综述

腹膜假黏液瘤的诊断与治疗研究进展

史冠军,夏　奥,蔚莉菲,马瑞卿,许洪斌
(航天中心医院,北京 100049)

摘　要:腹膜假黏液瘤是一种罕见的临床综合征,主要来源于阑尾黏液肿瘤,以腹腔内充满黏液或胶冻样腹水为特征。发病率每年约 3/100 万~4/100 万。由于其进展缓慢,无特异性临床症状,容易导致误诊。超声、CT 等影像学检查有助于诊断。目前细胞减灭术与腹腔热灌注化疗相结合的综合治疗策略已经成为腹膜假黏液瘤标准的治疗方法,然而术后高复发率仍是亟待解决的难题。肿瘤减灭程度是影响生存预后的重要因素,而全身化疗能否获益尚存在争议。
主题词:腹膜假黏液瘤;肿瘤细胞减灭术;腹腔热灌注化疗
中图分类号:R730.262　　**文献标识码**:A　　**文章编号**:1671-170X(2021)12-1034-07
doi:10.11735/j.issn.1671-170X.2021.12.B011

Research Progress in Diagnosis and Treatment of Pseudomyxoma Peritonei

SHI Guan-jun, XIA Ao, YU Li-fei, MA Rui-qing, XU Hong-bin
(*Aerospace Central Hospital*, *Beijing* 100049, *China*)

Abstract: Pseudomyxoma peritonei is a rare clinical syndrome with a incidence rate of $0.3/10^5$~$0.4/10^5$, which mainly comes from appendix myxoma and is characterized by mucus or jelly-like ascites in the abdominal cavity. Because of its slow progress and no specific clinical symptoms, it is often misdiagnosed. Ultrasonography, CT and other imaging examinations are helpful for diagnosis. At present, cytoreductive surgery combined with hyperthermic intraperitoneal chemotherapy is the standard therapeutic strategy for pseudomyxoma peritonei, but the high postoperative recurrence rate is still the problem to be solved urgently. The extent of tumor attenuation is an important factor affecting survival and prognosis, but whether systemic chemotherapy can benefit is still controversial.
Subject words: pseudomyxoma peritonei; cytoreductive surgery; hyperthermic intraperitoneal chemotherapy

腹膜假黏液瘤(pseudomyxoma peritonei,PMP)是一种肿瘤细胞通过“再分布现象”在腹腔内播散种植,并以产生大量黏液或胶冻样腹水为主要特征的临床罕见病[1-2]。1884 年 Werth 首次使用腹膜假黏液瘤的概念[3]。1937 年 Robert 提出一个假说,当阑尾黏液囊肿破裂后,逸出的上皮细胞在腹腔内种植,并继续分泌黏液,从而导致 PMP[3]。由于可用的数据有限,PMP 确切的发病率尚不清楚,最初提出的发病率约为 1/100 万~2/100 万,而最新一项研究预测发病率在每年 3/100 万~4/100 万,甚至可能更高[4]。虽然 PMP 主要来源于穿孔的阑尾黏液性肿瘤,但也可来源于其他器官,如卵巢、胰腺、脐尿管、结直肠以及畸胎瘤等[5-10]。PMP 生物学行为呈惰性生长,进展缓慢,缺乏特异性临床表现,早期发现较困难,多数患者出现症状时往往已经是肿瘤晚期。传统治疗方式以简单穿刺引流腹腔积液或反复手术减瘤为主,预后较差。20 世纪 90 年代提出的肿瘤细胞减灭术(cytoreductive surgery,CRS)与腹腔热灌注化疗(hyperthermic intraperitoneal chemotherapy,HIPEC)联合的综合治疗模式,使部分患者 10 年生存率从 30%提高至 63%~74%[11-13],目前已经成为 PMP 最有效的治疗方案。然而,如何减少术后复发,改善患者生存情况仍是一项难题。本文就 PMP 的诊断、治疗进展综述如下。

1　诊　断

1.1　临床表现

PMP 临床表现差异较大,通常与腹腔内黏液积

基金项目:首都卫生发展科研专项基金(20200522-CHDRP-03)
通信作者:许洪斌,主任,主任医师,本科;航天中心医院黏液瘤科,北京市海淀区玉泉路 15 号(100049);E-mail:bdhtyy721nyl@163.com
收稿日期:2021-02-20;**修回日期**:2021-05-04

聚的程度有关。由于其起病隐匿，进展缓慢，早期往往没有任何症状或表现出易误诊为胃肠道疾病的轻微症状。随着腹腔内黏液不断的增加，逐渐出现腹胀、腹痛、腹腔积液、腹部包块以及饮食量减少、恶心、呕吐、肠梗阻等症状。部分患者可能出现类似急性阑尾炎表现的右下腹疼痛，而在女性患者中可能以妇科症状或卵巢肿物就诊，此外还可能表现出腹部以外的症状如腹股沟疝、黏液尿等[14-15]。

查体时可以发现腹部膨隆，腹围增大，肠鸣音减弱。腹部触诊有揉面感，压痛、反跳痛等腹膜刺激征往往不明显，有时可触及质韧的包块。由于黏液性腹水流动性差，移动性浊音常表现阴性。

1.2 影像学检查

1.2.1 彩 超

超声简单便捷，可做为 PMP 的首选检查方式。典型超声表现为[16-18]：腹盆腔大量流动性差、透声差的液性暗区，内有絮状、点状强回声及分隔带，当探头加压冲击时，暗区中点状强回声呈“礼花样”改变；肝、脾等脏器周边呈锯齿样或扇贝样；腹膜、大网膜弥漫性增厚；肠管广泛粘连，蠕动力减弱；有时可见肿大增粗的阑尾，呈“腊肠”样改变，甚至可见阑尾壁连续性中断。

罗丹等[19]指出超声对囊实性病变分辨率较好，可以更清楚地显示腹腔内黏液性病变及 CT 上无法显示的微小病灶，通过计算超声腹膜癌指数可以预测肿瘤的可切除性，对临床治疗方案的选择以及预后评估有着重要价值。

1.2.2 计算机体层扫描(CT)

CT 相对超声检查有其独特的优势，是 PMP 术前诊断及术后随访最常用的检查方式。典型 CT 表现为[20-21]：腹盆腔大量积液或多发类圆形囊实性肿块，积液密度通常高于常见腹腔积液的密度；增强扫描时无明显强化或仅肿瘤边缘轻微强化；肝、脾周围可见“贝壳样”压迹；病变局限在腹腔内，不会累及腹膜后间隙。CT 可以清楚地显示病变分布范围以及与各脏器之间的相互关系，但是对小肠及肠系膜受累情况的敏感性较低[22]。贾红敏等[23]提出通过 CT 多平面重建及容积重建技术评判 PMP 患者有无小肠袢聚集，预判小肠系膜有无挛缩。

Bouquot 等[24]专门针对 PMP 提出了一个 CT 评分系统，基于肝周区域 5 个位置，预测肿瘤可切除术，灵敏度为 94%，特异度为 81%。

1.2.3 磁共振成像(MRI)

MRI 与 CT 影像特征基本类似，但在区分黏液和普通腹腔积液时更敏感。MRI 还可以更好地显示肠壁与肿瘤的边界，评估小肠尤其是胃小弯、十二指肠受累情况[1]。

1.2.4 正电子发射型计算机断层扫描(PET-CT)

PET-CT 在 PMP 诊断中的作用尚有争议，通常认为在诊断 PMP 时价值不大，但 PET-CT 有助于发现腹腔以外的转移病灶[25-26]。也有研究认为 PET-CT在评估病理级别、能否完全减瘤和预测肿瘤复发时有一定帮助[27-29]。

1.2.5 全消化道造影

通过碘剂消化道造影，进一步了解肠道运动情况、有无扩张、梗阻以及肠系膜有无挛缩。需要注意的是，消化道造影应避免使用钡剂，以免加重肠梗阻。

1.3 肿瘤标志物

肿瘤标志物在 PMP 诊断方面的研究较少，虽然多数 PMP 患者会出现血清 CEA、CA125、CA199 等肿瘤标志物升高，但由于缺乏特异性，诊断价值有限[30]。有研究认为，通过检测肿瘤标志物水平可以评估肿瘤侵袭程度、腹膜肿瘤负荷以及肿瘤细胞增生活性[31]。而当联合检测腹腔积液肿瘤标志物时，可能有助于 PMP 病理级别的判断[32]。

肿瘤标志物主要用于术后复查和早期发现复发[33]。研究发现，肿瘤标志物升高的患者，术后复发的风险明显增高[34-36]，而在治疗后复查过程中出现肿瘤标志物升高，往往提示肿瘤复发[33,37]。

1.4 腹腔穿刺

腹腔穿刺是一种常用的诊断方式，但是对于 PMP 来说，由于腹腔积液大多为黏液或胶冻样，很难通过穿刺针抽出。另外，穿刺液里可能没有或仅少量的上皮细胞，很难通过典型细胞学特征进行诊断。但在腹腔穿刺中能够抽出黏液或胶冻样物，对 PMP 的诊断有重大的提示价值[38]。

1.5 腹腔镜检查

腹腔镜探查可以在较小创伤下尽可能地全面观察腹腔内病变情况，评估肿瘤程度，为制定手术方案提供依据，避免开腹给患者造成的损伤，同时可以进行多处活检，提高诊断的准确率[39-40]。

1.6 病理学诊断

PMP 病理学分类及诊断标志一直都具有挑战

性且存在争议,先后有过多种分类标准,甚至有过一些相互混淆和重叠的概念。最早的病理分类是Ronnett等[41]在1995年提出的,将PMP分为三种类型:播散性腹膜黏液性腺瘤病(disseminated peritoneal adenomucinosis,DPAM)、腹膜黏液癌病(peritoneal mucinous carcinomatosis,PMCA)、中间类型(peritoneal mucinous carcinomatosis with intermediate features,PMCA-I)。2016年腹膜表面肿瘤学国际联盟(PSOGI)达成的专家共识[42],将PMP分为4类:无细胞性黏液、腹膜低级别黏液癌、腹膜高级别黏液癌和腹膜高级别黏液癌伴印戒细胞(Table 1)。

免疫组织化学特异标志物有助于PMP来源的判断,阑尾来源通常表现为:CK7(+)、CK20(+)、CEA(+)、CDX2(+)、MUC2(+)、MUC5AC(+)、ER(–)和PR(–)[43]。

2 治　疗

2.1 手术治疗

手术一直是PMP的主要治疗方式。传统手术以简单减瘤为主,虽然短时间内可改善患者症状,但术后复发率高,需要多次手术,预后较差。目前公认最有效的治疗方式是CRS与HIPEC相结合的综合治疗策略[44-46]。尽管缺少前瞻性研究证实,但越来越多的回顾性研究报道了该方式取得的良好的临床效果。肿瘤细胞减灭最终目的是尽可能清除包括原发病灶、腹膜、大网膜及受累脏器在内所有肉眼可见的肿瘤。研究结果显示,肿瘤减灭程度(completeness of cytoreduction,CC)是影响预后的重要因素,完全减瘤患者的生存时间明显优于姑息减瘤者[11-13]。腹膜癌指数(peritoneal cancer index,PCI)是描述腹膜肿瘤累及程度的一个定量指标,高PCI被认为是CRS相对禁忌证,因为它显著增加了并发症的发生率和死亡率[47],以及对生活质量有重大影响[48]。Trilling等[49]提出针对高PCI患者,可采用两次手术进行减瘤,以降低手术并发症发生率和死亡率。由于多数PMP患者就诊时已经是疾病晚期,很难达到完全减瘤。翟喜超等[50]提出将患者分为临床早期和临床晚期,对于临床早期的患者应按照CRS程序,尽可能切除所有肉眼可见肿瘤,包括原发病灶、腹膜、大网膜以及受累及的脏器;而对于临床晚期的患者,则以解决患者主要症状,改善生活质量的个体化治疗策略为主,避免盲目追求完全减瘤而扩大手术范围,导致术后并发症增加,影响患者预后。

2.2 化　疗

2.2.1 腹腔灌注化疗

腹腔灌注化疗是治疗PMP重要的辅助方式,包括术中腹腔热灌注化疗(HIPEC)和术后腹腔灌注化疗。术中HIPEC在CRS完成后进行,有开放式和封闭式,化疗药物通常使用丝裂霉素、顺铂、奥沙利铂、5-氟尿嘧啶、多西他赛等,循环速度控制在400~1 000 mL,加热温度至(43±0.5) ℃,治疗时间60~90 min[13,51]。术后腹腔灌注化疗目前主要有两种方式,一种是普通腹腔灌注化疗,将化疗药物灌入腹腔,保留24 h后放出,不进行加热和循环;另一种是与术中基本相同的方式。

2.2.2 新辅助腹腔内化疗

Prabhu等[52]报道了27例首次评估无法达到完全减瘤的PMP患者,在接受新辅助腹腔内化疗后,54.5%的患者达到了完全减瘤,但是该研究未说明新辅助腹腔内化疗疗程数目。1例PMP患者接受12个疗程新辅助腹腔内化疗后,同样达到了完全减瘤[53]。这似乎为PMP的治疗提供了一项新的方式,有待更

Table 1　Pathological classification of PMP

PMP grade	Current terminology	Histologic feature
Grade 1	Acellular mucin	Mucin with no evidence of epithelial cells
	Low-grade mucinous carcinoma peritonei/disseminated peritoneal adenomucinosis (DPAM)	Pseudostratified or flat strips of epithelium with mild nuclear atypia, pattern of pushing invasion across a broad front and overall maintenance of cellular polarity
Grade 2	High-grade mucinous carcinoma peritonei/peritoneal mucinous carcinomatosis (PMCA)	Vesicular nuclei with prominent nucleoli, cellular stratification, cribriform or micropapillary architecture, elevated mitotic activity, high cellularity (at least 20% epithelial cells within mucin pools), irregular infiltrative glands or single cell with desmoplasia
Grade 3	High-grade mucinous carcinoma peritonei with signet-ring cells/peritoneal mucinous carcinomatosis with signet-ring cells (PMCA-S)	High grade histologic features, as reported above, with more than focal areas with signet-ring cell morphology (>10% of cells)

大样本验证。

2.2.3 全身化疗

全身化疗不是推荐的必要治疗程序，是否实施主要依赖于各中心治疗经验。术前新辅助化疗不能改善总生存期[54-55]，甚至有结果显示术前新辅助化疗是 PMP 的不良预后因素[56-57]。研究发现，低级别阑尾来源 PMP 患者在 CRS+HIPEC 治疗后，接受全身化疗无法获益，但高级别 PMP 患者接受全身化疗有延长整体生存期的趋势[55,58-60]。对于无法手术的晚期 PMP 患者，全身化疗或许可改善生存质量[61]，联合新血管生成抑制剂或许可延长生存期[62-63]。

2.2.4 其他治疗

PMP 其他治疗包括放疗[64]、免疫治疗、生物治疗等，文献中仅见个案报道，临床应用较少。

3 预 后

目前研究发现，减瘤程度、病理级别、肿瘤标志物以及 HIPEC 是影响 PMP 患者预后的重要因素。Chua 等[11]报道了一项来自 16 个中心 2 298 例 PMP 患者的研究，结果显示接受 CRS+HIPEC 治疗后中位生存时间为 196 个月，中位无进展生存期为 98 个月，3、5、10 和 15 年生存率分别为 80%、74%、63%和 59%，其中 CC-0/1 的患者 5 年生存率高达 85%。2016 年 Ansari 等[12]在单中心 1 000 例患者的研究指出，接受 CC-0/1 的患者 5 年和 10 年总生存率分别为 87.4%和 70.3%，而 CC-2/3 的患者 5 年和 10 年生存率分别为 39.2%和 8.1%。2020 年夏奥等[13]报道了国内最大样本量的单中心研究结果，5 年、10 年生存率分别为 52.7%、44.8%，CC-0/1 患者和 CC-2/3 患者 5 年、10 生存率分别为 77.0%、64.3%和 45.8%、39.4%。虽然与国外研究结果存在偏差，但仍显示出 CRS+HIPEC 良好的预后，也显示出完全减瘤是影响 PMP 患者预后的重要因素。然而 CRS+HIPEC 在带来良好生存效果的同时，也伴有较高的手术并发症发生风险，因此选择适当的患者和在有经验的治疗中心接受治疗是非常重要的。

肿瘤标志物升高是 PMP 预后不良的独立相关因素，Taflampas 等[34]报道了 519 例接受 CRS+HIPEC 治疗的阑尾来源 PMP 患者，术前肿瘤标志物正常的患者无病生存率和总体生存率显著更高。而 van Eden 等[36]的研究发现，当 CEA、CA125、CA199 高出正常水平 3 倍以上，比组织学亚型更能预测更差的预后。Zhou 等[65]荟萃分析近 20 年发表的文献，结果发现在接受减瘤手术但不能接受完全细胞减灭术的 PMP 患者中，低级别 PMP 比高级别 PMP 预后更好。

近年来，新型基因测序技术逐渐成熟并应用于 PMP 的基础研究。与 PMP 相关的基因主要有 *K-RAS*、*GNAS*、*TP53* 等，基因突变是 PMP 预后不良的相关因素[66]。

4 随 访

在手术后 5 年内，建议 3~6 个月复查 1 次，包括肿瘤标志物、腹盆腔增强 CT。5 年以后每年复查 1 次。尽管在 CRS+HIPEC 综合治疗策略下，PMP 获得良好的效果，但术后复发率仍高达 28%。而对于肿瘤复发后如何治疗，尚无统一的标准。但普遍认为，肿瘤复发患者再次治疗必须个体化，再次接受手术仍能获益。

5 小 结

腹膜假黏液瘤发病机制尚不清楚，虽然其生物学行为表现为惰性，但缺乏有效的治疗，预后较差。目前公认最有效的治疗方式为 CRS+HIPEC，部分患者接受治疗后可显著延长生存期。同时这也一项比较复杂的手术方式，手术并发症发生率高，建议在有经验的中心进行。

参考文献：

[1] Rizvi SA, Syed W, Shergill R. Approach to pseudomyxoma peritonei[J]. World J Gastrointest Surg, 2018, 10(5): 49–56.

[2] Smeenk RM, Velthuysen ML, Verwaal VJ, et al. Appendiceal neoplasms and pseudomyxoma peritonei: a population based study[J]. Eur J Surg Oncol, 2008, 34(2): 196–201.

[3] Mittal R, Chandramohan A, Moran B. Pseudomyxoma peritonei: natural history and treatment[J]. Int J Hyperthermia, 2017, 33(5): 511–519.

[4] Patrick-Brown T, Carr NJ, Swanson DM, et al. Estimating the prevalence of pseudomyxoma peritonei in europe using a novel statistical method [J]. Ann Surg Oncol, 2021, 28(1): 252–257.

[5] Yan F,Shi F,Li X,et al. Clinicopathological characteristics of pseudomyxoma peritonei originated from ovaries[J]. Cancer Manag Res,2020,12:7569-7578.

[6] Kataoka A,Ito K,Takemura N,et al. Immunohistochemical staining as supportive diagnostic tool for pseudomyxoma peritonei arising from intraductal papillary mucinous neoplasm: a report of two cases and literature review[J]. Pancreatology,2020,20(6):1226-1233.

[7] Gohda Y,Noguchi R,Horie T,et al. Pseudomyxoma peritonei of a mature ovarian teratoma caused by mismatch repair deficiency in a patient with Lynch syndrome: a case report[J]. BMC Med Genet,2016,17(1):94.

[8] Liang L,Zhou N,Xu H,et al. Urachal mucinous adenocarcinoma with pseudomyxoma peritonei: a case report[J]. Medicine (Baltimore),2017,96(35):e7548.

[9] Gong Y,Wang X,Zhu Z. Pseudomyxoma peritonei originating from transverse colon mucinous adenocarcinoma: a case report and literature review [J]. Gastroenterol Res Pract,2020,2020:5826214.

[10] Wambura C,Jusabani A,Sherman O,et al. Pseudomyxoma pleurii and peritonei secondary to sigmoid colon adenocarcinoma: a rare clinico-pathologico-radiological presentation[J]. Oxf Med Case Reports,2018,2018(10):omy057.

[11] Chua TC,Moran BJ,Sugarbaker PH,et al. Early- and long-term outcome data of patients with pseudomyxoma peritonei from appendiceal origin treated by a strategy of cytoreductive surgery and hyperthermic intraperitoneal chemotherapy[J]. J Clin Oncol,2012,30(20):2449-2456.

[12] Ansari N,Chandrakumaran K,Dayal S,et al. Cytoreductive surgery and hyperthermic intraperitoneal chemotherapy in 1000 patients with perforated appendiceal epithelial tumours[J]. Eur J Surg Oncol,2016,42(7):1035-1041.

[13] 夏奥,马瑞卿,翟喜超,等. 细胞减灭术+腹腔热灌注化疗治疗腹膜假黏液瘤—单中心 854 例经验总结[J]. 中国肿瘤临床,2020,47(3):145-151.
Xia A,Ma RQ,Zhai XC,et al. Cytoreductive surgery combined with hyperthermic intraperitoneal chemotherapy for pseudomyxoma peritonei: a single-center experience with 854 patients[J]. Chin J Clin Oncol,2020,47(3):145-151.

[14] Esquivel J,Sugarbaker PH. Clinical presentation of the pseudomyxoma peritonei syndrome[J]. Br J Surg,2000,87(10):1414-1418.

[15] Liu Y,Ishibashi H,Hirano M,et al. Cytoreductive surgery plus hyperthermic intraperitoneal chemotherapy for pseudomyxoma peritonei arising from urachus [J]. Ann Surg Oncol,2015,22(8):2799-2805.

[16] 霍慧萍. 彩色多普勒超声诊断腹膜假性黏液瘤的临床应用[J]. 内蒙古医科大学学报,2019,41(2):155-157.
Huo HP. Clinical application of color Doppler ultrasound in diagnosis of peritoneal pseudomyxoma [J]. Journal of Inner Mongolia Medical University,2019,41(2):155-157.

[17] 周南,梁蕾,郭君. 超声在腹膜假黏液瘤诊断及临床分级中的应用价值[J]. 医学研究杂志,2016,45(3):167-170.
Zhou N,Liang L,Guo J. Value of sonography in the diagnosis and pathologic staging of pseudomyxoma peritonei before surgery[J]. J Med Res, 2016,45(3):167-170.

[18] 陈仙秋,张燕绒,许崇永,等. 腹膜假性黏液瘤的 B 超诊断[J]. 影像诊断与介入放射学,2014,(5):376-379.
Chen XQ,Zhang YR,Xu CY,et al. Ultrasound diagnosis of pseudomyxoma peritonei [J]. Diagnostic Imaging & Interventional Radiology,2014,(5):376-379.

[19] 罗丹,梁蕾,周南,等. 超声评估腹膜假黏液瘤的腹膜癌指数与肿瘤可切除的相关研究[J]. 中国超声医学杂志,2021,37(3):285-288.
Luo D,Liang L,Zhou N,et al. Correlation between peritoneal cancer index and respectability of pseudomyxoma peritonei evaluated by ultrasonography[J]. Chinese Journal of Ultrasound in Medicine,2021,37(3):285-288.

[20] 于兰英,王旭. 腹膜假性黏液瘤的 CT、MRI 表现[J]. 实用放射学杂志,2016,32(5):737-741.
Yu LY,Wang X. The CT and MRI features of pseudomyxoma peritonei [J]. Journal of Practical Radiology, 2016,32(5):737-741.

[21] Sulkin TV,O'Neill H,Amin AI,et al. CT in pseudomyxoma peritonei: a review of 17 cases[J]. Clin Radiol,2002,57(7):608-613.

[22] Leimkühler M,de Haas RJ,Pol VEH,et al. Adding diagnostic laparoscopy to computed tomography for the evaluation of peritoneal metastases in patients with colorectal cancer: a retrospective cohort study[J]. Surg Oncol,2020,33:135-140.

[23] 贾红敏,翟喜超,王志群,等. 小肠袢聚集对预测腹膜假黏液瘤患者小肠系膜挛缩的 CT 价值研究 [J]. CT 理论与应用研究,2021,30(2):183-191.
Jia HM,Zhai XC,Wang ZQ,et al. CT value of small intestinal loop aggregation in predicting mesenteric contracture in patients with peritoneal pseudomyxoma[J]. CT Theory and Applications,2021,30(2):183-191.

[24] Bouquot M,Dohan A,Gayat E,et al. Prediction of resectability in pseudomyxoma peritonei with a new CT score[J]. Ann Surg Oncol,2018,25(3):694-701.

[25] Kim TH,Kim J,Kang YK,et al. Identification of metabolic biomarkers using serial ^{18}F-FDG PET/CT for prediction of recurrence in advanced epithelial ovarian cancer[J]. Transl Oncol,2017,10(3):297-303.

[26] Rohani P,Scotti SD,Shen P,et al. Use of FDG-PET imag-

ing for patients with disseminated cancer of the appendix [J]. Am Surg, 2010, 76(12): 1338–1344.

[27] Dubreuil J, Giammarile F, Rousset P, et al. FDG-PET/ceCT is useful to predict recurrence of pseudomyxoma peritonei [J]. Eur J Nucl Med Mol Imaging, 2016, 43(9): 1630–1637.

[28] Passot G, Glehen O, Pellet O, et al. Pseudomyxoma peritonei: role of ^{18}F-FDG PET in preoperative evaluation of pathological grade and potential for complete cytoreduction[J]. Eur J Surg Oncol, 2010, 36(3): 315–323.

[29] Hotta M, Minamimoto R, Gohda Y, et al. Impact of a modified peritoneal cancer index using FDG-PET/CT (PET-PCI) in predicting tumor grade and progression-free survival in patients with pseudomyxoma peritonei[J]. Eur Radiol, 2019, 29(10): 5709–5716.

[30] 宋燕, 何美琳, 邵冬华, 等. 常用血清肿瘤标志物对腹膜假黏液瘤诊断价值的初步探讨 [J]. 医学研究杂志, 2016, 45(5): 126–129.
Song Y, He ML, Shao DH, et al. Investigation on the diagnosis value of serum tumor markers in patients with pseudomyxoma peritonei[J]. Journal of Medical Research, 2016, 45(5): 126–129.

[31] 于洋, 李鑫宝, 林育林, 等. 肿瘤细胞减灭术联合腹腔热灌注化疗治疗腹膜癌 1384 例疗效分析[J]. 中华胃肠外科杂志, 2021, 24(3): 230–239.
Yu Y, Li XB, Lin YL, et al. Efficacy of 1384 cases of peritoneal carcinomatosis underwent cytoreductive surgery plus hyperthermic intraperitoneal chemotherapy[J]. Chinese Journal of Gastrointestinal Surgery, 2021, 24(3): 230–239.

[32] 马福存, 柏明见, 何美琳, 等. 初步探讨三种腹水/血清肿瘤标志物对腹膜假粘液瘤的诊断价值 [J]. 标记免疫分析与临床, 2019, 26(5): 772–776.
Ma FC, Bai MJ, He ML, et al. A preliminary investigation of the diagnostic values of three ascites/serum tumor markers for peritoneal pseudomyxoma[J]. Labeled Immunoassays and Clinical Medicine, 2019, 26(5): 772–776.

[33] Wagner PL, Austin F, Sathaiah M, et al. Significance of serum tumor marker levels in peritoneal carcinomatosis of appendiceal origin[J]. Ann Surg Oncol, 2013, 20(2): 506–514.

[34] Taflampas P, Dayal S, Chandrakumaran K, et al. Pre-operative tumour marker status predicts recurrence and survival after complete cytoreduction and hyperthermic intraperitoneal chemotherapy for appendiceal pseudomyxoma peritonei: analysis of 519 patients[J]. Eur J Surg Oncol, 2014, 40(5): 515–520.

[35] Koh JL, Liauw W, Chua T, et al. Carbohydrate antigen 19-9 (CA 19-9) is an independent prognostic indicator in pseudomyxoma peritonei post cytoreductive surgery and perioperative intraperitoneal chemotherapy[J]. J Gastrointest Oncol, 2013, 4(2): 173–181.

[36] van Eden WJ, Kok NFM, Snaebjornsson P, et al. Factors influencing long-term survival after cytoreductive surgery and hyperthermic intraperitoneal chemotherapy for pseudomyxoma peritonei originating from appendiceal neoplasms[J]. BJS Open, 2019, 3(3): 376–386.

[37] Alexander-Sefre F, Chandrakumaran K, Banerjee S, et al. Elevated tumour markers prior to complete tumour removal in patients with pseudomyxoma peritonei predict early recurrence[J]. Colorectal Dis, 2005, 7(4): 382–386.

[38] Darr U, Renno A, Alkully T, et al. Diagnosis of Pseudomyxoma peritonei via endoscopic ultrasound guided fine needle aspiration: a case report and review of literature [J]. Scand J Gastroenterol, 2017, 52(5): 609–612.

[39] Seshadri RA, Hemanth Raj E. Diagnostic laparoscopy in the pre-operative assessment of patients undergoing cytoreductive surgery and hipec for peritoneal surface malignancies[J]. Indian J Surg Oncol, 2016, 7(2): 230–235.

[40] Sommariva A, Zagonel V, Rossi CR. The role of laparoscopy in peritoneal surface malignancies selected for hyperthermic intraperitoneal chemotherapy(HIPEC)[J]. Ann Surg Oncol, 2012, 19(12): 3737–3744.

[41] Ronnett BM, Zahn CM, Kurman RJ, et al. Disseminated peritoneal adenomucinosis and peritoneal mucinous carcinomatosis. A clinicopathologic analysis of 109 cases with emphasis on distinguishing pathologic features, site of origin, prognosis, and relationship to "pseudomyxoma peritonei"[J]. Am J Surg Pathol, 1995, 19(12): 1390–1408.

[42] Carr NJ, Cecil TD, Mohamed F, et al. A consensus for classification and pathologic reporting of pseudomyxoma peritonei and associated appendiceal neoplasia: the results of the Peritoneal Surface Oncology Group International (PSOGI) modified delphi process [J]. Am J Surg Pathol, 2016, 40(1): 14–26.

[43] Ramaswamy V. Pathology of mucinous appendiceal tumors and pseudomyxoma peritonei[J]. Indian J Surg Oncol, 2016, 7(2): 258–267.

[44] Sugarbaker PH. Peritonectomy procedures[J]. Ann Surg, 1995, 221(1): 29–42.

[45] Sugarbaker PH. New standard of care for appendiceal epithelial neoplasms and pseudomyxoma peritonei syndrome?[J]. Lancet Oncol, 2006, 7(1): 69–76.

[46] 李雁, 许洪斌, 彭正, 等. 肿瘤细胞减灭术加腹腔热灌注化疗治疗腹膜假黏液瘤专家共识 [J]. 中华医学杂志, 2019, 99(20): 1527–1535.
Li Y, Xu HB, Peng Z, et al. Expert consensus on the

treatment of peritoneal pseudomyxoma with tumor cytoreductive surgery and intraperitoneal hyperthermic perfusion chemotherapy[J]. Natl Med J China,2019,99(20):1527–1535.

[47] Saxena A,Yan TD,Chua TC,et al. Critical assessment of risk factors for complications after cytoreductive surgery and perioperative intraperitoneal chemotherapy for pseudomyxoma peritonei[J]. Ann Surg Oncol,2010,17:1291–1301.

[48] Leimkühler M,Hentzen JEKR,Hemmer PHJ,et al. Systematic review of factors affecting quality of life after cytoreductive surgery with hyperthermic intraperitoneal chemotherapy[J]. Ann Surg Oncol,2020,27(10):3973–3983.

[49] Trilling B,Brind´Amour A,Hamad R,et al. Two-step cytoreductive surgery and hyperthermic intraperitoneal chemotherapy for pseudomyxoma peritonei with high peritoneal carcinomatosis index[J]. World J Surg Oncol,2021,19(1):60.

[50] 翟喜超,夏奥,马瑞卿,等. 阑尾源性腹膜假黏液瘤单中心诊疗经验[J]. 中国肿瘤临床,2020,47(9):460–464.
Zhai XC,Xia A,Ma RQ,et al. Management of pseudomyxoma peritonei originating from the appendix:a single-center experience [J]. Chinese Journal of Clinical Oncology,2020,47(9):460–464.

[51] 李鑫宝,林育林,姬忠贺,等. 肿瘤细胞减灭术加腹腔热灌注化疗治疗腹膜假黏液瘤 182 例分析 [J]. 中国肿瘤临床,2018,45(18):943–949.
Li XB,Lin YL,Ji ZH,et al. Cytoreductive surgery plus hyperthermic intraperitoneal chemotherapy in patients with pseudomyxoma peritonei:analysis of 182 patients at a single center [J]. Chinese Journal of Clinical Oncology,2018,45(18):943–949.

[52] Prabhu A,Brandl A,Wakama S,et al. Neoadjuvant intraperitoneal chemotherapy in patients with pseudomyxoma peritonei-a novel treatment approach[J]. Cancers (Basel),2020,12(8):2212.

[53] Padmanabhan N,Ishibashi H,Nishihara K,et al. Complete pathological response of high grade appendicular neoplasm induced pseudomyxoma peritonei(PMP) after neoadjuvant intra-peritoneal chemotherapy: a case report [J]. Int J Surg Case Rep,2020,72:117–121.

[54] Munoz-Zuluaga CA,King MC,Ledakis P,et al. Systemic chemotherapy before cytoreductive surgery and hyperthermic intraperitoneal chemotherapy (CRS/HIPEC) in patients with high-grade mucinous carcinoma peritonei of appendiceal origin[J]. Eur J Surg Oncol,2019,45(9):1598–1606.

[55] Blackham AU,Swett K,Eng C,et al. Perioperative systemic chemotherapy for appendiceal mucinous carcinoma peritonei treated with cytoreductive surgery and hyperthermic intraperitoneal chemotherapy[J]. J Surg Oncol,2014,109(7):740–745.

[56] Austin F,Mavanur A,Sathaiah M,et al. Aggressive management of peritoneal carcinomatosis from mucinous appendiceal neoplasms [J]. Ann Surg Oncol,2012,19(5):1386–1393.

[57] Baratti D,Kusamura S,Nonaka D,et al. Pseudomyxoma peritonei:clinical pathological and biological prognostic factors in patients treated with cytoreductive surgery and hyperthermic intraperitoneal chemotherapy (HIPEC) [J]. Ann Surg Oncol,2008,15(2):526–534.

[58] Lu P,Fields AC,Meyerhardt JA,et al. Systemic chemotherapy and survival in patients with metastatic low-grade appendiceal mucinous adenocarcinoma [J]. J Surg Oncol,2019,120(3):446–451.

[59] Asare EA,Compton CC,Hanna NN,et al. The impact of stage,grade,and mucinous histology on the efficacy of systemic chemotherapy in adenocarcinomas of the appendix: analysis of the national cancer data base[J]. Cancer,2016,122(2):213–221.

[60] Hiraide S,Komine K,Sato Y,et al. Efficacy of modified FOLFOX6 chemotherapy for patients with unresectable pseudomyxoma peritonei[J]. Int J Clin Oncol,2020,25(4):774–781.

[61] Raimondi A,Corallo S,Niger M,et al. Metronomic capecitabine with cyclophosphamide regimen in unresectable or relapsed pseudomyxoma peritonei[J]. Clin Colorectal Cancer,2019,18(2):e179–e190.

[62] Hirano S,Gohda Y,Miyazaki H,et al. A case of pseudomyxoma peritonei successfully treated with trifluridine/tipiracil (TAS-102) and bevacizumab after palliative debulking surgery[J]. Chin Clin Oncol,2021,10(3):29.

[63] Choe JH,Overman MJ,Fournier KF,et al. Improved survival with anti-vegf therapy in the treatment of unresectable appendiceal epithelial neoplasms[J]. Ann Surg Oncol,2015,22(8):2578–2584.

[64] McGrath C,Linden K,Hube P,et al. Palliative radiation therapy for symptom control in an advanced case of pseudomyxoma peritonei[J]. Cureus,2017,9(6):e1407.

[65] Zhou S,Zhao H,He X. The Prognostic impact of pathology on patients with pseudomyxoma peritonei undergoing debulking surgery: a systematic review and meta-analysis of retrospective studies[J]. Front Surg,2020,7:554910.

[66] 顾依然,杨华,陆维祺,等. 腹膜假黏液瘤的基础研究进展[J]. 肿瘤学杂志,2020,26(10):907–913.
Gu YR,Yang H,Lu WQ,et al. Advance on the basic research of pseudomyxoma peritonei[J]. Journal of Chinese Oncology,2020,26(10):907–913.