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·临床病例讨论·

多发性虹膜痣综合征1例

肖婷婷，缪振中，李玉生，李国栋

(南昌大学第二附属医院眼科，南昌 330000)

[摘　要]　患者女，54岁，因“左眼视物模糊4个月余”入院，视力0.5，虹膜表面可见大量棕色色素结节，且多数带蒂。房角镜下见下方、鼻侧及颞侧房角大量色素沉着。超声生物显微镜检查：左眼虹膜表面欠光滑并见多个高反射点。光学相干断层扫描：左眼见上方及下方视神经纤维层厚度变薄。用药后眼压：34 mmHg。诊断为Cogan-Reese综合征并继发性青光眼，予多种药物以降低眼压，无效后行复合式小梁切除术。术后3个月内随访，视力、视野保持稳定，眼压8~20 mmHg。前房中深，滤过泡功能良好，视野显示MD=–10.77 dB，局部未用降眼压药。抗青光眼手术能控制眼压防止病情进一步进展，但长期有效性仍需继续随访观察。

[关键词]　虹膜角膜内皮综合征；青光眼；Cogan-Reese 综合征

Multiple iris nevus syndrome: A case report

XIAO Tingting, MIAO Zhenzhong, LI Yusheng, LI Guodong

(Department of Ophthalmology, Second Hospital Affiliated to Nanchang University, Nanchang 330000, China)

Abstract　A 54-year-old woman presented to our department with complaints of blurred vision for more than 4 months in her left eye. Upon examination, the patient had best corrected visual acuity of 0.5 OS, intraocular pressure (IOP) OS was 34 mmHg after medication. On slit-lamp examination, there were multiple iris nodules, most of them pedunculated and intensively distributed in the inferior direction. On gonioscopic examination, a large number of pigments were noticed at the inferior, nasal and temporal direction. Fundus examination revealed glaucomatous cupping of 0.7. Optical coherence tomography showed marked loss of thickness of retinal nerve fiber layer fibers in the inferior, and superior direction. Based on the clinical features, the diagnosis of secondary angle closure due to Cogan-Reese syndrome was reached. As the medical therapy was ineffective, it was planned to go for surgical management in the form of trabeculectomy with 5-FU. At the 3-month follow-up, best corrected visual acuity OS was 0.5, IOP OS was 8–20 mmHg, the anterior chamber depth was medium and the filtering bleb was functional. A visual field was performed, which showed MD=–10.77 dB and the patient wasn't given topical medication. A long-term follow-up is necessary because the disease itself is progressive in nature.

Keywords　iris corneal endothelial syndrome; glaucoma; Cogan-Reese syndrome

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通信作者 (Corresponding author)：李国栋，Email: daliguodong@sina.com

虹膜角膜内皮综合征(iris corneal endothelial syndrome，ICES)临床上常见于中青年女性，多单眼发病，可累及角膜、前房角、虹膜，其临床症状复杂，常伴随青光眼的发生。角膜内皮细胞结构异常和前房角的改变是ICES的共同特点，其主要临床类型有3种，其中Cogan-Reese综合征(虹膜痣综合征)：带蒂的色素性虹膜结节为其主要临床特征[1]。

1 病例资料

患者，女，54岁，因“左眼视物模糊4个月余”入院。4个月前无明显诱因致左眼视物模糊，偶伴眼胀，无明显眼红、眼痛，无畏光、流泪等。曾在当地就诊，眼压：59 mmHg (1 mmHg=0.133 kPa)，并予以马来酸噻吗洛尔、布林佐胺滴眼液及醋甲唑胺片降低眼压无效后入我院住院治疗。既往无眼部疾病及相似疾病家族史。入院专科体格检查：视力右眼0.6，左眼0.5，右眼专科体格检查未见明显异常。左眼结膜无充血，角膜尚清，前房轴深约4个角膜厚度(corneal thickness，CT)，下方、鼻侧及颞侧房角大量色素沉着(图1)。虹膜表面可见大量棕色色素结节，且多数带蒂(图2)。瞳孔约2 mm，对光反应存。晶状体皮质混浊，核淡黄色，前囊可见少量色素沉着。玻璃体絮状混浊。眼底视盘界清，色淡，视神经纤维杯盘比(optic nerve vertical cup disc ratio，C/D)约0.7，中央血管鼻侧移位，网膜平伏，黄斑亮点欠清(图3)。眼压：右眼13 mmHg，左眼34 mmHg(用药后)。辅助检查：超声生物显微镜检查(ultrasound biomicroscopy，UBM)：左眼虹膜表面欠光滑并见多个高反射点(图4)。视盘光学相干断层扫描(optic nerve head optical coherence tomography，ONH OCT)：左眼见上方及下方视神经纤维层厚度变薄(图5)。角膜内皮细胞密度、平均细胞面积、变异系数均在正常范围内[2](图6)。Humphrey视野检查仪示：环形暗点。平均缺损(mean deviation，MD)=−11.43 dB(图7)。经以上检查结合临床诊断为Cogan-Reese综合征并继发性青光眼。

左眼予布林佐胺滴眼液、马来酸噻吗洛尔滴眼液降眼压治疗无效后行“复合式小梁切除术”，术中巩膜瓣下置氟尿嘧啶棉片湿敷2 min，大量盐水冲洗。术后3 d视力0.5，眼压8 mmHg。结膜轻度充血，缝线在位，滤过泡轻度隆起，角膜尚清，前房轴深约3CT，虹膜纹理尚清，上方周切口通畅，虹膜表面可见大量棕黄色色素结节，且多数带蒂。瞳孔大小约4 mm，对光反应存，其余体格检查基本与治疗前相同。随访3个月时，左眼最佳校正视力0.5，眼压20 mmHg，前房中深，滤过泡功能良好(图8)，视野显示MD=−10.77 dB，局部未用降眼压药。本病例报告患者已签署知情同意书。

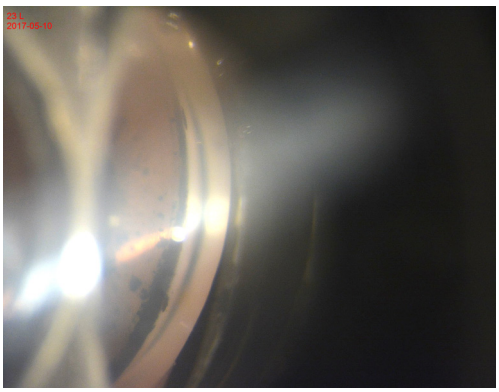

图1 房角镜下见房角见大量色素沉着

Figure 1 Gonioscopy revealed a large quantity of pigments in the angle

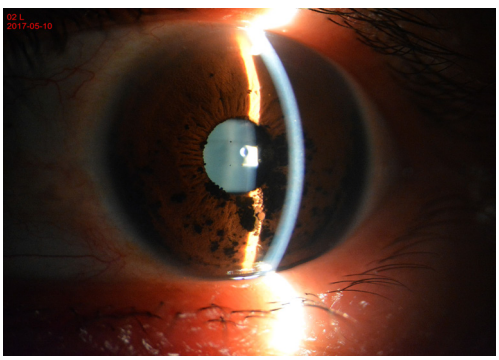

图2 眼前节照相示虹膜表面大量棕黄色色素结节，多数带蒂且以下方为密集

Figure 2 Anterior segment eye photograph: the photograph revealing the presence of multiple iris nodules, most of them pedunculated and the inferior is intensiveand intensively distributed in the inferior direction

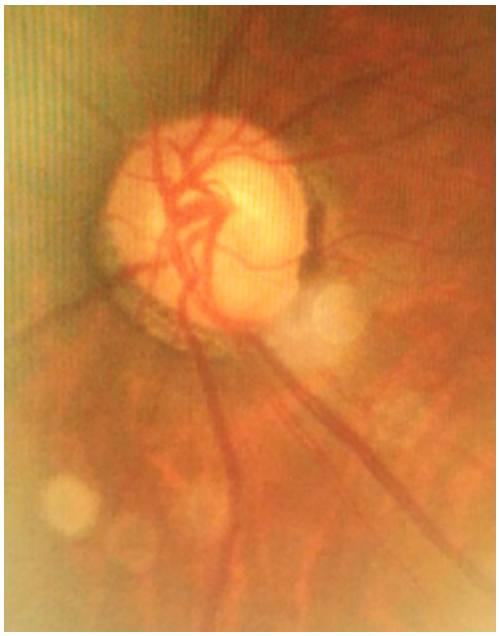

图3 眼底照相示：杯盘比约为0.7，伴中央血管鼻侧移位

Figure 3 Fundus photography confirmed the presence of the cup-disk ratio was 0.7 with nasal shifting of central vessels

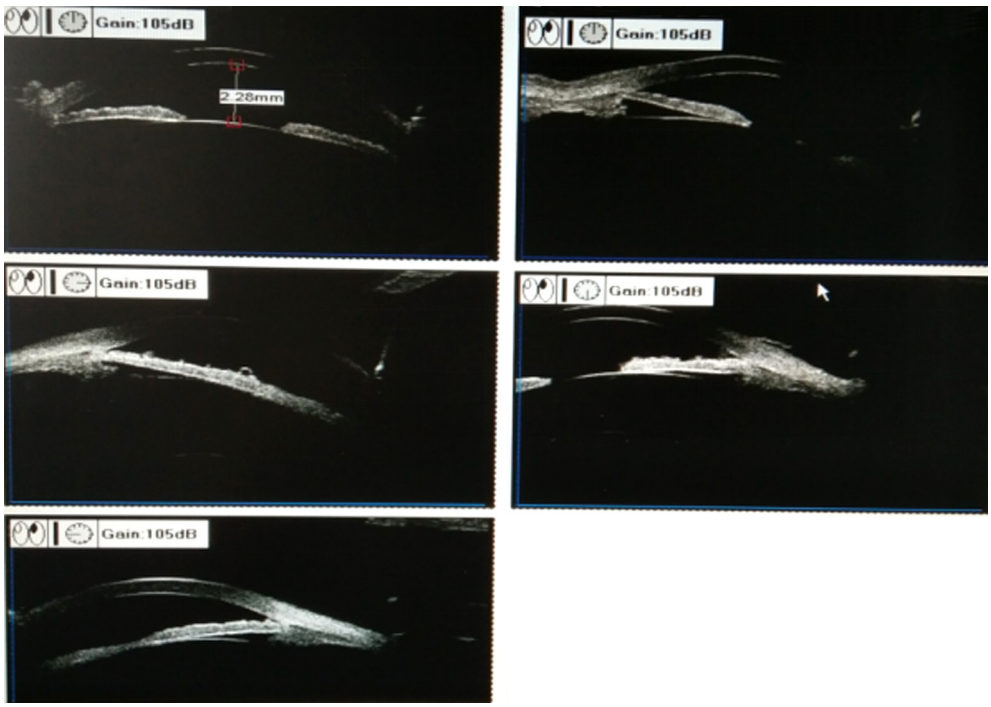

图4 UBM：虹膜表面欠光滑并见多个高反射点

Figure 4 UBM shows the iris surface was unsmooth and accompanied by multiple high reflectance points

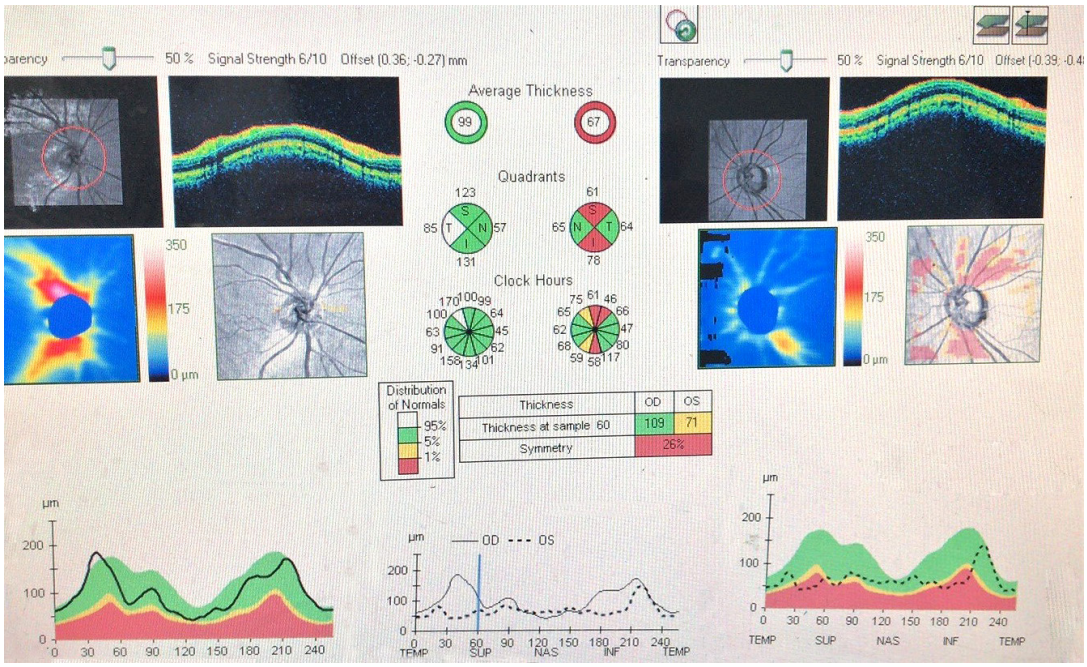

图5 ONH OCT：上方及下方视神经纤维层厚度变薄

Figure 5 ONH OCT shows marked loss of RNFL fibers in the inferior and superior sector

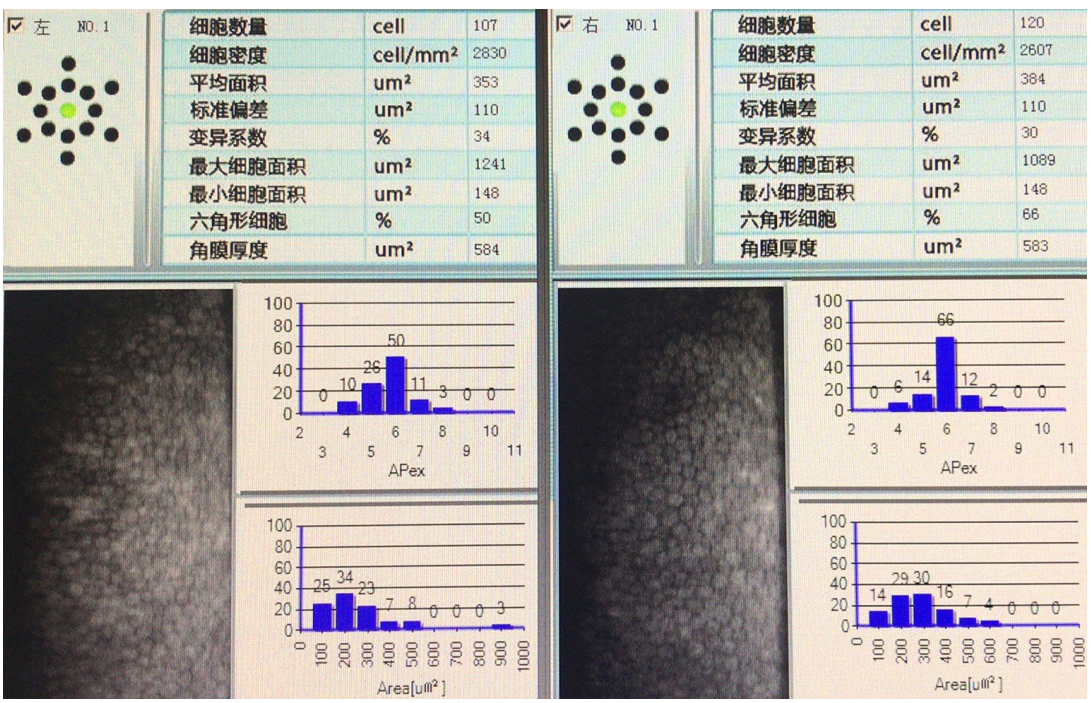

图6 角膜内皮镜示：角膜细胞密度、平均面积、变异系数均在正常范围内[2]

Figure 6 Peculiar microscopy shows endothelial cell density (MCD), mean cell area (MCA) and coefficient of variation (CV) in cell area were within normal ranges[2]

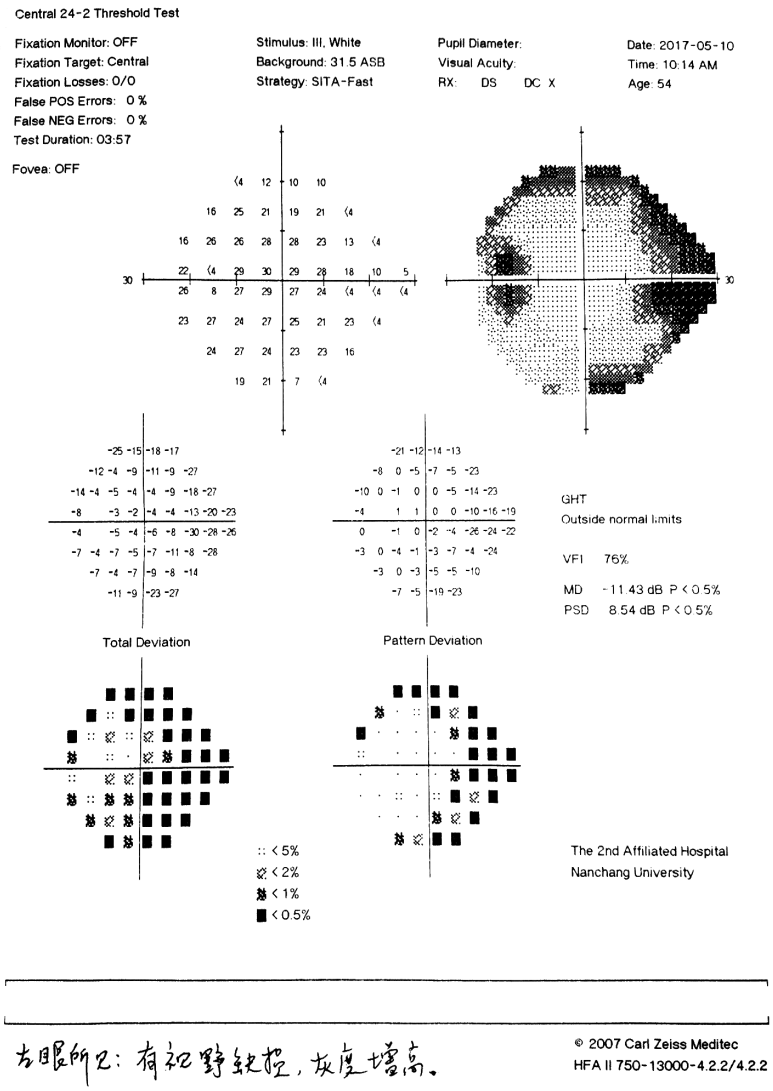

图7 Humphrey视野检查仪示：环形暗点，MD=–11.43dB

Figure 7 Humphrey visual field shows ring scotoma, MD=–11.43 dB

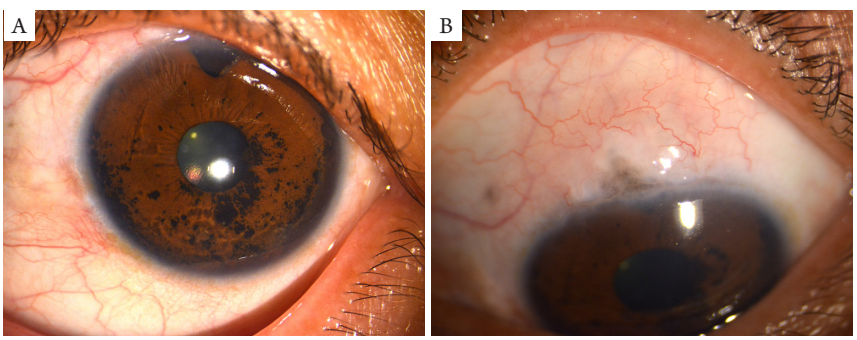

图8 术后3个月检查

Figure 8 Anterior segment eye photograph in the end of 3th month

(A)前房深度为中等；(B)滤过泡为功能性。

(A) Anterior chamber depth was medium; (B) Filtering bleb was functional.

2 讨论

ICES三型具有各自的虹膜特征，但角膜内皮异常增生是三者的基本病因，角膜内皮镜检查可发现角膜内皮呈弥漫性微细橘皮样外观，内皮细胞大小、形态及密度有不同程度的多型性改变。Cogan-Reese综合征诊断通常是由以下不同的特征观察：存在多个虹膜结节，通常有蒂，周围间质的虹膜呈现亏损的隐窝和蓬乱的外观。虹膜结节可能发生在疾病晚期，虹膜表面淡黄色结节可表现为早期表现，但病程后期，结节最终变成棕色且数量增加[1]。Cogan-Reese综合征还需要与其他条件：类似虹膜改变的如神经纤维瘤、黑色素瘤的鉴别诊断。与Cogan-Reese综合征不同，神经纤维瘤显示虹膜结节双边、平坦，更类似于虹膜痣。在Cogan-Reese虹膜结节的另一个重要的鉴别诊断是虹膜恶性黑色素瘤。主要的鉴别诊断方法是参考流行病学特征和临床特征：1)皮肤及虹膜颜色、强光接触等因素；2)巩膜表面显著扩张的“哨兵血管”；3)肿瘤迅速生长侵蚀晶状体和前房角导致自内障和青光眼的发生发展、晶状体脱位等；4)肿瘤的生长速度及范围[3]。

Chandran等[4]回顾性分析了1988年1月至2013年6月印度第三级眼科护理中心203个患ICES的患者，最常见的临床变异型为进行性虹膜萎缩(52%)，其次为Chandler综合征(39%)和Cogan-Reese综合征(9%)，70%以上与青光眼有关，一半具有角膜水肿表现，其中Cogan-Reese综合征角膜水肿占6%，其特征为呈圆形、不规则、高色素样虹膜结节。对于青光眼，50%予以药物得到控制，50%仍需要手术治疗：小梁切除术87例(33例术眼使用了丝裂霉素)，Ahmed青光眼阀植入7例，经睫状体光凝术20例，接受小梁切除术的27%患眼需要第二次手术来控制眼压，难治性患者行青光眼阀植入术。Chandran等[5]在另一篇关于小梁切除术联合丝裂霉素-c治疗ICES继发青光眼的疗效研究中发现其手术成功率6个月为94%，12个月为82%，36个月为71%，60个月为60%，Coviltir等[6]报道了小梁网切除及联合丝裂霉素对治疗ICES具有一定的疗效。王晓冰等[7]在10例ICES中有3眼行Ahmed青光眼阀植入术，其中1眼有1次小梁切除术史，2眼有2次小梁切除术史。术后浅前房低眼压(5 mmHg)1眼，考虑可能与穿刺口过大或术后引流盘周围未能形成有效组织包裹所致，经药物治疗术后14 d恢复，术后随访8~17个月，眼压控制良好．视力及视野均无进一步损害。由此可见根据患者临床表现及病情选择恰当的滤过手术对ICES眼压控制具有一定疗效。本次Cogan-Reese综合征病例的特别之处在于临床表现不典型：角膜水肿不显著，角膜内皮镜检查大致正常，房角镜下未见粘连、瞳孔领外翻，可能是由于房角大量色素附着遮蔽所致。本例在经药物治疗无效后行“复合式小梁网切除术”，术中进行抗纤维化处理，早期眼压控制尚可，但应长期随访动态监测角膜、视神经、视野变化。

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