

阴囊肿块切除术治疗阴囊腺样囊性癌： 一例病例报告

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摘 要

原发性皮肤腺样囊性癌(P_{ri}mary cutaneous adenoid cystic carcinoma, PCACC)是起源于皮肤附属器的罕见低度恶性肿瘤, 阴囊为极罕见发病部位, 临床极易误诊。本文报道1例60岁男性阴囊原发性腺样囊性癌患者, 患者阴囊无痛肿块病程4年, 初彩超误诊为附睾囊肿, 行局部肿物完整切除; 术后病理结合免疫组化确诊阴囊腺样囊性癌, 组织学以筛状、管状结构为主, 未见广泛实性区域。免疫组化肌上皮标志物p40、CK5/6弥漫阳性, 导管标志物CD10腔面阳性, CD117阴性。术后未行辅助放化疗, 截止至术后24个月随访, 患者未发现局部复发或远处转移的临床及影像学证据, 鉴于该肿瘤晚期复发的特性, 仍需进行长期乃至终身随访以监测其远期结局。结合现有文献总结本病临床、病理、免疫及分子特征, 阴囊皮下无痛结节需常规鉴别PCACC; 手术扩大切除为首选治疗, 伴神经侵犯、切缘阳性者推荐术后放疗; MYB基因重排为核心分子诊断指标, 该病复发潜伏期极长, 需终身随访。

关键词

皮肤腺样囊性癌, 阴囊, 免疫组化, MYB, 鉴别诊断

Treatment of Adenoid Cystic Carcinoma of Scrotum by Excision of Scrotal Masses: A Case Report

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Abstract

Primary cutaneous adenoid cystic carcinoma (PCACC) is a rare low-grade malignant tumor derived from cutaneous adnexa. The scrotum is an extremely rare primary site and easy to be misdiagnosed clinically. We report a 60-year-old male patient with primary scrotal PCACC who presented with a 4-year painless scrotal mass, initially misdiagnosed as epididymal cyst by ultrasonography. Complete tumorectomy was performed. Postoperative histopathology showed predominant cribriform and tubular architecture without extensive solid areas. Immunohistochemistry demonstrated diffuse positivity of myoepithelial markers p40 and CK5/6, luminal CD10 expression, and negative CD117. No adjuvant radiotherapy or chemotherapy was administrated. No clinical or imaging evidence of local recurrence or distant metastasis was found at 24 months postoperatively. Given the tumor's late-recurrence potential, continued long-term, even lifelong, follow-up is essential for monitoring ultimate outcomes. Combined with the available literature, we summarized the clinical, pathological, immunohistochemical and molecular features of PCACC. PCACC should be included in differential diagnosis for painless subcutaneous nodules of the scrotum. Wide surgical resection is the first-line therapy, and adjuvant radiotherapy is recommended for cases with perineural invasion or positive surgical margins. MYB gene rearrangement serves as a key molecular biomarker. Long-term lifelong follow-up is mandatory due to extremely long latent period of recurrence.

Keywords

Cutaneous Adenoid Cystic Carcinoma, Scrotum, Immunohistochemistry, MYB, Differential Diagnosis

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1. 病例介绍

1.1. 一般资料

患者男, 60 岁, 因左侧阴囊肿物渐进增大 4 年入院。4 年前偶然发现左侧阴囊皮下无痛硬结, 无瘙痒、破溃、出血, 无排尿不适、会阴部坠胀, 肿物缓慢增大, 无局部外伤、慢性炎症病史。既往无唾液腺、乳腺、外阴肿瘤病史, 无放化疗史, 无肿瘤家族遗传史。

1.2. 影像学检查

阴囊彩色多普勒超声: 左侧阴囊真皮层下实性小结, 内部见多发微小囊性腔隙; 附睾区见微小无回声灶, 超声初诊考虑附睾囊肿, 未重视皮肤附属器源性病变。完善全身浅表淋巴结彩超、胸部 CT 平扫, 未见淋巴结肿大及远处脏器转移病灶。

1.3. 手术与大体病理

局麻下行阴囊肿物完整切除术。大体标本: 灰粉色实性结节一枚, 大小 1.6 cm × 1.1 cm, 边界尚清, 切面质韧, 散在微小囊腔, 无出血、坏死, 与表层阴囊皮肤无粘连。镜下 HE 染色: 肿瘤细胞浸润真皮全层及皮下脂肪组织, 由导管上皮、肌上皮双相细胞构成, 呈典型筛状、管状排列, 囊腔内见 PAS 阳性嗜

酸性基底膜样物质；实性区域占比 < 5%，细胞异型性轻，核分裂稀少；局灶可见肿瘤细胞侵犯神经束，未见脉管癌栓(图 1)。

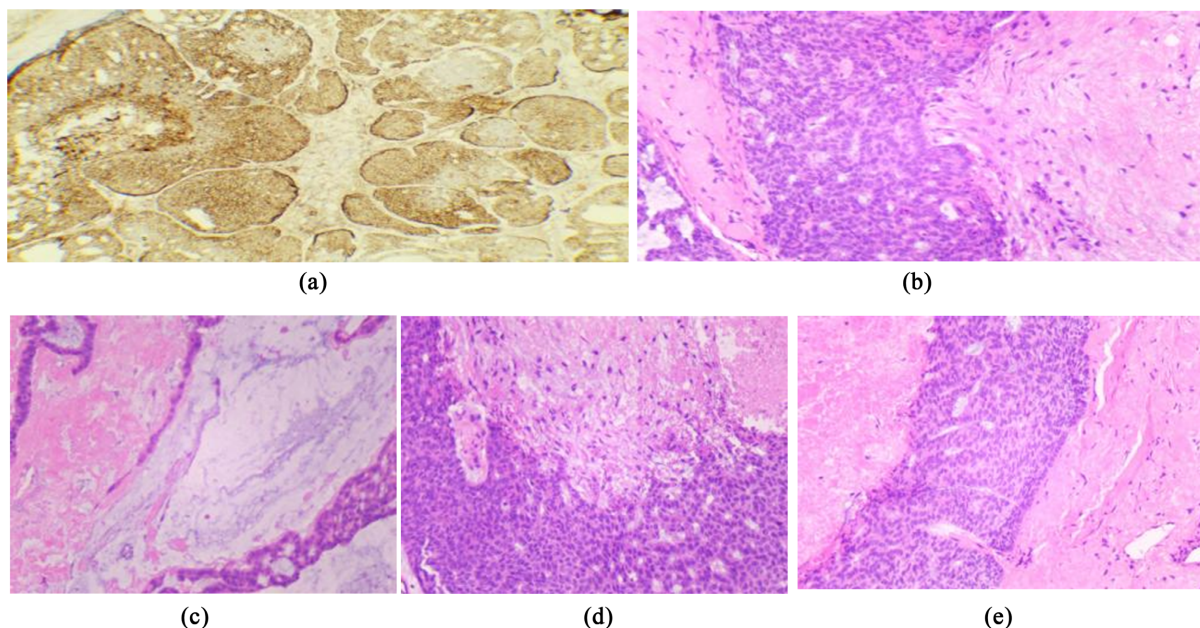


Figure 1. (a) and (b) show the characteristic tubular and cribriform pattern of basaloid cells, with round spaces containing mucinous material. (c)-(e) (low magnification) demonstrate that large areas of the tumor exhibit predominantly solid growth

图 1. (a)、(b) 可见特征性管状、筛状结构，基底样细胞围成圆形腔隙，腔内含黏液样物质。(c)-(e) 为低倍镜视野，肿瘤大片区域以实性生长方式为主

1.4. 免疫组化结果

肌上皮标志物：p40(弥漫强阳)、CK5/6(阳性)、SMA 局灶阳性；导管上皮标志物 CD10(腔面特异性表达)；CD117 阴性；S10 局灶弱阳性；PAX8、NKX3.1 均阴性，排除泌尿生殖道上皮源性转移癌，确诊阴囊原发性皮肤腺样囊性癌。

1.5. 术后随访

患者仅行单纯肿物切除，切缘宽度不足，存在局灶神经侵犯，拒绝辅助放疗、化疗。每 3 个月门诊复诊，每年复查阴囊超声、胸部 CT。截止至术后 24 个月随访，患者未发现局部复发或远处转移的临床及影像学证据，患者一般状态良好。鉴于该肿瘤晚期复发的特性，仍需进行长期乃至终身随访以监测其远期结局。

2. 讨论

2.1. 流行病学与临床特征

腺样囊性癌(ACC)最常起源于头颈部大小唾液腺，占唾液腺恶性肿瘤 10%~15% [1]。原发于皮肤附属器者称为 PCACC。PCACC 好发于头皮、胸壁，阴囊、外阴生殖器属于罕见发病部位，现有文献仅见 1 例阴囊复发性 PCACC 报道[2] [3]，本例为新增原发阴囊病例。PCACC 临床表现无特异性，多为缓慢生长、无痛坚实皮下结节，病程可达数年，极易被误诊为表皮囊肿、瘢痕疙瘩、基底细胞癌、附睾囊肿[4]。本例超声仅关注附睾病变，忽略真皮层原发附属器肿瘤，造成初诊误诊。PCACC 生物学行为较唾液腺 ACC

偏惰性, 但神经浸润发生率高达 76%, 局部复发率 51%, 复发间隔跨度极大, 短则 1 个月、长可达 40 年, 需长期随访[2] [3]。

2.2. 组织学分级标准与本例病理评估

Szanto 等提出的三级组织分级体系是目前 ACC 通用预后分级标准, 依据实性区域占比划分[5]: I级: 仅管状、筛状结构, 无实性成分; II级: 筛状为主, 实性区域 < 30%; III级: 实性区域 > 30%, 侵袭性强, 中位生存期仅 4 年。本例以筛状、管状结构为主, 实性区域极少, 归为I级低危亚型; 镜下双相细胞、筛状假囊内基底膜样物质是 ACC 特征性形态学标志, 需与多形性低度腺癌(PLGA)鉴别。

2.3. 免疫组化诊断与鉴别价值

1) CD117 (c-KIT)诊断局限性: 既往常将 CD117 作为 ACC 辅助标志物, 但系统综述及 Meta 分析证实其特异性较差, 多形性腺瘤、上皮肌上皮癌等多种唾液腺/皮肤肿瘤均可表达 CD117, 无法单独用于确诊 ACC [6]。本例 CD117 阴性, 与外阴生殖道 ACC 病例免疫表型一致, 提示 CD117 阴性不能排除本病, 需联合肌上皮标志物综合判断。

2) p63/p40 组合鉴别关键: PLGA 无成熟肌上皮分化, 特征免疫表型为 p63 (+)、p40 (-); 而 ACC 肌上皮细胞稳定共表达 p63、p40 (p63+/p40+), 该免疫组合鉴别二者特异度可达 99%, 是小活检标本的核心鉴别指标[7]。本例 p40 弥漫阳性, 可明确排除 PLGA。

3) 转移排除标志物: PAX8、NKX3.1 阴性, 排除前列腺、附睾等泌尿来源腺癌转移, 支持皮肤原发诊断[8]。

2.4. 分子病理特征

MYB 基因 t (6; 9) (q22-23)与 NFIB 融合是 ACC 特征性驱动突变, FISHMYB 断裂探针为分子确诊金标准; 头颈部 ACCMYB 重排阳性率约 56%, 皮肤、外阴生殖道 ACC 融合阳性率偏低, 部分仅存在 MYB 扩增而无经典 MYB-NFIB 融合[8] [9]。免疫组化 MYB 核表达 > 60%可 100%预判 MYB 基因重排, 可作为快速初筛手段[9]。本例未完善分子检测, 建议阴囊 PCACC 尤其是伴神经侵犯、切缘不足的高危病例补充 MYBFISH 检测, 指导远期靶向药物探索。

2.5. 治疗方案循证依据

1) 手术治疗: PCACC 标准术式为扩大切除术, 切缘至少 2 cm, 最大程度降低神经浸润相关局部复发; 头面部狭小皮损可选用 Mohs 显微外科, 阴囊部位优先传统扩大切除[4]。本例仅行单纯肿物摘除, 切缘不足, 存在远期复发风险, 是随访重点监测内容。

2) 辅助放疗价值: 一项纳入 3103 例患者的 Meta 分析证实, 术后辅助放疗可显著提升 5 年局部控制率(OR = 3.37, P = 0.009), 但对 5 年、10 年总生存期无统计学改善; 推荐适用人群: 切缘阳性、广泛神经侵犯、高分级实性型、肿瘤体积较大者[10]。本例存在局灶神经侵犯, 符合放疗指征, 但患者拒绝, 需强化长期影像学监测。

3) 全身治疗: PC 远处转移发生率仅 5%, 晚期转移患者可选用顺铂联合氟尿嘧啶方案; MYB、c-KIT 靶向药物仍处于临床试验阶段, 暂未常规应用于临床[8]。

2.6. 临床诊疗启示

阴囊原发性皮肤腺样囊性癌临床罕见、缺乏特征性表现, 影像学易误诊良性囊肿。对于阴囊病程数年、缓慢增大的无痛皮下结节, 不能仅依靠超声判断, 需尽早活检行病理 + 免疫组化检测。病理鉴别优

先采用 p63/p40 免疫组合区分 PLGA; 高危病例完善 MYB 分子检测。手术扩大切除为首选, 伴神经侵犯、切缘不足者建议术后辅助放疗; 本病复发潜伏期长达数十年, 需终身定期复查, 重点监测局部复发及肺部远处转移。

伦理声明

本研究报道了阴囊肿物切除术治疗阴囊腺样囊性癌。已获得患者及其家属的书面知情同意。该项目得到了昆明医科大学第二附属医院伦理委员会的批准。

作者贡献

陈春云: 撰写手稿、联系患者并获取知情同意。杨德林: 审阅稿件。所有作者均已阅读并同意最终版本。

利益冲突声明

所有作者声明不存在利益冲突。

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