

特发性腹膜后纤维化误诊为输尿管癌 1 例报告并文献复习

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摘要 目的:分析并掌握腹膜后纤维化的诊疗特点,避免对该病的误诊误治。方法:回顾性分析 1 例腹膜后纤维化患者分别因两侧肾积水先后两次住院并最终确诊的诊疗过程中的临床资料,包括临床表现、影像特点、病理结果、治疗方法及预后等,并结合相关文献进行复习。结果:患者因右肾积水首次入院诊断为右输尿管癌而行右侧肾输尿管切除术,后患者因左肾积水来我院住院诊断为腹膜后纤维化,行腹腔镜输尿管松解+腹腔内置术,术后长期随访疗效满意。结论:腹膜后纤维化作为一种少见病,缺乏对其认识极易导致误诊误治,掌握该疾病的临床特点及选择适宜的治疗方案,对本病的诊疗具有重要意义。

关键词 特发性腹膜后纤维化;误诊

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A Case of Idiopathic Retroperitoneal Fibrosis Misdiagnosis as Ureteral Carcinoma and Literature Review

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ABSTRACT Objective: To analyze and master the clinical characters of an idiopathic retroperitoneal fibrosis (IRPF) case to avoid the misdiagnosis and mistreatment. **Methods:** Analyzed the clinical data of an IRPF case who had been hospitalized twice because of unilateral hydronephrosis occurred on both sides successively, including clinical manifestations, radiographic evaluations, pathological results, treatment protocols and prognosis. Reviewed the literatures related to RPF in recent years. **Results:** The patient received the nephroureterectomy on right because she was misdiagnosed as ureteral carcinoma at the first hospitalization. The laparoscopic ureterolysis on left was performed on her because of the preoperative diagnosis of IRPF which was confirmed by the pathological result. The 3 years' follow up revealed the satisfied therapeutic effects. **Conclusions:** As a rare disease, the incidence of misdiagnosis on IRPF patients was relatively high, especially when the physician's knowledge of this disease was insufficient. It is meaningful to master the essentials of the diagnosis of this disease and choose the optimal treatment according to the individual conditions.

Key words: Idiopathic retroperitoneal fibrosis; Misdiagnosis

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腹膜后纤维化(RPF)是一种因腹膜后组织的非特异性炎症增生性反应引起腹膜后广泛纤维化的慢性进展性疾病,因临床发病率低,该病在发病早期极易引起误诊误治。现将一例被误诊的患者的诊疗经过回顾如下,并结合相关文献复习。

1 临床资料

患者女性,55岁,于2006年9月因右肾绞痛、乏力、呕吐到某院住院诊断为右侧输尿管尿路上皮癌,遂行右肾输尿管全长切除术,术后病理为右侧输尿管下段炎性病变。2008年8月11日患者突发左肾绞痛,伴纳差、恶心呕吐,就诊于当地医院行腹部B超、CT检查提示左肾输尿管积水,原因不明。因患者症状进行性加重,于8月14日转入我院治疗。查体:腹平坦,无腹壁静脉曲张,腹部柔软,无压痛、反跳痛。肝脏未触及,脾脏未触及,双侧腰部曲线对称,双肾区无隆起,右腰部皮肤可见一长约10cm的切口瘢痕,左肾区叩击痛(+),卧位及坐位双肾均未

触及。双侧输尿管行程区无压痛,膀胱区无膨隆,无压痛,未扪及包块。实验室检查:肌酐 $223\mu\text{mol/L}$,白细胞 $9.35\times 10^9/\text{L}$,血红蛋白 115g/L 。泌尿系超声:左肾集合系统分离,前后径约1.2cm,左侧输尿管扩张,上段内径约0.8cm。腹部CT平扫(图1)腰椎体前方腹主动脉、下腔静脉、髂总血管周围的腹膜后及盆腔间隙可见软组织影,同时包绕左侧输尿管。

鉴于患者为孤立肾合并肾功能不全,遂于入院当日急诊行左侧肾盂输尿管逆行造影(图2)发现左侧肾盂积水,近段输尿管向中线移位,输尿管内未见充盈缺损。急诊留置左侧输尿管导管(5F)后症状明显缓解,于当日复查肌酐降至 $140.1\mu\text{mol/L}$ 。后完善腹部MRI平扫+增强扫描(图3)腹膜后约腰3-4椎体前方可见稍长T1短T2异常信号影,最大层面约 $3.5\times 2.5\text{cm}$,上下累及长约5.5cm,其内包绕腹主动脉下段至髂血管分叉处,血管管腔轻度变窄,未见移位,动态增强扫描病变动脉期轻度增强,实质和延迟期明显强化。入院诊断:1、左肾输尿管积水;特发性腹膜后纤维化(idiopathic retroperitoneal fibrosis)可能性大;2、急性肾功能不全(肾后型);3、右侧肾输尿管全长切除术后。经讨论后于2008年8月26日行腹腔镜下左侧输尿管松解+腹腔内置术,术中所见:骶腰椎体范围的腹膜

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后组织增厚、僵硬,将左侧输尿管下段于髂髂关节附近包裹并向中线拉拢,致其上方输尿管及肾盂扩张积水。术中游离输尿管过程中去除髂髂关节附近包裹输尿管的僵硬致密组织送病理检验。术后病理结果(左输尿管周围组织)增生的纤维、血管及脂肪组织内见慢性炎细胞浸润。与术前诊断吻合。术后7天患者出院,继续口服泼尼松片共10个月(30mg/日2个月,逐渐减量至5mg/日2个月,5mg维持量6个月后停药),术后监测肌酐水平稳定在140-150 $\mu\text{mol/L}$,随访3年未出现左肾积水加重及肾功能减低。

2 讨论

腹膜后纤维化(RPF)是一种由于腹膜后纤维组织非特异性炎症反应纤维化导致腹膜后空腔脏器受压甚至梗阻的慢性进展性疾病,1948年Ormond第一次详细阐述了该疾病^[1],并于1960年提出该病为独立疾病。临床上较为少见,流行病学发生率大约在每年百万分之二至五^[2],好发于40-60岁,男:女为2-3:1^[3,4]。曾有报道52例患者平均随访85个月,28.12%的患者于随访期间死于尿毒症,10.3%发展为终末期肾功能不全,35.9%的患者为可代偿性肾功能不全,仅25.6%患者肾功能正常,提示本病的自然病程预后不佳。

根据病因是否明确,RPF可分为特发性和继发性两类,其中绝大部分为病因不明的特发性腹膜后纤维化(IRPF),尽管其病因及发病机制尚未明确^[5],但因其常伴发大动脉炎、硬化性胆管炎、硬化性纵隔炎等免疫性病变,目前学术界普遍倾向于RPF属于一类自身免疫反应性疾病^[6,7]。其组织病理学特点主要表现为非特异性炎症,可分为早期(亦称炎症活动期)和晚期(也叫慢性纤维化期)。早期病变组织中可见大量的炎细胞(以淋巴细胞、浆细胞和成纤维细胞为主,也常见一些嗜酸性细胞)分布于纤维胶原束中,并伴有丰富血管网,而晚期病变则以致密的纤维结缔组织为主,几乎没有血管和细胞浸润。

典型的IRPF病变部位常见于腰4、5椎体前方腹主动脉、下腔静脉、髂总血管周围的腹膜后及盆腔间隙,可引起输尿管受压或是影响输尿管正常蠕动,引起上尿路积水。RPF临床症状多为非特异性,可表现为腰背部或侧腹部钝痛,可向下腹、腹股沟放射。多数患者在症状出现4-6月后才能确诊,此时多已发展至肾积水、肾功能不全甚至尿毒症期,可表现为纳差、恶心、乏力、发热、高血压、少尿等症状。除此之外,病变组织还可能压迫下腔静脉导致下肢深静脉血栓及水肿形成,累及肾静脉可引起高血压、肉眼血尿的发生。而主动脉、纵隔、胆道系统及肾脏受累几率较小。实验室检查中多有血液沉降率(ESR)、C反应蛋白(CRP)及白细胞计数等炎症指标的升高,伴或不伴有贫血,后期可见肌酐、尿素氮水平升高、电解质紊乱等肾功能不全的表现。本例患者前后两次均以肾绞痛、乏力、恶心、呕吐起病,两次B超均提示患侧肾积水,其中第二次入院化验指标中除肌酐及尿素氮明显升高外,白细胞计数、ESR、CRP及Hb均在正常范围,可见炎症指标水平增高并不能作为诊断RPF的必要条件,急性炎症指标在正常范围亦有发生本病的可能,该患者的临床表现与其他病因导致的肾积水患者之间无明显差异。影像检查提示本病例的病变组织位于腰3-4水平,包绕了下腔静脉、腹主动脉下段至髂总血管分叉处以及左侧输尿管

管,属于典型的病变部位类型。

该病的临床表现及实验室检查均缺乏特异性,影像学检查成为其确诊的主要手段:1.B超:常表现为腹膜后的环绕主动脉、下腔静脉的低回声或等回声肿块,并且可发现因肿块包绕输尿管而导致的肾积水,结合彩色多普勒还可以了解肾血流、肾盂和输尿管积水情况。2.IVP(或逆行造影)表现:近段输尿管向中线移位,远端输尿管逐渐变细,多数累及双侧,偶尔累及单侧。3.CT表现:其中不同病理阶段的RPF其CT表现不同,病变在早期时,表现为腹膜后境界不清的弥漫低密度影(与肌肉类似),晚期时,表现为境界清楚的低、不均或等密度软组织影,呈对称或非对称性分布。增强后,早期病灶增强多较明显,晚期几乎无强化。因此CT是目前诊断该病及其病变进展的有效方法。4.MRI检查:T1加权相可见中低信号组织影(与肌肉相似或略低),血管信号清楚,T2加权相在大多数良性损害中表现为稍低信号区,而恶性表现为高信号不均质区^[8]。5.PET-CT因其具有全身显影及反映病变代谢活动状况等优势,评价其在RPF的诊断中的价值成为了近年来的讨论热点^[9-14],考虑到经济因素及实用性,该检查更适用于高度怀疑继发于恶性病变的腹膜后纤维化患者的筛查。本例患者的逆行肾盂输尿管造影表现与上述典型IVP基本一致,其CT表现为腹膜后低密度的不规则影,后经病理证实属于病变早期阶段,MRI影像在T2加权像上呈低信号,提示良性可能。该患者的逆行造影、CT及MRI三种影像学表现均符合典型的IRPF的特点,因此影像检查对于诊断RPF尤为重要。

RPF的治疗方法包括:1.急症处理:对已出现严重肾积水的患者实施输尿管支架管置入治疗后多数可获得满意效果,置入失败者可考虑行肾造瘘保护肾功能。2.药物治疗:对于确诊为IRPF者,尤其是处于疾病早期的患者,应尽早用药治疗^[15],目前常用的药物有激素类(泼尼松)、免疫抑制剂类(硫唑嘌呤、环磷酰胺)及雌激素调节剂(他莫昔芬),三类药物之间亦可联用。临床观察显示高剂量用药持续时间>6个月或低剂量维持用药时间>2年均有显著疗效^[16]。此外,有文献表明术后应用激素治疗较单纯手术治疗的复发率明显降低^[17],因此激素经常作为外科治疗的辅助手段,其适应症^[18]包括:术前准备、纤维化累及主动脉、腔静脉的术后治疗、术后疾病复发、高龄及体弱的患者。药物治疗的停药指征包括影像学及临床症状缓解、血沉稳定^[19]等。停药后可能会出现病情反复,因此须密切随访患者。3.外科治疗:对药物疗效不佳的患者可行手术治疗。手术可分为开放性手术和腹腔镜手术两种,手术一般选取腹正中经腹腔入路,从而可以同时双侧输尿管进行探查,对患侧进行输尿管松解,同时为保护输尿管不再受压,可选择输尿管腹膜侧边化或腹腔化处理。对于术前诊断不明确的患者术中需取活检^[20]。本文中的病例第二次住院时因输尿管梗阻导致了急性肾后性肾功能不全,给予置入输尿管支架管的急诊处理后肾功能恢复至接近正常水平,考虑到患者为孤立肾,因此我们选择了激素联合腹腔镜下输尿管松解+腹腔内置化手术的处理方案,术后继续应用激素10个月后停药,长期随访观察疗效稳定。

本病例的诊疗经验总结:RPF属少见疾病^[21],早期误诊率较高,国内曾有报道^[22]指出在RPF早期的误诊率高达26.8%

(115/429), 本病例于 2006 年外院住院期间未能就右肾及输尿管积水的病因做出正确的诊断, 误诊为右侧输尿管癌后错误地行右侧肾输尿管切除术, 造成了患者不必要的损失, 究其根本原因就在于忽视发生 RPF 的可能及缺乏对该疾病的认识。本

病例资料显示: 肾盂输尿管逆行造影、CT 及 MRI 检查对诊断腹膜后软组织异常、肾积水和输尿管扩张均有价值。对于肾积水的患者, 必需完善检查, 明确诊断后方可决定治疗方式, 这是我们对该病例的治疗体会。

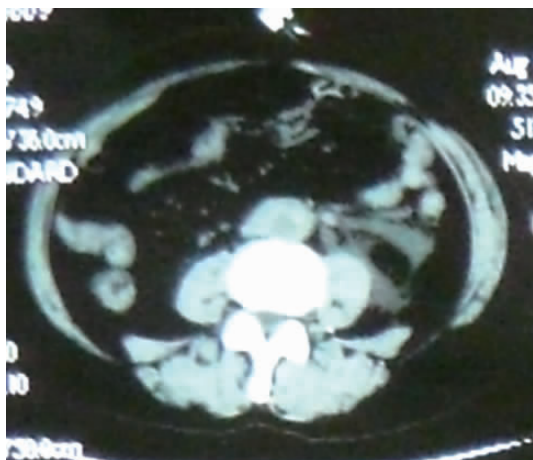


图 1 腹部 CT 平扫
Fig.1 Abdominal CT scan



图 2 肾盂输尿管逆行造影(左)
Fig.2 Retrograde angiography of renal pelvis and ureter (Left side)

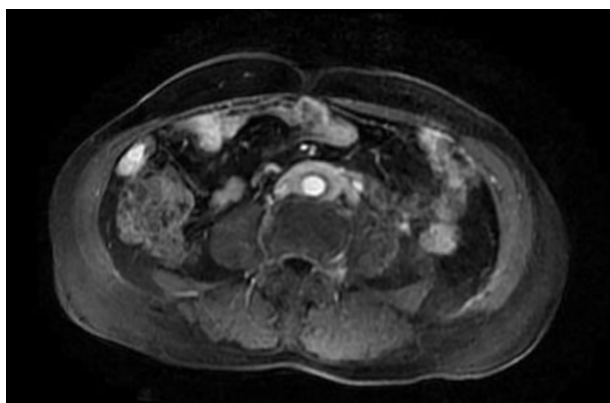


图 3 腹部 MRI 增强
Fig.3 Abdominal MRI enhanced scan

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转开腹,是避免胆漏等并发症发生的关键^[16-17]。对于 LPC 术后发生的胆漏,保持通畅的引流非常重要,本组 1 例术后出现胆漏,腹腔引流管胆汁样渗液第一天引流量为 100-250 mL,3 天后自止,未经特殊治疗,可能为急性炎症渗出以及胆囊管缝合不严密所致,我们常规放置引流管于网膜孔,位置一定放置妥当。

结果显示,腹腔镜胆囊部分切除术治疗慢性萎缩性胆囊炎是安全可行。如果术中发现胆管变异畸形,胆囊动脉异常出血以及萎缩性胆囊炎合并 Mirizzi 综合征,腹腔镜下处理非常困难,为了手术的安全,应及时中转开腹切除。同传统剖腹胆囊部分切除一样,腹腔镜胆囊大部分切除术仅用于标准胆囊切除施行困难或不能耐受的慢性萎缩性胆囊炎,尤其适用于基层医院,但严格掌握手术指征,不能作为常规应用^[18-20]。

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