· 短篇 ·

胰腺尾部炎性肌纤维母细胞瘤，并术后转移至双肺一例

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【摘要】炎性肌纤维母细胞瘤是相对少见的一种肿瘤，一般多见于成人或儿童的肺部，但发生在胰腺的很罕见。由于它不典型的临床表现及影像学征象，该肿瘤的确诊相对困难。这是一种良性的肿瘤性病变，但它具有侵袭性行为，也就是说它有恶性潜能[1]。我们在此报道的病例，在胰腺尾部发现了炎性肌纤维母细胞瘤，手术切除后定期随访，发现双肺新出现肿块，进行双肺肿块穿刺后确诊为胰腺肿瘤转移灶。

【关键词】炎性肌纤维母细胞瘤；胰腺；肺
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Case Report: Postoperative Inflammatory Myofibroblastic Tumor in the Tail of the Pancreas, Metastases to Both Lungs

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Abstract: Inflammatory myofibroblastic tumor (IMT) is rare tumor, It usually occurs in lung, and is extremely rare in the pancreas. Due to its atypical clinical and imaging manifestations, its diagnosis is relatively difficult. In our case, the man with an IMT in the tail of the pancreas, during follow-up after surgery, new lesions in both lungs were found and were confirmed.

Keywords: Inflammatory Myofibroblastic Tumor, Pancreas, Lungs

患者男性，34岁，无明显诱因腹部疼痛，呈阵发性，近期疼痛加剧，且发作频繁，患者否认恶心、呕吐症状。随后行腹部增强CT检查(图1)，发现胰腺尾部一个近10cm的低密度肿块，增强检查肿块不均匀强化。

患者于2020年8月进行手术治疗，术后病理结果为：低级别炎性肌纤维母细胞瘤。术后进行常规随访。2021年4月的CT扫描显示双肺出现新的肿块(图2)。2021年5月，进行右肺肿块穿刺。术后病理结合免疫组化显示：低级别炎性肌纤维母细胞瘤，结合既往病史，考虑转移瘤。免疫组化结果：CK (-), ALK lymphoid (-), SMA (weak +), Desmin (-), caldesmon (+), S-100 (-), CD34 (vascular +), Ki-67 (30 %), B-Catenin (membrane+), STAT6 (-), CD117 (-)。(图2)。

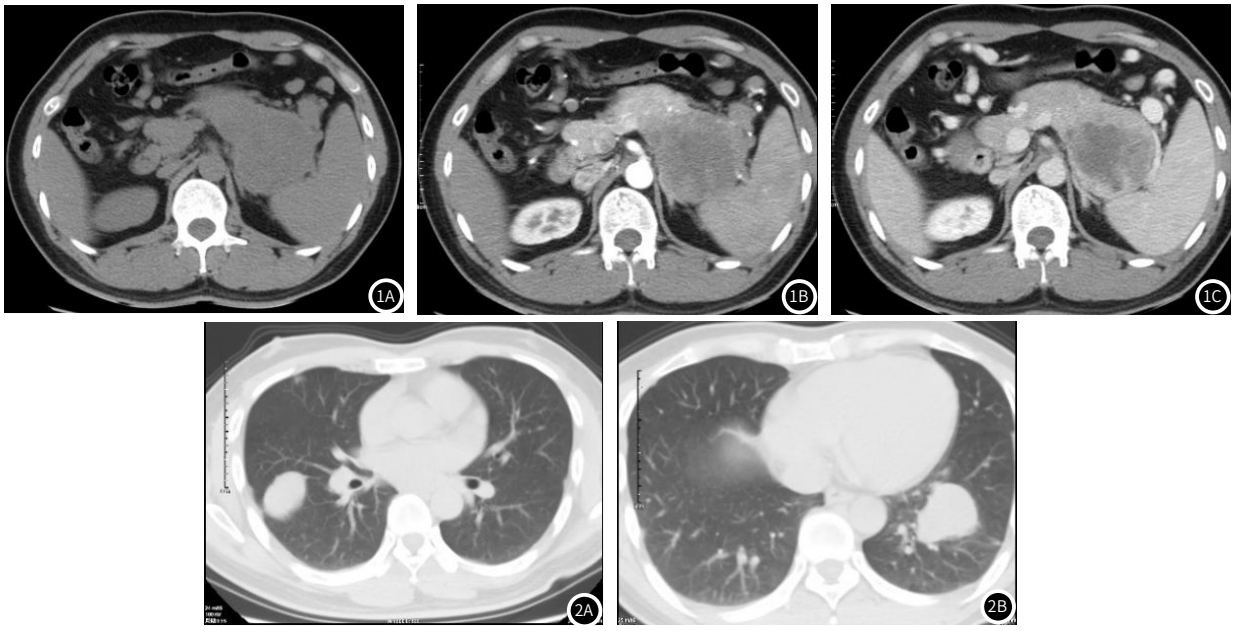

图1 增强CT，胰腺尾部可见圆形软组织肿块，内部密度不均匀(图1A)，增强检查显示不均匀增强，一些低密度区域未明显增强。(图1B-图1C)。**图2** 双肺可见多个肿块影，边界尚清晰。(图2A-图2B)。

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1 讨 论

胰腺炎性肌纤维母细胞瘤(Inflammatory myofibroblastic tumor, IMT)是一种罕见的间充质肿瘤，多数表现为良性，但具有恶性分化潜能，IMT可发生在身体的任何部位，最常见于肺、肝和胃肠道，也曾在胰腺中报道过，但相对罕见[2]。文献表明，男性更容易患胰腺炎性肌纤维母细胞瘤，而且主要发生在颈部[3]。胰腺IMT的症状是非特异性的，该患者也不例外，仅表现为腹痛。不同部位的IMT也可引起相应的症状，如胰头梗阻性黄疸；胰腺尾部脾脏破裂[4-5]。

胰腺炎性肌纤维母细胞瘤在影像学上也是不典型的，在这种情况下，它最初被认为是一种实性假乳头状肿瘤。胰腺炎性肌纤维母细胞瘤的最终诊断需要进一步的检查。胰腺炎性肌纤维母细胞瘤的CT通常显示低密度病变，增强后，表现为不均匀的延迟增强[6]。胰腺炎性肌纤维母细胞瘤的MRI通常在T1WI上表现为异质性低信号，在T2WI上则表现为等信号、轻度高信号和高信号。增强MRI显示不均匀强化。然而，也有病例报告显示T1WI和T2WI均为低信号[7]。

本例的特点是患者在常规随访中发现双肺肿块，病理穿刺后证实为炎性肌纤维母细胞瘤。炎性肌纤维母细胞瘤最常见于肺部，但本例肺部病变为转移灶。尽管患者的双肺都有炎性肌纤维母细胞瘤病灶，但他们没有症状，是在常规复查期间发现，这与文献报道一致[8]。同样，肺部病例的影像学特征也不明确[9]。

该患者在随访过程中出现双侧肿块，联想到先前的胰腺肿瘤，考虑双肺的病灶为胰腺炎性肌纤维母细胞瘤转移并不困难，而最终的病理活检证实肺部病变确实是炎性肌纤维母细胞瘤。以前的文献中从未报道过这种转移性病变。我们希望通过本病例报告，临床医生将更好地了解炎性肌纤维母细胞瘤。

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（上接第9页）

罕见病是发病率很低的一大类少见疾病，其中很多疾病为慢性、严重的遗传病，常常危及生命，多数为基因缺陷导致，缺乏有效的治疗方法[19]。关于Radio-Tartaglia 综合征的治疗方法的研究，目前有限，以对症治疗为主，可及早对患儿的发育障碍程度进行康复评价以制定相应的综合康复计划。由于其病例比较罕见，国内尚无相关报道，关于其临床表现和预后的信息仍有待补充，对现有病例的随访对于其预后尤为重要。鉴于Radio-Tartaglia 综合征患者可存在行为和精神异常，临床医生应叮嘱家长对患儿的行为和精神状态多加关注，以便及时介入心理疏导和行为管理。同时，不排除癫痫发作和先心病的可能性。对于女性患儿，应考虑到性早熟和肥胖倾向，对患儿体重和性成熟情况予以关注。此外，临床上也需要鉴别全面性发育迟缓(global developmental delay，GDD)为康复干预方式选择、预测康复干预结局提供了重要的线索[20]。

本例研究发现了Radio-Tartaglia综合征的新的SPEN 基因变异点c.6398T>A (p.Leu2133*)，明确了疾病诊断，给后期随访指引了方向。

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