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· 病例报告 ·

肝脏原发性 Rosai-Dorfman 病一例并文献复习

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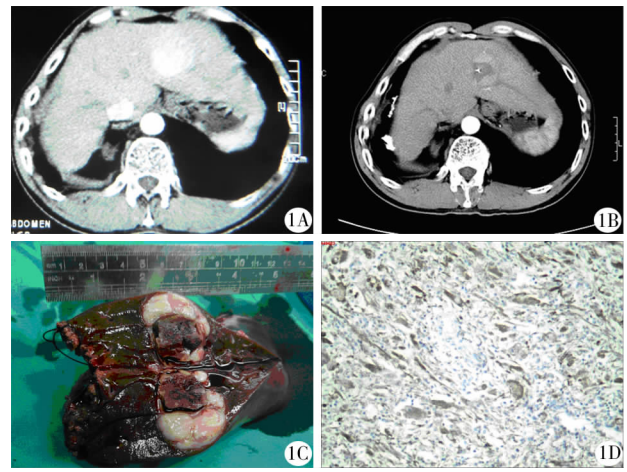
Rosai-Dorfman 病是一种特发性伴巨大淋巴结的窦组织细胞增生症。该病发生于淋巴结外者少见,发生在肝脏的罕见。本中心最近收治 1 例肝脏原发性 Rosai-Dorfman 病,手术治疗效果良好,报道如下。

患者,男,50 岁。因"发现肝脏占位 2 个月余"于 2014 年 11 月 17 日入院。12 年前因"胃癌"在当地医院行胃癌根治术,术后未行化疗。否认乙型病毒性肝炎、饮酒等肝病病史。入院查体未见明显阳性体征。

患者 2 个多月前在外院行上腹部 CT 检查提示左肝实性占位,考虑肝癌(图 1A)。乙肝标志物及 AFP、CEA 结果均正常。于 2014 年 9 月 18 日在外院行左肝肿瘤微波消融治疗。术后 1 个月复查 CT 提示左肝肿瘤部分存活,为进一步诊治收住我科。我院上腹部 CT 检查提示肿瘤残留或复发可能(图 1B)。于 2014 年 11 月 25 日行左半肝切除术(图 1C),术后恢复顺利出院。

我院病理科诊断意见为肝原发性 Rosai-Dorfman 病(图 1D)免疫组化 S-100(+),Vim(+),余均阴性。中山大学附属肿瘤医院会诊意见也考虑为肝脏 Rosai-Dorfman 病。现术后随访 2 个月余,患者一般情况良好,无肿瘤复发。



1A:微波射频治疗前上腹部 CT 结果,动脉期病灶明显强化;1B:上腹部 CT 动脉期结果,动脉期边缘强化;1C:左半肝外叶切除标本图片,见左肝肿瘤周边质硬,肿瘤中央坏死灶,包膜完整,切缘距肿瘤边缘约 4 cm;1D:S-100 免疫组化的结果,胞浆染色阳性(×100)

图 1 患者男,50 岁,诊断:肝脏原发性 Rosai-Dorfman 病

讨论 Rosai-Dorfman 病于 1969 年首先被 Rosai 和 Dorfman 报道。该病发生于淋巴结外者少见,多为个案报道。根据报道该病可发生于中枢神经系统[1]、眼眶[2]、鼻[3]、上呼

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吸道[4]及支气管[5]等部位,发生于肝脏者罕见。Lauwers 等[6]报道 5 例、Di Tommaso 等[7]和 Maheshwari 等[8]分别报道 1 例累及肝脏的 Rosai-Dorfman 病。检索国内文献未见肝脏原发性 Rosai-Dorfman 病的报道。由于临床罕见加之临床表现缺乏特异性,故肝脏原发性 Rosai-Dorfman 病的术前诊断十分困难。本病的 CT 表现与原发性肝癌相似,最终诊断需结合病理学免疫组化方可作出。本例患者射频肿瘤残留,后行左半肝切除术,术后病理学诊断为肝原发性 Rosai-Dorfman 病。其病理诊断依据如下:(1)纤维间质见大量胞体巨大,胞质丰富、核大、有明显嗜酸性核仁的大细胞;(2)可见吞噬小淋巴细胞现象;(3)大量淋巴浆细胞浸润;(4)免疫组化 S-100(+),Vim(+),其余均为阴性。其中免疫组化特征是 Rosai-Dorfman 病的特征性表现,其与原发于淋巴结内的 Rosai-Dorfman 病结果一致。另外结合腹部 CT 及病理结果,该病例不支持胃癌术后转移到肝脏的诊断。

Rosai-Dorfman 病通常被认为是一种自限性良性疾病。由于肝脏原发性 Rosai-Dorfman 病临床罕见、诊断困难,故是否需行手术切除尚未定论。本例患者手术切除后已随访 2 个月余,未见肿瘤复发,但仍需长期随访,以进一步明确手术治疗的效果。

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