

静脉内平滑肌瘤病长入右心1例 并文献复习

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摘要

目的: 通过分析静脉内平滑肌瘤病(intravenous leiomyomatosis, IVL)的临床特点、临床表现、诊断及治疗与预后, 同时阅读最新文献, 总结最新诊疗方法, 以提高该疾病的诊疗水平。方法: 分析青岛大学附属医院收治的1例静脉内平滑肌瘤病长入右心的诊治经过, 并复习国内外文献。结果: 患者吕某, 因“胸闷、憋气20天”入院, 入院后完善相关辅助检查, 排除禁忌后行手术, 术后病理: 血管内平滑肌瘤。结论: 静脉内平滑肌瘤病是一种罕见的、组织学表现为良性的子宫肿瘤, 特征是平滑肌细胞在血管内增生, 起源于子宫肌层, 以其最广泛的形式, 可以通过盆腔静脉和下腔静脉到达心脏, 造成血液动力学并发症。子宫平滑肌瘤侵入血管长入右心室极少, 病情严重, 早发现、早诊断、早治疗尤为重要。

关键词

静脉内平滑肌瘤病, 平滑肌瘤, 子宫

Intravenous Leiomyomatosis Growing into the Right Heart: A Case Report and Literature Review

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Abstract

Objective: To analyze the clinical features, diagnosis, treatment and prognosis of intravenous leiomyomatosis, read the latest literature, and summarize the latest diagnosis and treatment methods in order to improve the diagnosis and treatment of the disease. **Methods:** The diagnosis and treatment of 1 case of intravenous leiomyomatosis growing to the right heart treated in the Affiliated Hospital of Qingdao University were analyzed, and the literature at home and abroad was reviewed. **Results:** The patient, Lv, was admitted to the hospital because of “chest tightness and breath holding for 20 days”. After admission, he improved relevant auxiliary examinations and performed surgery after excluding contraindications. The postoperative pathology was intravascular leiomyoma. **Conclusion:** Intravenous leiomyomatosis is a rare and histologically benign uterine tumor, characterized by the proliferation of smooth muscle cells in blood vessels, which originated from the myometrium of uterus. In its most extensive form, it can reach the heart through pelvic vein and inferior vena cava, resulting in hemodynamic complications. Uterine leiomyoma rarely invades blood vessels and grows into the right ventricle, and its condition is serious. Early detection, diagnosis and treatment are particularly important.

Keywords

Intravenous Leiomyomatosis, Leiomyoma, Uterus

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1. 引言

静脉内平滑肌瘤病(intravenous leiomyomatosis, IVL)是一种罕见的良性平滑肌肿瘤,起源于子宫或子宫血管,它的特点是持续的腔内生长,可通过髂静脉和下腔静脉延伸到右心室和肺血管系统,导致危及生命的并发症[1] [2]。自 1896 年首次记录以来,其有着类似于恶性肿瘤的生物学行为,其沿着静脉腔生长,形成可延伸至心肺系统的瘤栓[3]。IVL 的发病率较低,早期患者的临床表现是非特异性的,大多数患者没有症状或特异性肿瘤标志物。IVL 的首选治疗是根治性肿瘤切除手术,以降低复发风险。肿瘤完全切除后不易复发,不完全切除后的术后复发率很高,约为 16.6%~30.0% [4]。基于其罕见性,多以病例报道的形式呈现,临床医师经验不足,影响疾病的诊治。现分析青岛大学附属医院收治的静脉内平滑肌瘤病 1 例的诊治经过,并复习国内外文献,总结最新诊疗方法,以提高该疾病的诊疗水平。

2. 病历资料

患者,女,42 岁,因“胸闷、憋气 20 天”就诊于我院心外科,伴有双下肢及颜面部水肿,伴有腹胀、纳差,无咳嗽、咳痰、咯血,无夜间端坐呼吸,病情反复发作,渐加重,行心脏超声提示右室内占位,考虑血管平滑肌瘤;三尖瓣口、主肺动脉血流梗阻;三尖瓣反流(重度);右房扩大;右室扩大;包积液(少量)。既往病史:2021 年 9 月于当地行子宫肌瘤切除术,术后病理提示肌瘤细胞丰富,部分静脉内增长,目前复查复发。入院后完善相关检查,予速尿等药物积极利尿、抗凝治疗,病情稳定,未见手术禁忌。经多学科评估报重大手术审批后于 2023-6-12 行心脏病损切除术 + 三尖瓣成形术 + 下腔静脉病损切除术 + 全子宫切除术 + 双附件切除术 + 右侧输尿管支架植入术 + 体外循环开放性心脏手术,术

后病理提示符合血管内平滑肌瘤,(下腔静脉肿物及心脏肿物)结合形态及免疫组化,符合血管内平滑肌瘤累及右心室,部分区域伴退变、梗死,周边肿瘤细胞增生活跃。免疫组化结果:子宫平滑肌瘤 SMA(+), Caldesmon(部分+), FH(+), Ki-67(+,约3%), CD10(-);心脏肿物 SMA(+), Caldesmon(部分+), FH(+), Ki-67(+,约3%);下腔静脉肿物 SMA(部分+), Caldesmon(灶+), FH(+), Ki-67(+,约3%)。术中出血量约1300 mL,输血6单位红细胞、700 mL血浆、1000 mL自体血,手术顺利,创伤大,术后恢复时间长。后给予多巴胺、米力农、无水头孢等积极强心、利尿、抗凝、祛痰、营养心肌、预防感染等对症支持治疗,术后患者出现反复发热不适,经泌尿外科会诊,积极予以行右侧肾盂穿刺引流,术后体温稳定。后复查CT,未见残余病灶。

3. 讨论

3.1. 发病机制

IVL的发病机制尚未明确,发生来源现有两种学说,一是起源于广泛侵入静脉内的子宫肌瘤,二是子宫静脉壁内的平滑肌组织[5]。肿瘤可以游走走在血管腔内,也有少部分情况是附着在血管或心房壁上。IVL主要通过子宫髂静脉和卵巢静脉侵入全身静脉循环,其中髂静脉为主要途径[6]。发生IVL的危险因素有诊断为子宫肌瘤、既往子宫手术史(比如子宫切除术或子宫肌瘤切除术)以及既往IVL史[7]。

3.2. 临床表现

IVL患者的临床表现是不典型的,特别是在初期,主要与肿瘤受累部位有关[3]。IVL的早期诊断是困难的。因为大多IVL者是无症状的,且初期肿瘤仍在子宫肌层的小血管内,故任何成像方法都难以检测到肿瘤。只有当患者出现子宫肌瘤的一些临床症状,如月经改变、腹痛和腹部包块时,IVL的初步诊断才得以实现[8]。当肿瘤侵入静脉时,会引起下肢肿胀、腹水和肝脏肿大。随着IVL继续进展,可演变为心脏内平滑肌瘤病,从而影响三尖瓣启闭,导致充血性心力衰竭、晕厥,甚至猝死[9]。IVL术中所见多为不规则形态的子宫,伴外突结节,部分可见沿血管走形的蠕虫样结节。

3.3. 诊断

术前诊断IVL的最佳方法仍然是一个存在争议的话题。IVL的成像方法包括超声、CT和磁共振成像(MRI)。超声检查,尤其是心脏超声,在评价血管和心脏病变方面具有良好用途。CT和MRI用于更好地描述复杂的腹部肿块。两者都可以显示肿瘤的起源及静脉受累的部位。CT更快且更便宜,但MRI能够更好地识别软组织成分的差异,这可能有助于识别静脉内的附着点。冰冻病理准确率低,石蜡病理是诊断IVL最可靠的方法。IVL同子宫平滑肌瘤表现出相似的良性细胞学特征与免疫组化特点,即均为平滑肌细胞分化、无核异型性,ER和PR阳性,CD34阴性,Ki-67指数低[10]。

3.4. 治疗

多学科会诊能为IVL患者提供更好的治疗方案,让患者受益最大化。手术切除是IVL最有效的治疗方式,尽量一期完成包括子宫全切除+双侧附件切除+血管内肿瘤及心脏内肿瘤的切除。对于无子宫外血管受累的年轻、未育患者,可暂行保守手术,但术后应密切监测,一旦复发,尽早手术切除;对于反复发作,广泛累及子宫外大血管及心脏的患者,即使未生育,也应切除子宫及双侧附件,以减少复发,延长生存时间;对于无法手术、术后有肿瘤残留或复发的IVL患者,可采用抗雌激素治疗[11]。

4. 总结

综上所述,静脉内平滑肌瘤病(IVL)是一种罕见的良性平滑肌肿瘤,沿静脉血管生长,多起源于盆腔

静脉,可沿静脉系统蔓延,累及髂静脉、下腔静脉甚至心脏。IVL好发于生育期及围绝经期妇女,起病隐匿,临床表现缺乏特异性,取决于IVL肿瘤累及的部位,伴有下腔静脉等大血管和心脏受累的IVL的临床表现更是不尽相同。临床上应增强对IVL的辨别能力,对于子宫肌瘤病史或有子宫手术史的患者,如发现盆腔包块、下腔静脉或心脏内肿物时,考虑有可能IVL所致。子宫肌瘤患者如出现胸闷、憋气、下肢肿胀等症状时应疑诊伴有下腔静脉和心脏病变。超声、CT及磁共振成像(MRI)等影像学检查有助于发现IVL,并评估肿瘤的大小、范围和严重程度。IVL的需要多学科共同诊疗,全面和细致的术前评估有利于IVL的早期诊断、手术方案的制订和良好治疗结局的获得。术后积极监测是否出现复发,从而减少不良预后的发生。

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