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急性轮状外层视网膜病变1例

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[摘要] 患者女，60岁，因“右眼前黑影飘动1月，视力下降8天”就诊。视力：右眼0.1，不能矫正；左眼0.6矫正0.9。右眼眼底见视盘周围边界清晰不规则灰白色区，并波及中心凹。视野检查：右眼对应眼底病灶的视野缺损；左眼正常。光学相干断层成像术(optical coherence tomography, OCT)显示灰白色区域椭圆体带不规则、缺失，视网膜色素上皮(retinal pigment epithelium, RPE)层见数个指状隆起。眼底自发荧光(autofluorescence, AF)示：受影响区域内呈高荧光和部分不规则低荧光区。荧光素眼底血管造影(fundus fluorescein angiography, FFA)示：早期见荧光渗漏，晚期荧光着染、蓄积。吲哚菁绿血管造影(indocyanine green angiography, ICGA)示：见以视乳头为中心，边界清晰的低荧光区。诊断：右眼急性轮状外层视网膜病变。治疗：给予抗炎和改善血液循环4周，眼底灰白色环状带消失，视力明显好转。随访6个月，患者病情控制良好。

[关键词] 外层视网膜病变；视野缺损；光学相干断层成像术；椭圆体带

Acute annular outer retinopathy: A case report

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Abstract A 60-year-old woman was admitted to Chengdu Aidi Eye Hospital because of “dark shadow fluttering in the right eye for 1 month and vision loss for 8 days”. Visual acuity—with a myopic correction—was 0.1 with the right eye and 0.9 with the left eye. The right eye fundus presented a well-defined, irregular, grayish white area around the optic disc, and affected the fovea, corresponding to the visual field defect of the fundus lesion. Optical coherence tomography (OCT) showed that the ellipsoid bands in this region were irregular and absent, and several finger-like ridges were seen in the retinal pigment epithelium (RPE) layer. Fundus autofluorescence (AF): High fluorescence and some irregular low fluorescence in the affected area. Fundus fluorescein angiography (FFA): Fluorescence leakage was seen in the early stage, fluorescence staining and accumulation in the late stage. Indocyanine green angiography (ICGA): A well-defined low-fluorescence area centered on the optic nipple was observed. Diagnosis:

Acute annular outer retinopathy. Treatment: Anti-inflammatory and improved blood circulation for 4 weeks, the gray and white ring of fundus disappeared and the visual acuity improved obviously.

Keywords outer retinopathy; visual field defect; optical coherence tomography; ellipsoid band

急性轮状外层视网膜病变(acute annular outer retinopathy, AAOR)的病例报道较少, 其致病机理、发展过程中的伴发症状、治疗方案及视力预后方面也并不统一。成都爱迪眼科医院门诊发现1例如下。

1 临床资料

患者, 女, 60岁, 因“右眼前黑影飘动1月, 视力下降8天”就诊。否认外伤、高血压、糖尿病史。全身查体未见异常。血糖4.6 mmol/L, 血压118/68 mmHg (1 mmHg=0.133 kPa)。化验室检查无特殊。专科检查: 视力右眼0.1, 不能矫正; 左眼0.6, 矫正0.9。眼压: 右眼14.0 mmHg, 左眼15.3 mmHg。右眼前节(-), 玻璃体混浊(++) , 细胞(++) , 眼底见视盘周围边界清晰不规则灰白色区, 并波及中心凹(图1)。视网膜血管形态正常, 无出血、渗出。光学相干断层成像术(optical coherence tomography, OCT)显示灰白色区域椭圆体带不规则、缺失, 视网膜色素上皮(retinal pigment epithelium, RPE)层见数个指状隆起(图2)。视野检查: 右眼对应眼底病灶的视野缺损(图3)。左眼正常。

眼底自发荧光(autofluorescence, AF)示: 受影响区域内呈高荧光和部分不规则低荧光区(图4)。荧光素眼底血管造影(fundus fluorescein angiography, FFA)示: 早期见荧光渗漏, 晚期荧光着染、蓄积(图5)。吲哚菁绿血管造影(indocyanine green angiography, ICGA)见以视乳头为中心、边界清晰的低荧光区(图6)。

诊断: 右眼AAOR。

治疗: 强的松30 mg, 1次/d, 血栓通1.5 g, 3次/d, 胰激肽原酶肠溶片, 3次/d。减轻组织水肿和改善血液供应。

治疗4周后复查, 自觉视物明显清晰。专科检查: 视力: 右眼0.4, 不能矫正; 左眼0.8, 矫正0.9。右眼前节(-), 玻璃体混浊(+), 眼底未见

明显异常, 视网膜血管形态正常, 无出血、渗出(图7)。AF示: 视盘周围不规则高荧光区和部分不规则颗粒状低荧光(图8)。OCT显示中心凹鼻侧椭圆体带不规则、缺失(图9)。诊断同前。继续服用药物。



图1右眼眼底视盘周围见灰白色环形带, 波及黄斑区

Figure 1 Grey-white annular band was seen around Fundus optic disc, affecting the macular area

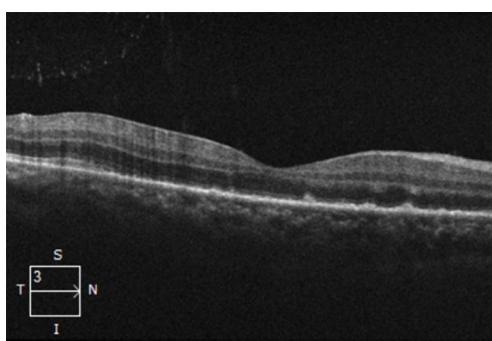


图2 OCT显示灰白色区域椭圆体带不规则、缺失, RPE层见数个指状隆起

Figure 2 OCT showed the ellipsoid bands in this region were irregular and absent, and several finger-like ridges were seen in the RPE layer

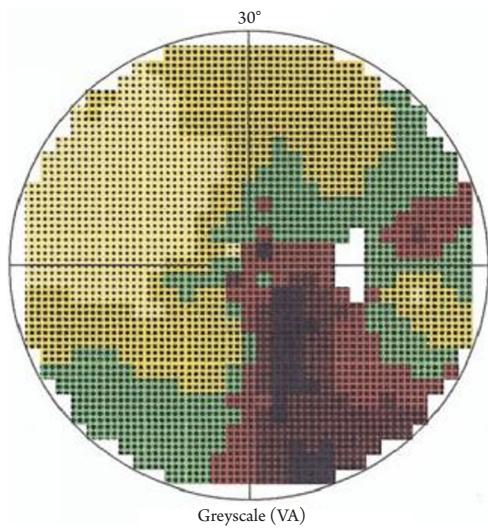


图3 与眼底病灶相对应的生理盲点扩大

Figure 3 Enlarge of the physiological blind spot corresponding to the fundus lesion

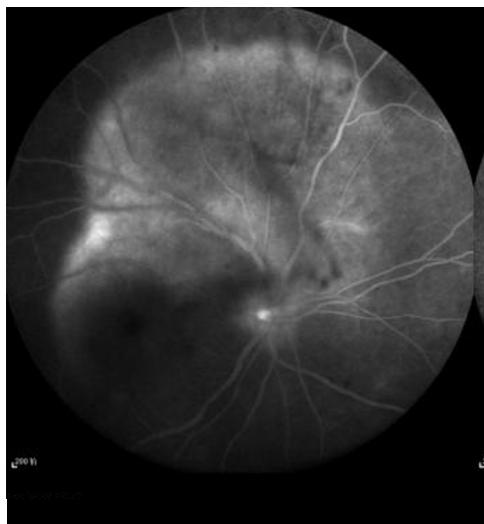


图5 FFA示病灶区域高荧光，荧光渗漏。

Figure 5 FFA showed fluorescence leakage was seen in the early stage, fluorescence staining and accumulation in the late stage



图4 右眼AF示与病灶相对应的区域显示高荧光，其间可见颗粒状低荧光

Figure 4 Right eye AF showed high fluorescence and some irregular low fluorescence in the affected area

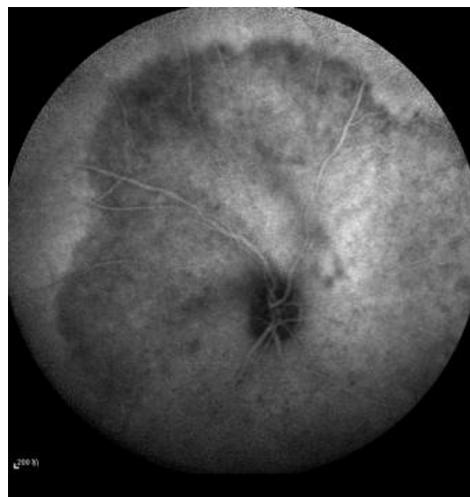


图6 ICGA见以视乳头为中心、边界清晰的低荧光区

Figure 6 In ICGA, a well-defined low-fluorescence area centered on the optic nipple was observed



图7 眼底未见明显异常

Figure 7 No obvious abnormality in fundus

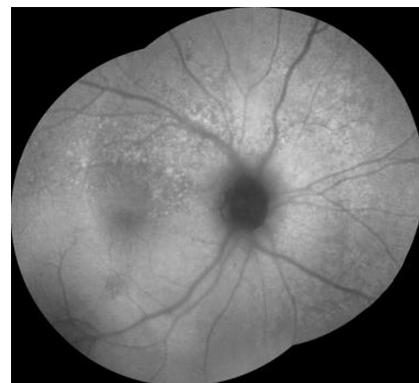


图8 AF示视盘周围不规则高荧光区和部分不规则颗粒状低荧光

Figure 8 AF showed irregular high fluorescence area and partial irregular granular low fluorescence around optic disc

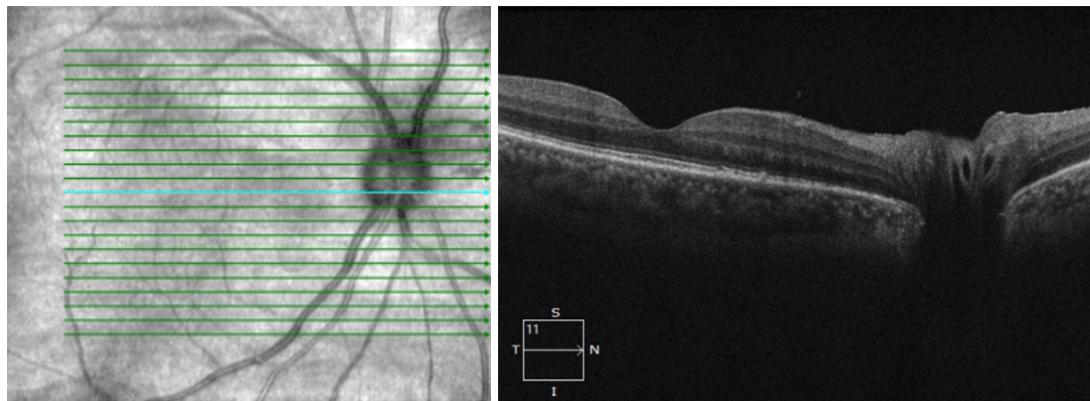


图9 OCT显示中心凹鼻侧椭圆体带不规则、缺失

Figure 9 Irregular and absent ellipsoid bands on the foveal nasal side

2 讨论

1995年Gass和Stern^[1]首先对AAOR作了描述,认为此病是急性区域性隐匿性外层视网膜病变(acute zonal occult outer retinopathy, AZOOR)的一种亚型,病因尚不明确,推测病灶可能是应对病毒感染介导的免疫反应。而后的几例个案报道^[2-3]中也强调了病毒感染及自身免疫反应对此病的重要性。因此,有人认为视网膜对病毒感染的免疫反应会导致视网膜形成一个肉眼可见的白环。还有作者^[4]发现该病发展过程中可伴发脑膜炎、听力下降等全身症状。目前此病发病机理仍不十分明确。

AAOR在已经报道的病例中,常见特征主要是:单眼发病,症状性视野缺损,眼底为一种特

征性不规则的灰白色、深层视网膜不透明的环形带,环内视网膜动脉和静脉变窄。病灶内椭圆体带结构紊乱,RPE层结构受损^[5]。频域光学相干断层扫描(spectral domain optical coherence tomography, SD-OCT)示:病灶区视网膜外核层(outer nuclear layer, ONL)变薄,光感受器复合体(外界膜, 椭圆体带, RPE尖端)模糊不清。眼底视网膜灰白色环形带对应区域为高反射率的ONL和Henle纤维层(Henle fiber layer, HFL)^[6]。FFA中视网膜循环时间延长,受影响区域内普遍高荧光,散在椒盐状、颗粒状或斑片状低荧光。这个环所包围的视网膜区域与它的绝对视野缺损相对应。眼底自发荧光显示病灶区域高荧光。ICGA晚期像见边界清晰的低荧光区^[7]。视网膜电图(electroretinogram, ERG)检查视网膜振幅降低,

表明病变部位应当在视网膜层面^[4]。

AAOR在疾病发展过程中，一般1~2周后眼底病灶会逐步消失^[5]，个别病例可延缓到2年，部分患者眼底病灶可完全消失，而部分患者的RPE脱色素环可能会遗留。Gass和Stern^[1]假设，如果感光细胞恢复其功能，就不会有眼科检查的后遗症。然而，如果感光细胞永久失去功能，可能会出现进行性视网膜外层变性和色素上皮萎缩。目前尚无明确治疗方案。患有此病的眼睛不经治疗也可自行缓解，视力改善^[6-7]。经过药物治疗的眼睛可能也会出现RPE层色素环遗留，无明显视力改善^[8-9]。本例患者给予抗炎和改善血液循环4周，眼底灰白色环带消失，视力明显好转。

目前为止，AAOR的病例报道较少，其致病机理、发展过程中的伴发症状、治疗方案及视力预后方面也并不统一。

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