

成人右侧侧脑室原发性上皮样肉瘤 1 例

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【摘要】 上皮样肉瘤(ES)是一种少见且具有高度侵袭性的软组织恶性肿瘤,其特征为生长缓慢的无痛性肿块,通常发生在四肢远端,常常误诊为良性疾病;但局部复发倾向高,易发生淋巴和血液转移扩散,预后较差。中枢神经系统原发性ES极为罕见,临床表现及影像学表现无特异性,术前诊断困难,容易造成误诊而延误病情。本文报道 1 例 60 岁男性,因记忆力减退 1 周入院,入院体格检查未见阳性体征;颅脑 MRI 发现右侧侧脑室三角区大小约 1.8 cm×1.6 cm 结节状长 T₁、稍长 T₂ 信号影,增强后呈明显强化。完善术前准备,经颞叶皮层入路手术全切除右侧侧脑室三角区病灶,术后病理诊断为 ES。术后未接受放化疗,术后 1.5 年随访,肿瘤无复发及远处转移,无神经系统阳性体征。

【关键词】 侧脑室肿瘤;上皮样肉瘤;显微手术

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A primary epithelioid sarcoma of the right lateral ventricle in an adult

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【Abstract】 Epithelioid sarcoma (ES) is a rare and highly aggressive malignant soft tissue neoplasm characterized by indolent, painless growths typically manifesting in the distal extremities of the limbs. These lesions are frequently misdiagnosed as benign entities. However, ES exhibits a pronounced predilection for local recurrence and propensity for lymphatic and hematogenous dissemination, resulting in an unfavorable prognosis. Primary ES originating within the central nervous system (CNS) is exceedingly uncommon, with clinical and radiological features lacking specificity, thereby posing diagnostic challenges that may lead to misidentification and delayed intervention. This report details a 60-year-old male presenting with a one-week history of declining memory function. Physical examination revealed no discernible abnormalities. Brain MRI showed a nodular lesion measuring approximately 1.8 cm × 1.6 cm in the trigone of the right lateral ventricle, displaying prolonged T₁ and slightly protracted T₂ signal characteristics with conspicuous enhancement following contrast administration. Following thorough preoperative preparations, a complete resection of the lesion in the right lateral ventricle was achieved via the temporal lobe cortex approach. The postoperative pathological diagnosis confirmed ES. The patient did not undergo adjuvant radiotherapy or chemotherapy postoperatively. A follow-up was conducted 1.5 years after the surgery, and no tumor recurrence or distant metastasis was observed. Additionally, there were no positive neurological signs.

【Key words】 Ventricular tumors; Epithelioid sarcoma; Microsurgery

上皮样肉瘤(epithelioid sarcoma, ES)是一种罕见的恶性间充质肿瘤^[1], 占有肉瘤的 1% 以下^[2], 其特征为生长缓慢的无痛性肿块, 通常发生在四肢远端^[3, 4]。准确诊断 ES 具有挑战性, 常常误诊为良性疾病; 但局部复发倾向高, 易发生淋巴和血液转移扩散, 预后较差^[5]。颅内原发性 ES 极为罕见^[6]。本文报道 1 例侧脑室原发性 ES, 为临床提供参考。

1 病例资料

60 岁男性, 因记忆力减退 1 周于 2022 年 6 月 16 日入院, 既往体健, 无家族遗传史。入院体格检查未见神经系统阳性体征。颅脑 MRI: 右侧侧脑室三角区见大小约 1.8 cm×1.6 cm 结节状长 T₁、稍长 T₂ 信号影, 其内信号不均匀, FLAIR 像呈稍高信号, 形态不规则, 与侧脑室壁关系密切(图 1A~C), 增强

后呈明显强化, 邻近侧脑室三角区周围脑实质呈毛刺状、星芒状强化, 沿右侧侧脑室管膜亦见条状强化(图 1D)。MRS: 右侧侧脑室三角区病灶 NAA 峰减低, Cho 峰升高, Cr 峰未见明显异常。MRA: 颈内动脉颅内段、大脑中动脉、大脑前动脉、基底动脉及大脑后动脉主干及其主要分支无明显狭窄及分支减少。DTI: 右侧大脑半球脑白质纤维束较对侧减少, 较对侧稀疏, 双侧分布不对称。PET/CT: 右侧侧脑室三角区见结节状软组织密度影, 伴 FDG 摄取增高, 最大 SUV 为 9.4; 骨骼未见骨质破坏征象, 未见异常 FDG 浓聚灶, 考虑为原发性病变。排除手术禁忌后, 经颞叶皮层入路手术切除右侧三角区病变。术中切开蛛网膜沿边界逐步分离、探查, 可见肿瘤位于三角区, 呈灰白色鱼肉样, 质地偏硬, 瘤周组织黄变, 边界不清, 血供丰富, 镜下全切除肿瘤。术后复查颅脑 CT 示中线偏移较前好转(图 1H)。术后病理(图 1E~G): 右侧侧脑室恶性肿瘤, 免疫组化结果: 肿瘤细胞呈 CK(-), Vim(+), GFAP(-), IDH1(-), Olig-2(-), ATRX(局灶弱+), P53(野生型), MGMT(+), EMA(局灶+), PTEN(局灶弱+), EGFR(-), S100(-), PR(-), SSTR2(-), Nestin(部分+), CD31(局灶+), ERG(-), CD34(-), FVIII(-), E-cd(-), INI-1(-/+), CK19(-), CK7

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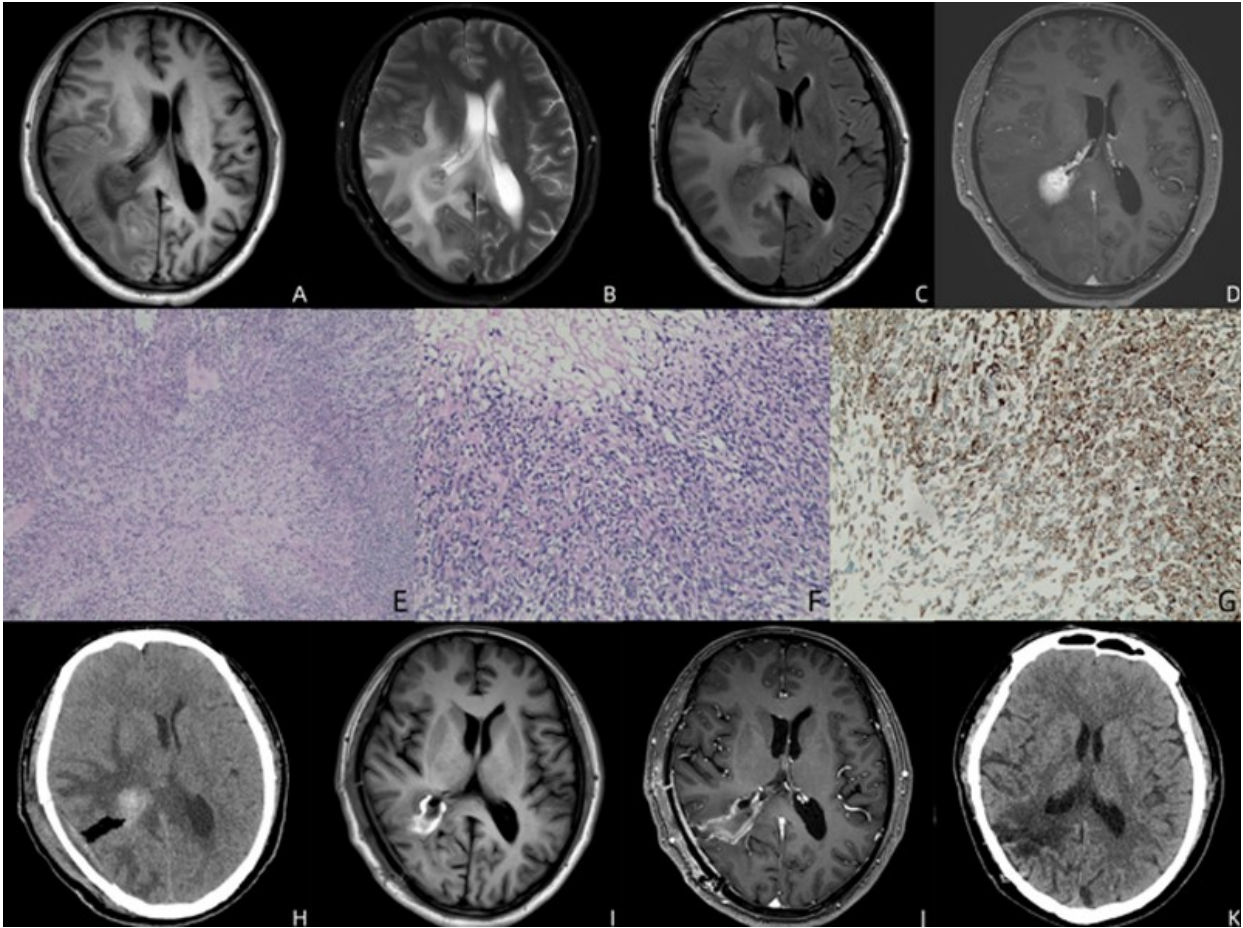


图1 右侧侧脑室原发性上皮样肉瘤手术前后影像及术后病理表现

A、B. 术前颅脑 MRI T₁WI、T₂WI 示右侧侧脑室三角区大小约 1.8 cm×1.6 cm 结节状长 T₁、稍长 T₂ 信号影,其内信号不均匀;C. 术前颅脑 MRI T₂ Flair 示肿瘤呈稍高信号,形态不规则,与侧脑室壁关系密切;D. 术前 MRI 增强扫描示肿瘤明显强化,邻近侧脑室三角区周围脑实质呈毛刺状、星芒状强化,沿右侧侧脑室管膜亦见条状强化;E. 术后病理检查见散在凝固性坏死(HE,×40);F. 术后病理检查发现瘤细胞含丰富的嗜酸性细胞质及偏心性空泡状核,核仁明显(HE,×100);G. 术后 Vimentin 免疫组化染色呈阳性(×100);H. 术后 1 周复查颅脑 CT 见右侧侧脑室三角区周围脑组织仍有水肿,中线偏移程度较术前好转;I、J. 术后 1 个月复查 MRI 平扫+增强扫描示术区胶质增生,周围脑组织水肿明显消退,未见肿瘤复发征象;K. 术后 1 年复查颅脑 CT 示中线偏移不明显,无新增占位改变

Figure 1 Pre- and post-operative imaging, as well as postoperative pathological findings of a primary epithelioid sarcoma in the right lateral ventricle

A-B: Preoperative T₁-weighted and T₂-weighted MR images of the brain reveal a nodular lesion measuring approximately 1.8 cm×1.6 cm in the trigone of the right lateral ventricle, characterized by long T₁ and slightly longer T₂ signal intensity with heterogeneous internal signals. C: Preoperative T₂-FLAIR MR images demonstrate a tumor with slightly elevated signal intensity and irregular morphology closely associated with the lateral ventricle wall. D: Preoperative contrast-enhanced MR images show marked enhancement of the tumor, along with spiculated and star-shaped enhancement in the surrounding brain parenchyma around the trigone of the lateral ventricle, as well as linear enhancement along the membrane of the right lateral ventricular. E: Postoperatively, histopathological examination reveals scattered coagulative necrosis (HE, ×40). F: Histological analysis following surgery identified tumor cells containing abundant eosinophilic cytoplasm and eccentric vacuolated nuclei with prominent nucleoli (HE, ×100). G: Positive Vimentin immunohistochemical staining is observed (×100). H: One-week post-surgery follow-up CT scan indicates persistent edema around the right-sided trigone area, accompanied by improved midline shift compared to pre-surgery. I-J: One-month post-surgery follow-up MR flat scan + enhanced scan suggests gliosis in the operative area and significantly reduced edema in surrounding brain tissue without signs of tumor recurrence. K: One-year post-surgery follow-up CT demonstrated no significant midline shift or new lesion formation.

(-),CK5/6(-),Calretinin(-),P63(个别细胞弱+),Ki-67(约10%+),特殊染色结果:网染(网状纤维+)。结合免疫组化结果诊断为ES。术后未进行规律放化疗,仅口服丙戊酸钠缓释片预防癫痫。术后1个月复查头颅MRI未见肿瘤复发迹象(图1I、1J)。2023年9月21日复查颅脑CT(图1K):右侧侧脑室三角区占位术后改变,右侧侧脑室后角旁片状稍低密度软化灶,未见肿瘤复发及颅内占位效应。2023年12月5日随访,无神经系统无阳性体征。

2 讨论

侧脑室ES诊断较为困难,鉴别诊断尤为重要^[7]。根据文献报道,侧脑室三角区好发的肿瘤为脑膜瘤、脉络丛乳头状瘤、转移瘤^[8,9],也可见星形细胞瘤、室管膜瘤^[10]。①转移瘤年龄较大,颅内多发占位,大部分可以找到原发灶。②侧脑室良性脑膜瘤分叶较规则,不典型和恶性脑膜瘤呈不规则分叶状,MRI平扫呈等信号,信号较均匀,多呈显著均匀强化,

MRS 表现为 NAA 峰缺乏, Cr 峰下降, Cho 峰增高, Cho/Cr 呈显著升高, 可出现脑膜瘤相对特征性的 Ala 峰^[10]。③脉络丛乳头状瘤起源于脉络丛上皮, 发生在侧脑室多为儿童, 年龄不超过 10 岁, 囊变少见, 增强扫描呈明显强化, 由于肿瘤分泌脑脊液旺盛, 易引起交通性脑积水^[10]。④侧脑室三角区星形细胞瘤发病年龄较大, 囊变坏死多见, 增强后多为不均匀强化^[11]。⑤室管膜瘤 MRI 表现为 T₁WI 等或低信号, T₂WI 高信号, 由于囊变、钙化及出血, 信号大部分不均匀; 囊变区 T₂WI 和 FLAIR 呈高信号, 病灶多与侧脑室室壁广基相连, 沿脑室塑形生长是其特点^[12]。由于 ES 的病理组织形态学与多种其他病变类似, 因此明确诊断需依赖免疫组织化学染色等辅助手段^[2]。

ES 因具有侵袭力强、易复发等特点, 且常以较近侧继发病灶及最后发生转移。广范围的切除被认为是目前最好的治疗方法^[2,3]。因肿瘤与周围脑组织边界不清, 在保证神经功能的情况下, 全切除肿瘤手术难度较高。文献报道, ES 的 5、10 年生存率分别在 50%~70%、42%~55%。与预后有关的因素有: 肿瘤的部位, 肿瘤的大小, 年龄, 性别等^[13]。由于发病率低, 到目前为止, 尚无前瞻性研究来评价全身化疗的疗效。大多回顾性研究是针对原发性 ES。研究表明切除术后辅助化疗能一定程度上降低远端型 ES 的复发风险, 提高生存率^[14]。本文病例术后未接受放化疗, 术后随访 1.5 年, 仍存活且无原位复发及远处转移迹象。

总之, ES 是一种发病率很低的软组织恶性肿瘤, 无明显特异性表现, 诊断困难, 容易造成误诊而延误病情。原发性侧脑室三角区 ES 的发生率极低, 需与好发于侧脑室三角区的转移瘤、脑膜瘤、室管膜瘤、脉络丛乳头状瘤、星形细胞瘤相鉴别, 明确诊断仍依赖于病理、免疫组织化学染色等方法。

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【作者共贡献声明】: 罗登建负责收集病例资料、分析数据、撰写论文; 韩锋提供写作思路、修订论文; 杨明、杨华、万豪杰、黄睿、陈伟协助收集数据, 参与文章讨论, 提出修改意见。

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