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点状内层脉络膜病变一例

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摘　要

点状内层脉络膜病变(punctate inner choroidopathy, PIC)是一类较罕见的眼底疾病，包括脉络膜内层和视网膜色素上皮层(retina pigment epithelium, RPE)。它在年轻的近视女性中更常见，患者主要表现为视力下降、眼睛前黑点和视物变形。它的特征是在视网膜色素上皮和脉络膜内层出现散发性类圆形的黄白色病灶，眼前节和玻璃体无炎症。脉络膜新生血管(choroid neovascularization, CNV)形成的主要原因也是本病视力丧失的主要原因。早期发现、早期治疗PIC继发性CNV，对于患者视力的保护极其重要。

关键词

点状内层脉络膜病变，脉络膜新生血管，光学相干断层扫描

One Case of Punctured Inner Choroid Disease

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Abstract

The punctate inner choroidopathy (PIC) is a rare fundus disease. The lesion consists of the inner choroid and retina pigment epithelium (RPE). It is more common in young women with myopia,

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with reduced vision, dark spots in front of the eye and distorted vision. It is characterized by sporadic round yellowish-white lesions in the retinal pigment epithelium and choroid inner layer, without inflammation in anterior segment and vitreous. Choroid neovascularization (CNV) is the main cause of vision loss in this disease. Early detection and early treatment of PIC secondary CNV are extremely important for visual protection of patients.

Keywords

Punctured Inner Choroid Disease, Choroidal Neovascularization, Optical Coherence Tomography

1. 引言

PIC，是一种多发生在近视眼女性的炎症性脉络膜视网膜疾病，本病例患者为青年女性，有高度近视史，曾在外院初诊为多造性脉络膜炎，后期就诊于我院，通过长期的随访、治疗、分析考虑为点状内层脉络膜病变，特此报道。

2. 临床资料

患者，女，15 岁，因“自觉看不清事物、变形半年左右”于 2020-11-15 日来我院检查。既往双眼有高度近视病史，曾在外院就诊过，为双眼多灶性脉络膜炎(MC/MFC)、右眼继发性 CNV，曾给予右眼抗 VEGF (康柏西普) 3 次，否认其他家族病史。眼科查体：视力：右眼 0.01，−7.0 DS = 0.5，左眼：0.05，−7.0 DS/−1.5 × 180˚ = 1.0。双眼角膜透明，前房正常深度，房水清澈，圆形瞳孔，直径约 3.0 mm，存在对光反射，晶状体及玻璃体透明，看不到炎症细胞。眼底查看：双眼颞侧可见近视弧形斑，颞下近端血管弓和黄斑处可见黄白色病灶，视盘旁及视网膜周围弥漫性视网膜脉络膜萎缩斑(见图 1)。光学相干断层扫描(SD-OCT)检查：右眼颞下近端血管弓处的椭圆体区(EZ)和外界膜(ELM)被破坏，病变处瘢痕形成，黄斑外丛状层中的“驼峰状”中等反射结节病变(见图 1)，左眼黄斑区形态大致正常。眼底视网膜的荧光素血管造影(FFA)检查：双眼黄斑中心凹附近的荧光点状增强，后期渗透，周边血管稍微渗透。眼底脉络膜的吲哚青绿血管造影(ICGA)检查：右眼黄斑区早期点状强荧光，周围透见荧光，后期低荧光，左眼黄斑区多灶低荧光(见图 2)。化验：血常规(−)、肝肾功(−)、尿常规(−)、HIV-Ab(−)、梅毒(−)、补体 C3C4 正常、抗核抗体(−)、血沉(−)、免疫球蛋白 A/G/M、CRP(−)、结核杆菌－干扰素释放试验(−)、抗中性粒细胞胞浆抗体(−)，影像学：胸部 CT 未见明显异常。诊断：1) 双眼 PIC，右眼继发性 CNV；2) 双眼屈光不正。治疗：建议患者行泼尼松 1 mg/(kg·d)药物口服及糖皮质激素球旁注射治疗、右眼行抗 VEGF 治疗、患者拒绝糖皮质激素治疗，于 2020-11-26 日给予患者右眼抗 VEGF (康柏西普)治疗。随后，患者于 2021-04-22 日出现左眼视力逐渐下降，矫正视力：右眼：1.0，左眼：0.6，SD-OCT：左眼视盘到中心凹区域，有一个凸起的中度高层反射灶在外核状层下，IS/OS 层模糊、部分缺失、RPE 层模糊、部分中断。近颞下血管弓处的椭圆体区和外界膜被破坏，病灶疤痕形成(见图 3)。分别于 2021-04-23、2021-05-28、2021-06-25 日给予患者双眼抗 VEGF (雷珠单抗)治疗，随后双眼矫正视力稳定在 1.0。左眼的随访检查如图 4 所示。该病例报道已经获得病人及家属的知情同意。

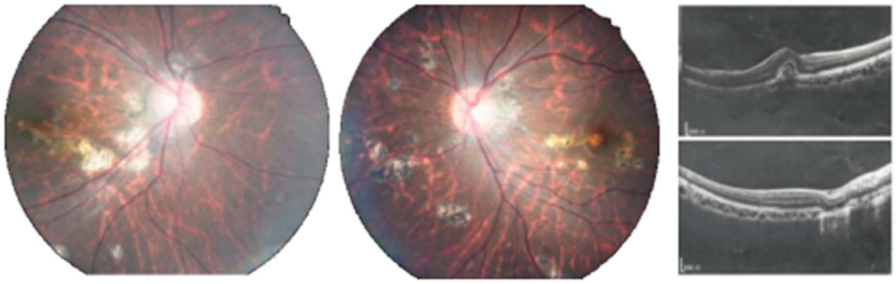

Figure 1. (a) Myopic arc-shaped plaque was seen in the temporal side of the right eye, and 3 yellow and white lesions were observed in the proximal vascular arch of the inferior temporal and the subtemporal macular, about 1/3PD in size; (b) There were seven yellow and white lesions (about 1/6PD in size) on the subnasal side of macular fovea. There were diffuse choroidal atrophy spots beside the optic disc and around the retina; (c) In the right eye, the ellipsoid area and the outer membrane at the proximal subtemporal vascular arch were destroyed, and scar was formed at the lesion. The "humped" moderate reflective nodules in the outer macular plexus layer were observed

图 1. (a) 右眼颞侧见近视弧形斑，颞下近端血管弓和黄斑颞下可见 3 处黄白色病灶，约 1/3PD 大小；(b) 左眼颞侧见视盘萎缩弧，黄斑中心凹鼻下侧见 7 处黄白色病灶，约 1/6PD 大小，视盘旁及视网膜周围弥漫性视网膜脉络膜萎缩斑；(c) 右眼颞下近端血管弓处的椭圆体区和外界膜被破坏，病变处瘢痕形成，黄斑外丛状层中的"驼峰状"中等反射结节病变

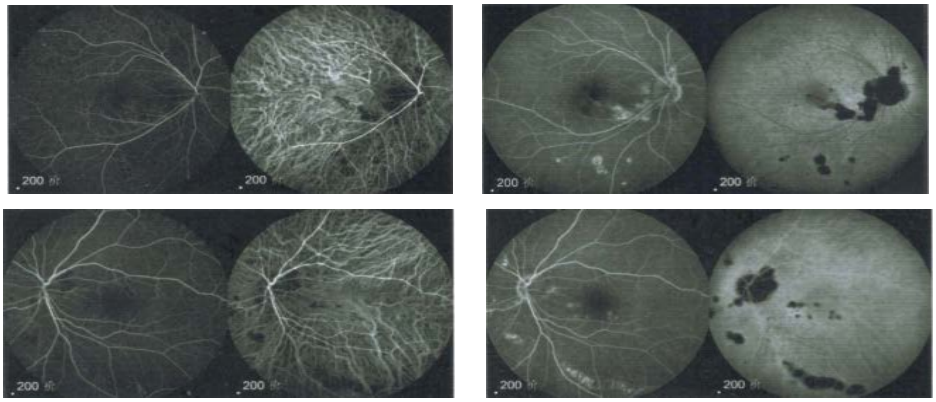

Figure 2. FFA: Spot-enhanced fluorescence near macular fovea in both eyes, late leakage, and slight fluorescence leakage in peripheral retinal vessels; ICGA: spot-like strong fluorescence in the macular area of the right eye in the early stage, transparent fluorescence in the surrounding area, low fluorescence in the late stage, multi-focal low fluorescence in the macular area of the left eye

图 2. FFA：双眼黄斑中心凹附近的荧光点状增强，后期渗漏，周边视网膜血管轻度荧光渗漏；ICGA：右眼黄斑区早期点状强荧光，周围透见荧光，后期低荧光，左眼黄斑区多灶低荧光

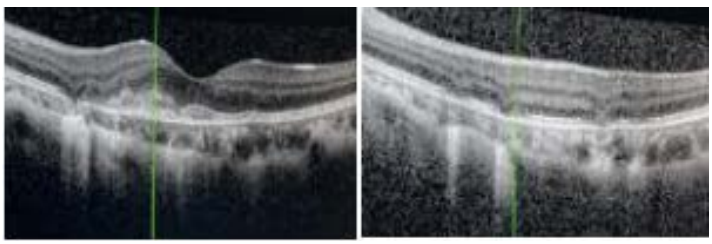

Figure 3. OCT examination on 2021-04-22. From the optic disc of the left eye to the fovea, there IS a raised medium-high reflex foci below the outer nuclear layer. The IS/OS layer IS blurred and partially missing, and the RPE layer IS blurred and partially interrupted. The ellipsoid area and outer membrane of the proximal subtemporal vascular arch were destroyed and the lesion scar was formed

图 3. 2021-04-22 日 OCT 检查。左眼视盘到中心凹区域，有一个凸起的中度高层反射灶在外核状层下，IS/OS 层模糊、部分缺失、RPE 层模糊、部分中断。近颞下血管弓处的椭圆体区和外界膜被破坏，病灶疤痕形成

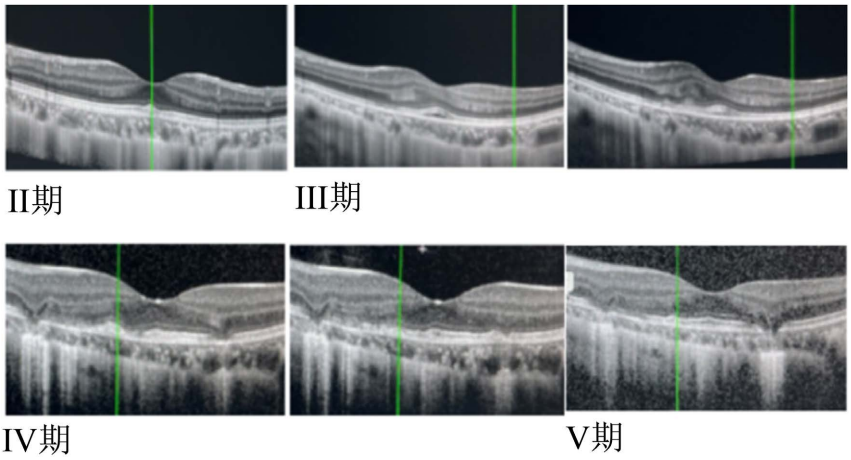

Figure 4. Follow-up examination of the left eye. Stage II (2020-10-25): RPE local uplift, ellipsoid (IS/OS) zone damage; Stage III (2021-4-19, 2021-4-22): The lesion breaks outward and destroys the RPE layer; the lesion breaks inward and destroys the outer membrane and outer nuclear layer; Stage IV (2021-5-5, 2021-5-13): Retrograde regression of outer retinal nodules with V-shaped appearance; Stage V (2021-10-31): Photosensitive cells around the lesion were destroyed with multi-level RPE proliferation to repair RPE rupture

图 4. 左眼随访检查。II 期(2020-10-25)：RPE 局部隆起、椭圆体(IS/OS)带受损；III 期(2021-4-19、2021-4-22)：病灶向外突破，破坏了 RPE 层，病灶向内突破，破坏了外界膜和外核层；IV 期(2021-5-5、2021-5-13)：外层视网膜结节病变逆行消退、呈 V 形外观；V 期(2021-10-31)：病变周围的感光细胞被破坏伴多层次的 RPE 增殖修复 RPE 破裂

3. 讨论

点状内层脉络膜病变是一种炎症性疾病。首先由 Watzke 等[1]于 1984 年提出，它在年轻的近视女性中更常见，常常因视力减退、模糊视线和闪光感为主要临床特征就诊。表现为类圆形黄白色病灶或局灶脉络膜凹陷，出现在乳斑束，常出现在 Brunch 膜、脉络膜毛细血管及视网膜色素上皮层中，眼前节和玻璃体无炎症[2]。近年来，Zhang 等[3]将 SD-OCT 用于对 PIC 疾病的检查，据观察，该病的进展可分为五个阶段。基于上述研究，刘文等[4]人描述了以下五点：I 期：外层核膜表现正常或轻微不规则；II 期：RPE 局部隆起、椭圆体(IS/OS)带受损；III 期：中等反射的结节向外破坏，突破 RPE 层，向内破坏，突破外界膜和外核层，大小不等的中度反射性结节的“骆驼状”病灶出现在外丛状层；IV 期：外层视网膜结节病变逆行消退、光感受器消退、脉络膜组织丢失；V 期：病变逐渐损伤病变周围的感光细胞，并随之破坏丢失伴增殖的 RPE，同期，RPE 增殖(伴后影)修复 RPE 破裂，缓解视网膜疝，使视网膜外壁轮廓重现。临床上，该病常与其他眼底疾病相鉴别。MFC 常双眼发病，在吉尔伯特等人[5]的研究中，证实 MFC 以周围绒毛膜视网膜病变为特征，常伴有眼内炎症反应且与 B 淋巴细胞的浸润有关，尤其是 CNV，这在 PIC 中没有观察到。本病例中，患者未检测到炎性细胞出现在前房和玻璃体腔中，这是与 MFC 的主要鉴别点[6]。多灶性脉络膜炎伴葡萄膜炎病灶通常位于赤道部和乳斑束，直径一般比 PIC 大，但没有 PIC 深。伴有眼内炎症反应[7]，特发性脉络膜新生血管主要病变为黄斑区的渗出性视网膜脉络膜，常伴有黄斑部出血和瘢痕形成[8]。

该患者第一次在我院 OCT 检查显示右眼黄斑区已继发 CNV，黄斑区病变对应 III 期；颞下血管弓处 RPE 增殖修复 RPE 破裂，缓解视网膜疝，使视网膜外壁轮廓重现对应 V 期；随后左眼进行性发展，视乳头至黄斑中心凹外层视网膜结节逆行消退对应 IV 期；黄斑处 RPE 局部隆起、IS/OS 带受损对应 II 期；颞下血管弓附近椭圆体区和外界膜被破坏，形成视网膜内疝(呈 V 形外观)对应 IV 期。抗 VEGF 药物治疗后，2021-10-31 日再查 OCT 显示双眼病灶萎缩，左眼黄斑区病灶由 II 期演变到 V 期形态。即 RPE 局部隆起演变成 RPE 增殖修复 RPE 破裂。当下 PIC 继发 CNV 的治疗标准不明确，主要是应用抗血管内皮生

长因子药物，张华和杨晓红[9] [10]等人证明治疗 PIC 引发的 CNV 治疗效果较好。它的作用是有效减少视网膜下 CNV 的生成，降低单位时间内透过血管壁的物质量，阻止血管渗漏、防止新生血管的形成，有效降低视网膜中央厚度[11]。若患者仅发生 PIC，无新生血管的生成，可应用糖皮质激素进行治疗，其作用是，有效防止 PIC 病灶的形成，抑制血管的新生，从而降低 CNV 发生的风险[12]。

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