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· 病例报道 ·

Ultrasonic diagnosis of Abernethy malformation with pulmonary hypertension: a case report

超声诊断 Abernethy 畸形合并肺动脉高压 1 例

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患者男, 18 岁, 因“活动后气促、心悸 10 余年, 晕厥 2 次”入院。心电图检查: P2 亢进, P2>A2, 心前区未闻及杂音。超声心动图检查: 右心稍大, 三尖瓣轻度反流, 肺动脉及其分支增宽, 肺动脉瓣轻度反流, 肺动脉高压(中度), 肺动脉收缩压 71 mm Hg (1 mm Hg=0.133 kPa), 舒张压 42 mm Hg(图 1A)。上腹部超声检查: 脾静脉内径约 7 mm, 脾静脉与肠系膜上静脉汇合成门静脉, 门静脉于肝门下方直接汇入下腔静脉, 入下腔静脉处扩张, 内径约 35 mm, 未见入肝门静脉血流(图 1B)。超声提示: 先天性肝外门-腔静脉分流。上腹部增强 CT 检查: 肠系膜上静脉与脾静脉汇合成门静脉, 门静脉大部分汇入下腔静脉, 汇入处直径约 37 mm, 另外分出一纤细分支, 直径约 2 mm, 走行至肝脏, 未见左、右属支显影; 脾静脉、左肾静脉扩张, 左肾静脉直径约 14 mm, 脾静脉直径约 15 mm; 肝门处门静脉明显纤细; 下腔静脉明显增粗, 直径约 31 mm(图 2)。CT 诊断: Abernethy 畸形(肝外门-腔静脉分流, II 型), 合并门静脉性肺动脉高压。

讨论: Abernethy 畸形即先天性肝外门-腔静脉分流, 是一种罕见的先天畸形, 为胚胎期脐静脉和卵黄静脉发育异常, 导致门静脉与腔静脉之间异常分流所致。临床表现为门静脉畸

形, 如门静脉干及其分支闭塞或变细, 并可见一迂曲扩张的肝外门-腔静脉分流通路。1994 年 Morgan 和 Superina^[1] 将 Abernethy 畸形分为: I 型, 肝脏完全无门静脉血灌注, 即肝内门静脉缺如, 胃肠道静脉血流完全汇入腔静脉; I a 型为肠系膜上静脉与脾静脉无汇合; I b 型为肠系膜上静脉与脾静脉汇合。II 型, 门静脉血部分向肝脏灌注, 肝外门-腔静脉间存在先天性分流; 本例即为此型, 以肺动脉高压为首发症状, 属于先天性门-腔静脉分流导致的肺动脉高压。CT 提示有纤细的门静脉供应肝脏组织, 超声于第一肝门区未扫及门静脉及血流频谱, 仅探查到肝动脉血流频谱, 肠系膜上静脉与脾静脉增粗并直接汇入下腔静脉, 故诊断为 Abernethy 畸形, 但未能进一步准确分型, 分析原因为门静脉往往纤细、发育不良, 较正常汇入肝脏的位置高, 在第一肝门上方汇入肝脏, 肝内又无法探查到明确左、右属支, 因此并未扫查到此支纤细的门静脉。

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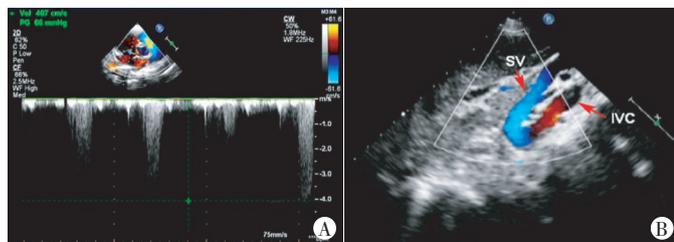


图 1 Abernethy 畸形合并肺动脉高压声像图(IVC: 下腔静脉; SV: 脾静脉)

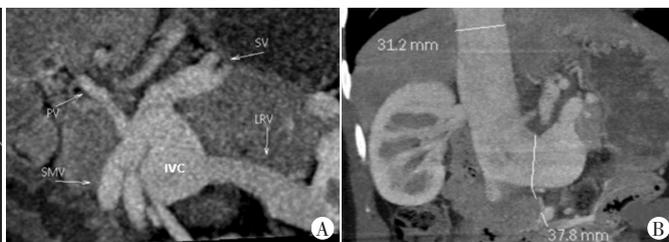


图 2 Abernethy 畸形合并肺动脉高压增强 CT 图(PV: 门静脉; IVC: 下腔静脉; SV: 脾静脉; SMV: 肠系膜上静脉; LRV: 左肾静脉)

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