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成人弯刀综合症 1 例并相关文献复习 *

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摘要 目的:探讨我院 1 例成人弯刀综合症患者的临床表现、诊断手段及治疗方法。**方法:**简要陈述我院 1 例成人弯刀综合症患者的入院基本情况及诊疗过程,调研并结合相关文献进行复习。**结果:**该患者结合临床表现,通过超声心动图、胸部后前位 X 片及右心导管造影等检查,结果证实该患者右下肺静脉畸形引流,诊断为弯刀综合症,并进行相关对症治疗及后续生活和治疗指导。**结论:**弯刀综合症为一种罕见而复杂的先天性心肺血管疾病,属于肺静脉畸形引流的一个类型(心下型),病因不明,此病在成人中更为少见,该病的诊断主要依赖于右心导管术和心血管造影以及一些影像学手段,成人型多以保守对症治疗为主,若出现严重症状者则选择外科手术纠正畸形引流治疗。

关键词:弯刀综合症;先天性心肺血管疾病;成年;诊断

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Scimitar Syndrome of an Adult: A case Report and Review of Literature

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ABSTRACT Objective:To study the clinical manifestation, diagnosis and treatment of Scimitar Syndrome. **Methods:** Basic conditions and the treatment process of one case diagnosed with Scimitar Syndrome were briefly described, and its relevant literatures were surveyed and reviewed. **Results:** Right inferior anomalous pulmonary venous was confirmed by ultrasound, plain chest radiographs and angiocardiography combined with clinical manifestations, and Scimitar Syndrome was diagnosed. She had accepted symptomatic treatment and guidance of follow-up life. **Conclusion:** Scimitar syndrome is a rare and complex congenital cardio-pulmonary vascular disease, and one type of anomalous pulmonary venous drainage (infracardiac type). Its etiology is unclear. The disease is more rare in adults. Its diagnosis is mainly dependent on angiocardiography and imaging methods. The adult combined with Scimitar Syndrome are usually given symptomatic treatment. If severe symptoms appear, its primary treatment is surgery to correct anomalous drainage.

Key words: Scimitar Syndrome; Congenital cardio-pulmonary vascular disease; Adult; Diagnosis

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前言

肺静脉异位引流分为两种类型,即完全型肺静脉以为引流和部分型肺静脉异位引流,其发病率约占先天性心脏病的 2%^[1]。部分肺静脉异位引流为 4 支肺静脉中任何 1-3 支未与左心房正常连接,而与右心房或腔静脉相连接。根据引流位置的不同,可分为心上型、心内型及心下型^[2]。

弯刀综合症,即“Scimitar 综合症”,此命名由 Neill 等人首次提出^[3],是一种罕见的先天性心肺血管疾病,属于部分型肺静脉异位引流的心下型。它的病因尚不清楚,有文献报道可能与胚胎发育早期,肺芽发育障碍有关^[4],其发病率大概占部分型肺静脉异位引流的 3-5%^[5]。本文结合 1 例成人型弯刀综合症患者的临床资料合并相关文献进行复习,探讨弯刀综合症的临床症状及目前的诊断手段及治疗情况。

1 病例资料

1.1 一般资料

患者,女,33 岁,主因“胸闷、气短 8 个月余”入院。患者于 8 个月前出现活动后胸闷、气短明显加重,口唇紫绀,活动能力明显下降,但无胸痛、心悸等症状,休息时症状缓解。患者于 5 岁时行房间隔缺损修补术,入院查体:血压 119/74mmHg,心率 88 次/分,律齐,双肺呼吸音清,胸骨左缘第 4 肋间闻及 III 级吹风样杂音,叩诊右心界扩大,双下肢无水肿。

1.2 辅助检查

患者于外院行肺动脉 CTA 检查示有肺静脉汇入下腔静脉,后进入右心房;另于右心房左外侧可见一血管分支影,其远端显示欠清晰,肺动脉及下腔静脉增宽;右心增大;胸壁软组织及胸骨呈术后改变。入院后行心脏超声检查示左室射血分数

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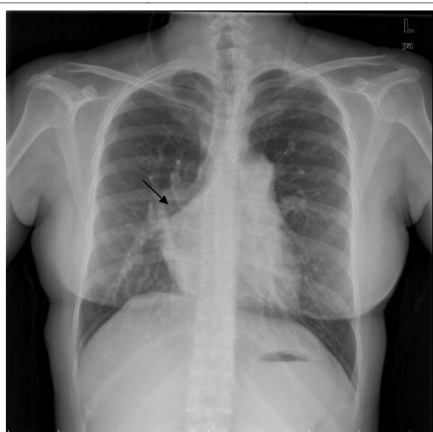


图1 胸部X片*注:箭头所示为“弯刀征”
Fig. 1 Chest X-rays *Note: The arrow shows the “scimitar sign”

56%,估测肺动脉收缩压约为 114mmHg,重度肺动脉高压,右心扩大,主肺动脉及右肺动脉增宽,三尖瓣大量反流,肺动脉瓣少-中量反流,下腔静脉增宽。胸部X线检查可见心脏右侧向下朝向膈有一个弯曲的血管影,右心扩大,如图所示(图1)。行右心导管及肺血管扩张试验检查结果示:吸入伊洛前列素前,右房压 11/11/10mmHg,右室压 88/3/11mmHg,肺动脉压 95/35/54mmHg,肺毛细血管楔压 9/10/9mmHg,主动脉压 127/76/94mmHg,全肺阻力 2.5wu,全肺阻力指数 4.5wu/m², Qp/Qs:0.50;吸入伊洛前列素 20 微克 10 分钟后,右房压 12/7/12mmHg,右室压 62/7/12mmHg,肺动脉压 62/22/36mmHg,主动脉压 110/63/77mmHg,血氧饱和度为 98%。各部分血红蛋白含量、氧含量及氧饱和度测得结果(表1)。左右肺动脉主干增宽,肺动脉各分支屈曲,血流尚可,未见充盈缺损及“截断征”;肺循环时间延长,右下肺静脉引流下腔静脉至右心房,选

表1 右心导管检查各部血红蛋白、氧含量及氧饱和度

Table 1 Hemoglobin,blood oxygen content and oxygen saturation measured by right heart catheterization

Position	Hemoglobin(g/dl)	Oxygen content (ml/dl)	Oxygen saturation (%)
Aorta	12.6	16.11	94
Pulmonary artery	13.3	15.01	83
Right ventricle	13.6		81
Superior vena cava	12.9		64
Right atrium(upper)	12.9		71
Right atrium(middle)	13.3	15.56	86
Right atrium(lower)	13.3		81
Inferior vena cava	12.9		79
Right inferior pulmonary vein	12.6		96
Pulmonary vein	12.6	16.11	94



图2 A 右心导管造影*注:箭头所示为异常的右下肺静脉
Fig. 2A Angiography of right heart catheterization *Note: The arrow shows the anomalous right inferior pulmonary vein



图2 B 右心导管造影*注:箭头所示为异常的右下肺静脉
Fig. 2B Angiography of right heart catheterization *Note: The arrow shows the anomalous right inferior pulmonary vein

择性右下肺静脉造影,证实右下肺静脉以为引流(图2A、B)。

1.3 诊断依据

该患者胸闷气短,且胸骨左缘第4肋间闻及Ⅲ级吹风样杂音,叩诊心浊音界右移,似右位心。胸片可见“弯刀征”,根据胸片此特征多可作出诊断;外院行肺动脉CTA检查也证实有肺静脉汇入下腔静脉;心导管检查右下肺静脉引流下腔静脉至右心房,且右心房的血氧含量高于腔静脉。心脏和纵膈稍右移可

能与右肺容积缩小有关,为继发性改变。胸闷气短的症状可与肺动脉高压有关。该患者目前的症状及体征符合弯刀综合症的特征,且各项检查证实该患者诊断明确。

2 讨论及文献复习

弯刀综合症,主要是指右肺静脉(通常所涉及的为异常的右下肺静脉^[6])流入下腔静脉,前后位胸片示异位引流的静脉

汇合成一条大血管,成凸面向右的新月状或弯刀状,与右心缘大致平行,X线上称为弯刀征(scimitar sign)。本应流入左房的右肺静脉经下腔静脉流入右房,造成了房性水平的左向右分流,造成右房肥大,长期的结果会导致右心衰竭。目前弯刀综合症的临床谱很广泛^[8],通常分为三类:一类是在婴幼儿期出现症状^[9],通常是在出生后两个月内出现呼吸困难,反复肺部感染和心衰的症状,一般很难成年,对于在急诊过程中婴儿呈现出现严重哮鸣音要高度怀疑一些罕见病如弯刀综合症等^[10];另一类在成年出现症状的患者,通常表现为呼吸窘迫,右肺发育不全和心脏右移^[11];还有很多成人或年龄稍大患者未有明显临床症状,在由其他症状导致的诊治或者例行体检中发现。弯刀综合症作为一种复杂的静脉回流异常病症,目前用于它的诊断多为螺旋CT和MRI^[12],但传统意义上讲,心导管检查仍然是诊断的金标准,而且可以定量Qp/Qs,同时也是异常血管线圈闭塞治疗的选择手段^[13]。该病的心肺症状和体征取决于继发的左向右分流的血流动力学变化,肺全身血流比(Qp/Qs),以及相关的心脏异常的存在(如房室间隔缺损等),若Qp:Qs>2,有可能会发展成为继发性的肺动脉高压。目前对于此病的保守治疗主要是对症治疗,包括进展性心力衰竭的控制、肺动脉高压的治疗和适当的抗感染治疗^[10]。由于本病变复杂,故无固定治疗方法,对于成人的手术指征没有明确的指南^[14],手术修复的指征包括弯刀综合症伴房间隔缺损,肺动脉高压,异常的肺静脉主干狭窄,而在婴幼儿期,肺动脉高压是评价手术的一个指标,伴随肺动脉高压在逐渐进展,患者后续有效的医治,可能会延缓手术时间^[12]。由于异常肺静脉开口位置不定及肺内血管和心血管畸形各异,目前的手术纠正方式各异^[15]。目前研究报告不同类型的弯刀综合症的死亡率报道不尽相同,范围从64%到16%不等^[16]。

该患者入院后经右心导管检查证实为右下肺静脉-右心房异位引流,重度肺动脉高压,X线检查可见“弯刀征”,确诊为成人弯刀综合症1例。弯刀综合症患者伴有房缺和肺动脉高压,成人中很少见^[17],该患者因其在5岁时行房缺修补术,对其后期的症状控制和生活质量有很大的改善作用。肺血管扩张试验结果表明患者的肺血管功能较好,心外科会诊意见,给予手术治疗。目前出院给予家庭氧疗,波生坦、贝前列素钠片等治疗降低肺动脉压治疗,待患者病情稳定,给予手术治疗,纠正肺静脉的畸形引流。

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