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小肠肠系膜韧带样纤维瘤病1例

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摘　要

目的：小肠肠系膜韧带样纤维瘤病是一种罕见病，临床术前诊断通常容易误诊。本研究通过病理报道分析该疾病临床症状、影像特点及病理特征，回顾性分析并进行总结，旨在增加对该病的影像和临床认识，促进疾病的治疗及改善预后。方法：报道1例发生在腹腔内的小肠肠系膜处的韧带样纤维瘤病。结果：根据临床表现、CT检查及病理结果，诊断为小肠肠系膜韧带样纤维瘤病。结论：韧带样纤维瘤病发生率低，腹腔内发病者罕见，起病隐匿，肿瘤生长缓慢但腹腔内型一般体积较大，术前诊断通常容易误诊。

关键词

小肠肠系膜，韧带样纤维瘤病，1例

Mesenteric Desmoid-Type Fibromatosis of the Small Intestine

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Abstract

Objective: Ligamentoid fibromatosis of small intestine mesentery is a rare disease, which is usually misdiagnosed before clinical operation. This study analyzed the clinical symptoms, imaging features, and pathological features of the disease through pathological reports, and made a retrospective analysis and summary, aiming to increase the imaging and clinical understanding of the disease, promote the treatment of the disease and improve the prognosis. Methods: A case of desmoid fibromatosis of small intestine mesentery in the abdominal cavity was reported. Results:

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According to the clinical manifestation, CT examination, and pathological results, it was diagnosed as small intestinal mesenteric ligament-like fibromatosis. Conclusion: The incidence of desmoid fibromatosis is low, the incidence in the abdominal cavity is rare, the onset is hidden, and the tumor grows slowly, but the intraperitoneal type is generally large, and the preoperative diagnosis is usually easy to be misdiagnosed.

Keywords

The Mesentery of Small Intestine, Desmoid-Type Fibromatosis, 1 Case

1. 引言

小肠肠系膜韧带样纤维瘤病是指发生在小肠肠系膜的韧带样纤维瘤病(desmoid-type fibromatosis, DTF)，临床比较罕见，临床症状出现较晚且不典型，术前临床诊断容易误诊。本文报道 1 例小肠肠系膜韧带样纤维瘤病的临床、CT 及病理学特点并复习相关文献，旨在增加对该病的影像和临床认识。现对我院收治的 1 例小肠肠系膜韧带样纤维瘤病报道如下。

2. 临床资料和结果

2.1. 一般资料及查体

患者男性，60 岁。因上腹部疼痛不适 2 年余，加重 1 个月余入院。查体：右下腹触及一类圆形肿块，质地较硬，活动度差，伴有压痛，无反跳痛。实验室检查：血、尿常规及肾功能未见明显异常，便隐血试验阴性。

2.2. 辅助检查

2.2.1. 影像学检查及诊断

腹部 CT 平扫示右下腹腔内见一类圆形略低密度软组织肿块，密度大致均匀，边界较清晰，局部肠管受压移位，与肠管关系密切，局部肠管管壁增厚，肿块周围脂肪间隙模糊欠清；增强扫描示动脉期强化不明显，门脉期呈轻度强化，延迟期呈轻至中度的延迟性强化，平扫－动脉期－静脉期－延迟期平均 CT 值约为：39 Hu-42 Hu-48 Hu-59 Hu。如图 1(a)~(d)所示。CT 诊断：右下腹腹腔内肠系膜肿物，淋巴瘤可能。

2.2.2. 手术所见

于右下腹腔小肠肠系膜根部查见体积约 8.5 * 7.5 * 5 cm 的肿物，切面灰白，质韧，边界尚清，局部与小肠关系密切，似侵犯肠壁固有肌层，小肠肠管切除长度约 36 cm，最大周长约 4.5 cm，肠粘膜尚光滑，灰白色，质韧，距肿块浸润一端 6.5 cm，另一端切线 7 cm。于肿瘤周围肠系膜处查见淋巴结 16 个，大者长径约 0.6 cm，灰白色，质韧。

2.2.3. 光镜所见及病理结果

光镜下见肿瘤由梭形纤维母细胞和胶原纤维构成，呈束状、编织状排列，如图 2 所示。病理诊断：

小肠肠系膜韧带样纤维瘤病，体积 8.5 * 7.5 * 5 cm，肿瘤侵犯肠壁固有肌层，未累及两端切线。肠系膜(16个)淋巴结呈反应性增生。免疫组化：B-catenin (核+)，LEF-1 (−)、CD117 (−)、DOG1 (−)、CD34 (−)、SMA (−)、S-100 (−)。

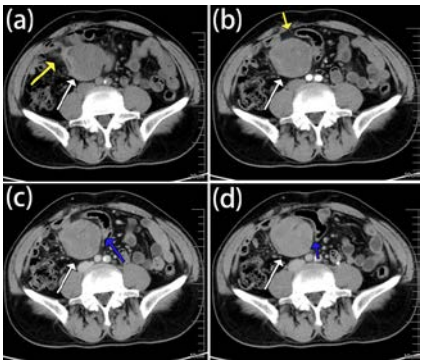

Figure 1. As indicated by the white arrow, (a) A kind of round slightly low-density mass was seen in the right lower abdominal cavity on the plain scan, the density was generally uniform, the boundary was clear, the local intestinal tube was pressed and displaced, closely related to the intestinal tube, the local intestinal tube wall was thickened; (b) The enhancement was not obvious in the arterial phase; (c) The portal phase of enhanced scanning showed mild enhancement (d) the delayed phase of enhanced scanning showed mild to moderate delayed enhancement. The yellow arrow indicated that the fat space around the focus was blurred. As pointed out by the blue arrow, the tumor pushed and pressed the local small intestinal tissue, which was closely related to the small intestine, and the local intestinal wall thickens

图 1. 如图中白色箭头所指，(a) 平扫右下腹腔内见一类圆形略低密度肿块，密度大致均匀，边界较清晰，局部肠管受压移位，与肠管关系密切，局部肠管管壁增厚；(b) 增强扫描动脉期强化不明显；(c) 增强扫描门脉期呈轻度强化；(d) 增强扫描延迟期进一步强化，呈轻至中度的延迟性强化。如黄色箭头所指，病灶周围脂肪间隙模糊。如蓝色箭头所指，肿瘤推压局部小肠组织，与小肠关系密切，局部肠壁增厚

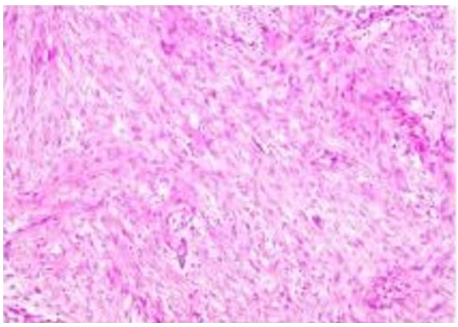

Figure 2. Pathological examination showed that the tumor was composed of proliferated fibroblasts and collagen fibers. The tumor cells were slender and long fusiform, arranged in bundles and weaves

图 2. 病理学检查显示肿瘤由增生的纤维母细胞和胶原纤维构成。瘤细胞呈纤细的长梭形，呈束状、编织状排列

3. 讨论

韧带样纤维瘤病(desmoid-type fibromatosis, DTF)又称侵袭性纤维瘤病(aggressive fibromatosis, AF)、硬纤维瘤、韧带样瘤等，最早是在 1948 年由 Stout 提出的[1]。DTF 是一种无性纤维母细胞肿瘤性增生症，占所有肿瘤的 0.03%，在所有软组织肿瘤中占比不到 3% [2]。DTF 是一种罕见的结缔组织良性肿瘤，起

源于深层肌肉筋膜、腱膜、肌腱和瘢痕组织[3] [4] [5]，其特征是浸润性生长和没有转移潜力。虽然组织学上是良性的，即无坏死、核多形性和不典型有丝分裂，但具有局部侵袭性的特点，有压迫局部结构的倾向，如肠道、服盆腔神经和大血管、输尿管、膀胱、阴道等，因此被归为中度恶性肿瘤[6] [7] [8]。发病部位多见于四肢及腹壁，腹腔内发病少见，尤其是肠系膜韧带样纤维瘤病少见。DTF 的病因可能是创伤后、既往腹部手术史、激素因素等[9]，病灶可以零星发生，也可以出现在先天性综合征的背景下，如家族性腺瘤性息肉病等[10]。然而，到目前为止，还没有相关文献表明髋关节假体和硬纤维瘤之间的联系[11]。DTF 发病年龄在 16 岁至 60 岁之间，高发病率年龄约为 30 岁[9]。DTF 女性发病率较高(男女比例为 1:1.5~2.5) [12]。DTF 临床上比较罕见，加上其临床病程的多变性，没有任何报道证明特定临床干预措施是有效的[13]。DTF 主要的治疗方法是根治性手术切除，安全切除范围为 2~3 cm。然而，由于肿瘤边界不清，“真正的”根治性切除手术通常很难实现[4] [14]。在一项有史以来病例数量最大的 DTF 调查中，大多数放射肿瘤学家认为放射疗法是一种值得和有用的侵袭性纤维瘤病的治疗适应症[2]。本篇病例报告是一位 60 岁男性患者，在腹腔内小肠肠系膜处发现一类圆形肿块，术后病理证实为小肠肠系膜韧带样纤维瘤病。

在我们的病例中，DTF 的最终诊断是术后通过手术标本的组织学检查获得的。患者术后恢复良好，术后 5 个月内无不适，复查未见复发征象。DTF 的罕见发生和不同的临床表现限制了影像科医生的术前诊断的能力。腹腔内 DTF 临床体征和影像学特征无特异性，大多数肠系膜纤维瘤位于肠系膜的根部，生长缓慢，临床症状明显时肿瘤体积多较大，多数为单发，偶尔可为多发，呈圆形或卵圆形病灶，周围无明显肿大的淋巴结及腹腔积液。CT 平扫呈较均匀稍低软组织密度，增强扫描显示肿瘤体内部多数可见轻到中度的延迟性强化(镜下所见肿瘤内的纤维母细胞和大量变性的胶原纤维是延迟强化的病理基础)。

肠系膜 DTF 的鉴别诊断主要包括 1) 胃肠道间质瘤，CD34 和 CD117 染色为阳性，多数瘤体位于胃肠道腔外，也可向腔内生长，良性肿瘤可出现斑点状、环形或弧形钙化，肿瘤较大时多见低密度坏死、囊变区，增强实性部分呈明显、持续强化，坏死囊变区不强化，肿瘤可经蒂与胃肠道壁相连或局部壁增厚，有助于确定肿瘤起源；2) 胃肠道淋巴瘤，四十岁以上中老年多见，男多于女，多发生肠壁环形增厚，一般呈较均匀等密度，粘膜面可出血多发坏死，多伴有淋巴结增多增大，密度均匀，增强扫描呈不强化或轻度强化。3) 孤立性纤维瘤，可发生于身体的任何部位，包括肠系膜[15] [16]，多呈膨胀性生长，钙化少见，肿瘤较大时内可见钙化，肿瘤内可见多发坏死囊变区，增强扫描呈明显不均匀强化，富血管区和肿瘤细胞密集区明显强化，致密胶原纤维区强化相对较弱、呈延迟强化，坏死囊变区无强化。

本文考虑到小肠肠系膜韧带样纤维瘤病临床发病率低，术前诊断困难，误诊率高，对于疑似 DTF 的病例，应结合组织病理学检查。因此，本报道旨在通过罕见病例报道以及相关文献的复习来引起临床医师的重视。

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