

## · 论著 · 罕见病 ·

## 特发性肠系膜静脉硬化性结肠炎一例报道及文献复习

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【摘要】目的 探讨特发性肠系膜静脉硬化性结肠炎的临床特征、诊断要点及治疗策略。方法 回顾性分析徐州市中医院2024年4月收治的1例IMP患者的临床资料，结合文献复习，总结其临床表现、影像学与内镜特征、病理表现及治疗方法。结果 患者为68岁女性，长期服用含栀子苷的中成药，临床表现为间断性腹泻，影像学显示结肠壁增厚、肠系膜血管钙化，内镜下见黏膜紫蓝色改变、多发溃疡，病理提示慢性活动性炎症伴纤维化。经对症治疗病情好转出院。结论 IMP是一种与长期服用含栀子苷中草药相关的罕见结肠疾病，诊断需综合病史、影像、内镜及病理，治疗应个体化，包括停药、对症处理或手术治疗，并加强长期随访。

【关键词】静脉硬化性结肠炎；特发性结肠炎；栀子苷；肠系膜静脉钙化；病例报告

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## Idiopathic Mesenteric Phlebosclerotic Colitis: a Case Report and Literature Review

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**Abstract: Objective** To investigate the clinical features, diagnostic points, and treatment strategies of idiopathic mesenteric phlebosclerotic colitis (IMP). **Methods** The clinical data of one IMP patient admitted to Xuzhou Hospital of Traditional Chinese Medicine in April 2024 were retrospectively analyzed. Literature review was conducted to summarize its clinical manifestations, imaging and endoscopic features, pathological findings, and treatment approaches. **Results** The patient was a 68-year-old female with a long-term history of taking Chinese patent medicines containing geniposide. Clinical presentation included intermittent diarrhea. Imaging showed colonic wall thickening and mesenteric vascular calcification. Endoscopy revealed bluish-purple mucosal discoloration and multiple ulcers. Pathology indicated chronic active inflammation with fibrosis. The patient improved after symptomatic treatment and was discharged. **Conclusion** IMP is a rare colonic disease associated with long-term use of geniposide-containing herbal medicines. Diagnosis requires comprehensive evaluation of history, imaging, endoscopy, and pathology. Treatment should be individualized, including drug cessation, symptomatic management, or surgical intervention, with emphasis on long-term follow-up.

**Keywords: Phlebosclerotic Colitis; Idiopathic Colitis; Geniposide; Mesenteric Venous Calcification; Case Report**

特发性肠系膜静脉硬化性结肠炎(idiopathic mesenteric phlebosclerosis, IMP)临床发病少见，最初由日本学者首先报道<sup>[1-2]</sup>。患者临床初诊时，由于本病罕见，往往给接诊医师在诊疗上带来一定困难。本院于2024年4月收治IMP确诊患者一例，现笔者结合该患者临床诊疗经过及既往文献，讨论特发性静脉硬化性结肠炎的临床特征及诊疗要点。

## 1 病例报告

患者女性，68岁，主诉“间断性腹泻不适1年。”入院。患者近1年来无明显诱因出现间断性腹泻不适，家属未知晓，未予重视，未行特殊治疗。后因家属知晓上述症状后急送至我院诊治。追问病史，既往曾长期服用多种中成药。查体：患者面色呈青蓝色，眶周皮肤见色素沉着；腹平腹软，无腹痛；直肠指检阴性。急诊全腹CT平扫示：结肠、直肠壁弥漫性增厚、密度减低，局部边缘及肠系膜血管见散在致密影，周围脂肪间隙模糊；腹膜见散在小淋巴结影。腹腔内见少量液性密度；腹主动脉较细(见图1)。

急诊予以收住入院，实验室检查结果示：WBC  $9.57 \times 10^9$ /

L, HGB 103g/L, AMYL 149U/L,  $K^+$  3.0mmol/L, 瘤标(-)，乙、丙、戊肝病毒肝炎(-)，梅毒、艾滋(-)。入院后给予对症治疗，并完善电子结肠镜检查。结肠镜检查镜下示：(升结肠及横结肠)粘膜广泛水肿并多发溃疡，并呈广泛紫蓝色改变，肠壁硬化性改变(见图2)。于回盲部溃疡边缘行多处内镜下活组织钳夹取样，病理示：粘膜重度慢性活动性炎，肉芽组织形成伴坏死，局灶间质纤维组织增生伴玻璃样变性(见图3)。

结合患者病史、临床症状、放射影像学及内镜、活检病理结果，诊断IMP。予患者行对症治疗后一般情况良好，要求出院，予以出院。嘱出院后：温软饮食，三月后我院复查肠镜，暂停逍遥丸服药，不适随诊。

## 2 文献复习

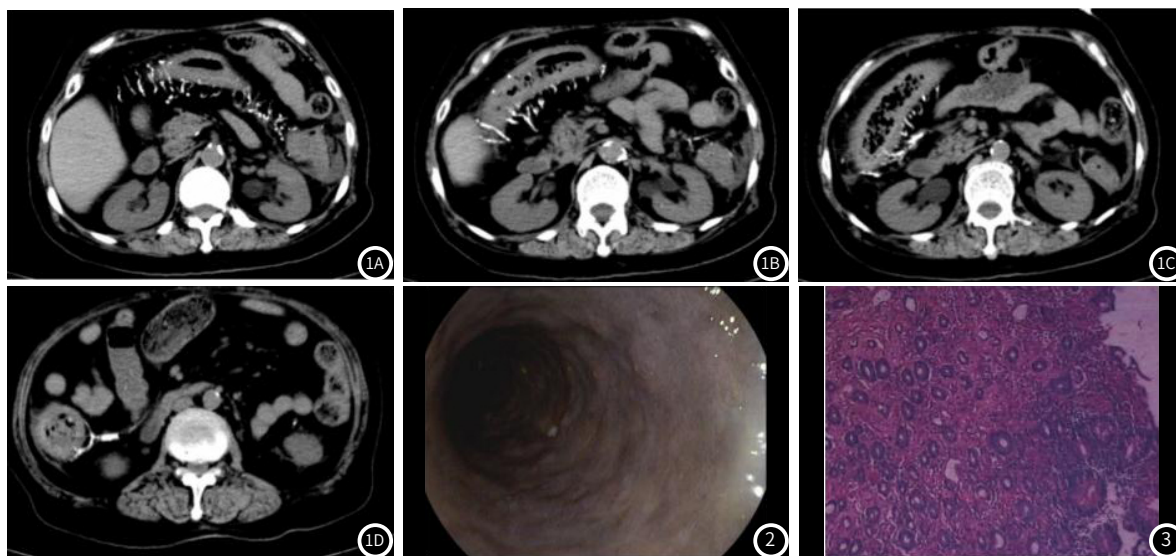
IMP是一种罕见的临床疾病，1991年由日本学者Koyama等予以首次报道，2003年由Iwashita等正式命名“特发性肠系膜硬化性肠炎”。本病好发于中老年人群，女性发病率较男性高。国内当前病例报道病例男性多于女性，可能与患者虽接受相关治疗但并未被报道有关。

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本病的病因尚不清楚,既往文献及研究认为本病可能与患者长期服用含有栀子苷成份的中草药相关<sup>[3]</sup>,本文报道病例曾长期口服中成药“丹栀逍遥丸”,其成份包含“焦栀子”,与文献报道吻合。相关研究认为,栀子苷经肠道细菌分解后形成京尼平,当其吸收后与肠系膜血管中大蛋白质等物质形成蓝色颗粒,进而引起肠系膜静脉色素沉着、硬化或钙化<sup>[3-4]</sup>;亦有研究显示,栀子苷可通过SCF/c-kit信号通路促进黑素生成,其中c-kit受体在去

甲肾上腺素的作用下表皮黑素细胞明显增多<sup>[5]</sup>,这可能与患者面部皮肤及结肠粘膜呈青蓝色改变相关<sup>[6]</sup>。文献报道有患者面部有蓝色色素沉着,经停药半年后面部青蓝色沉着改善。本文报道病例面部呈明显青蓝色改变,伴眶周皮肤明显沉着,应与上述因素相关。当前文献报道本病的发病主要集中于东亚地区,因而考虑有基因遗传相关性,但笔者认为介于中草药的应用主要集中于东亚地区,可能构成东亚地区的高发病率要素之一<sup>[7-8]</sup>。



**图1** 全腹CT平扫:结肠、直肠壁弥漫性增厚、密度减低,局部边缘及肠系膜血管见散在致密影,周围脂肪间隙模糊;腹膜见散在小淋巴结影。腹腔内见少量液性密度;腹主动脉较细。**图2** 结肠镜:粘膜广泛水肿并多发溃疡,并呈广泛紫蓝色改变,肠壁硬化性改变。**图3** 病理HE染色:粘膜重度慢性活动性炎,肉芽组织形成伴坏死,局灶间质纤维组织增生伴玻璃样变性。

IMP的病变主要累及右半结肠<sup>[9]</sup>,因其主要累及静脉系统致肠壁吸收回流障碍,肠壁水肿慢性炎症以致硬化,故而CT等影像学表现亦以此为基础。CT平扫除可提示上述解剖学改变外,其以血管受累为基础的点状、线状、弹簧样高密度钙化影,沿静脉分布的广泛均匀钙化,肠系膜慢性充血所致的较为均匀的肠壁增厚是其影像学特征<sup>[10]</sup>;增强CT扫描可见肠壁分层、肠壁周围系膜增厚、以及肠管周围反应性淋巴结增生等改变<sup>[11]</sup>。肠系膜血管疾病的影像谱系分析为IMP的鉴别提供了更广阔的视角<sup>[12]</sup>。

本文所报道的肠镜下表现亦与本文所报道病例相契合,肠镜检查多可见病变结肠粘膜色素沉着、粘膜充血水肿并多发溃疡或糜烂灶。既往文献认为紫蓝色粘膜为本病的特征性内镜下表现<sup>[10]</sup>,胃肠道黏膜异常变色的诊断与鉴别诊断思路对于识别此特征具有重要参考价值<sup>[13]</sup>。本文所报道的肠镜下表现亦与本次所报道病例相契合,但同时该病应与动脉硬化所致缺血性肠病、门脉高压、血吸虫肠病等相鉴别<sup>[14]</sup>。

本病的病理表现多为:粘膜炎症改变,可伴出血,肠血管壁增厚、玻璃样变及钙化,肉芽肿形成,间质纤维组织增生,肠壁坏死灶等,同时可伴有炎症反应性淋巴结增生<sup>[15]</sup>。然而类似的病理表现亦可见于其它炎性肠病<sup>[16]</sup>,因而并不具备特异性,故笔者认为该病的确诊应综合病史、影像学、病理学检查,从而为后续治疗提供指向。

有关IMP的治疗相关文献较辅助诊断文献为少,且由于本病发病少见,且其下消化道症状表现多样,可无症状,或有腹

痛、腹胀、便秘、腹泻、便血等临床症状<sup>[17]</sup>,笔者结合所报道病例及既往外文献<sup>[15]</sup>对其治疗手段归纳如下,对于无明显症状或无明显并发症的病例可予解除发病因素并行定期复查;合并明显消化道症状患者在解除发病因素外可予对症治疗;对于合并严重便血、穿孔等急腹症患者应行紧急外科手术干预,其手术方式应结合围手术期实验室及影像学检查做细致规划,行右半结肠切除、次全结肠切除、全结肠切除或结肠造瘘等<sup>[18]</sup>;当该病的致病诱因解除后,文献报道面部色素沉着得到改善,但相关结肠及血管病变是否可逆未见明确文献报道及相关研究,因而加强该类疾病患者的长期随访应被纳入临床诊疗的一项重要。另由于本病多发生于老年人,多数长期服用中成药的老年人常合并其他慢性基础疾病,因而应着重加强相关患者围手术期管理<sup>[19]</sup>。

### 3 总结

IMP发病少见,多集中于长期服用包含栀子苷成份中草药的老年人,临床诊疗工作中,尤其对于消化科和普外科医师,应加强对该病的认识。影像学、内镜、病理学检查是其确诊的必要手段<sup>[21]</sup>。治疗应结合患者病变情况给予恰当的方式,包含临床随诊的患者和行相应手术治疗的,在去除该病致病因素的同时,应加强患者随访,以便患者得到及时、妥善的临床干预<sup>[19-20]</sup>。临床实践中可参考最新的炎症性肠病诊疗质量控制评估体系,以规范此类罕见病的长期管理流程<sup>[21]</sup>。

(参考文献下转第 14 页)

时应警惕该疾病的存在,建议给予及时有效的黏膜活检以早期发现、早期治疗,避免疾病进展。虽然目前多数文献表明该疾病预后良好,笔者查阅文献发现报道的案例多为中老年人,能够接受手术及后期的放化疗来改善预后,但对于高龄无法耐受手术风险的患者,治疗方案的选择相对棘手,单一的化疗被认为是首选方案,但该例患者最终出现了疾病进展,新的化疗方案仍是原发性胃肠道EMP亟需探索的关键。由于病例的罕见,更多的数据需要被收集分析,以期为临床医师提供一定的参考价值。

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