌沿食管右前方向上蔓延，查胸、上腹部CT示：可见穿过膈肌之处至纵隔造影剂进入，并见右主支气管与其分支显影。

于2009年4月9日全麻下经右胸后外侧切口行支气管胆管瘘切除结扎缝合术。术中见右下肺表面有胆汁样沉着。食管前并行一管状结构，上端与右主支气管近隆突处相连，向下经食管裂孔与左肝内相连，长约15 cm，上管口直径约1 cm，有软骨环，下管口直径约1.5 cm，肌样管状结构（图1）。上端与右主支气管相连处根部用双7号线结扎，下端与膈肌下1 cm处双重结扎，切除瘘管，两残端再用3-0可吸收线缝扎。

术后病理：近端见支气管性组织伴炎症改变，远端见肌样组织伴炎症改变（图2）。诊断：1. 支气管胆管瘘（先天性）；2. 胆汁性肺炎。

术后患者症状消失，无并发症，痊愈出院。

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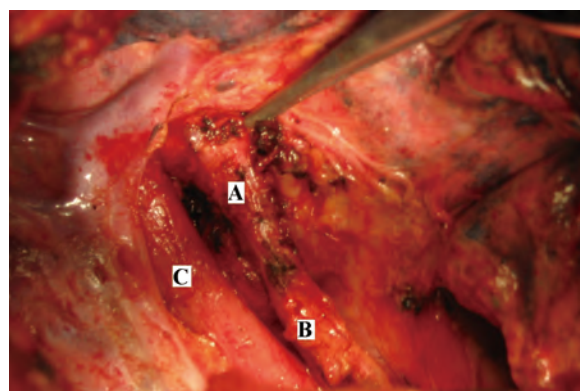


图1 术中所见，支气管胆管瘘

A: 瘘管近端；B: 瘘管远端；C: 食管。

Fig 1 The bronchobiliary fistula in the operation

A: Proximal end of the fistula; B: Remote end of the fistula; C: Esophagus.



图2 切除的支气管胆管瘘管

A: 近端，见支气管组织；B: 远端，见肌样组织。

Fig 2 The excisional bronchobiliary fistula

A: Proximal end; it can be seen bronchial tissues; B: Remote end; it can be seen muscle tissues.

2 讨论

先天性支气管胆管瘘是一种极其罕见的畸形，女孩较男孩常见，大多数因呼吸道症状和反复的肺部感染被发现^[1]。成人先天性支气管胆管瘘更为罕见，文献报告仅5例^[2,3]。在成人病例中，胆汁流向肺组织对肺产生损伤^[3]，但此患者没有明显的肺损伤，可能因患者能够自主地把支气管内胆汁咯出。

诊断主要依靠患者长期咳胆汁样痰及肺部感染症状病史。辅助检查包括支气管镜、ERCP或MRCP、胸上腹CT、便常规、胆汁样痰化验等。支气管镜能够很好地显示瘘管在气管内开口，并能够确定瘘管流出成分。但是ERCP或MRCP检查在确定有无胆道畸形中占有重要地位，先天性支气管胆管瘘并存胆道发育不全或胆总管闭锁者占36.8%^[4,5]。胸、上腹部CT检查对于继发性支气管胆管瘘有鉴别作用。

诊断明确即应手术治疗，介入栓塞疗效尚不明确。患者胆汁长期流向肺组织所致肺损伤，多伴肺部感染，术前需应用抗生素，如患者为先天性支气管胆管瘘无胆道畸形，可行右胸后外侧入路，暴露右肺和瘘管，上端

尽量靠近支气管结扎瘘管或用支气管闭合器闭合，以防止残存组织继续分泌液体，对呼吸道刺激引起术后呼吸道刺激症状。下端贴近食管裂孔结扎。如患者为先天性支气管胆管瘘伴胆道畸形，开胸结扎上端瘘管同时开腹行胆道成形术。未治疗的支气管胆管瘘，可能会导致进行性呼吸功能障碍和死亡，但是如果治疗正确，预后良好，总死亡率为25%^[4]。

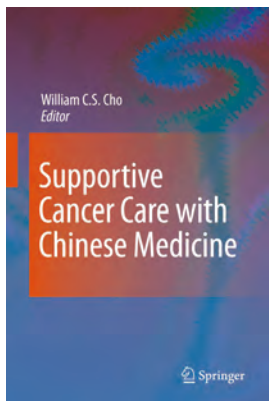
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(收稿: 2009-09-21 修回: 2009-10-19)

(本文编辑 南娟)

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2010. Approx. 400 p. Hardcover

ISBN 978-90-481-3554-7

W.C.S. Cho, Queen Elizabeth Hospital, Hong Kong (Ed.)

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