孤立性肾髓外浆细胞瘤 1 例报告并文献复习

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[摘要] **目的**:提高肾脏浆细胞瘤的诊治水平。**方法**:肾脏浆细胞瘤患者 1 例,女,55 岁。体检 B 超偶然发现左肾前唇实质内可见一范围约 4.7 cm×2.6 cm 的低回声包块;CT 检查左肾见一大小约 3.0 cm×4.0 cm×3.5 cm 低密度影;增强 CT 示左侧肾脏占位性病变,考虑肾肿瘤性病变不除外。行后腹腔镜左肾根治性切除术。**结果**:病理报告示左肾上极可见囊实性肿物,大小约 4.0 cm×3.0 cm,腔内含淡黄色的浆液性液体,切面淡黄色,其余肾脏皮髓质分界清楚。镜下肿瘤细胞有大片浆细胞样改变,密集成片,细胞大小不一,多为单核,也可见双核细胞,细胞核为圆形或椭圆形,位于细胞一端,染色质粗糙,呈"车辐"状排列,胞浆成双嗜性,核与胞浆之间可见"月牙状"淡染区。病理诊断:髓外浆细胞瘤。术后随访 6 个月未见复发和转移。**结论**:髓外浆细胞瘤是一种发生于骨髓之外的恶性肿瘤,此类患者没有多发骨髓瘤的临床表现,发病率低,组织活检发现单片状浆细胞并且未发现任何多发性骨髓瘤的迹象是诊断的惟一途径;浆细胞瘤对放疗相对敏感,外科手术和放疗联合治疗效果最好。

[关键词] 肾脏浆细胞瘤;临床研究;免疫组织化学
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Isolated extramedullary plasmacytoma of the kidney: a case report and literature review

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Abstract **Objective**: To optimize the diagnosis and treatment of plasmacytoma of the kidney. **Method**: The clinical data of 1 case of plasmacytoma of the kidney including manifestation, imaging, pathology and therapy were reviewed. The female patient aged 55 was incidentally found that a low-echo mass measuring 4.7 cm×2.6 cm was in left kidney by ultrasound detection. Computer tomography (CT) and enhanced CT examination revealed an occupying lesion measuring with low density 3.0 cm×4.0 cm×3.5 cm in left kidney. The patient then underwent radical nephrectomy. **Result**: The pathological characteristics were seen as follows. A solid-cystic mass measuring 4.0 cm×3.0 cm was detected at the upper pole of left kidney, with yellow serous fluid inside and yellowish cross-section of the mass. The boundary between cortex and medullary is clear. Microscopically, intense arrangement of plasmocyte-like tumor cells with varying size was seen. These cells presented mono or binucleated, with round or oval nucleus locating in the cell at one end. The rough chromatin demonstrated spoke-like arrangement, and the cytoplasm exhibited dual tropism. Crescent-shaped olistherozone were seen between nucleus and cytoplasm. The pathological diagnosis was extramedullary plasmacytoma. A 6-month follow-up displayed no sign of relapse or metastasis. **Conclusion**: Extramedullary plasmacytoma is a rare malignant neoplasm typically arising outside bone marrow. Patient with this kind of disease show no clinical manifestation of multiplemyeloma. The diagnosis relies on the finding of single-sheet-like plasma cells in biopsy detection as well as no sign of multiple myeloma. Due to its sensitivity to radiotherapy, a combination of surgery and radiotherapy could be a promising treatment for extramedullary plasmacytoma.

Key words plasmacytoma of the kidney; clinical research; immunohistochemistry

髓外浆细胞瘤(extramedul-lary plastmacytoma, EMP)是罕见发生于骨髓外软组织内的浆细胞瘤,不伴有多发性骨髓瘤的全身特征表现。EMP 男女发病比例为 3∶1,多集中于 50～70 岁之间[1]。孤立性肾髓外浆细胞瘤极为罕见,我院于 2011 年 7 月收治 1 例发生在肾上的髓外浆细胞瘤患者,现报告如下。

1 病例报告

患者,女,55 岁,查体发现左肾上极肿物 1 周余入院,查体未见阳性体征。既往糖尿病病史 5 年余,给予皮下注射胰岛素控制血糖,血糖控制比较稳定。超声示左肾前唇实质内可见一范围约 4.7 cm×2.6 cm 的低回声包块;CT 检查左肾见一大小约 3.0 cm×4.0 cm×3.5 cm 低密度影,CT 值约 20 HU(图 1A);增强 CT 示左侧肾脏占位性病变,

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CT 值约 27 HU(图 1B)。随后我们为患者进行了后腹腔镜左肾根治性切除术,术后大体标本见左肾前唇可见大小约 4.0 cm×3.0 cm 囊实性肿物,中心腔内含淡黄色的浆液性液体,周边切面淡黄色,其余肾脏皮髓质分界清楚,淋巴结未受到肿瘤浸润(图 1C)。患者术后恢复顺利。切除标本的组织病理学显示,高倍镜(400×)肿瘤细胞有大片浆细胞样改变,密集成片,细胞大小不一,多为单核,也可见双核细胞,细胞核为圆形或椭圆形,位于细胞一端,染色质粗糙,呈"车辐"状排列,胞浆成双嗜性,核与胞浆之间可见"月牙状"淡染区(图 1D)。

免疫组织化学:浆细胞抗体(+),髓过氧化物酶(+),PAS 染色法(+),波形蛋白(+),CD38(+),CD45(−),CD20(−),CK(−),κ 链(+),λ 链(−),淀粉样物质(+)(图 2)。病理报告为左肾髓外浆细胞瘤。术后查尿本周蛋白阴性,血清蛋白电泳未见 M 带,骨髓活检也未见异常。

以上都表明本例为原发性肾髓外浆细胞瘤。患者术后每 3 个月复查 1 次,包括体检、胸部 X 线片、B 超、CT、本周蛋白尿及血清蛋白电泳试验等检查。患者在半年内的随访中无肿瘤复发迹象。

2 讨论

根据 WHO(2001)标准,浆细胞源性肿瘤分为多发性骨髓瘤浆细胞瘤和浆细胞瘤。浆细胞瘤包括孤立性骨浆细胞瘤和孤立性髓外浆细胞瘤[2]。髓外浆细胞瘤被定义为发生于骨髓外的由浆细胞分化的单克隆增殖细胞构成的孤立性肿瘤。髓外浆细胞瘤为不常见肿瘤,其世界范围内的发生率为 3/100 000 人口。孤立性髓外浆细胞瘤在所有浆细胞肿瘤中所占比例小于 3%～10%[3]。髓外浆细胞瘤更好发于男性,男女发生比例约为 3∶1,中位发病年龄为 55 岁,与多发性骨髓瘤的中位发病年龄相比大约早 10 年[4～6]。

EMP 80% 发生于上呼吸道,以鼻腔、鼻窦和鼻咽部最常见,大约占该部位所有类型肿瘤的 3%～4%[7]。EMP 发生于肾脏更为罕见,原发肾髓外

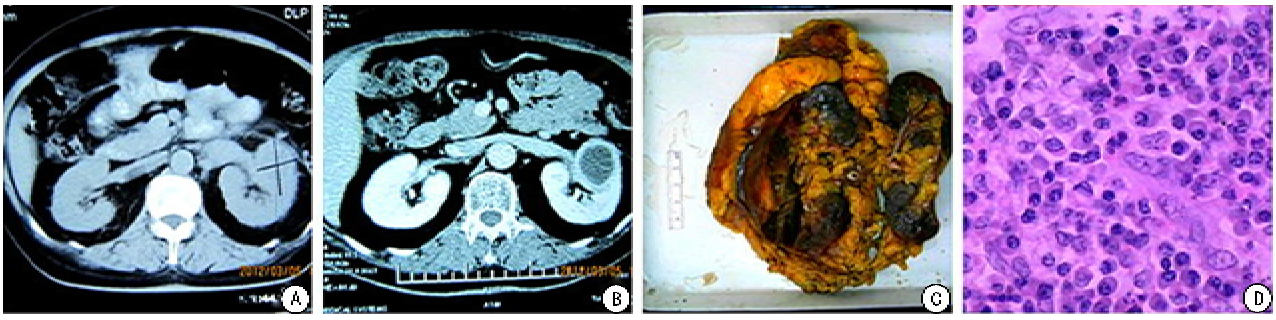
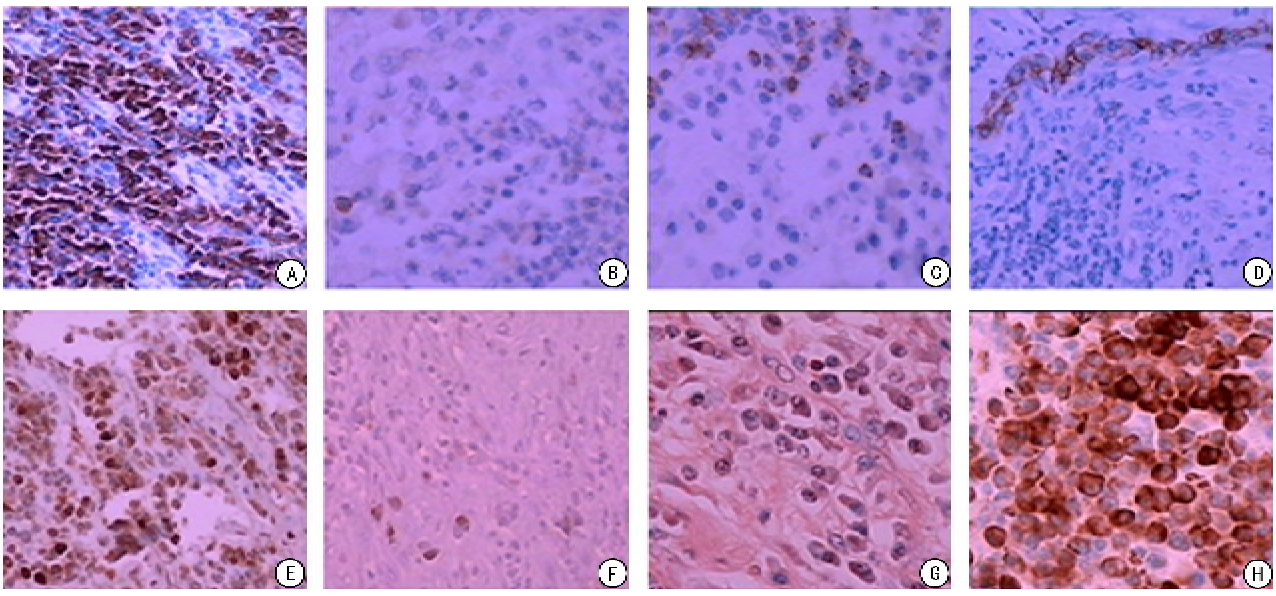


图 1 A、B:双肾 CT 平扫及强化;C:个体标本;D:肿瘤组织病理切片(HE,400×)



A:CD38 表达阳性(200×);B:CD45 表达阴性(400×);C:CD20 表达阴性(400×);D:CK 表达阴性(200×);E:κ 轻链表达阳性(200×);F:λ 轻链表达阴性(100×);G:淀粉样物质表达阳性(400×);H:浆细胞抗体表达阳性(400×)

图 2 肿瘤组织免疫组织化学

浆细胞瘤在术前凭影像学不能与其他肾肿瘤相鉴别，组织活检发现单片状浆细胞并且未发现有任何多发性骨髓瘤的临床迹象是诊断的惟一途径。一旦组织学诊断出浆细胞瘤，患者需要进行一系列检查来排除其他系统的恶性浆细胞瘤受累（多发骨髓瘤）[8]。浆细胞瘤的诊断应要求确认浆细胞表达 CD38 显型与确定细胞质 KAPPA 或 Lambda 轻链[9]。骨髓活检、尿本周蛋白和血清蛋白电泳可以排除系统性疾病并证实原发性肾浆细胞瘤的诊断。有报道称 CD45 与 CD20 表达阴性也是浆细胞肿瘤的显著特征[10]。本例患者所有免役组化结果完全符合上述报道所描述，故本病例诊断无误。

肾髓外浆细胞瘤的治疗方案选择包括手术、化疗和放疗，单独或联合治疗[8]。浆细胞瘤通常对放疗非常敏感，手术与放疗联合疗法是一种已被接受的基于对可治愈性病变的处理方法，事实上联合治疗可以得到最好的治疗效果。局部治疗的最佳照射剂量为 40～50 Gy（取决于肿瘤大小），在 4～6 周内完成[11]。化疗适用于未分化的髓外浆细胞瘤、复发或难治性肿瘤患者[12]。在我们的病例中，我们为患者进行了后腹腔镜下根治性肾切除术，组织学结果显示为肾浆细胞瘤，因为术后患者及家属暂不接受放疗，故未给予放疗。但有病例报道肾髓外浆细胞瘤术后未给予放疗，半年后局部复发，因此，肾浆细胞瘤手术切除后仍需辅助治疗。

肾髓外细胞瘤预后相对较好，10 年总体生存率为 70%，发展为多发性骨髓瘤后 10 年总体生存率较孤立性骨浆细胞瘤差，为 11%～30%不等[5]。虽然髓外细胞瘤预后相对较好，但仍有 30%的患者有原位的复发，40%的患者有转移[13]。此外，虽然髓外浆细胞瘤被认为和多发骨髓瘤属于不同类型的疾病，但报道中有 17%～33%的病例进展为了后者，因为这种进展严重影响预后，所以临床过程中应严密的关注患者[14]。所以肾髓外细胞瘤术后复查也显得至关重要，CT 异常和血清蛋白的升高尤其是 M 蛋白，可以作为标记物，监测这些指标是很有必要的。我们的病例在半年的随访中未见局部复发及系统性病变。

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