

腹膜后局限性 CastLeman 病 4 例报告并文献复习

杨冬园¹ 周四维²

摘要 目的: 报告偶然发现的 4 例腹膜后局限性 CastLeman 病, 并结合文献复习, 以提高对该疾病的认识及诊疗水平。方法: 回顾性分析 4 例术前未能明确诊断的后腹膜占位性病变病例, 经手术切除, 术后病理证实为较罕见的局限性 CastLeman 病。结果: 4 例均经手术完整切除肿瘤, 未行辅助治疗, 随访 7 个月~4 年 2 个月, 4 例患者均无瘤存活, 未见肿瘤复发。结论: 腹膜后 CastLeman 病临床症状不典型, 术前实验室及影像学检查无特异性, 常易误诊。手术探查切除肿瘤为首选治疗方法, 疗效较好, 最终明确诊断有赖于病理学检查。

关键词: CastLeman 病; 诊断; 治疗

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Retroperitoneal local CastLeman's disease: 4 case reports and literature review

YANG Dongyuan¹ ZHOU Siwei²

(¹Department of Urology, Sichuan Province No. 4 People's Hospital, Chengdu, 610016, China; ²Department of Urology, Tongji Hospital, Tongji Medical College, Huazhong University of Science and Technology)

Corresponding author: YANG Dongyuan, E-mail: yangdy6611@163.com

Abstract **Objective:** We reported the incidentally found 4 cases of retroperitoneal the local CastLeman's disease and undertook a literature review to improve the diagnosis and treatment of the disease. **Method:** Retrospectively analyzed 4 cases with retroperitoneal mass which failed to confirm the diagnosis before surgery. After surgical resection, the pathological results were local CastLeman's disease. **Result:** All 4 cases received complete surgical resection of the tumor. No adjuvant therapy was administrated. Follow-up ranged from 4 months to 4 years and 2 months. No recurrence was observed. **Conclusion:** Retroperitoneal CastLeman's disease has no typical clinical findings. It may pose a diagnose dilemma because of its lack of specific lab test and distinguish radiographic features. Surgical exploration to remove the tumor is the preferred treatment with fair outcome. Eventually confirming diagnosis depends on the pathological examination.

Key words CastLeman disease; diagnosis; treatment

CastLeman 病(CastLeman's disease, CD)是一种罕见的淋巴增生性疾病,其不同于肿瘤或转移瘤的淋巴结增生,又称巨大淋巴结增生症、滤泡淋巴结网状内皮细胞瘤、淋巴错构瘤等,一直未能统一,该瘤由 CastLeman 于 1954 年首次报告,故统称 CastLeman 病。该病的发病原因不明,由于临床症状不典型,实验室及影像学检查均无特异性,因此临床上很容易误诊^[1,2],我科 2003~2010 年收治腹膜后占位性病变 4 例经手术切除后病理证实为局限性 CastLeman 病。为加强对该疾病的认知,获取经验,提高诊疗水平,现结合文献复习报告如下。

1 资料与方法

1.1 临床资料

2003~2010 年,我科收治 4 例常规体检中偶然发现的腹膜占位性病变患者,均为男性,年龄 44~52 岁,平均 45 岁。身体健康,均否认有重要疾病及遗传性疾病,无任何相关临床症状。3 例 B 超发现左肾上极区域内低回声肿块,其内部回声欠均

匀,边界欠清晰,体积分别为 3.5 cm×4.5 cm、3.5 cm×5.0 cm、4.0 cm×5.2 cm,1 例 B 超发现左肾肾门区一低回声肿块,内部回声欠均匀,边界尚清晰,体积约 2.4 cm×4.5 cm。4 例肿块内均可见少许散在的点状或片状钙化灶。4 例均行 CT 平扫加增强扫描,肿块位置及大小同 B 超发现,CT 值平均 44 HU,增强后为 76 HU,4 例肿瘤内部密度较高,尚均匀,肿瘤边界基本清晰(图 1~4),肿块无局部浸润,腹膜后均未发现增大的淋巴结,亦未发现远处转移灶。B 超及 CT 均报告为“腹膜后占位性病变,性质待查,考虑肾上腺无功能性嗜铬细胞瘤可能(?)”。实验室检查:4 例患者肝、肾功能、水电解质、血常规均正常;血皮质醇、促肾上腺皮质激素、醛固酮、血浆肾上腺素、血浆去甲基肾上腺素均在正常范围内。

1.2 治疗方法

4 例均在全麻下行剖腹探查术,术中见肿瘤均有完整包膜,与周围组织无紧密粘连,分界较清,肿块呈实性,质地柔韧,易于分离,故均完整摘除肿瘤。纵行剖开肿瘤,见切面呈鱼肉样,乳白色,质地

¹四川省第四人民医院泌尿外科(成都,610016)

²华中科技大学同济医学院附属同济医院泌尿外科
通信作者:杨冬园, E-mail: yangdy6611@163.com



图 1 病例 1CT 图



图 4 病例 4CT 图

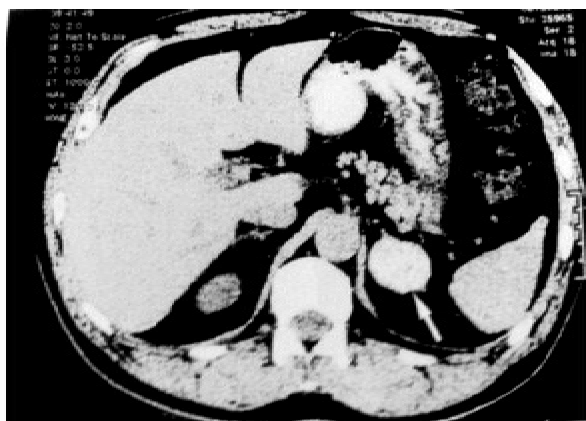


图 2 病例 2CT 图

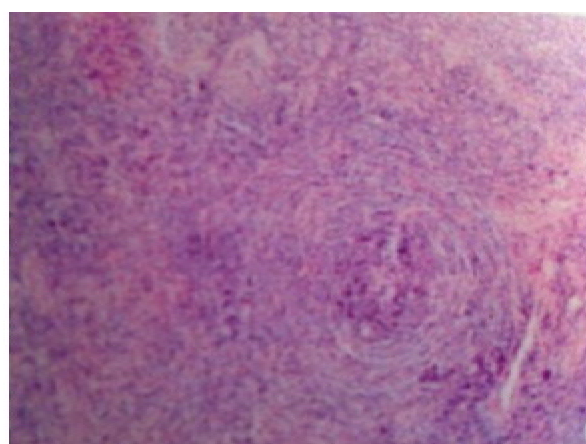


图 5 病理诊断:腹膜后透明血管型 CastLeman 病



图 3 病例 3CT 图

尚均匀,未见灶性坏死灶,似肉瘤样变。

2 结果

术后病理检查示:4 例病理标本皮髓质内充满大量均匀分布的滤泡及小淋巴细胞,呈环型整齐排列,多层分布呈“葱皮”样,未见核分裂,滤泡间隙内充满增生小血管,管壁呈玻璃样变,病理诊断:腹膜后透明血管型 CastLeman 病(图 5)。免疫组化结果:CD₃(+),CD₂₀(+),Bcl-2(-),CD₂₁(+),CD₃₅

(+),未给予处理。术后随访 7 个月~4 年 2 个月,均未见肿瘤复发,无瘤存活。

3 讨论

CastLeman 病较罕见,多在常规体检或因其他疾病检查时偶然发现,因其临床症状不典型,实验室检查及影像学检查无特异性,因此常易误诊,且多误诊为肾上腺嗜铬细胞瘤^[3],本组 4 例均误诊可疑肾上腺无功能性肾上腺嗜铬细胞瘤,术后病理检查明确诊断为腹膜后透明血管型 CastLeman 病。

根据病理学特点,CastLeman 病可分为 3 型:①透明血管型,最为常见,约占全部病例的 90%,主要由增生的淋巴细胞及丰富的血管组成^[4]。②浆细胞。③混合型。在最常见的透明血管型 CastLeman 病中,临床上又可分为局限性 CastLeman 病(Localized CastLeman's disease, LCD)和多中心型 CastLeman 病(Multicentric CastLeman's disease, MCD),均具有淋巴结结构基本完整、滤泡增生及血管丰富的特点。LCD 多无自觉症状,病理表现的特点是:淋巴结滤泡内生发中心呈萎缩或退行性变,滤泡树突状细胞断裂、区间可见除树突状细胞外,还可见成纤维网状内皮细胞、小 T 淋巴

细胞等,并可见小血管呈玻璃样变并穿过生发中心,生发中心周围环绕增宽的“葱皮”样套区,呈同心圆样排列,此型多属良性进程^[5],本组4例均属此型。MCD中浆细胞通常有明显的严重症状,如发热、乏力、食纳不佳、消瘦、肝脾肿大、贫血、高免疫球蛋白血症,常伴有痛性、多灶性淋巴结病,本组未发现该类型 Castleman 病。有较多文献指出, Castleman 病与疱疹病毒8(HHV-8)感染有关,HHV-8的基因组中有IL-6基因类似物^[6],发生 Castleman 病的HIV感染者,HHV-8的检测率可高达100%,可见免疫缺陷或免疫调节异常是 Castleman 病发生的重要因素^[7]。另外,有文献报告,炎性介质IL-6、表皮细胞生长因子、 α -干扰素等,在 Castleman 病的病理发生中也起着重要的作用^[8]。

一般来讲,LCD经手术切除后,疗效十分满意,若因其他原因不能手术或仅仅只能行部分切除的减瘤手术,可采用放射治疗,其有效率也可达72%^[9],国外学者报告13例LCD,手术完整切除肿瘤10例,3例行减瘤手术,术后辅以放疗,随访9~76个月,无一例死亡^[10]。本组4例均为LCD,手术完整切除肿瘤,随访7个月~4年2个月,均无瘤存活。关于MCD,文献报告^[8],手术切除肿瘤只能暂时缓解症状,应采用综合治疗,化疗(如CHOP方案),辅以糖皮质激素,对大多数MCD有效;利妥昔单抗、托珠单抗也有明显的缓解症状,缩小受累淋巴结的作用^[11]。

参考文献

- 1 赵晨晖,周文龙,张志伟,等.腹膜后局限型 Castleman 病2例及文献复习[J].现代泌尿生殖肿瘤杂志,2011,3(2),77-79.
- 2 宋科,蓝儒竹. Castleman 病1例报告[J].现代泌尿生

殖肿瘤杂志,2011,3(6),360-360.

- 3 肖峻,陈凌武,郑伏甫.腹膜后局限性 Castleman 病误诊嗜铬细胞瘤4例并文献复习[J].现代泌尿生殖肿瘤杂志,2012,4(1):10-12.
- 4 Powles T, Stebbing J, Montoto S, et al. Rituximab as retreatment for rituximab pretreated HIV-associated multicentric Castleman disease[J]. Blood, 2007, 110: 4132-4133.
- 5 Chen C H, Liu H C, Tung K Y, et al. Surgical outcome of superficial and deep Castleman disease[J]. ANZ J Surg, 2007, 77: 339-343.
- 6 Kawabata H, Tomosugi N, Kanda J, et al. Anti-interleukin 6 receptor antibody tocilizumab reduces the level of serum hepcidin in patients with multicentric Castleman's disease[J]. Haematologica, 2007, 92: 857-858.
- 7 Beck J T, Hsu S M, Wijdenes J, et al. Brief report: alleviation of systemic manifestations of Castleman's disease by monoclonal anti-interleukin-6 antibody[J]. N Engl J Med, 1994, 330: 602-605.
- 8 Dham A, Peterson B A. Castleman disease[J]. Curr Opin Hematol, 2007, 14: 354-359.
- 9 Chronowski G M, Ha C S, Wilder R B, et al. Treatment of unicentric and multicentric Castleman disease and the role of radiotherapy[J]. Cancer, 2001, 92: 670-676.
- 10 Bestawros A, Michel R, Séguin C, et al. Multicentric Castleman's disease treated with combination chemotherapy and rituximab in four HIV-positive men: a case series[J]. Am J Hematol, 2008, 83: 508-511.
- 11 Matsuyama M, Suzuki T, Tsuboi H, et al. Anti-interleukin-6 receptor antibody (tocilizumab) treatment of multicentric Castleman's disease[J]. Intern Med, 2007, 146: 771-774.

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- 2 Jemal A, Siegel R, Ward E, et al. Cancer statistics [J]. CA Cancer J Clin, 2008, 58: 71-96.
- 3 叶定伟.前列腺癌的流行病学和中国的发病趋势[J].中华外科杂志,2006,44(6):362-364.
- 4 马斌荣. SPSS for Windows Ver. 11.5 在医学统计中的应用[M].北京:北京科学出版社,2004:57-141.
- 5 Tombran-Tink J, Johnson L V. Neuronal differentiation of retinoblastoma cells induced by medium conditioned by human RPE cells [J]. Invest Ophthalmol Visual Sci, 1989, 30, 1700-1707.
- 6 Tombran-Tink J, Chader G G, Johnson L V. PEDF: a pigment epithelium-derived factor with potent neuronal differentiative activity [J]. Exp Eye Res, 1991, 53: 411-414.

- 7 Halin S, Wikstrom P, Rudolfsson S H, et al. Decreased pigment epithelium-derived factor is associated with metastatic phenotype in human and rat prostate tumors [J]. Cancer Res, 2004, 64: 5664-5671.
- 8 Doll J A, Stellmach V M, Bouck N P, et al. Pigment epithelium-derived factor regulates the vasculature and mass of the prostate and pancreas[J]. Nat Med, 2003, 9: 774-780.
- 9 Byrne J C, Downes M R, O'Donoghue N, et al. 2D-DIGE as a strategy to identify serum markers for the progression of prostate cancer [J]. J Proteome Res, 2009, 8: 942-957.

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