Advances in Clinical Medicine 临床医学进展, 2025, 15(2), 1258-1262
Published Online February 2025 in Hans. https://www.hanspub.org/journal/acm
https://doi.org/10.12677/acm.2025.152469

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胰腺癌肉瘤误诊为胰腺滑膜肉瘤1例报道并文献回顾

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收稿日期：2025年1月26日；录用日期：2025年2月19日；发布日期：2025年2月27日

摘　要

胰腺癌肉瘤(Pancreatic carcinosarcoma)是一种极其罕见的疾病，由于相关文献报道及病理数据较少，极易造成误诊。患者最初因梗阻性黄疸入院，入院后完善患者相关辅助检查后发现胰腺钩突占位，结合实验室检查结果诊断为胰腺恶性肿瘤并行手术治疗，术后病理见：胰腺钩突处结节样肉瘤，肿瘤细胞弥漫分布，呈梭形或卵圆形，异型性明显，肿瘤未累及切缘及淋巴结。最初根据病理及免疫组化结果将其诊断为胰腺滑膜肉瘤，后经基因检测并回顾免疫组化结果后确诊为胰腺癌肉瘤，患者术后行化疗并门诊随访6个月，未见复发及转移。

关键词

胰腺癌肉瘤，文献复习，临床病理分析

A Case Report of Pancreatic Carcinosarcoma Misdiagnosed as Pancreatic Synovial Sarcoma and Literature Review

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Received: Jan. 26th, 2025; accepted: Feb. 19th, 2025; published: Feb. 27th, 2025

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文章引用: 常思远, 类成刚, 周饮际, 史光军. 胰腺癌肉瘤误诊为胰腺滑膜肉瘤 1 例报道并文献回顾[J]. 临床医学进展, 2025, 15(2): 1258-1262. DOI: 10.12677/acm.2025.152469

Abstract

Pancreatic carcinosarcoma is an extremely rare disease. Due to the scarcity of case reports and pathological data, it is highly prone to misdiagnosis. The patient initially presented with obstructive jaundice and was admitted to the hospital. Further diagnostic workups revealed a mass in the pancreatic uncinate process. Combining these findings with laboratory test results, the diagnosis of a malignant pancreatic tumor was made, and the patient underwent surgical treatment. Postoperative pathology revealed a nodular sarcoma in the pancreatic uncinate process, characterized by diffusely distributed tumor cells, which were spindle-shaped or oval with significant atypia. The tumor did not involve the surgical margins or lymph nodes. Initially, based on pathological and immunohistochemical findings, the tumor was diagnosed as pancreatic synovial sarcoma. However, genetic testing and a review of the immunohistochemical results ultimately confirmed the diagnosis of pancreatic carcinosarcoma. The patient underwent postoperative chemotherapy and was followed up in outpatient clinics for six months, during which no recurrence or metastasis was observed.

Keywords

Pancreatic Carcinosarcoma, Clinicopathological Analysis, Literature Review

1. 引言

胰腺癌肉瘤是一种极其罕见的疾病，其预后较传统胰腺导管腺癌(简称 PDAC)更差。胰腺癌肉瘤由具有梭形细胞形态的细胞组成，有或没有腺体成分。根据世界卫生组织(World Health Organization, WHO)对消化系统肿瘤的分类，胰腺癌肉瘤归为胰腺未分化癌。由于相关报道较少，导致临床医生对该疾病的了解较为有限。本文希望通过这份误诊病例，提高临床医生对这种疾病的警惕性。

2. 病例资料

患者为老年男性，71 岁，因腹痛伴皮肤巩膜黄染半月入院，入院体格检查发现：患者全身皮肤粘膜及巩膜黄染，左上腹有压痛，无反跳痛。实验室检查发现：总胆红素 208.90 umol/L (0~26 umol/L)直接胆红素 118.90 umol/L (0~4 umol/L)间接红素 90.00 umol/L (0~17 umol/L)碱性磷酸酶 472.23 U/L (45~125 U/L)糖类抗原 19~9127.1 U/ml (0~30 U/ml)。行超声检查提示：胰头区见低回声结节，大小约 36 mm × 27 mm，形态不规则，边界欠清，未见明显血流信号。后行胰腺强化 CT 检查：胰头区见类圆形低密度影，较大截面约 28 mm × 28 mm，增强扫描可见轻度强化，三期 CT 值约 34 HU，42 HU，45 HU (见图 1)。根据辅助检查结果，诊断为胰头恶性肿瘤，遂行根治性胰十二指肠切除术 + 门静脉部分切除伴吻合术。术中见肝脏呈淤胆改变，胰腺钩突可见占位性病变，大小约 30 mm × 30 mm，侵及门静脉及肠系膜上静脉右后壁，长度约 20 mm。术后病理见：胰腺钩突处结节样肉瘤，大小 30 mm × 25 mm × 25 mm，切面灰黄质地稍韧，肿瘤细胞弥漫分布，呈梭形或卵圆形，异型性明显，核分裂像多见，可见脉管内瘤栓及侵犯神经现象，肿瘤未累及切缘及淋巴结。免疫组化结果：HMB45−，S100−，CD56+，SOX-10−，Vimentin 弥漫+，p53，Ki67 50%+，EMA−，CD99+，STAT6−，CD34−，SMA−，β-catenin+，Synt−，CgA−，CD117−，SSTR2−，INSM1−，Desmin−，H-cald−，符合滑膜肉瘤表现(见图 2)。然而双相型滑膜肉瘤与癌肉瘤、纤维肉瘤在组

织学上具有重叠性，且免疫组化并非其决定性诊断证据。为明确滑膜肉瘤诊断，我们对患者的病理组织样本进行了 SYT 基因重排检测(荧光原位杂交，FISH)，结果未检测到 SYT 基因分离重排(见图 3)，由此诊断排除滑膜肉瘤。为进一步明确病理性质，我们针对 CK5/6、D2-40、WT-1、MC、CR 指标进行免疫组化检测，结果除了 CK5/6 部分阳性其他指标均为阴性(见图 2)，除外纤维肉瘤后最终确诊为癌肉瘤。患者康复出院后行化疗并门诊随访 6 个月，未见复发及转移。

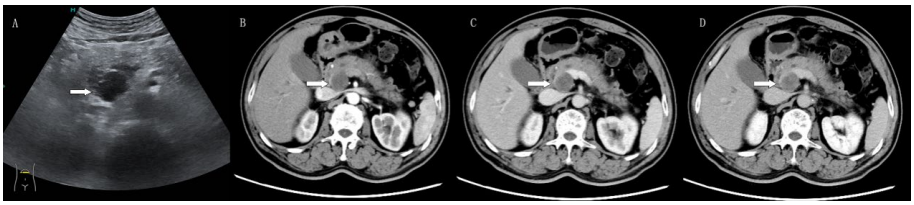

Figure 1. (A): Ultrasound Imaging indicates a 36 mm × 27 mm hypoechoic mass. (B): CT Arterial Phase Imaging. (C): CT Venous Phase Imaging. (D): CT Delayed Phase Imaging. B, C, D three-phase CT scans reveal a hypodense lesion in the head of the pancreas

图 1. (A)：超声成像显示 36 mm × 27 mm 的低回声肿块。(B)：CT 动脉期成像。(C)：CT 静脉期成像。(D)：CT 延迟期成像。B、C、D 三相 CT 扫描显示胰腺头部的低密度病变

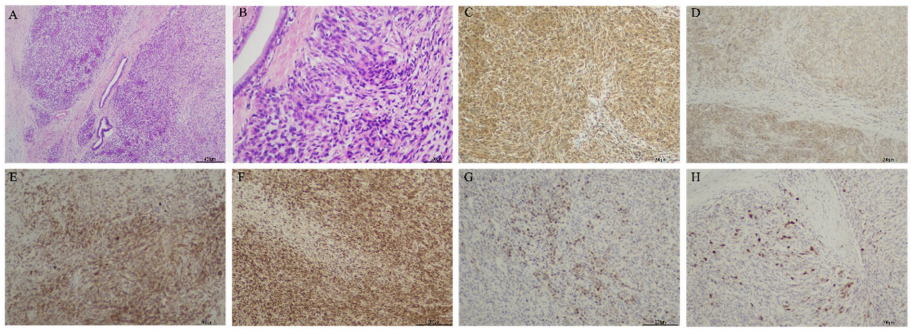

Figure 2. (A), (B) are H&E stained sections of the tumor. (C): Immunohistochemistry shows CD99 positivity. (D): Immunohistochemistry shows *β*-catenin positivity. (E): Immunohistochemistry shows CD56 positivity. (F): Immunohistochemistry shows vimentin diffuse nuclear staining pattern. (G): Immunohistochemistry shows CK positivity. (H): Immunohistochemistry shows CR positivity

图 2. (A)、(B)为肿瘤的 H&E 染色切片。(C)：免疫组化示 CD99 (+)。(D)：免疫组化示 *β*-catenin (+)。(E)：免疫组化示 CD56 (+)。(F)：免疫组化示 Vimentin (弥漫+)。(G)：免疫组化示 CK (+)。(H)：免疫组化示 CR (+)

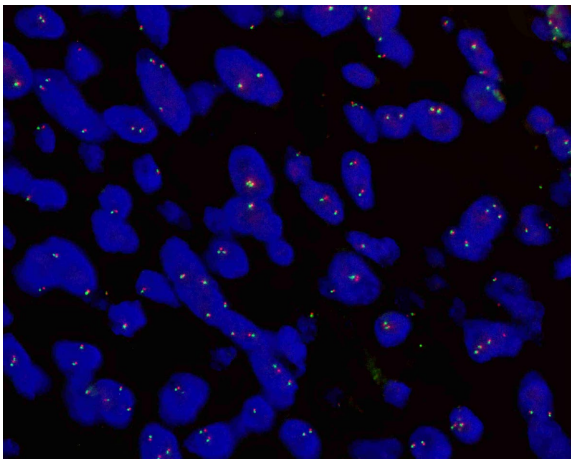

Figure 3. Genetic testing did not detect SYT gene rearrangement

图 3. 基因检测未发现 SYT 基因重排

3. 讨论

癌肉瘤是一种极其罕见的中胚层恶性肿瘤，多见于子宫及卵巢，在消化系统、肝脏和泌尿道也可有少量出现[1]。胰腺原发的癌肉瘤极为罕见，它是由两种不同恶性组织组成的肿瘤，同时具有上皮样细胞及间充质细胞成分。其中上皮样细胞成分可以是腺癌、鳞状细胞癌或基底细胞癌等。间充质细胞成分可以是梭形细胞肉瘤、横纹肉瘤、骨肉瘤、软骨肉瘤或未分化肉瘤，其免疫组化中间充质和上皮标志物均有表达[2] [3]。关于其发病机制尚无定论。目前上皮间充质转化(EMT)理论和单克隆理论是学术界较为认可的两种理论。上皮间充质转化(EMT)理论认为，癌肉瘤来源于胚胎上皮细胞转化成间充质细胞所导致[4]。而单克隆理论认为癌肉瘤来源于具有多能分化潜力的多能干细胞[5]。尽管已有术前影像学特征的相关文献与报道，但这些研究及报道仍未寻找到特异性的影像学特征可以在术前可靠地诊断此病[6]。胰腺癌肉瘤并无特异性临床表现，最常表现为与占位效应相关的腹痛、梗阻等症状，其次是体重减轻，以及胰腺外分泌功能受损导致的腹泻，与其他胰腺肿瘤并无明显差异。这导致其诊断主要依赖术后病理学检查，然而部分癌肉瘤的组织学特征与双向滑膜肉瘤存在相似性，只有通过 SYT-SSX 基因融合检测才能排除滑膜肉瘤诊断，极易导致误诊[7]。本案例中，患者就诊时并无特异性表现，仅表现为与占位效应相关的症状，其强化 CT 影像与胰腺癌强化 CT 影像并无显著差异。术后结合病理切片及免疫组化发现其肿瘤细胞呈双相性表现，符合双相性滑膜肉瘤表现，但基因检测却表明无 SYT 基因分离重排，从而排除胰腺滑膜肉瘤。

胰腺癌肉瘤的预后极差，中位生存期为 6 个月，3 年生存率低于 3% [8]。手术是胰腺癌肉瘤的首选治疗方式，但因其起病隐匿、症状不具有特异性，影像学检查经常被延迟，患者经常出现转移性或局部晚期疾病，导致血管系统受累而无法根治性切除从而导致治疗效果较差。M. Lee 等人对既往报道过的 48 例胰腺癌肉瘤患者数据进行了总结并分析。研究团队发现，在纳入的 48 名患者中，除 2 例无法查询患者的特征数据，46 名患者中有 26 名(56.5%)是女性。且胰腺癌肉瘤多发于胰头部(69.6%)，胰体/尾部仅占(30.4%) [9]。对于无法耐受手术或已经发生远处转移的患者，全身化疗可以控制疾病的发展并将分期降低到可行手术切除的程度[10]。已有文献记录表明，接受辅助化疗或术后辅以放化疗可延长患者的总生存期，但二者术后生存时间并无显著统计学差异，即使是常规的 PDAC 化疗方案也能提高胰腺癌肉瘤患者的生存率[9] [11]。

综上所述，胰腺癌肉瘤是一种恶性程度极高且极易造成误诊的恶性肿瘤，临床医生应提高对这种疾病的警惕性。当病理学检查及免疫组化提示为双相肉瘤且无法判断肿瘤类别，应及时行基因检测明确诊断。给予患者新辅助化疗并术后化学治疗，可以预防癌肉瘤的复发，并改善患者的预后。

声　明

该病例报道已获得患者知情同意。

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