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# IRVAN综合征治疗一例及文献复习

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## 摘　要

**目的：报道1例特发性视网膜血管炎、动脉瘤、视神经视网膜炎(IRVAN)综合征治疗过程的临床特征和治疗要点，分析该病形成原因及治疗预后。方法：回顾该病例的诊疗过程，并结合相关文献分析该病表现及治疗预后。结果：根据患者病史、症状、体征及眼底荧光素血管造影结果，诊断为“IRVAN综合征III期”，行玻璃体腔注射及激光光凝，随访10个月，病情稳定。结论：IRVAN综合征较为罕见，如果不及早进行诊断和治疗，预后通常较差。因此，提高对该疾病的认识对于提高患者的生存质量至关重要。**

## 关键词

**特发性视网膜血管炎，动脉瘤，视神经视网膜炎，IRVAN综合征**

# A Case of Idiopathic Retinal Vasculitis, Aneurysms, and Neuroretinitis (IRVAN) Syndrome and a Literature Review

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## Abstract

**Objective: This study reports the clinical features and treatment of a case of Idiopathic Retinal Vasculitis, Aneurysms, and Neuroretinitis (IRVAN) Syndrome and analyzes the cause and treatment prognosis of the disease. Methods: The diagnosis and treatment of this case were reviewed, and the manifestations and prognosis of this disease were analyzed combined with related literatures. Results: According to the patient's medical history, symptoms, signs and fundus fluorescein angiography results, we diagnosed "IRVAN Syndrome Stage III". We successfully treated with intravitreal injection and Laser photocoagulation. After 10 months of follow-up, the condition was stable. Conclusion: IRVAN syndrome is rare, and the prognosis is usually poor if it is not diagnosed and treated early. Therefore, it is very important to improve the understanding of the disease for improving the quality of life of patients.**

## Keywords

**Idiopathic Retinal Vasculitis, Aneurysms, Neuroretinitis, IRVAN Syndrome**

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## 1. 背景

特发性视网膜血管炎、动脉瘤、视神经视网膜炎综合征(idiopathic retinal vaseulitis, aneurysms and neuroretinitis, IRVAN)是一种罕见的视网膜血管炎，主要以眼底后极部多发视网膜动脉瘤样扩张，视网膜血管炎以及周边毛细血管大量无灌注区为特点。该病好发于年轻女性，有报道的最小患者是 7 岁，最大 65 岁[1]，多累及双眼，但也有少量病例报道单眼发病[2] [3]。最早是 Karel [4]在 1973 年报道了一例荧光素下显示视网膜血管炎及多发视网膜小动脉瘤的年轻患者。1983 年 Kincaid 等[5]进一步报道了两例患者，但当时并没提出明确的疾病名称，一直到 1995 年 Chang 报道了 10 例相关病例[6]，总结临床表现并命名为 IRVAN 综合征。其后 Samuel [7]建议将 IRVAN 综合征分为 5 期，以便于指导临床诊断及治疗。本文报道了 1 例 32 岁女性，结合临床及 FFA 表现，考虑为双眼 IRVAN 综合征(III 期)的病例，分析病因、表现及治疗预后，以提高对该病的认识。

## 2. 临床资料

患者女，32 岁，因“右眼视物不清 1 年余，左眼黑影飘动 7 天。”于 2022 年 5 月 11 号来诊。眼科检查：右眼矫正视力 0.1，左眼矫正视力 1.0；双眼眼压正常；双眼前节检查未见异常；双眼玻璃体腔混浊，见少量炎性细胞；右眼底视乳头边界不清，表面及盘周动脉瘤样扩张，鼻侧及颞上、颞下半环形硬性渗出，黄斑鼻侧大量硬性渗出，动静脉管径粗细不均，视网膜见大量激光斑；左眼底视乳头充血水肿，边界不清，视盘及黄斑鼻侧见硬性渗出，视网膜见大量激光斑(见图 1)。

追问病史发现，患者 1 年前在当地医院就诊，考虑“视网膜血管炎”，未治疗。4 个月前右眼视物不清加重，外院行双眼眼底荧光素血管造影(fundus fluoresce in angiography, FFA)，见双眼视乳头周围及视网膜上有多个动脉瘤样扩张，周边毛细血管无灌注区及新生血管(见图 2)，诊断“1. 视网膜新生血管(双)

2. IRVAN 综合征(双)？)”，予以双眼激光及玻璃体腔药物注射术治疗。期间因疫情未定期复查。7 天前患者左眼出现黑影飘动，来我院就诊，行 FFA 见双眼视网膜动、静脉粗细不均，充盈迟缓，视乳头表面及盘周动脉瘤样扩张，静脉管径不均匀节段壁染，散在荧光渗漏灶，晚期视盘高荧光。右眼后极部颞上、颞下血管分叉处瘤样扩张，左眼鼻上方血管瘤样扩张(见图 3)。光相干断层扫描(OCT)示：右眼黄斑囊样水肿；左眼黄斑中心凹形态尚可，鼻侧硬渗。

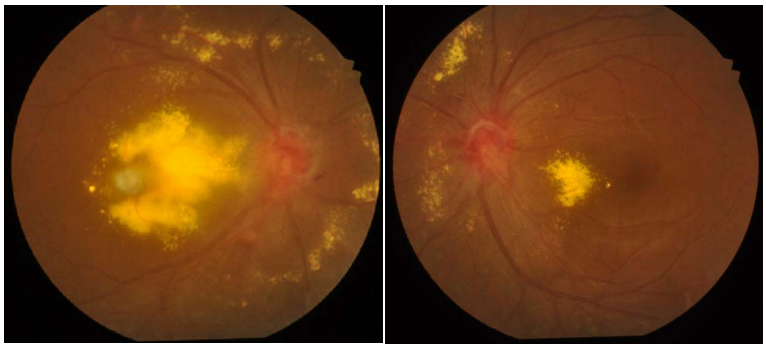


**Figure 1.** Fundus image of the IRVAN patient at the first diagnosis
**图 1.** IRVAN 患者首诊时眼底图像

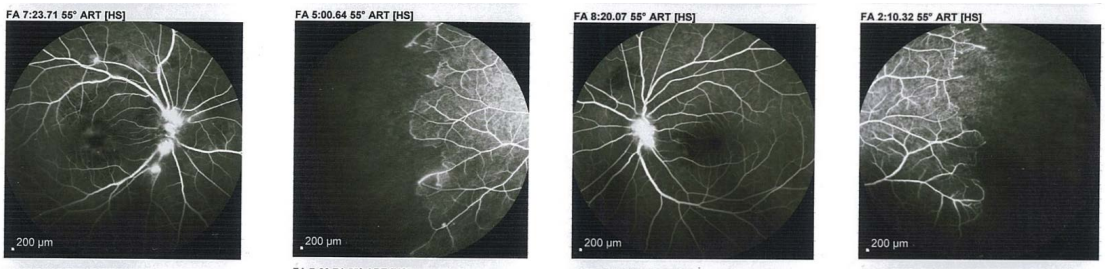
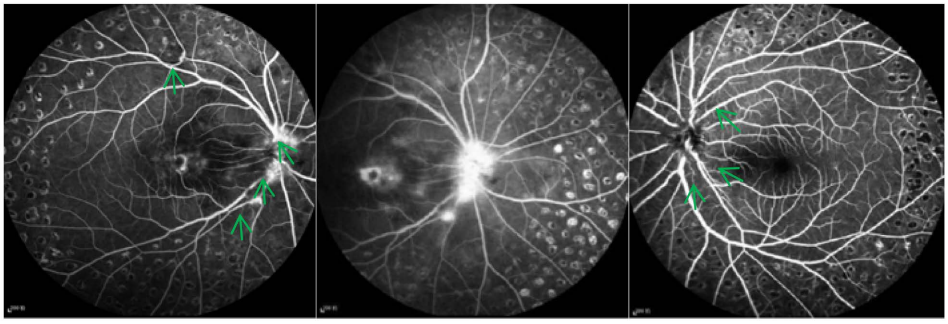



**Figure 2.** FFA image of the IRVAN patient at other hospital
**图 2.** IRVAN 患者外院 FFA 图像



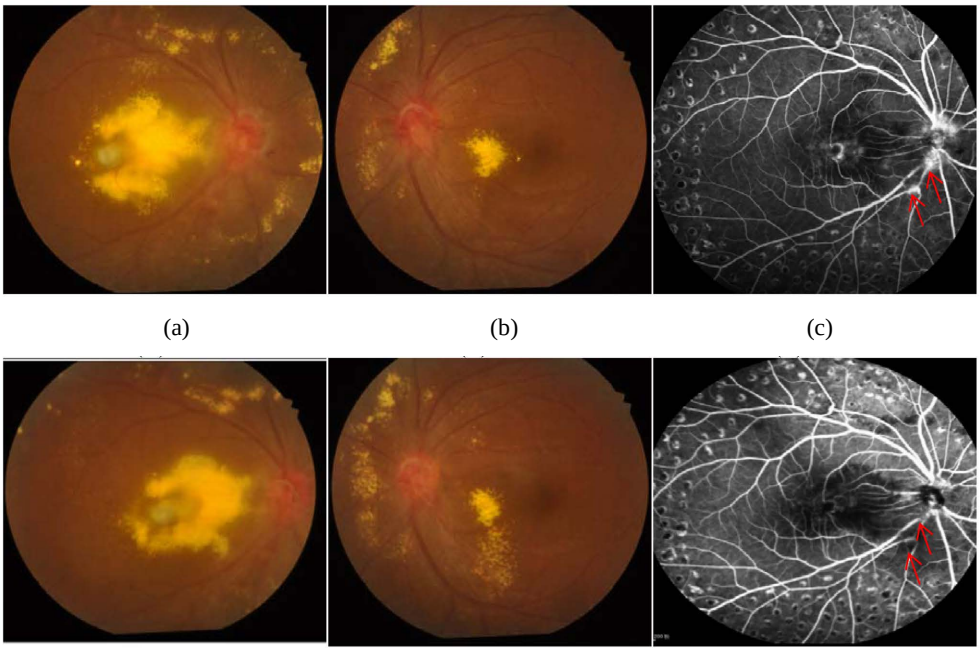
(a) (b) (c)

(a)，(b)：右眼 50˚ FFA 图像见视网膜动、静脉粗细不均，充盈迟缓，视乳头表面及盘周动脉瘤样扩张，后极部颞上、颞下血管分叉处瘤样扩张，晚期视盘高荧光。(c)：左眼 50˚ FFA 图像见视乳头表面及盘周动脉瘤样扩张，左眼鼻上方血管瘤样扩张。

**Figure 3.** FFA image of right of the IRVAN patient at the first diagnosis
**图 3.** IRVAN 患者首诊时右侧 FFA 图像

患者入院诊断：1) 双眼 IRVAN 综合征(III 期)；2) 双眼黄斑水肿。给予患者双眼玻璃体腔药物注射

术及补充激光治疗。患者定期复诊，最近一次于 2023 年 3 月 29 日复诊，眼科检查：右眼矫正视力 0.1，左眼矫正视力 1.0；眼底硬渗较前减少(见图 4)




(a)，(b)，(d)，(e)：对比最近一次复查(d)，(e)和第一次就诊(a)，(b)时的眼底图像，可以发现脂质渗出明显减少。(c)，(f)：对比右眼最近一次复查(f)和第一次就诊(c)时的 ffa 图像，颞上、颞下及视盘周围的动脉瘤的大小及形态改变。

**Figure 4.** The fundus and FFA images of IRVAN patients at the last reexamination
**图 4.** IRVAN 患者最近一次复查时眼底及 FFA 图像

## 3. 讨论

不虽然目前对于该病的病因没有明确的定论，但许多学者认为该病与眼内炎症、血管炎症相关。动脉管壁的炎症，引起管壁渗出，动脉壁变薄，易于脂质渗出及形成动脉瘤[8]。长期随访发现，动脉瘤的位置、大小等会改变，甚至消失，这也说明动脉瘤具有移动性[9] [10] [11]。部分病例里前房或玻璃体腔出现的炎性细胞，给予类固醇治疗后，恢复良好，如 Vichare 等针对一位玻璃体腔出现炎症细胞，右眼最佳矫正指数，左眼最佳矫正 6/24 的患者，应用静脉注射甲基泼尼松龙 1 mg/天治疗三天后改为泼尼松龙口服 1 mg/kg，后缓慢减量，患者最佳矫正视力右眼 6/24，左眼 6/9，双眼玻璃体腔炎症细胞消失，这些都支持疾病与炎症相关[12] [13] [14]。虽然患者大多无全身系统性疾病，但仍有少量病例报道伴有 p-ANCA 升高、真菌过敏性鼻窦炎、高同型半胱氨酸血症、颅内压升高[15] [16] [17] [18] [19]。在我们的病例中最后一次复查 FFA 与首诊时比较，可见动脉瘤大小、形态发生了变化(见图 4(e)，图 4(f))，这也支持动脉瘤移动性的可能。

目前主要是系统治疗和眼部治疗。系统治疗主要是口服或静脉给予药物。眼部治疗主要包括激光光凝，玻璃体腔内注射抗血管内皮生长因子、类固醇植入物，玻璃体切割术等。根据 Samuel 制定的分期，一些人主张 I 期主要以观察为主[20]，另一些则认为可以给予一些激素，改善炎症及渗出。II 期和 III 期患者多建议采用激光或联合全身激素治疗，避免因无灌注区导致的视力丧失及并发症。Rouvas 等[21]建

议对无灌注区大于两个象限的患者行全视网膜激光光凝，对小于两象限的给予局部光凝。近几年报道对于出现黄斑脂质渗出及水肿的患者，予以玻璃体腔注射抗血管内皮生长因子、曲安奈德或地塞米松植入剂可以明显的降低水肿，减少渗出[22] [23] [24] [25]。熊小燕等报道[26]的一例伴有视网膜有髓神经纤维的 IRVAN 患者，在及时全视网膜光凝后渐进性消退，提示及时干预和视网膜光凝治疗是改善眼底缺血，保留较好视力的关键环节之一。Cheema 等[27]使用英夫利昔单抗治疗 IRVAN 综合征患者，患者视力得到提高。伍志琴[1] [28]等报道了一例双眼视力 0.3 的 IRVAN 综合征患者，予以患者局部激光光凝，随访 2 年后，患者视力稳定右眼 0.4，左眼 0.6。本病例，因考虑患者在外院已行激光及玻璃体药物治疗，我们给予补充激光及连续三个月的玻璃体腔药物注射术，术后患者视力稳定。

在我们的病例中患者双眼初诊时考虑 III 期，复诊中视力虽仍为右眼矫正 0.1，左眼矫正 1.0，但对比眼底和 FFA，可见黄斑处渗出减少，动脉瘤大小及形态变化。预后左眼优于右眼，我们考虑是因为右眼患者前期未重视未治疗，激光光凝后因疫情未规律复诊补充激光，而左眼早期予以激光，并在 7 天前出现症状后及时补充激光及抗 VEGF 治疗，所以右眼预后不如左眼。但本病例仍需继续长期随访。鉴于本病较为罕见，现在大多文献均为病例报道及病例系列报道，患者预后相差较大，缺乏大样本、多中心的观察。

## 4. 总结

综上所述，本病的自然预后差，现有治疗不能消除病因，多是减少渗出、降低水肿及毛细血管无灌注区等对症治疗，防止玻璃体积血、新生血管性青光眼等严重并发症的发生。

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## 参考文献

[1] DiLoreto, D.J. and Sadda, S.R. (2003) Idiopathic Retinal Vasculitis, Aneurysms, and Neuroretinitis (IRVAN) with Preserved Perfusion. *Retina*, **23**, 554-557. https://doi.org/10.1097/00006982-200308000-00022

[2] Chawla, R., Kumar, A., Ravani, R., *et al*. (2018) Multimodal Imaging Questions Etiology of Idiopathic Retinal Vasculitis, Aneurysms and Neuroretinitis Syndrome (IRVAN Syndrome). *Medical Hypotheses*, **111**, 12-14. https://doi.org/10.1016/j.mehy.2017.10.031

[3] Moosavi, M., Hosseini, S.M., Shoeibi, N., *et al*. (2015) Unilateral Idiopathic Retinal Vasculitis, Aneurysms, and Neuroretinitis Syndrome (IRVAN) in a Young Female. *Journal of Current Ophthalmology*, **27**, 63-66. https://doi.org/10.1016/j.joco.2015.10.006

[4] Karel, I., Peleska, M. and Divisova, G. (1973) Fluorescence Angiography in Retinal Vasculitis in Children's Uveitis. *Ophthalmologica*, **166**, 251-264. https://doi.org/10.1159/000306822

[5] Kincaid, J. and Schatz, H. (1983) Bilateral Retinal Arteritis with Multiple Aneurysmal Dilatations. *Retina*, **3**, 171-178. https://doi.org/10.1097/00006982-198300330-00005

[6] Chang, T.S., Aylward, G.W., Davis, J.L., *et al*. (1995) Idiopathic Retinal Vasculitis, Aneurysms, and Neuro-Retinitis. Retinal Vasculitis Study. *Ophthalmology*, **102**, 1089-1097. https://doi.org/10.1016/S0161-6420(95)30907-4

[7] Samuel, M.A., Equi, R.A., Chang, T.S., *et al*. (2007) Idiopathic Retinitis, Vasculitis, Aneurysms, and Neuroretinitis (IRVAN): New Observations and a Proposed Staging System. *Ophthalmology*, **114**, 1526-1529. https://doi.org/10.1016/j.ophtha.2006.11.014

[8] Ali, K.H., Ali, K.Q., Shahzad, M.A., *et al*. (2022) Comprehensive Overview of IRVAN Syndrome: A Structured Review of Case Reports and Case Series. *Therapeutic Advances in Ophthalmology*, **14**, 1-12. https://doi.org/10.1177/25158414211070880

[9] Yeshurun, I., Recillas-Gispert, C., Navarro-Lopez, P., *et al*. (2003) Extensive Dynamics in Location, Shape, and Size of Aneurysms in a Patient with Idiopathic Retinal Vasculitis, Aneurysms, and Neuroretinitis (IRVAN) Syndrome. Idiopathic Retinal Vasculitis, Aneurysms, and Neuroretinitis. *American Journal of Ophthalmology*, **135**, 118-120. https://doi.org/10.1016/S0002-9394(02)01823-8

[10] Zhang, Y., Chang, T., Ge, G., *et al*. (2022) Extensive Dynamics of Aneurysms and Long-Term Prognosis in IRVAN Syndrome: A Case Series. *Ocular Immunology and Inflammation*, **30**, 623-627. https://doi.org/10.1080/09273948.2020.1821902

[11] 韩梅, 王兰惠, 时冀川, 等. 双侧视网膜动脉炎伴多发性瘤样动脉扩张激光光凝治疗疗效观察[J]. 中华眼底病杂志, 2012, 28(2): 173-175. https://doi.org/10.3760/cma.j.issn.1005-1015.2012.02.019

[12] Mohd, A.A., Wan, A.H.W., Yaakub, M., *et al*. (2022) Idiopathic Retinitis, Vasculitis, Aneurysms, and Neuroretinitis (IRVAN): Early Treatment Saves Sight. *Cureus*, **14**, e23049.

[13] Vichare, N., Bhargava, N., Brijmohan, *et al*. (2015) A Rare Case of Idiopathic Retinal Vasculitis, Aneurysms and Neuroretinitis (IRVAN) Syndrome. *Medical Journal Armed Forces India*, **71**, S257-S260. https://doi.org/10.1016/j.mjafi.2014.07.011

[14] Empeslidis, T., Banerjee, S., Vardarinos, A., *et al*. (2013) Dexamethasone Intravitreal Implant for Idiopathic Retinal Vasculitis, Aneurysms, and Neuroretinitis. *European Journal of Ophthalmology*, **23**, 757-760. https://doi.org/10.5301/ejo.5000301

[15] Hammond, M.D., Ward, T.P., Katz, B., *et al*. (2004) Elevated Intracranial Pressure Associated with Idiopathic Retinal Vasculitis, Aneurysms, and Neuroretinitis Syndrome. *Journal of Neuro-Ophthalmology*, **24**, 221-224. https://doi.org/10.1097/00041327-200409000-00008

[16] Soheilian, M., Nourinia, R., Tavallali, A., *et al*. (2010) Idiopathic Retinal Vasculitis, Aneurysms, and Neuroretinitis (IRVAN) Syndrome Associated with Positive Perinuclear Antineutrophil Cytoplasmic Antibody (P-ANCA). *Retinal Cases and Brief Reports*, **4**, 198-201. https://doi.org/10.1097/ICB.0b013e3181a59c07

[17] Parchand, S., Bhalekar, S., Gupta, A., *et al*. (2012) Primary Branch Retinal Artery Occlusion in Idiopathic Retinal Vasculitis, Aneurysms, and Neuroretinitis Syndrome Associated with Hyperhomocysteinemia. *Retinal Cases and Brief Reports*, **6**, 349-352. https://doi.org/10.1097/ICB.0b013e31823c1289

[18] Abu El-Asrar, A. (2004) Regression of Aneurysmal Dilatations in a Case of Idiopathic Retinal Vasculitis, Aneurysms and Neuroretinitis (IRVAN) Associated with Allergic Fungal Sinusitis. *Eye* (*London*), **18**, 197-201. https://doi.org/10.1038/sj.eye.6700554

[19] Nourinia, R., Montahai, T., Amoohashemi, N., *et al*. (2011) Idiopathic Retinal Vasculitis, Aneurysms and Neuroretinitis Syndrome Associated with Positive Perinuclear Antineutrophil Cytoplasmic Antibody. *Journal of Ophthalmic and Vision Research*, **6**, 330-333.

[20] Basha, M., Brown, G.C., Palombaro, G., *et al*. (2014) Management of IRVAN Syndrome with Observation. *Ophthalmic Surgery, Lasers and Imaging Retina*, **45**, e18-e22. https://doi.org/10.3928/23258160-20140331-02

[21] Rouvas, A., Nikita, E., Markomichelakis, N., *et al*. (2013) Idiopathic Retinal Vasculitis, Arteriolar Macroaneurysms and Neuroretinitis: Clinical Course and Treatment. *Journal of Ophthalmic Inflammation and Infection*, **3**, Article No. 21. https://doi.org/10.1186/1869-5760-3-21

[22] Chong, Y.J., Logeswaran, A., Sarmad, A., *et al*. (2019) Idiopathic Retinal Vasculitis, Aneurysms and Neuroretinitis (IRVAN): Case Series of Three Patients with Multimodal Imaging. *Graefe's Archive for Clinical and Experimental Ophthalmology*, **257**, 1013-1018. https://doi.org/10.1007/s00417-019-04291-5

[23] Saatci, A.O., Ayhan, Z., Takeş, Ö., *et al*. (2015) Single Bilateral Dexamethasone Implant in Addition to Panretinal Photocoagulation and Oral Azathioprine Treatment in IRVAN Syndrome. *Case Reports in Ophthalmology*, **6**, 56-62. https://doi.org/10.1159/000375481

[24] Karasu, B., Özkan, D., Erdoğan, G., *et al*. (2019) The Fluorescein Angiographic Photodiagnosis of Idiopathic Retinal Vasculitis, Aneurysms, and Neuroretinitis (IRVAN) Syndrome: Outcome of Combined Therapy. *Photodiagnosis and Photodynamic Therapy*, **27**, 336-339. https://doi.org/10.1016/j.pdpdt.2019.06.013

[25] Terrence Kwong-Weng Soong, Visvaraja Subrayan, Choo May May. 玻璃体内注射曲安奈德治疗继发于 IRVAN 的黄斑水肿(英文) [J]. 国际眼科杂志, 2010, 10(11): 2058-2059.

[26] 熊小艳, 赵菊莲. IRVAN 全视网膜光凝后视网膜有髓神经纤维渐进性消退一例[J]. 中华眼视光学与视觉科学杂志, 2020, 22(3): 233-235. https://doi.org/10.3760/cma.j.cn115909-20190814-00226

[27] Cheema, R.A., Al-Askar, E. and Cheema, H.R. (2011) Infliximab Therapy for Idiopathic Retinal Vasculitis, Aneurysm, and Neuroretinitis Syndrome. *Journal of Ocular Pharmacology and Therapeutics*, **27**, 407-410. https://doi.org/10.1089/jop.2010.0189

[28] 伍志琴, 聂尚武, 吳明, 等. 激光光凝治疗双侧视网膜动脉炎伴多发性瘤样动脉扩张一例[J]. 应用激光, 2021, 41(5): 1138-1142. https://doi.org/10.14128/j.cnki.al.20214105.1138