

• 病例报告 •

## 矫正型大动脉转位合并内脏反位及卵圆孔未闭并发脑梗死 1 例

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[关键词] 矫正型大动脉转位;卵圆孔未闭;脑梗死

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### Corrected transposition of great arteries complicate with visceral inversion, patent foramen ovale and cerebral infarction: a case report

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**Summary** Corrected transposition of great arteries (CTGA) is exceptionally rare, totaling approximately 0.4%–1% of all congenital heart diseases. Studies have demonstrated that 80%–95% of CTGA patients had situs solitus, while 5%–20% had situs inversus. We describe a patient with congenitally corrected transposition and situs inversus, complicated with patent foramen ovale and stroke. How should we treat such complicated patients? In this paper, we review a large number of relevant literatures combined with the patient's treatment experience to provide some suggestions.

**Key words** corrected transposition of great arteries; patent foramen ovale; stroke

矫正型大动脉转位 (corrected transposition of great arteries, CTGA) 是一种罕见的复杂性先天性

心脏病, 占先天性心脏病的 0.4%~1%<sup>[1-2]</sup>。有研究表明, 80%~95% 的 CTGA 患者内脏正位, 5%~20% 存在内脏反位<sup>[3]</sup>。本例患者 CTGA 合并内脏反位, 同时存在卵圆孔未闭 (patent foramen ovale, PFO), 并发生脑梗死。对于这一特殊类型

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的患者,临床该如何处理? 本文通过查阅大量文献,结合患者的具体情况进行阐述。

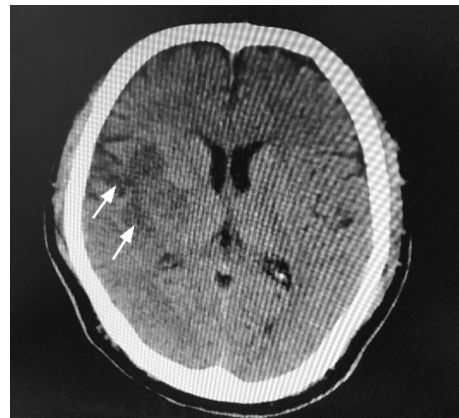
## 1 病例资料

患者,男,52岁。因“突发意识障碍 2 h 余”入院。体温 36.8℃、脉搏 67 次/min、呼吸 18 次/min、血压 98/65 mmHg(1 mmHg=0.133 kPa)。入院体检:神志不清;双侧瞳孔等大、圆形,直径 3 mm,对光反射迟钝;肌力(0~5 级):左上肢 0 级、左下肢 0 级、右上肢 5 级及右下肢 5 级。考虑急性脑梗死。遂行头颅 CT 示:右侧额颞顶叶及右侧基底节区梗死(图 1)。急诊行“脑血管造影+机械取栓术”,术中行猪尾巴主动脉弓上造影见患者心脏位于右侧胸腔,主动脉弓上各大血管开口均起自右侧,5F 单弯造影管插入右侧颈内动脉起始部造影见颈内动脉 C2 以远不显影。取栓微导管取出栓子,造影见颈内动脉、大脑中动脉、大脑前动脉各分支均显影良好,血流速度正常。患者返回病房后继续治疗。造影显示患者心脏位于右侧胸腔,行经胸超声心动图检查示:肝脏位于左上腹,心脏位于右侧胸腔,心房反位,心室右襟,主动脉与肺动脉呈右前左后排列。主动脉与解剖右室(功能左室)相连,肺动脉与解剖左室(功能右室)相连(图 2a)。解剖右室增大(52 mm),室壁增厚(9 mm),左房、右房内径增大,解剖左室内径正常。静息状态下室壁收缩未见明显异常。三尖瓣(右侧房室瓣)左房侧收缩期可见少量反流束,二尖瓣(左侧房室瓣)右房侧收缩期可见微量反流束(图 2b)。房水平可见异常左向右穿隔血流束。考虑为 CTGA(IDD 型)、PFO。食管超声心动图明确为 PFO,左房及左心耳内未见自发显影及附壁血栓形成(图 2c、d)。增强 CT 示:内脏反位:肝脏、脾脏、胃反位;CTGA;右位主动脉弓、胸腹主动脉未见明显异常。心电图示:窦性心律,PR 间期延长,右位心。患者病情稳定,神志清楚,双瞳正常,左上肢肌力 4 级,左下肢肌力 4 级,右侧肌力正常。出院后继续康复治疗。

## 2 讨论

CTGA 是指心房、心室连接不协调合并心室动

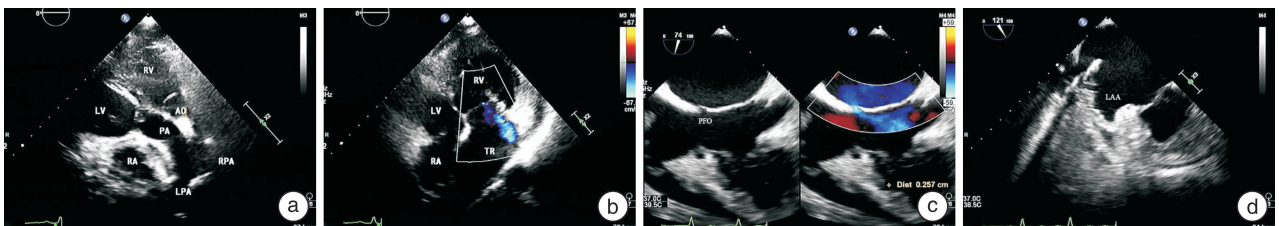
脉连接不协调。CTGA 患者可存在内脏正位或内脏反位,合并内脏正位的 CTGA 患者占 80%~95%<sup>[3]</sup>。在 CTGA 患者中,90% 合并室间隔缺损,65% 合并肺动脉狭窄,46% 合并三尖瓣反流<sup>[4]</sup>。有 1% 的 CTGA 患者不存在上述合并症(室间隔缺损、肺动脉狭窄)而被较晚诊断<sup>[5]</sup>。本例患者无室间隔缺损和肺动脉狭窄,平常无不适表现,因脑梗死就诊才被诊断出存在 CTGA。有研究发现,CTGA 内脏反位的患者室间隔缺损、肺动脉狭窄的发生率较 CTGA 内脏正位的患者高<sup>[6]</sup>,但也有研究认为两者发生率无差异<sup>[7]</sup>。上述结论均基于小样本的统计结果,其真实发生率可能还需要更多样本的研究予以确认。但越来越多研究支持 CTGA 内脏反位患者房室传导阻滞和三尖瓣关闭不全发生率较 CTGA 内脏正位的患者低<sup>[6-7]</sup>。一方面因为 CTGA 内脏反位患者形态学三尖瓣的发育较正常,出现 *abstain* 畸形较少见。另一方面,CTGA 内脏反位患者其房室结位于比较正常的后下部,而 CTGA 内脏正位患者房室结位于前上部,其房室传导系统排列错乱<sup>[8]</sup>。本例患者心电图提示 PR 间期延长,需长期随访房室传导阻滞有无进展。



注:箭头示右侧额颞顶叶及右侧基底节区梗死。

图 1 患者头颅 CT 图像

Figure 1 A head CT image of the patient



a: 心房反位,心室右襟,主动脉与肺动脉呈右前左后排列,主动脉与解剖右室相连,肺动脉与解剖左室相连;b: 三尖瓣少量反流束;c: PFO;d: 左房及左心耳内未见自发显影及附壁血栓形成。

图 2 患者超声心动图

Figure 2 Echocardiography images of the patient

对于没有合并室间隔缺损及肺动脉狭窄的 CTGA 患者,虽然无症状,但长期随访解剖右室的功能及三尖瓣关闭不全的程度很重要。有研究表明,25%没有合并心脏畸形的 CTGA 患者会在 45 岁发生解剖右室功能衰竭,而 67%合并其他心脏畸形(室间隔缺损及肺动脉狭窄)的 CTGA 患者会发生解剖右室功能衰竭<sup>[9]</sup>。一般认为解剖右室功能衰竭与严重三尖瓣关闭不全或进行性传导系统异常有关<sup>[5]</sup>。有研究报道 1 例 82 岁无其他心脏畸形的 CTGA 内脏反位患者,在不存在严重三尖瓣功能不全或心律失常的情况下,因解剖右室功能衰竭就诊,随后心脏磁共振发现解剖右室存在纤维化,推测解剖右室纤维化可引起右室功能衰竭<sup>[5]</sup>。右室壁瘢痕或纤维化与较大年龄、解剖右室低射血分数、解剖右室高压力和心律失常有关。本例患者目前虽无解剖右室衰竭表现,三尖瓣反流也较少,但仍需要长期随访解剖右室功能。心脏磁共振被认为是能早期准确判断解剖右室纤维化相关右室功能不全的一项检查。

CTGA 患者目前有两大类手术方式<sup>[10]</sup>。一类是生理性修补术,包括室缺修补、左室流出道梗阻疏通术及三尖瓣修补或置换。另一类是解剖性修补术,目的是使体静脉的血回流到右房,经解剖右室到达肺循环,肺静脉的血入左房,经解剖左室至体循环,减少解剖右室功能衰竭和三尖瓣功能不全的发生率。但是,解剖性修补术复杂、难度高,患者也存在较多并发症,需要更多的经验总结。对于存在解剖右室功能衰竭的 CTGA 合并心脏反位患者,也可进行双心室起搏+埋藏式除颤器治疗<sup>[11]</sup>。

CTGA 可合并 PFO。CTGA 合并 PFO 并发脑梗死鲜有报道。有研究得出矛盾栓塞危险(risk of paradoxical embolism, RoPE)系数评分在 7、8 和 9 分时,PFO 占脑梗死原因的 72%、84% 和 88%<sup>[12]</sup>。根据 RoPE 系数评分,本例患者评分为 7 分。本例患者无大血管动脉粥样硬化及血栓,无脑卒中常见危险因素,无深静脉血栓史,无近期旅行史,无肺栓塞。结合病史和 RoPE 评分考虑脑梗死与 PFO 有关。有研究报道在隐源性脑卒中患者中,有 40% 以上没有明确卒中病因的患者存在 PFO<sup>[13]</sup>。对于 PFO 相关脑梗死的二级预防,大量的研究证明,PFO 封堵治疗在降低二次脑梗死上优于抗血小板治疗,与抗凝药物治疗无差异<sup>[14]</sup>。PFO 封堵治疗可能会导致一部分人出现暂时性心房颤动,抗凝药物存在长期出血风险,本例患者应选择哪种治疗方法,因根据患者的具体情况和更为详细的检查,做出更有利患者的决策方案。

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