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临床经验 CLINICAL PRACTICE

腹腔内促纤维增生性小圆细胞肿瘤4例

章 颖, 马向涛, 付 静, 余力伟

■背景资料
腹腔内促纤维增生性小圆细胞肿瘤(IDSRCT)是一种罕见腹膜恶性肿瘤.

章颖, 北京海淀医院妇产科 北京市 100080
马向涛, 余力伟, 北京海淀医院外科 北京市 100080
付静, 北京海淀医院病理科 北京市 100080
通讯作者: 马向涛, 100080, 北京市海淀区中关村大街29号, 北京海淀医院外科. xiangtao_ma@pku.org.cn
电话: 010-62639764 传真: 010-62583027
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Intra-abdominal desmoplastic small round cell tumor: an analysis of 4 cases

Ying Zhang, Xiang-Tao Ma, Jing Fu, Li-Wei Yu

Ying Zhang, Department of Obstetrics and Gynecology, Beijing Haidian Hospital, Beijing 100080, China
Xiang-Tao Ma, Li-Wei Yu, Department of Surgery, Beijing Haidian Hospital, Beijing 100080, China
Jing Fu, Department of Pathology, Beijing Haidian Hospital, Beijing 100080, China
Correspondence to: Dr. Xiang-Tao Ma, Department of Surgery, Beijing Haidian Hospital, 29 Zhongguancun Avenue, Haiding District, Beijing 100080, China. xiangtao_ma@pku.org.cn
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Abstract

AIM: To investigate the clinical features of intra-abdominal desmoplastic small round cell tumor (IDSRCT).

METHODS: We retrospectively analyzed 4 cases of IDSRCT in our hospital from June 1996 to October 2006.

RESULTS: All the 4 cases were admitted with symptoms of ascites and/or abdominal mass. The patients underwent exploratory laparotomy, revealing multiple peritoneal and mesenteric masses with an extensive seeding. The diagnosis was made by pathology after operation.

CONCLUSION: IDSRCT is a very rare peritoneal malignant neoplasm, and it is a disease with high metastasis and poor prognosis.

Key Words: Peritoneum neoplasm; Desmoplastic small round cell tumor; Metastasis

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摘要

目的: 探讨腹腔内促纤维增生性小圆细胞瘤(intra-abdominal desmoplastic small round cell tumor, IDSRCT)的临床特点.

方法: 对我院于1996-06/2006-10收治的4例IDSRCT患者诊疗及预后进行回顾性分析.

结果: 本研究报道4例患者, 均表现为腹水和/或腹部肿物收入院, 剖腹探查术发现肿瘤腹腔内广泛种植, 术后病理证实为IDSRCT.

结论: 腹腔内促纤维增生性小圆细胞瘤是一种罕见腹膜恶性肿瘤, 诊断时患者已经出现广泛转移且预后不良.

关键词: 腹膜肿瘤; 促纤维增生性小圆细胞肿瘤; 转移

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0 引言

腹腔内促纤维增生性小圆细胞瘤(intra-abdominal desmoplastic small round cell tumor, IDSRCT)是一种罕见腹膜恶性肿瘤[1-3]. 患者发病年龄3-68(平均22)岁. 我院于1996-06/2006-10收治4例IDSRCT患者, 术后病理证实为IDSRCT. 我们对其诊疗及预后进行了回顾性分析.

1 材料和方法

1.1 材料 我院1996-06/2006-10收治4例腹腔内促纤维增生性小圆细胞瘤患者, 其中女2例, 男2例, 年龄18-83岁, 临床表现为腹水和/或腹部肿物, 病程4-12 wk.

1.2 方法

1.2.1 入院查体 腹略胀, 未见胃肠形, 下腹轻压痛, 未及明确肿物, 下腹部可触及包块, 表面不

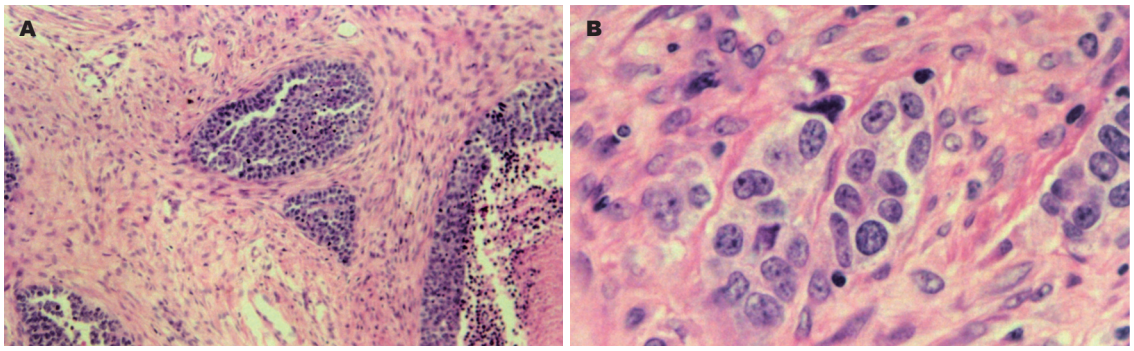

图 1 腹腔内促纤维增生性小圆细胞肿瘤病理. A: 苏木精伊红染色(×100); B: 苏木精伊红染色(×400).

表 1 IDSRCT患者实验室检查结果

编号	性别	年龄(岁)	病程	临床表现	CEA (μg/L)	CA199 (kU/L)	CA125 (kU/L)
1	女	83	4 wk	腹胀腹痛	0.88	80.37	1210.6
2	女	26	7 wk	腹胀腹痛	1.22	117.2	583.41
3	男	18	3 wk	腹痛	4.31	293.61	129.62
4	男	53	12 wk	腹胀呕吐	10.80	945.81	51.23

光滑, 可以推动, 边界不清. 肛门指诊直肠黏膜光滑, 未及肿物.

1.2.2 实验室检查(表1) 肿瘤标记物中癌胚抗原(carcinoembryonic antigen, CEA) 0.88-10.80 μg/L (正常值0-3.4 μg/L), CA19-9 80.37-945.81 kU/L (正常值0-35 kU/L), CA125 51.23-1210.60 kU/L (正常值0-35 kU/L), AFP在正常范围内(正常值<5.8 kU/L).

1.2.3 影像学检查 胸片均未见异常. 腹部B超: 中等-大量腹水. 妇科B超: 子宫壁回声不均匀, 子宫后方不规则肿物, 内呈实性不均匀中低回声, 双侧附件不清.

2 结果

2.1 手术方式 全部患者均行剖腹探查术, 术中发现腹腔内约淡黄色腹水400-3000 mL, 腹膜表面遍布粟粒样结节, 直径0.5-1.5 cm, 大网膜呈饼状增厚最大约15 cm×12 cm×4 cm, 肝脏表面可及大小不等多发结节, 直径1-2 cm; 2例女性患者子宫后方可见大小约5-8 cm灰白色质硬肿物. 2例患者行大网膜切除术, 另外2例患者行大网膜活检术.

2.2 术后病理 术后病理报告: 腹腔内促纤维增生性小圆细胞瘤(图1). 光镜下见肿瘤细胞聚集成界限清楚、大小不等、形状不一的巢状或梁索状结构, 位于致密的纤维间中. 肿瘤细胞小或中等大, 呈圆形或卵圆形, 少数呈梭形. 肿瘤细胞排列紧密, 细胞核呈圆形或卵圆形, 核分裂象多见. 免疫组织化学染色: CK(+), Vimentin(+), Desmin(+), NSE(+), CgA(+), Syn(-), Calretinin(-).

2.3 生存情况 所有病例术后恢复好, 无手术并发症发生, 未行化疗, 均有完整随访资料, 随访时间3-9 mo, 患者最终均死于肿瘤广泛转移.

3 讨论

腹腔内促纤维增生性小圆细胞瘤是一种罕见腹膜恶性肿瘤, 也有文献报道促纤维增生性小圆细胞瘤可以发生于腹膜外部位如: 颅内、眼眶、舌根、胸膜与睾丸等部位[4-7]. 患者发病年龄3-68(平均22)岁. IDSRCT患者多以腹部肿物就诊, 伴有腹痛、腰痛、恶心、呕吐、便秘、消瘦等或有腹水征[8-10]. 由于肿瘤细胞被纤维组织分隔伴有坏死, B超与CT可显示单个或多个大小不一、形态不规则、非均质的肿块[11-12]. 本研究报道病例均表现为腹水和/或腹部肿物收入院, 病程4-12 wk, 其中1例83岁女性患者, 为国内外文献报道中年龄最大的患者.

光镜下可以见到IDSRCT细胞聚集成界限清楚、大小不等、形状不一的巢状或梁索状结构, 位于致密的纤维间中, 有些区域瘤细胞密集而间质少, 有些区域微小的瘤细胞团位于丰富的纤维间质中[13-15]. 肿瘤细胞小或中等大, 呈圆形或卵圆形, 少数呈梭形. 肿瘤细胞排列紧密, 胞质少, 嗜酸性, 呈透明或空泡状. 细胞核呈圆

■应用要点
姑息性切除能减轻机体对肿瘤的负荷, 有利提高术后化疗等综合治疗的疗效.

■同行评价
腹腔内促纤维增生性小圆细胞瘤(IDSRCT)是一种罕见的、恶性度高的腹膜肿瘤, 作为病例报道有一定的临床意义.

形或卵圆形, 染色深, 染色质细颗粒状或斑点状, 核仁不明显, 核分裂象多见. 瘤巢周边细胞有的可呈栅栏状或阅兵样排列. 电镜下IDSRCT肿瘤细胞体积小, 绝大多数呈圆形或卵圆形, 少数为短梭形, 相互紧密排列. 瘤细胞无基板, 偶见细胞间短小的微绒毛或指状突起, 细胞间小腔形成, 胞突相互缠绕. 核浆比例大, 细胞器少, 含量多少不等, 主要有线粒体、糖原颗粒、脂滴及溶酶体[6]. 瘤细胞巢外的间质细胞埋没在致密的胶原性基质内, 这些间质细胞包括纤维母细胞、肌纤维母细胞及处于二者之间的中间型细胞[16-18].

大多数IDSRCT肿瘤细胞表现为Cytokeratin, Desmin, Vimentin, EMA与NSE阳性. 其中Cytokeratin与EMA呈胞质弥漫型强阳性, Desmin多为核旁小球样点状阳性, Vimentin呈弥漫或点状弱阳性, NSE呈灶性弥漫胞质型弱阳性[13-14,16,19]. Ladanyi *et al*[20]发现大多数IDSRCT中存在染色体易位t(11; 22)(p13; q12), 而位于22号染色体q12的EWS基因与位于11号染色体p13的WT1基因可以融合形成EWS/WT1融合基因, EWS/WT1融合基因可诱导血小板源性生长因子A表达是导致IDSRCT表现为质硬肿物的原因之一[21-22].

腹腔内促纤维增生性小圆细胞瘤是一种主要发生于腹膜的高度恶性小圆细胞肿瘤, 青少年好发, 免疫组化显示瘤细胞具有多向分化的特点. 根据这些特点, 结合光镜、电镜与免疫组化研究, IDSRCT与转移性小细胞癌, 恶性横纹肌样瘤, 小细胞恶性间皮瘤, 原始神经外胚叶瘤, 胚胎性横纹肌肉瘤, 非霍奇金淋巴瘤, 神经母细胞瘤, Ewing瘤等小圆细胞恶性肿瘤之间不难鉴别[23-25].

IDSRCT以手术治疗为主, 但是由于多数患者就诊时已经出现肿瘤腹腔内广泛转移, 很少能获得根治性效果, 多数情况下手术作为肿瘤细胞减数或者活检的一种手段. 术中见肿瘤多位于腹腔浆膜面相应的脏器表面和/或脏器之间的单个、多个形状不规则的灰白色质硬肿块, 肿瘤可累及大网膜. 由于肿瘤多发往往手术难以彻底切除, 因此多配合化疗, 但是远期效果差, IDSRCT对放疗不敏感[26-28]. Fizazi *et al*[29]发现血清CA125与NSE在60%的IDSRCT患者中升高, 给与有效化疗后患者腹水减少, 肿瘤缩小, CA125与NSE水平可降至正常.

由于IDSRCT早期无特异性症状, 就诊时50%的患者已经出现淋巴结转移, 常见远处转移部位为肝脏. 肿瘤可以压迫腹腔内器官而引起肠梗阻及尿路梗阻等症状. IDSRCT恶性程度高, 多数患者于1 a内死亡, 5 a生存率为15%[30-32]. 本研究报道4例患者均于术后9 mo内死亡, 与国外文献报道相符合.

4 参考文献

1 Gerald WL, Miller HK, Battifora H, Miettinen M, Silva EG, Rosai J. Intra-abdominal desmoplastic small round-cell tumor. Report of 19 cases of a distinctive type of high-grade polyphenotypic malignancy affecting young individuals. *Am J Surg Pathol* 1991; 15: 499-513

2 Cheung NY, Khoo US, Chan KW. Intra-abdominal desmoplastic small round-cell tumour. *Histopathology* 1992; 20: 531-534

3 Yeoh G, Russell P, Wills EJ, Fleming S. Intra-abdominal desmoplastic small round cell tumor. *Pathology* 1993; 25: 197-202

4 Parkash V, Gerald WL, Parma A, Miettinen M, Rosai J. Desmoplastic small round cell tumor of the pleura. *Am J Surg Pathol* 1995; 19: 659-665

5 Devaney K. Intra-abdominal desmoplastic small round cell tumor of the peritoneum in a young man. *Ultrastruct Pathol* 1994; 18: 389-398

6 Resnick MB, Donovan M. Intra-abdominal desmoplastic small round cell tumor with extensive extra-abdominal involvement. *Pediatr Pathol Lab Med* 1995; 15: 797-803

7 Takahira K, Ohi S, Fujii N, Matsuura Y, Sano M, Hanai H, Kaneko E. Intra-abdominal desmoplastic small round cell tumor (IDSRT). *J Gastroenterol* 2000; 35: 712-716

8 Nathan JD, Gingalewski C, Salem RR. Intra-abdominal desmoplastic small round cell tumor. *Yale J Biol Med* 2001; 74: 13-20

9 Dorsey BV, Benjamin LE, Rauscher F 3rd, Klencke B, Venook AP, Warren RS, Weidner N. Intra-abdominal desmoplastic small round-cell tumor: expansion of the pathologic profile. *Mod Pathol* 1996; 9: 703-709

10 马向涛, 付静, 余力伟, 王杉, 杜如昱. 腹腔内促纤维增生性小圆细胞肿瘤一例. 中华普通外科杂志 2006; 21: 119

11 Chouli M, Viala J, Dromain C, Fizazi K, Duvillard P, Vanel D. Intra-abdominal desmoplastic small round cell tumors: CT findings and clinicopathological correlations in 13 cases. *Eur J Radiol* 2005; 54: 438-442

12 Bellah R, Suzuki-Bordalo L, Brecher E, Ginsberg JP, Maris J, Pawel BR. Desmoplastic small round cell tumor in the abdomen and pelvis: report of CT findings in 11 affected children and young adults. *AJR Am J Roentgenol* 2005; 184: 1910-1914

13 Froberg K, Brown RE, Gaylord H, Manivel C. Intra-abdominal desmoplastic small round cell tumor: immunohistochemical evidence for up-regulation of autocrine and paracrine growth factors. *Ann Clin Lab Sci* 1999; 29: 78-85

14 McCluggage WG, Wilkinson N. Metastatic neoplasms involving the ovary: a review with an emphasis on morphological and immunohistoche-

mical features. *Histopathology* 2005; 47: 231-247
15 Lippe P, Berardi R, Cappelletti C, Massacesi C, Mattioli R, Latini L, Cellerino R. Desmoplastic small round cell tumour: a description of two cases and review of the literature. *Oncology* 2003; 64: 14-17
16 Takekawa Y, Ugajin W, Koide H, Nishio S, Yamamoto T, Sawada T. Pathologic, cytologic and immunohistochemical findings of an intra-abdominal desmoplastic small round cell tumor in a 15-year-old male. *Pathol Int* 2000; 50: 417-420
17 Smith ME, Pelletier JP, Daniels R. Pathologic quiz case. Intra-abdominal desmoplastic small round cell tumor. *Arch Pathol Lab Med* 2000; 124: 1839-1840
18 Reich O, Justus J, Tamussino KF. Intra-abdominal desmoplastic small round cell tumor in a 68-year-old female. *Eur J Gynaecol Oncol* 2000; 21: 126-127
19 Chambonniere ML, Mosnier-Damet M, Mosnier JF. Expression of microtubule-associated protein tau by gastrointestinal stromal tumors. *Hum Pathol* 2001; 32: 1166-1173
20 Ladanyi M, Gerald W. Fusion of the EWS and WT1 genes in the desmoplastic small round cell tumor. *Cancer Res* 1994; 54: 2837-2840
21 Katz RL, Quezado M, Senderowicz AM, Villalba L, Laskin WB, Tsokos M. An intra-abdominal small round cell neoplasm with features of primitive neuroectodermal and desmoplastic round cell tumor and a EWS/FLI-1 fusion transcript. *Hum Pathol* 1997; 28: 502-509
22 Argatoff LH, O'Connell JX, Mathers JA, Gilks CB, Sorensen PH. Detection of the EWS/WT1 gene fusion by reverse transcriptase-polymerase chain reaction in the diagnosis of intra-abdominal desmoplastic small round cell tumor. *Am J Surg Pathol* 1996; 20: 406-412
23 Gil A, Gomez Portilla A, Brun EA, Sugarbaker PH. Clinical perspective on desmoplastic small round-cell tumor. *Oncology* 2004; 67: 231-242
24 McCluggage WG. Ovarian neoplasms composed of small round cells: a review. *Adv Anat Pathol* 2004; 11: 288-296
25 Kim JH, Goo HW, Yoon CH. Intra-abdominal desmoplastic small round-cell tumour: multiphase CT findings in two children. *Pediatr Radiol* 2003; 33: 418-421
26 Suehara Y, Yazawa Y, Hitachi K. Intraabdominal desmoplastic small round cell tumor: results of ifosfamide-based chemotherapy. *Int J Clin Oncol* 2004; 9: 134-138
27 De Lena M, Caruso ML, Marzullo F, Mancarella S, Armentano R, Ventrella V, Guida M. Complete response to chemotherapy in intra-abdominal desmoplastic small round cell carcinoma. A case report. *Tumori* 1998; 84: 412-416
28 Jahraus CD, Glisson SD, St Clair WH. Treatment of desmoplastic small round cell tumor with image-guided intensity modulated radiation therapy as a component of multimodality treatment. *Tumori* 2005; 91: 253-255
29 Fizazi K, Farhat F, Theodore C, Rixe O, Le Cesne A, Comoy E, Le Chevalier T. Ca125 and neuron-specific enolase (NSE) as tumour markers for intra-abdominal desmoplastic small round-cell tumours. *Br J Cancer* 1997; 75: 76-78
30 Lal DR, Su WT, Wolden SL, Loh KC, Modak S, La Quaglia MP. Results of multimodal treatment for desmoplastic small round cell tumors. *J Pediatr Surg* 2005; 40: 251-255
31 Hassan I, Shyyan R, Donohue JH, Edmonson JH, Gunderson LL, Moir CR, Arndt CA, Nascimento AG, Que FG. Intraabdominal desmoplastic small round cell tumors: a diagnostic and therapeutic challenge. *Cancer* 2005; 104: 1264-1270
32 Livaditi E, Mavridis G, Soutis M, Papandreou E, Moschovi M, Papadakis V, Stefanaki K, Christopoulos-Geroulanos G. Diffuse intraabdominal desmoplastic small round cell tumor: a ten-year experience. *Eur J Pediatr Surg* 2006; 16: 423-427

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