

IRVAN综合征一例

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摘要

14岁青少年女性, 左眼视力缓慢下降伴眼前黑影遮挡1年就诊, 根据眼底表现及荧光素眼底血管造影(FFA)表现诊断为特发性视网膜血管炎、动脉瘤和神经视网膜炎(IRVAN)综合征二期, 给与激素冲击治疗和抗VEGF药物玻璃体注射后行视网膜激光光凝术, 患者术后病情稳定。

关键词

特发性视网膜血管炎, 动脉瘤, 视神经视网膜炎综合征

One Case of IRVAN Syndrome

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Abstract

A 14-year-old female adolescent with slow visual acuity decline in her left eye and dark shadow occlusion in front of her eyes for 1 year was diagnosed as idiopathic retinal vasculitis, aneurysm and neuroretinitis (IRVAN) syndrome phase II based on fundus manifestations and fluorescein fundus angiography (FFA). She received hormone shock therapy and vitreous injection of anti-VEGF drugs and underwent retinal laser photocoagulation. The patient's condition was stable after operation.

Keywords

Idiopathic Retinal Vasculitis, Aneurysm, Optic Neuroretinitis Syndrome

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1. 引言

IRVAN 综合征(idiopathic retinal vasculitis, aneurysms, and neuroretinitis)是一组特发性视网膜血管炎、动脉瘤和视神经视网膜炎综合症的简称,以眼底特征性视网膜改变为病变特点。我院于 2022 年 3 月份诊治 1 例双眼发病年轻女性患者,双眼眼经过积极治疗后病情稳定。

患者女,14 岁,因发现左眼是视力缓慢下降伴眼前黑影遮挡 1 年于 2022 年 03 月 29 日来诊。眼科检查:右眼视力:0.3,矫正: -2.00 DS = 0.8,左眼视力:0.02 (OS 颞侧视力)矫正无助,双眼眼压:17 mmHg 14 mmHg。双眼眼前节未见明显异常,右眼眼底可见视盘充血,边界模糊,黄斑区下方大片黄白色渗出,周边视网膜可见不易数的动脉白鞘,呈节段状串珠样改变,散在点片状出血。左眼眼底可见视盘充血,边界模糊,黄斑区大片类圆形厚重黄白色渗出,后极部散在点片状渗出,周边视网膜可见不易数的动脉白鞘,呈节段状串珠样改变,散在点片状出血(见图 1)。双眼黄斑 OCT 检查:右眼黄斑区可见点片状高反射信号,左眼黄斑中心凹下致密硬性渗出沉积,隆起的高反射信号,深部组织遮蔽信号,少量网膜下液(见图 2)。入院诊断:双眼视网膜血管炎。系统检查未发现全身异常,否认家族史。入院后行双眼荧光素眼底血管造影,见双眼视盘及视网膜动静脉充盈时间稍延迟,动脉分支处见瘤样膨大,晚期渗漏,周边视网膜见大片毛细血管无灌注区,晚期视盘呈高荧光(见图 3)。生化检查:肝肾功能、血清蛋白、ANCA 正常,ANA (-)、HLA-B27 正常,C3、C4 正常。类风湿因子测定等血液免疫学和生物化学检查未见异常。头颅磁共振成像(MRI)检查无异常。结合患者病史、临床表现和 FFA 检测特点,最后诊断为 IRVAN 综合征。诊疗计划:激素冲击治疗甲强龙 500 mg 静滴*3 d,我院于 2022 年 4 月 5 日局部麻醉下行双眼玻璃体

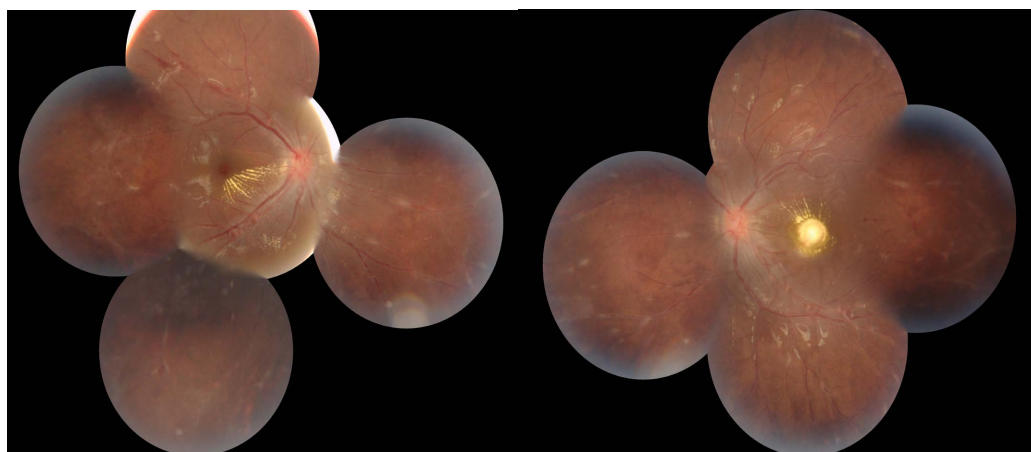


Figure 1. In the fundus of the right eye, hyperemia of the optic disc was observed, the boundary was blurred, a large yellow-white exudation was observed below the macular area, and the white sheath of the artery was not easy to number in the peripheral retina, with segmental beaded changes and scattered patchy bleeding. In the fundus of the left eye, the optic disc was hyperemia with blurred boundaries, and the macular area was a large round thick yellowish-white exudation, with scattered patchy exudation in the posterior pole. The peripheral retina showed an infrequent white sheath of arteries, which presented segmentary beaded changes and scattered patchy bleeding

图 1. 右眼眼底可见视盘充血,边界模糊,黄斑区下方大片黄白色渗出,周边视网膜可见不易数的动脉白鞘,呈节段状串珠样改变,散在点片状出血;左眼眼底可见视盘充血,边界模糊,黄斑区大片类圆形厚重黄白色渗出,后极部散在点片状渗出,周边视网膜可见不易数的动脉白鞘,呈节段状串珠样改变,散在点片状出血

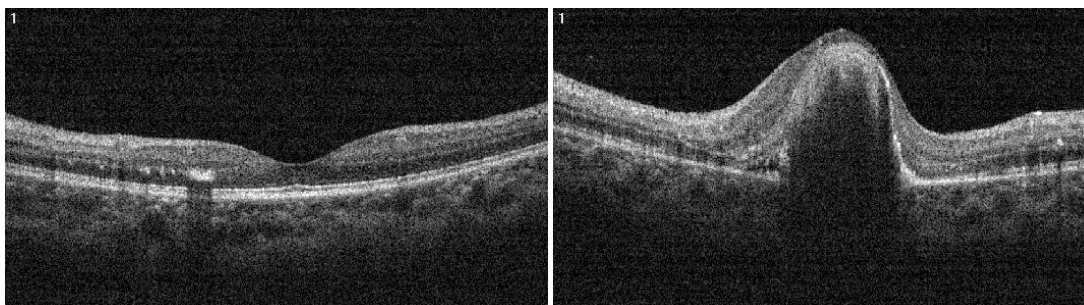


Figure 2. Patchy high-reflection signals can be seen in the macular area of the right eye. Dense hard exudation and deposition in the fovea of the macula of the left eye, elevated high reflection signal, deep tissue masking signal

图 2. 右眼黄斑区可见点片状高反射信号；左眼黄斑中心凹下致密硬性渗出沉积，隆起的高反射信号，深部组织遮蔽信号

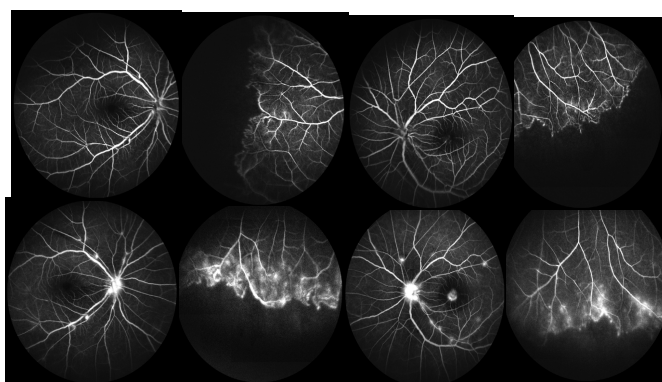


Figure 3. The arteriovenous filling time of optic disc and retina in both eyes was slightly delayed, with tumorous enlargement and late leakage in the branch of the artery, large capillary non-perfusion area in the peripheral retina, and high fluorescence in the late optic disc

图 3. 双眼视盘及视网膜动静静脉充盈时间稍延迟，动脉分支处见瘤样膨大，晚期渗漏，周边视网膜见片毛细血管无灌注区，晚期视盘呈高荧光

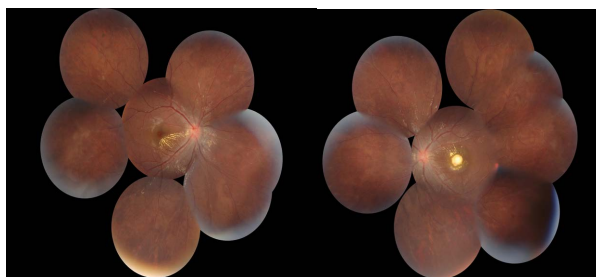


Figure 4. Most of the white sheath of peripheral retinal artery disappeared in both eyes, and superficial retinal bleeding was absorbed

图 4. 双眼周边视网膜动脉白鞘大部分消失，视网膜浅层出血吸收

腔药物注射(康柏西普)，术后妥布霉素滴眼液点眼，1周后行双眼视网膜激光光凝治疗。术后一周治疗效果：双眼视力：VOD：0.5，矫正视力：-2.00 DS = 1.0，VOS：0.06(颞侧视力)，矫正无助，双眼前节未见明显异常，右眼底可见视盘充血，边界基本清晰，黄斑区下方大片黄白色渗出，周边视网膜动脉白鞘大部分消失，视网膜浅层出血吸收，左眼底见视盘充血，边界基本清晰，黄斑区大片状类圆形厚重黄白色渗出，后极部散在点片状渗出较前减少，周边视网膜动脉白鞘大部分消失，视网膜浅层出血吸收(见图4)。双眼眼压：Tn。双眼黄斑 OCT：右眼黄斑区点状高反射数量较前减少，信号反射减弱；左眼黄斑区

致密硬性渗出较前减轻,网膜下液部分吸收(见图5)。行激素冲击治疗和抗 VEGF 药物玻璃体注射后行视网膜激光光凝术,术后 1 周患者视力提高,眼底硬性渗出较前减少,周边视网膜动脉白鞘大部分消失,视网膜浅层出血吸收,病情稳定。

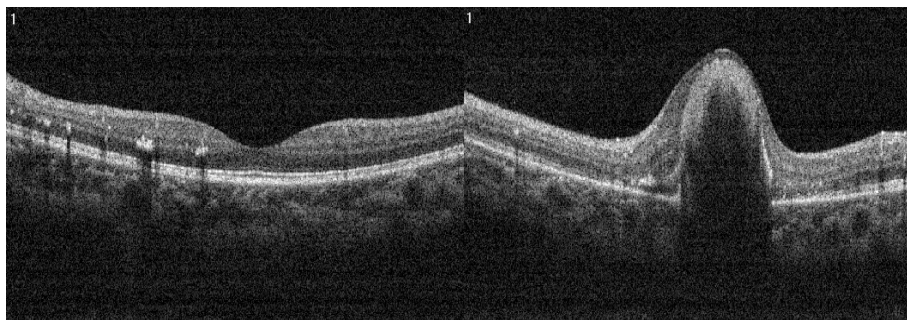


Figure 5. The number of punctate hyperreflexes in the macular area of the right eye decreased, and the signal reflexes weakened. The dense and hard exudation of the left eye was less than before, and the subomental fluid was partially absorbed
图 5. 右眼黄斑区点状高反射数量较前减少,信号反射减弱;左眼致密硬性渗出较前减轻,网膜下液部分吸收

2. 讨论

IRVAN 综合征是眼科罕见的眼底疾病,1983 年由 Kincaid 和 Schatz 首次发现,最先由 Chang 等[1]提出命名,我国最先由王光璐等[2]系统报道此病。它是一种病因未明的可能严重导致视力下降的疾病,该病以年轻女性多见,发病时平均年龄为 30~40 岁[3]。它通常是一种无症状的疾病,但如果不及时治疗,可能会导致严重的视力丧失。它通常是双侧的,但也有一些单侧受累的病例,还有一些病例是从单侧开始的,最终累及双眼。一般多无明显自觉症状,视力逐渐下降就诊,多不伴全身系统性疾病,据报道[4],在 IRVAN 患者中,炎症可能由对结核抗原或真菌成分的过敏反应激活。许多研究表明血管炎与结核病因之间有很强的联系。尽管 IRVAN 综合征的发病机制尚不清楚,但人们普遍认为该谱系是由于轻度眼内炎症、血管炎和视网膜前膜形成而引起的眼内炎症的结果。人们认为,随着时间的推移,迁移的、节段性的视网膜血管炎症会导致动脉壁减弱,从而形成动脉瘤、血管渗漏和毛细血管不灌注。VEGF 在视网膜缺血反应中的表达可能进一步增加血管通透性,导致视网膜渗出和水肿[5] [6] [7]。

目前报道实验室各项检查及其他辅助检查多无明显异常。该综合征的主要诊断依据眼底表现和 FFA 检测,IRVAN 综合征的诊断有 3 个主要标准(视网膜血管炎、动脉分叉处动脉瘤扩张和神经视网膜炎三者缺一不可)和 3 个可选的次要标准(毛细血管不灌注、视网膜新生血管和黄斑渗出)。Samuel 等[8] [9]将 IRVAN 综合征临床共分为五期:一期:微小动脉瘤、黄白色硬性渗出、视神经视网膜炎;二期:视网膜周边毛细血管无灌注区的形成;三期:视盘、视网膜新生血管或者玻璃体积血;四期:眼前节新生血管;五期:新生血管性青光眼。临床上需与视网膜静脉周围炎(Eales 病)、视网膜大动脉瘤及外层渗出性视网膜病变(Coats 病)相鉴别[10] [11]。Eales 病:常见于青年男性,双眼反复发病,炎症明显累及视网膜静脉,可有结核病史,结核菌素试验呈阳性反应;视网膜大动脉瘤:常单眼发病,多为患有高血压及动脉硬化的老年人,眼底表现为单发的视网膜动脉瘤,瘤体呈纺锤形,偶有 2 个以上,大小直径小于 1/4 PD; Coats 病:好发于青少年男性,主要在周边静脉和毛细血管的动脉瘤样扩张,囊样扩张或串珠状且数量较多,合并大量的脂类物质渗出。

因为病因不明确,所以暂无特效治疗方案。其临床特点主要为视网膜血管炎、动脉瘤及神经视网膜炎,疾病发展至中晚期会出现前部葡萄膜炎、玻璃体炎、渗出性视网膜脱离,广泛的周边毛细血管无灌注将导致大面积的视网膜缺血缺氧,引起后极部视网膜大量硬性渗出、黄斑水肿、视网膜与视盘新生

血管生成、玻璃体积血甚至新生血管性青光眼等严重并发症。全身治疗[12]包括口服和静脉注射皮质类固醇、改善疾病的抗风湿药物(DMARDs)和生物制剂。眼部治疗包括对动脉瘤和毛细血管非灌注区(CNP)进行激光光凝、玻璃体腔内注射抗血管内皮生长因子(VEGF)、玻璃体腔内类固醇植入、玻璃体切除术和新血管性青光眼的内科和外科治疗。目前共有以下几种治疗方案。(1) 视网膜激光光凝术: 运用视网膜激光光凝术治疗后,可以降低视网膜的耗氧量,从而减少视网膜新生血管的生成、周边视网膜无灌注区的形成以及视网膜血管的渗漏,对于一期、二期病变,维持稳定视力,动脉瘤变小或者消失,三期及以后的病变,即使行光凝术,视力不同程度的下降。所以当确诊该综合征时,应尽早行视网膜光凝术;(2) 糖皮质激素治疗: 虽然 IRVAN 综合征中存在炎症因子发挥作用,但是口服糖皮质激素治疗本病并没有明显改善眼底血管炎症和阻止病情进展,有报道在 PRP 术后,患者出现视网膜渗出和视神经水肿时,激素冲击,视力能快速提高,后续无明显病情反复;(3) 抗 VEGF 药物治疗: 三期及以后的患者采用激光干预效果不佳,可联合抗 VEGF 药物治疗注射,能够有效控制视网膜新生血管和视网膜渗出,预后视力明显提高,病情稳定,未见复发倾向;(4) 并发症的治疗: 当出现反复玻璃体积血牵拉视网膜脱离时可行玻璃体切割术,当出现新生血管性青光眼时先行抗 VEGF 治疗后再行抗青光眼手术等。早期干预这些患者的形式是及时的激光治疗,而不是等待新血管的发展是重要的,因为这种疾病的性质比其他缺血性视网膜病变更具侵袭性。抗 vegf 和英夫利昔单抗已被用作 IRVAN 与激光治疗的辅助治疗,以减少疾病进展。然而,类固醇的作用仍然不确定。虽然与 p-ANCAs、结节病、颅内压升高、抗磷脂抗体、结核菌素过敏和同型半胱氨酸升高有关,但通常不能确定全身性病理性[13] [14]。在 Massicotte Erika 等人[15]的回顾性研究中,建议对所有诊断为 IRVAN 综合征的患者进行抗磷脂抗体筛查。这种全身性疾病可能产生严重的后果,早期诊断可以避免。缺血事件与 IRVAN 综合征之间的关系尚不清楚,但肯定需要进一步研究。同时,建议对所有诊断为 IRVAN 综合征的患者进行凝血因子异常筛查,因为适当的治疗可能会预防严重的视网膜缺血事件。尽管存在这些联系,但仍主张进行有针对性的实验室检查,而不建议进行全面调查。鉴于 IRVAN 的临床特点,当出现症状后就诊时应完善眼底检查以及 FFA 检查,诊断该综合征后尽早根据疾病分期完成相应治疗,保证患者的视觉质量。

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