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急性旁中心中层黄斑病变1例

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摘 要

急性旁中央中层黄斑病变(Paracentral acute middle maculopathy, PAMM)是Sarraf等人首先通过光学相干断层扫描发现，是位于视网膜中部的中间毛细血管丛(ICP)水平缺血的结果。已经确定了各种病因，包括局部血管闭塞性疾病，如视网膜动脉闭塞和视网膜中央静脉闭塞(CRVO)，以及全身性疾病，如糖尿病视网膜病变、远达性视网膜病变和镰状细胞视网膜病变。此外，一些特定药物的使用或代谢紊乱，如高同型半胱氨酸血症，也被认为是可能的致病因素。本文讲述1例视力、体征治疗后均明显好转的PAMM。

关键词

急性旁中心中层黄斑病变，光谱域光相干断层扫描，扫频源光学相干断层扫描血管照影，病因

Paracentral Acute Middle Maculopathy: A Case Report

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Abstract

Paracentral acute middle maculopathy (PAMM) is a tomographic finding firstly described by Sarraf *et al.* as the results of ischemia at the level of the intermediate capillary plexus (ICP) located in the middle retina. Various aetiologies have been identified which include local vascular occlusive disorders such as retinal arterial occlusion and central retinal vein occlusion (CRVO), as well as systemic diseases such as diabetic retinopathy, purtscher retinopathy and sickle cell retinopathy. This article describes a case of PAMM, whose visual acuity and physical signs improved significantly after treatment.

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Keywords

Paracentral Acute Middle Maculopathy (PAMM), Swept-Domain Optical Coherence Tomography (SS-OCT), Swept-Source Optical Coherence Tomography Angiography (SS-OCTA), Pathophysiology

1. 病例介绍

患者，女，43 岁，于 2024 年 7 月 23 日因“左眼突然视视力下降 5.5 小时”来我院眼科门诊就诊。患者就诊时最佳矫正视力为右眼 0.8 左眼 0.1。裂隙灯显微镜检查双眼前节未见明显异常；散瞳间接检眼镜检查眼底双眼未见明显异常。NCT：右眼 15 mmHg，左眼 18 mmHg。

既往史：患者否认眼部手术及外伤史，否认糖尿病、高血压、冠心病等慢性病史，否认乙肝、结核等传染病史，否认输血及血制品输入史，否认药物滥用和吸烟史，否认近期疫苗接种史。

辅助检查：光谱域光相干断层扫描(swept-domain optical coherence tomography, SS-OCT)可见左眼内核层(inner nuclear layer, INL)条带状强反射(图 1)；扫频源光学相干断层扫描血管照影(swept-source optical coherence tomography angiography, OCTA)右眼各层未见明显异常；左眼浅层毛细血管丛未见明显异常，深毛细血管丛可见黄斑旁中心多发大小不等的蕨类植物样强反射病灶(图 2)；视野检查左眼多个旁中心暗点(图 3)。颈动脉彩色多普勒超声提示双侧颈总动脉斑块形成；头部 MRA 未见异常。

化验提示：同型半胱氨酸升高(12.6 umol/L)，血常规、炎性指标、脑脊液常规、生化及抗核抗体谱未见明显异常。

结合患者病史及各项检查，考虑诊断为左眼急性旁中心中层黄斑病变(paracentral acute middle maculopathy, PAMM)，予以下治疗：舌下含服硝酸甘油片 0.5 mg，90% O_2 + 10% CO_2 高流量持续吸氧，左眼球后注射山莨菪碱注射液 + 地塞米松磷酸钠注射液 + 盐酸利多卡因注射液，每日 1 次，连续 3 天。3 天后复查，患者左眼矫正视力为 0.8。

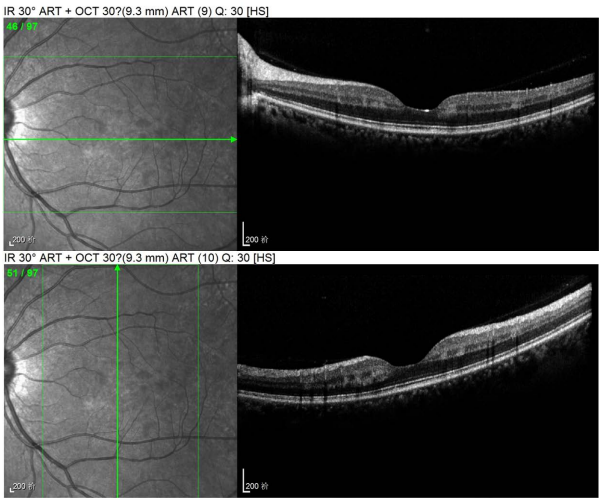

Figure 1. Spectral domain optical coherence tomography (SD-OCT)
图 1. 光谱域光学相干断层扫描(SD-OCT)

描述：扫频源光相干断层扫描可见左眼黄斑旁内核层条带状强反射。

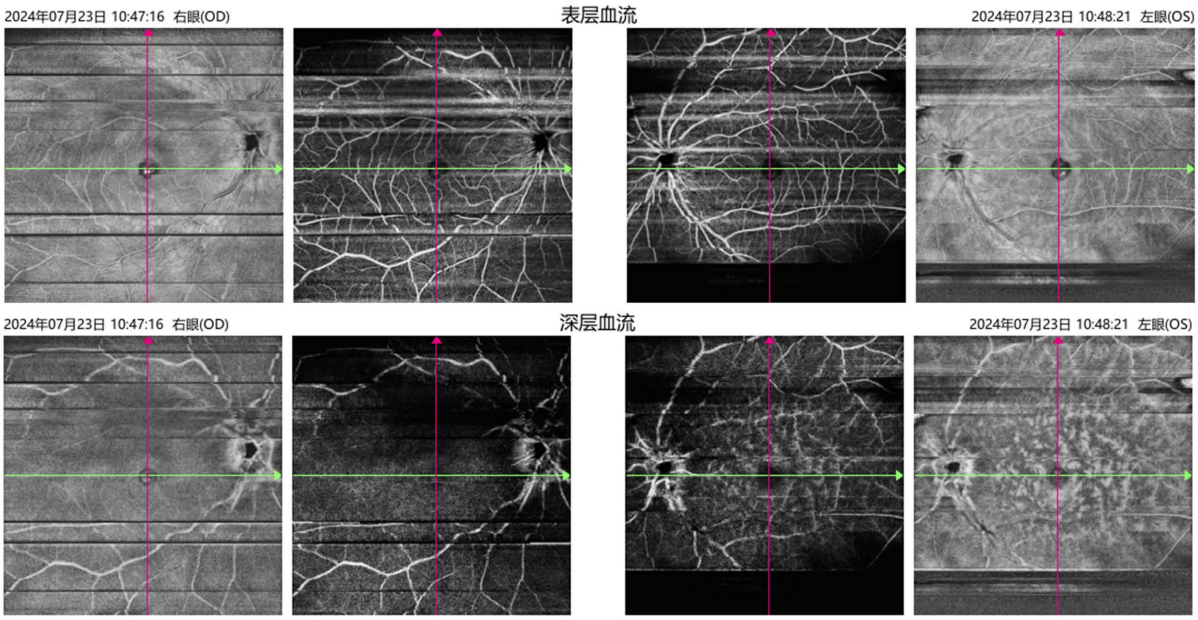

Figure 2. Swept-source optical coherence tomography angiography (SS-OCTA)
图 2. 扫频源光相干断层扫描(SS-OCTA)

描述：双眼表层血流未见明显异常；右眼深层血流未见明显异常，左眼深层血流可见黄斑旁中心多发大小不等的蕨类植物样强反射病灶。

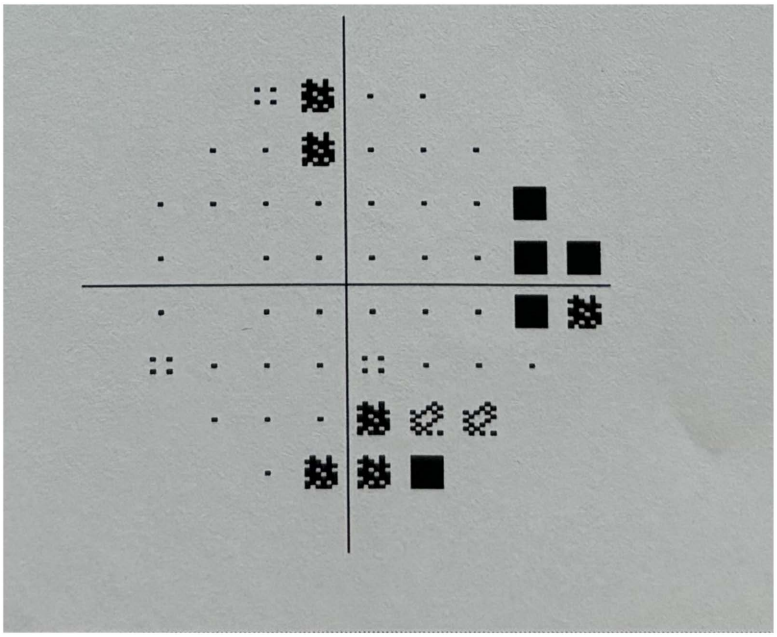

Figure 3. Visual fields
图 3. 视野

描述：可见左眼旁中心暗点。

2. 讨论

急性旁中心中层黄斑病变(Paracentral acute middle maculopathy, PAMM)是一种通常由光学相干断层扫描(OCT)发现的疾病，发生在广泛的视网膜血管疾病中，表现为内核层(INL)的高反射带，可累及或不累及到邻近的内、外丛状层(IPL, OPL) [1] [2]。PAMM 提示深血管复合体，特别是深视网膜毛细血管丛(DCP)内灌注不足引起的视网膜梗死[2]。随着 PAMM 的解决，INL 变薄的遗留问题随之会出现[1] [3]。该患者的 OCTA 深层血流灌注异常，与多项研究一致。

一些眼部疾病已有报道与 PAMM 相关。鸟枪弹样脉络膜视网膜病变的 PAMM 可能是由于视网膜毛细血管丛的短暂闭塞[4]；内眼手术如超声乳化手术[5] [6]、睫状体部玻璃体切除术治疗牵引性视网膜脱离[7]、玻璃体积血[8]、视网膜前膜剥除[9]，外眼手术如翼状胬肉切除术[10]，有报道 PAMM 与以上这些内、外眼手术相关。正常超声乳化手术后的 PAMM 可能由于视网膜中央动脉(CRA)痉挛或短暂闭塞而发生[5] [6]。可能的影响因素包括血管病变危险因素或眼周麻醉导致眼压升高和通过 CRA 的血流减少[11]以及术后即刻早期眼压升高。在最近的一项研究中，增殖性糖尿病视网膜病变玻璃体切除术后 PAMM 的发生率为 3.8% [7]。最后，眼受压导致 CRA 和睫状体后动脉的血流损伤引起全眼缺血也被发现是 PAMM 的原因[12]。翼状胬肉切除术使用的眼周麻醉可能是外眼手术中黄斑灌注不足的原因[10]。PAMM 在全身性疾病中也有被发现，如特发性颅内高压[13]、脑膜炎[14]。颈动脉疾病也与 PAMM 有关，包括颈内动脉夹层和高流量颈动脉海绵窦瘘[15]。典型的 PAMM 体征也被描述为与 Susac 综合征[16]、网状脂肪肝[17]、抗磷脂综合征[18] [19]和巨细胞动脉炎[20] [21]有关。血管病变危险因素可能与 PAMM 有关，包括蛛网膜下腔出血或低血压、血脂异常[22]、药物(包括口服避孕药)、偏头痛、贫血和咖啡因摄入[1] [23] [24]。药物性 PAMM 的报道越来越多。服用舒马曲坦治疗偏头痛与 PAMM 有关[25]。这种情况下的发病机制可能是多因素的，包括偏头痛引起的血管痉挛和药物通过选择性 5-羟色胺 1d 受体激动剂活性诱导的 DCP 血管收缩[25]。该例患者无特殊相关病史，仅检查发现同型半胱氨酸升高、双侧颈总动脉内中膜增厚，推测其发病可能与上述原因有关。

PAMM 可为单侧或双侧[26]。PAMM 患者通常表现为突发性的一个或多个中心旁暗点[23] [26]。患者可主诉中心视力模糊或难以聚焦[26]。视敏度(VA)可能正常或略有下降[1] [23] [26]。平均发病年龄 49~53 岁，无性别偏好[27] [28]。该患者单侧突发性发病，视野检查多个中心旁暗点。

必须与 PAMM 区分开来的是急性黄斑区神经视网膜病变(acute macular neuroretinopathy, AMN)。虽然 AMN 与 PAMM 有共同的方面，比如危险因素(高凝状态、口服避孕药、吸烟或与前期病毒综合征的关联) [29]和临床表现(伴有或不伴有视力改变的中央旁暗点)，但是它们具有不同的形态学特征。在眼底检查中，AMN 通常表现为扁平的红棕色楔形病变，顶点指向中央凹，而 PAMM 则表现为深灰色病变[3]。这两种疾病都可能没有明显的眼底病变[30]。在 OCT 成像上，AMN 急性期的超反射带比 PAMM 深，位于外丛状/外核层交界处演变为椭球带和外限带的衰减[31]。

目前，PAMM 的治疗方向主要通过以下这两点，一是识别和治疗相关的血管病变，二是控制系统性危险因素。眼局部外用前列腺素 E1 后视力有提升[32]和舌下含服硝酸甘油后出现的盲点消退[33]近年来有被报道过，也有研究认为对于由视网膜动脉不完全阻塞引起的 PAMM 患者予以经验性的低剂量阿司匹林治疗可减少其进展为视网膜动脉完全阻塞进而视力丧失的可能性[34]。该例患者经连续 3 天改善微循环后，自觉患眼视力较前明显提升，提示对于 PAMM，早期诊断并及时予以改善微循环、营养神经等治疗对患者视力预后可能有积极作用。

诊断检查应个体化，并考虑患者既往病史、心血管危险因素的存在以及眼部和全身检查结果。在一些患者中，即使经过广泛的检查，也无法确定病因。急性旁中心中层黄斑病变是一种高度依赖 SD-OCT

扫描的疾病，可能代表视网膜内缺血的潜在过程。SD-OCT 和 OCT-A 技术的发展使人们更容易发现PAMM。本病例突出了眼多模态成像在诊断视网膜疾病和不明原因的视觉症状患者中的重要性。

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