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假性剥脱综合征合并年龄相关性白内障病例报告1例并文献复习

张君丽[1]，万晓梅[1]，杜珂婕[1]，罗文娟[2*]

[1]青岛大学青岛医学院，山东 青岛
[2]青岛大学附属医院眼科，山东 青岛

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摘 要

本文报道1例我院收治的假性剥脱综合征合并年龄相关性白内障病例，并结合文献进行复习。患者于我院行左眼白内障超声乳化吸除联合人工晶状体植入术，病人术后视力显著改善，预后较好。假性剥脱综合征是一种与年龄相关的全身性细胞外基质异常疾病，眼部表现以晶状体前囊膜剥脱物质沉积为主要特征，常合并白内障、青光眼等并发症，因此，眼科医师应提高对该病的认识，术前充分评估并选择恰当的手术策略，以降低术中及术后并发症产生的风险。

关键词

假性剥脱综合征，年龄相关性白内障，青光眼

The Pseudoexfoliation Syndrome Complicated with Age-Related Cataract: A Case Report and Literature Review

Junli Zhang[1], Xiaomei Wan[1], Kejie Du[1], Wenjuan Luo[2*]

[1]Qingdao Medical College of Qingdao University, Qingdao Shandong
[2]Department of Ophthalmology, The Affiliated Hospital of Qingdao University, Qingdao Shandong

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Abstract

This paper reports a case of pseudoexfoliation syndrome complicated by age-related cataract

*通讯作者。

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admitted to our hospital and conducts a literature review. The patient underwent phacoemulsification and intraocular lens implantation in the left eye at our hospital. Post-operatively, the patient's visual acuity improved significantly, and the prognosis was favorable. Pseudoexfoliation syndrome is an age-related systemic disorder characterized by extracellular matrix abnormalities. The main ocular manifestation is the deposition of exfoliative substances on the anterior lens capsule, and it is often complicated by cataracts, glaucoma and other conditions. Therefore, ophthalmologists should enhance their understanding of this disease, conduct a comprehensive pre-operative assessment, and select appropriate surgical strategies to reduce the risks of intra-operative and post-operative complications.

Keywords

Pseudoexfoliation Syndrome, Age-Related Cataract, Glaucoma

1. 引言

假性剥脱综合征(Pseudoexfoliation syndrome, PEX)被认为是一种与年龄相关的全身性细胞外基质异常疾病[1]，由 Lindberg 等于 1917 年首次提出[2]。该病的特征性表现为眼内组织表面灰白色纤维样物质沉积，最常累及晶状体前囊膜、虹膜、小梁网等结构[3]。PEX 患者常合并年龄相关性白内障，其白内障的发生率较同龄人群显著增高，且进展更快[4]，更严重的并发症比如角膜内皮病变、瞳孔功能障碍、晶状体脱位或半脱位，晚期也能合并视神经病变，即假性剥脱性青光眼(PEXG)，最终导致失明[5]。本文对我院收治的 1 例 PEX 合并年龄相关性白内障进行分析，并结合相关文献，探讨其发病机制、临床特点、诊断要点、鉴别诊断及治疗策略，旨在为眼科医师对 PEX 及其并发症的诊疗提供指导。

2. 病历资料

患者女性，88 岁，因“双眼无痛性、渐进性视力下降 7 年”于 2025 年 03 月 18 日于我院就诊。患者既往体健，否认糖尿病、高血压、青光眼等病史，家族中无类似眼病患者。全身检查未见明显异常。眼科检查：视力(国际标准视力表)：右眼 0.2，左眼 0.12，矫正视力无提高；眼压：右眼 15 mmHg，左眼 18 mmHg；双眼球结膜无明显充血，角膜透明，前房深度可，房水清，瞳孔圆，直径约 3 mm，对光反应正常，晶状体前囊膜中央区透明，瞳孔缘及周边部晶体前囊膜表面见灰白色碎屑样膜状物附着，晶状体皮质不均匀混浊，右眼核硬度分级III，左眼核硬度分级II级，双眼眼底模糊可见视盘边界清，色可，C/D 约 0.3，视网膜平伏，黄斑中心凹反光未见，血管走形大致正常。

辅助检查：眼前节照相示双眼晶体前囊膜表面见灰白色碎屑样膜状物附着(图 1)；角膜内皮细胞计数示右眼 2899 个/mm^2，左眼 2591 个/mm^2 (图 2)；前房角镜检查示宽角，未见明显色素沉着；超声生物显微镜(Ultrasound Biomicroscopy, UBM)检查显示双眼晶状体前可见大量细小膜状物附着(图 3)；激光扫描检眼镜检查(Scanning Laser Ophthalmoscopy)未见明显异常(图 4)。综合分析后诊断为：年龄相关性白内障(双 未成熟期)、假性囊膜剥脱综合征(双)。

治疗经过：术前对患者眼部情况及全身情况进行充分评估，于 2025 年 3 月 20 日与局部麻醉下行左眼白内障超声乳化吸除联合人工晶体植入术，采用表面麻醉，术中可见晶状体前囊膜表面大量灰白色

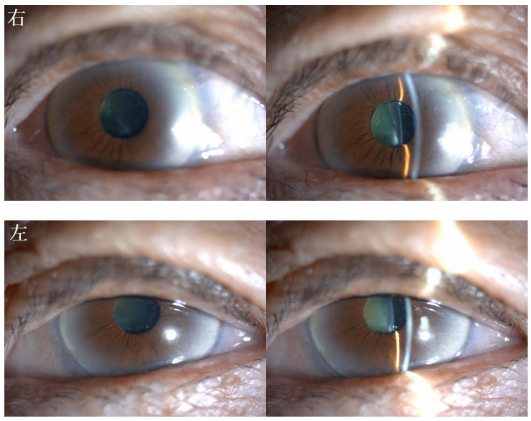
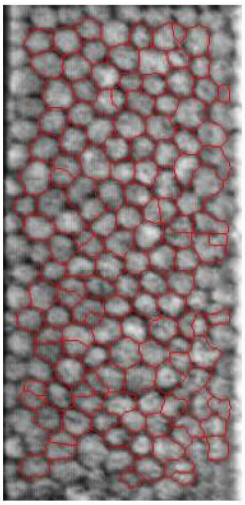


Figure 1. Anter segment photography
图 1. 眼前节检查图像

OD
右眼

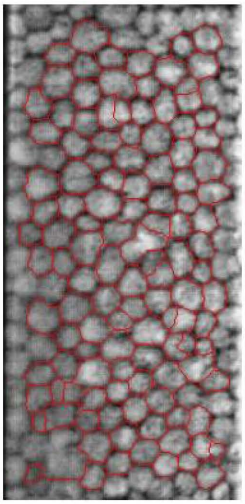


Corneal Thickness 512(UM)	
Number	139
CD(mm²)	2899
AVG(um²)	345
SD	130
CV	38
Max(um²)	905
Min(um²)	105

POLYMEGATHISM		
um2	%	50
0-100	0	
100-175	9	
175-250	14	
250-325	18	
325-400	30	
400-475	21	
475-550	4	
550-625	1	
625-700	0	
700-	3	

PLEOMORPHISM		
	%	50
3	1	
4	5	
5	20	
6A	49	
7	21	
8	4	
9	0	
10	0	

OS
左眼



Corneal Thickness 512(UM)	
Number	120
CD(mm²)	2591
AVG(um²)	388
SD	119
CV	31
Max(um²)	678
Min(um²)	132

POLYMEGATHISM		
um2	%	50
0-100	0	
100-175	5	
175-250	6	
250-325	22	
325-400	25	
400-475	20	
475-550	10	
550-625	10	
625-700	3	
700-	0	

PLEOMORPHISM		
	%	50
3	0	
4	4	
5	23	
6A	45	
7	23	
8	6	
9	0	
10	0	

Figure 2. Corneal endothelial examination
图 2. 角膜内皮检查图像

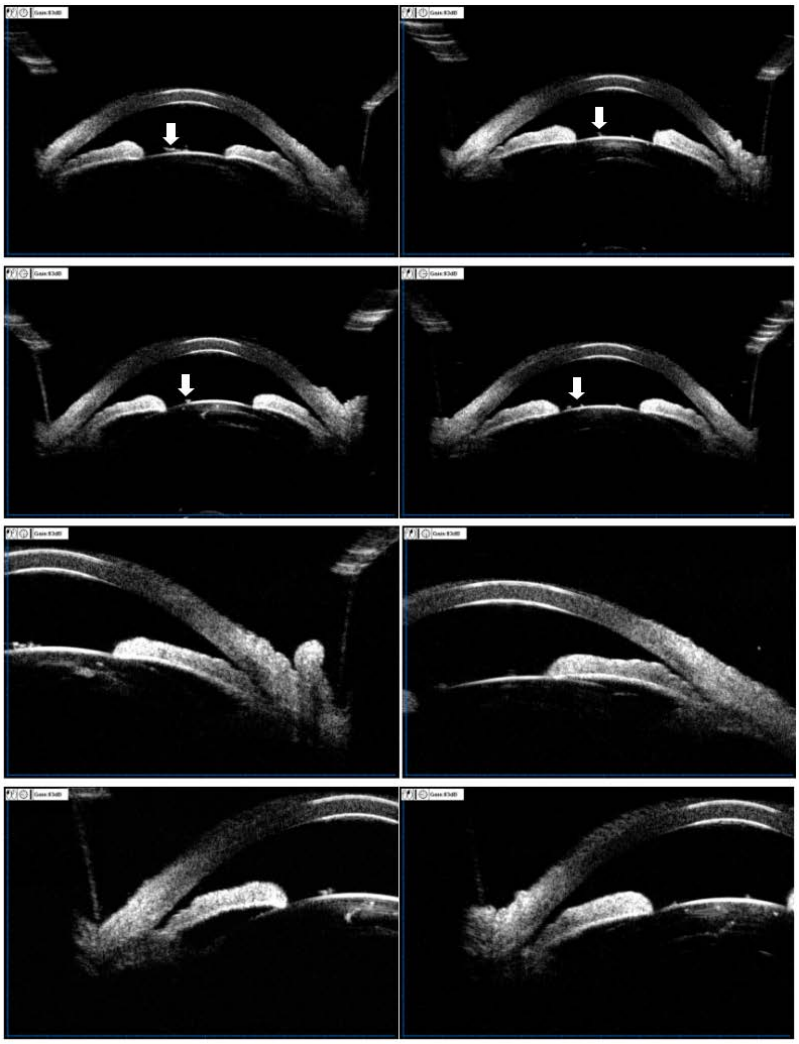


Figure 3. Ultrasound biomicroscopy (White arrows indicate the presence of debris-like membranous attachments)
图 3. 超声生物显微镜(白色箭头示碎屑样膜状物附着)

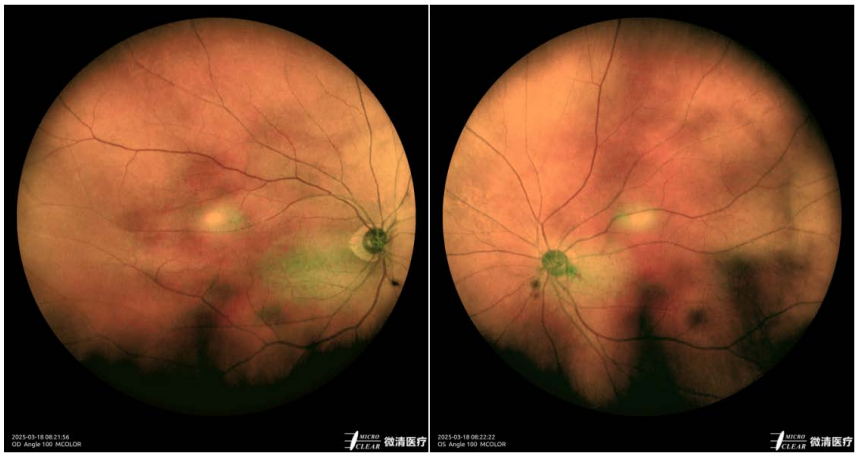


Figure 4. Laser scanning ophthalmoscope fundus examination image
图 4. 激光扫描检眼镜眼底检查图像

碎屑样物质，连续环行撕前囊，用核分块切除技术超声粉碎晶体核，换注吸头清除周边皮质及残留灰白色物质，注入粘弹剂后行后囊抛光，植入眼力健 ZCB00 + 24.5D 人工晶体一枚，清除粘弹剂后冲洗前房，封闭切口后地塞米松注射液 2.5 mg 球结膜下注射，典必殊眼膏包眼，手术顺利，安全返回病房。

术后第一天，眼科检查：左眼：视力：0.5，眼压：18 mmHg，球结膜轻度充血，角膜透明，前房中央深度正常，房水细胞(+)，瞳孔圆，对光反应正常，人工晶体位正。给予左氧氟沙星滴眼液预防感染，典必殊滴眼液、普南扑灵滴眼液，每天四次，减轻炎症反应。

出院后于 2025 年 4 月 1 日与我院门诊复查，眼科检查：视力：左眼 0.5，矫正视力：0.7，眼压：左眼 Tn，双眼结膜无充血水肿，角膜透明，前房中深，瞳孔圆，直径 3 mm，对光反应正常，右眼瞳孔缘及晶体前表面见白色碎屑样膜状物附着，晶状体皮质不均匀混浊，左眼人工晶体位正。暂未出现后发性白内障、青光眼等并发症。

3. 讨论

假性剥脱综合征(PEX)作为一种与年龄密切相关的全身性细胞外基质性疾病，其合并白内障或青光眼的临床随访一直是眼科医师面临的重大挑战。本例患者的诊治过程引发了我们对 PEX 合并白内障或青光眼等相关问题的深入思考，现结合最新文献进行如下讨论。

3.1. PEX 的病因学

PEX 的进展缓慢，其特征为异常纤维性物质在眼内及全身多个器官的进行性沉积。据调查显示，在不同地区的 PEX 患病率有所差异，芬兰、希腊、冰岛等地发病率高达 29% [6] [7]，我国 60 岁或以上患者的患病率为 0.4% [8]。PEX 的发病机制尚未完全明确，但研究表明其与遗传、环境及代谢等因素密切相关[9]-[11]。近年来，细胞外基质代谢异常被认为是该病的核心机制，尤其是 LOXL1 基因突变导致弹性纤维异常沉积，通过影响悬韧带及晶状体囊膜的稳定性从而产生各种并发症[12]。此外，氧化应激和紫外线暴露能加速异常纤维蛋白的聚集，尤其在高海拔或强光照地区的患者中更为显著[13]。同时该病具有明显的全身性特征，有调查显示，该类患者心脑血管疾病等全身疾病的发生率显著高于普通人群[1] [14]。

3.2. PEX 的临床表现及相关检查

PEX 的诊断主要依靠眼部特征性临床表现：1) 晶状体改变：最具诊断价值的是晶状体前囊膜的“靶心样”外观，由中央透明盘区(直径约 1.0~2.5 mm)、中间透明带及周边颗粒区组成，约 18%~20%患者可无中央盘区表现。早期可表现为均匀的毛玻璃样或草席样外观，随病程进展发展为典型的放射状条纹；2) 虹膜与瞳孔改变：瞳孔缘可见诊断性的灰白色碎屑样物质沉积，虹膜括约肌区域色素沉着显著而周边部少见。虹膜透照检查可见下方象限呈“蛾食状”或“漩涡状”色素脱失；3) 房角改变：房角镜检查可见小梁网色素沉着呈“沙砾状”分布不均，Schwalbe 线前方可能出现 Sampaolesis 线(色素线) [15]，散瞳后常见前房内色素颗粒大量释放[16]；4) 悬韧带改变：悬韧带常被剥脱物质包裹而变得脆弱，易发生断裂，导致晶状体震颤、半脱位甚至全脱位；5) 其他眼部改变：角膜内皮后可见剥脱物质沉积，内皮细胞计数下降或多形性改变[17]；前玻璃体膜也可有剥脱物积聚。

PEX 虽然主要表现为眼部体征，但其本质为一种全身细胞外基质性疾病，大量的流行病学及病理学研究表明，PEX 的物质沉积并不局限于眼部，它可以发生在身体多个器官和组织的血管壁和结缔组织中，因此，PEX 患者合并某些全身性疾病的风险增高。最常见的是心脑血管系统：多个研究发现 PEX 患者中高血压(尤其是难治性高血压)的患病率明显高于非 PEX 人群，发生心肌梗死和冠状动脉粥样硬化性心脏病的风险增加，该类疾病可能与物质沉积于血管壁导致其结构和功能改变有关[18] [19]；PEX 患者主动脉

壁中膜弹性纤维的 PEX 物质沉积导致其结构薄弱，导致腹主动脉瘤的患病率增加[20]。其次是神经系统病变：PEX 患者中阿尔兹海默病的患病率较非 PEX 患者更高，这可能与 PEX 物质沉积于脑膜血管和脑实质中有关，且二者具有相似的病理生理学基础(如 LOX1 基因的多态性、氧化应激、细胞外基质蛋白沉积等) [21]。另外有研究表明，PEX 患者还可以合并感音神经性耳聋、肝硬化、慢性肾脏病等全身各器官疾病[22]。

同时，全面而细致的术前检查对于评估风险、制定安全计划和术后管理至关重要。基础的检查包括：最佳矫正视力 BCVA、眼压 IOP、裂隙灯显微镜检查、前房角镜检查、OCT、眼底检查等。PEX 患者需重点关注的检查主要有：1) 超声生物显微镜：UBM 处于核心地位，重点评估晶状体悬韧带状态、前房角结构、晶状体是否脱位及睫状体状态等，并且 UBM 提供的关键信息直接决定了手术方式的选择(是否需要囊袋张力环、悬吊装置及手术技巧调整等)；2) 眼前节照相：该检查不仅可以客观清晰地记录 PEX 物质在晶状体前囊膜、瞳孔缘的沉积，也可以观察是否有虹膜色素缺失、萎缩等改变，同时在房角镜下拍摄还可能观察到 Sampaolesis 线(色素线)；3) 角膜内皮细胞计数：该检查主要用于评估术前内皮储备，对手术具有指导意义；4) 视野检查：若患者合并青光眼，进行该检查可评估相关视功能损害情况。因此，对于任何怀疑或确诊为 PEX 的患者，尤其是择期行白内障手术时，必须将 UBM 和高质量眼前节照相等检查列为术前检查的重中之重，它们提供的特异性信息直接关系到手术安全、预后及并发症的防范。

3.3. PEX 的鉴别诊断

PEX 需与真性剥脱综合征、色素播散综合征、虹膜炎继发青光眼、原发性家族性淀粉样变性等疾病鉴别。假性剥脱综合征与真性剥脱综合征是两种临床表现相似但病因、病理机制及预后具有显著差异的眼部疾病[23]。由于二者均可导致严重的视力损害，精准的鉴别诊断对制定治疗方案至关重要。真性剥脱综合征是一种更为罕见的疾病，主要由极端环境暴露或眼部创伤等引起，例如长期暴露于高温环境(如玻璃吹制工人)、红外线辐射、严重眼外伤或重度葡萄膜炎等[24]-[26]。与 PEX 不同的是，真性剥脱综合征并非系统性病变，而是局限性的晶状体囊膜损伤，发病机制主要是物理性损伤导致晶状体前囊分层。临床表现：1) 晶状体改变：裂隙灯检查可见晶状体前囊有边缘卷曲的透明薄片，剥脱的囊膜薄片可漂浮于前房内，与 PEX 的毛玻璃样或草席样外观不同；2) 常可发现与病因相关的体征，如高温暴露所致者可能有眼睑皮肤热烧伤的表现；外伤所致者可见相应外伤表现；3) 缺乏系统性表现：不伴有虹膜特征性色素改变、房角色素异常沉积等 PEX 的典型表现[27]。真性剥脱综合征患者合并白内障时手术难度及继发并发症的风险通常低于 PEX。

色素播散综合征的患者发病年龄相对较轻，多见于 30~40 岁近视患者，其中男性更易伴发青光眼；虹膜透照检查显示中周部放射状色素缺失，小梁网色素带致密且平滑，可见 Krukenberg 梭形角膜内皮色素沉着[28]。虹膜炎继发性青光眼的患者通常较年轻，眼科检查：房水闪辉(+)、可见炎症细胞，可伴有虹膜前后黏连[29]。原发性家族性淀粉样变性的患者伴有全身性疾病，超微结构显示淀粉样蛋白沉积，与 PEX 的纤维性物质不同，家族史呈阳性[30] [31]。

3.4. PEX 的并发症

PEX 患者合并白内障：有研究表明，患有 PEX 的白内障患者进展速度快于普通白内障[32]。PEX 合并白内障手术较普通白内障更困难，术中和术后并发症的风险更高[33]，因此全方位的围手术期管理及评估极为重要。术前重点评估：1) 通过 UBM 详细检查悬韧带状态，本例患者 UBM 检查示全周房角开放，晶状体悬韧带未见明显异常，睫状体及前部脉络膜未见脱离回声(图 3)；2) 评估角膜内皮细胞数量及质量，本例患者角膜内皮计数左眼 2591 个/mm^2，低于正常值(2899 ± 410 个/mm^2)，且 CV 值(细胞面积变异

系数)稍高于正常值(30%)、6A (六边形细胞百分比)低于正常值(50%) (图 2)，分析该患者角膜内皮状态稍差可能与高龄及 PEX 有关；3) 检测前房深度及房角结构，本例患者前房深度及房角结构正常(图 3)；4) 充分散瞳进行详细检查等。术中重点注意：合并 PEX 的白内障患者手术过程中面临更多挑战，包括悬韧带脆弱易断裂、瞳孔难以散大、浅前房，术后存在炎症反应重、前节色素膜反应、人工晶状体表面沉着物等并发症[15] [34]，针对这些风险，手术策略需要更加完善，例如角膜切口避开剥脱物质沉积区；避免祥悬韧带薄弱区施力；超声乳化采用低流量、低负压模式，缩短超声时间，增加冷却时间；采用囊膜张力环稳定囊袋；使用虹膜拉钩处理小瞳孔[35]；人工晶状体优先选择一片式疏水性丙烯酸人工晶体，旨在减轻术后反应及减少并发症发生，提高手术效果[36]。结合本例患者具体情况，术中我们采用了足量的具有高粘弹、高内聚性的粘弹剂，使其更好的维持前房，形成坚实的“软壳”保护内皮，抵御水流、器械和核块的冲击，并在手术结束前彻底清除粘弹剂，避免术后早期高眼压等问题；并且将超声乳化能量控制在低超声功率、低流量状态，并且轻柔操作，避免幅度过大扰动前房相关结构；植入的人工晶体 ZCB00 属于一片式疏水性丙烯酸人工晶体，更加适合 PEX 患者。最终，PEX 患者不管是否行手术治疗，都需长期随访，关注其全身情况，密切监测眼压、前房、囊带、晶体形态及位置等情况。

PEX 患者合并青光眼：PEX 患者青光眼的发生率显著高于普通人群[37]，其机制涉及多因素交互作用：1) 机械性房水引流障碍：剥脱物质在小梁网的沉积是核心病理特征。PEX 患者小梁网间隙被纤维碎屑和色素颗粒双重阻塞，小梁细胞数量减少 30%~50%，细胞外基质重构异常[38]。这种结构改变使房水流出阻力增加，眼压(IOP)进行性升高；2) 血－房水屏障破坏：PEX 患者虹膜血管通透性增高，房水中炎症因子(如 IL-6、TGF-β1)水平升高，导致小梁网炎症反应和纤维化[39]；3) 血管调节异常：视神经微循环障碍是 PEX 青光眼进展迅速的重要原因。研究表明，PEX 患者眼血流速度降低 40% [40]，视网膜神经纤维层(RNFL)厚度丢失速率是原发性开角型青光眼(POAG)的 2~3 倍[41]。值得注意的是，PEX 青光眼具有独特临床特征：昼夜眼压波动大；对药物治疗反应差，尤其晚期患者；视神经损害更重，即使 IOP 控制良好，RNFL 仍持续变薄[42]。PEX 合并青光眼的患者诊治更具多元化，需全面评估后进行个体化设计。药物治疗可选用一线药物(前列腺素类似物、β 受体阻滞剂)、二线药物(α2 受体激动剂、碳酸酐酶抑制剂)。激光治疗：选择性激光小梁成形术(SLT)，初期效果较好，但长期疗效有限[43]；激光周边虹膜切除术(Laser peripheral iridectomy)，用于合并房角狭窄者。手术治疗：小梁切除术、微创青光眼手术(MIGS)如小梁抽吸术，可清除小梁网剥脱物质、青光眼合并白内障时，可考虑超声乳化联合小梁切除术[43]。

4. 结论

综上所述，假性剥脱综合征合并年龄相关性白内障在老年人群中并不罕见，其特征性临床表现和手术挑战需要眼科医师充分掌握。详细的术前评估、个体化的手术方案和严谨的术后管理是获得良好预后的关键。本例患者通过精准治疗获得满意效果，但需长期随访监测可能出现的并发症。本文总结了 PEX 的常见病因、临床表现、检验检查、诊断、鉴别诊断及合并不同并发症的治疗特点，未来需要更多研究探讨 PEX 的发病机制和更优化的治疗策略。

声　明

本研究遵循《赫尔辛基宣言》原则，充分尊重了患者的自愿参与和知情同意权。该病例由患者或其法定代理人知情同意后参与。在病例资料收集过程中，已采取匿名处理，确保患者个人信息的安全及隐私。

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