Case Report

Posterior Segment Pathologies in Leprosy Patients with Visual Impairment: A Case Series

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Abstract

Leprosy is an important infectious disease, which is still prevalent in developing countries including Indonesia. This disease may affect the skin, extremities, peripheral nerves, and the eyes, causing disabilities of the patient. Leprosy is known to cause various ocular disorders, however, posterior segment abnormalities which involve the retina, choroid dan vitreous, have rarely been reported. Therefore, we present a case series of patients with posterior segment pathologies in visually impaired leprosy patients. These patients were identified during a community-based screening program on 99 leprosy patients, held in July 2019 in Alverno Hospital, Singkawang West Kalimantan. Patient comprised of inpatients and former patients who lived within the city and nearby cities who were invited for the program. All patients underwent comprehensive ophthalmology examination, performed by general ophthalmologists. Visual impairment was found in 15 patients, and their pupils were dilated, followed with posterior segment examination with indirect ophthalmoscopy and fundus photograph, using a handheld fundus camera, done by a vitreoretina specialist. Out of all patients screened, we did not find any leprosy-related posterior segment abnormalities. However, posterior segment pathologies were found in six patients, including peripapillary atrophy, myopic crescent, drusen and chorioretinal atrophy. One patient showed extensive chorioretinal atrophy with pigment clumping, which may be caused by leprosy-related chronic inflammatory process.
Keywords: *leprosy, posterior segment of the eye, visual impairment.*

Patologi Segmen Posterior Mata Pasien Lepra dengan Gangguan Penglihatan: Sebuah Serial Kasus

Abstrak

Kusta atau lepra merupakan penyakit infeksi penting yang masih banyak dijumpai di negara berkembang termasuk Indonesia. Penyakit ini dapat mengenai kulit, ekstremitas, saraf tepi, dan mata sehingga menyebabkan kecacatan pada pasien kusta. Kusta dapat menyebabkan berbagai gangguan mata, namun, kelainan segmen posterior yang melibatkan retina, koroid dan vitreous, jarang dilaporkan. Oleh karena itu, kami menyajikan case series yang melaporkan patologi segmen posterior pada pasien kusta yang mengalami gangguan penglihatan. Pasien-pasien ini kami temukan dalam program skrining berbasis komunitas pada 99 pasien kusta, yang diselenggarakan pada Juli 2019 di Rumah Sakit Alverno, Singkawang, Kalimantan Barat. Pasien terdiri atas pasien rawat inap dan mantan pasien yang tinggal di dalam kota dan dari kota-kota terdekat, yang diundang untuk mengikuti program tersebut. Seluruh pasien kusta yang datang menjalani pemeriksaan oftalmologi komprehensif yang dilakukan oleh dokter spesialis mata. Pada 15 pasien dengan gangguan penglihatan, dilakukan dilatasi pupil, dilanjutkan pemeriksaan segmen posterior dengan oftalmoskopi indirek serta foto fundus menggunakan kamera fundus hand-held, yang dilakukan oleh spesialis vitreoretina. Dari semua pasien yang diskrining, kami tidak menemukan kelainan segmen posterior terkait kusta. Namun, patologi segmen posterior ditemukan pada enam pasien, termasuk atrofi peripapiler, sabit rabun, drusen, dan atrofi korioretina. Satu pasien menunjukkan atrofi chorioretinal yang luas dengan gumpalan-gumpalan pigmen, yang mungkin disebabkan oleh proses inflamasi kronis terkait kusta.
Kata kunci: *gangguan penglihatan, lepra, segmen posterior mata.*

Introduction

Leprosy is a chronic infectious disease caused by *Mycobacterium leprae* that mainly affects the skin and peripheral nervous system. Complications from leprosy may cause permanent damage to the organs affected and cause disabilities of the leprosy patient, even after the disease has been cured.[1] Globally, 202.185 new cases of leprosy were detected, with a new case detection rate of 25.9 per million population. The highest proportion was found in South-east Asian Region, accounting for 71.3% of cases. Three countries with the highest incidence rates are India, Brazil, and Indonesia, respectively reporting more than 10.000 leprosy cases.[2]

Leprosy can lead to debilitating ocular complications, making it even more challenging for patients to care of themselves.[1] Ocular involvement found in 70-75% of patients with leprosy and blindness occurs in 5% of patients. Predominantly, leprosy affects the anterior segment of the eye, including lagophthalmos, entropion, ectropion, trichiasis, corneal hypesthesia, corneal ulcer, uveitis, scleritis, and glaucoma.[3] Conversely, there are only few reports regarding leprosy-related posterior segment lesions. Therefore, this case series describes the posterior segment pathologies found in visually impaired leprosy patients in a community-based screening program in Singkawang, West Kalimantan, Indonesia. The main study has been granted ethical approval from the University of Indonesia ethics committee with protocol number 19-05-0633.

Case Illustration

These cases were found during a community-based healthcare program for leprosy patients. Details of the program had been reported previously.[4] In brief, a joint team of ophthalmologists, dermato-venereologists, and medical rehabilitation experts, titled "Katamataku (*identifikasi tanda-tanda kelainan mata, kulit dan ekstremitas pada kusta)"* of the University of Indonesia, collaborated to deliver comprehensive examination and health promotion in large leprosy patient communities in Indonesia. The program lasted for two days in one of Indonesia's largest leprosy patient communities in Singkawang, West Kalimantan, on July 19 and 20, 2019. Patients comprised of inpatients, and former patients living within Singkawang and nearby cities, who were invited to participate in the program. All patients received explanation about the program and gave written consent prior to the examination. Each patient was subjected to detailed history taking, followed by examinations involving the eye, skin, and extremities.

Ophthalmology examinations consisted of best-corrected visual acuity (BCVA), corneal sensitivity test using cochet bonet aesthesiometry, tear production test using schirmer's test, intraocular pressure measurement, and slit-lamp biomicroscopy. Posterior segment examination was performed if BCVA was worse than 6/18 or discordant with the anterior segment findings. Posterior segment was evaluated following pupil dilation with 1% tropicamide eye drop, using 78D condensing lens and binocular indirect ophthalmoscope. Fundus photography using Visuscout100® handheld fundus camera (Carl Zeiss Meditec AG, Jena, Germany) was also performed. Of the 99 leprosy patients in the program, 14 patients underwent posterior segment examination to determine the cause of decreased visual acuity. Patients' characteristics are presented in Table 1.

Table 1. Demographic and Clinical Characteristics of Patients Receiving Posterior Segment Examination

Demographic and Clinical Characteristics	Frequency	%
Male gender	11	79
Mean age ± SD, years	62 ± 4.32	
Mean duration of leprosy ± SD, years	18.5 ± 20.25	
Visual acuity category (n = 28 eyes)		
Mild visual impairment (≥ 6/18)	11	39
Moderate visual impairment (< 6/18 to ≥ 6/60)	6	22
Severe visual impairment (< 6/60 to ≥ 3/60)	5	17
Blindness (<3/60 to NLP)	6	22
Eyelid and anterior segment ocular manifestations (n = 14)		
Lagophthalmos	5	36
Madarosis	10	71
Ectropion and entropion	5	36
Pterygium	5	36
Corneal scar	2	14
Cataract	7	50
Posterior segment involvement (n = 14)		
Unilateral	4	29
Bilateral	2	14
None	8	57

NLP: no light perception; SD: standard deviation.

Most patients were male (79%), with mean age of 62 years (34-90 years). Eight patients (57%) had multibacillary leprosy, one patient (7%) had paucibacillary leprosy, and leprosy type of the other five patients was unknown. While every patient exhibited multiple ocular pathologies, the most common one was madarosis, found in 71% of patients. Almost half of the eyes suffered mild visual impairment (39.2%). Out of 14 patients (28 eyes), abnormal fundus conditions were detected in six patients (eight eyes). Details of patients with posterior segment abnormalities are elaborated in Table 2. Fundus photographs of three leprosy cases, which were selected to demonstrate the variation of posterior segment pathologies are displayed in Figure 1-3.

Table 2. Clinical Features of The Eyes with Posterior Segment Pathologies

Number	Age (years)	Leprosy type	Duration of leprosy (years)	BCVA	Posterior segment findings	Other vision-threatening ocular problems
1*	57	PB	3	5/60	Peri-papillary atrophy	Aphakia
2*	57	PB	3	5/60	Peri-papillary atrophy, minimal macular drusen	Aphakia
3†	64	MB	40	1/60	Extensive chorioretinal atrophy, pigment clumping	Corneal scar, senile immature cataract
4†	64	MB	40	6/24	Extensive chorioretinal atrophy, pigment clumping	Senile immature cataract
5	63	MB	35	6/18	Extramacular drusen	Senile immature cataract
6	67	U	60	6/30	Drusen	Pseudophakia, mild corneal edema
7	90	U	10	1/300	Peri-papillary atrophy, myopic crescent	Aphakia
8	90	U	8	6/30	Drusen	Senile immature cataract

BCVA: best-corrected visual acuity, PB: paucibacillary, MB: multibacillary, U: unknown type of leprosy, *belong to the same patient, †belong to the same patient

Patient 1

The first patient was 57-year-old male with paucibacillary leprosy of three years (case no. 1 and 2 in Table 2). BCVA was 5/60 on both eyes. Both eyes were aphakic. Posterior segment examination revealed bilateral peri-papillary atrophy and minimal macular drusen on the left eye (Figure 1).

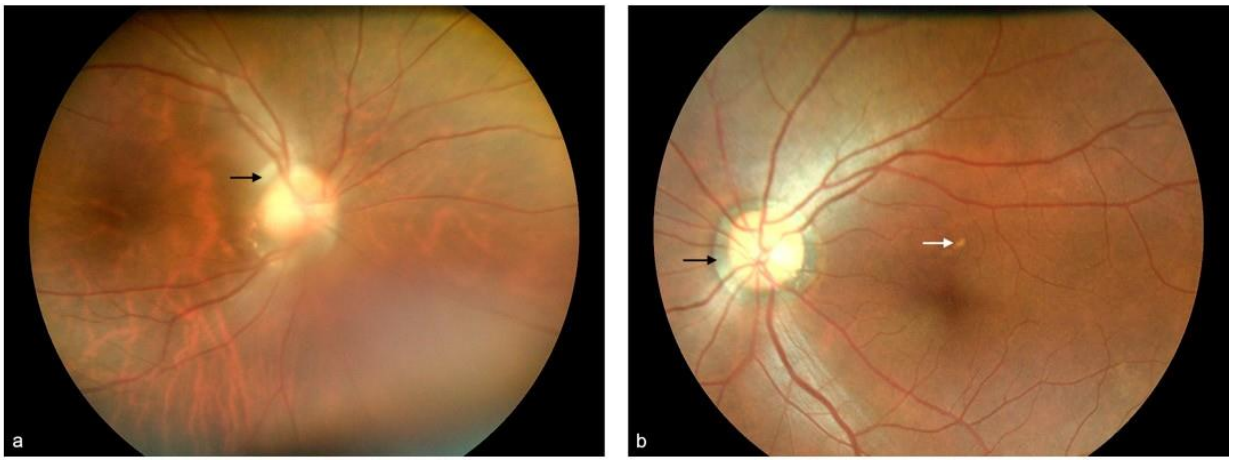

Figure 1. Fundus photograph of the right eye (a) and the left eye (b) from the first patient, showing bilateral peri-papillary atrophy (black arrow) and minimal macular drusen on the left eye (white arrow).

Patient 2

The second patient was a 64-year-old male with multibacillary leprosy of 40 years (case no. 3 and 4 in Table 2). BCVA was 1/60 on the right eye and 6/24 on the left eye. There was corneal scar on the right eye and cataract in both eyes. Posterior segment examination revealed extensive chorioretinal atrophy with retinal pigment epithelium (RPE) clumping (bone spicule) along the retinal arcade vessels almost symmetrical on both eyes (Figure 2).

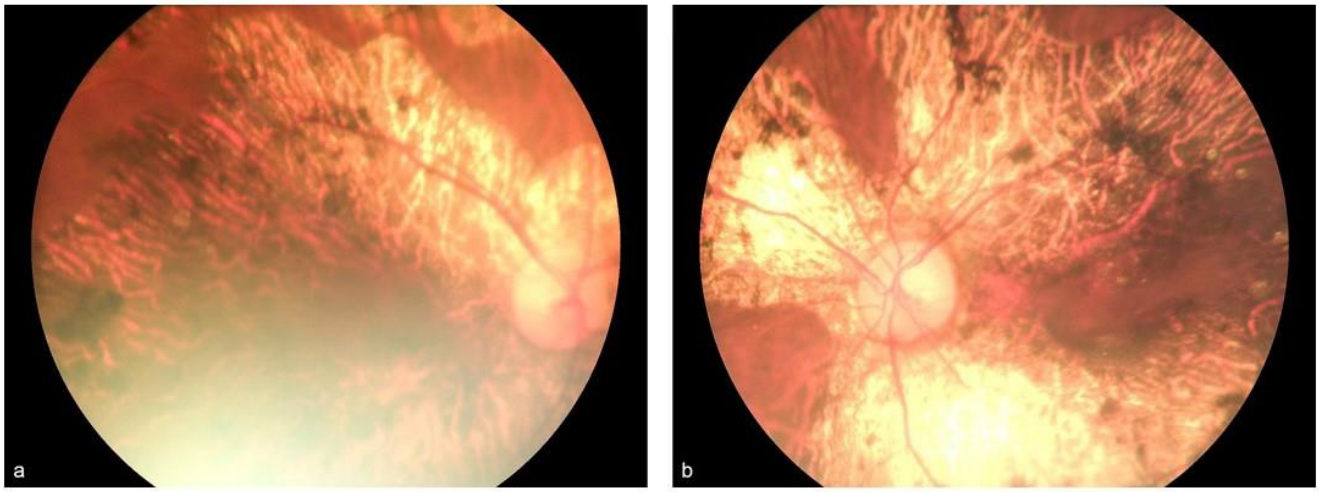

Figure 2. Fundus photograph of the right eye (a) and the left eye (b) from the second patient, showing bilateral extensive chorioretinal atrophy with RPE clumping along the retinal arcade vessels.

Patient 3

The third patient was a 63-year-old male with multibacillary leprosy of 35 years (case no. 5 in Table 2). BCVA was 6/18 on the right eye and 6/12 on the left eye. There were bilateral corneal scar and cataract on the right eye. The left eye was pseudophakic with posterior capsular opacification. Faint extramacular drusen was found on the right eye (Figure 3a).

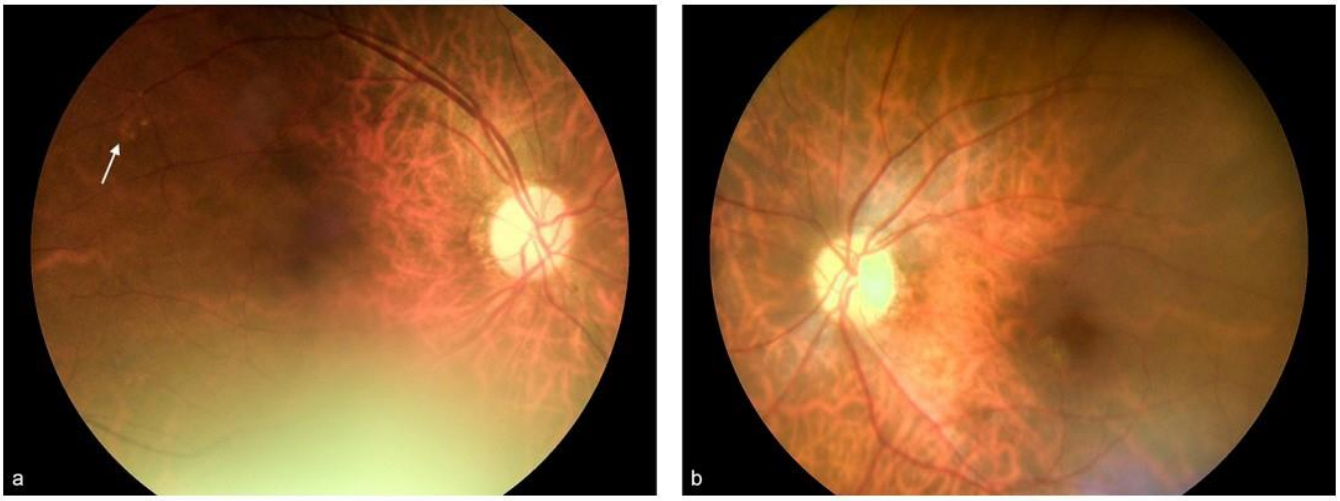

Figure 3. Fundus photograph from the third patient, revealing faint extramacular drusen was found on the right eye (a), pointed by white arrow.

Discussion

There are scant reports regarding posterior segment involvement in leprosy and the mechanism is poorly understood, previously mentioned is the possibility of direct spread from the ciliary body.[5] Sensitized uveal tract can lead to a few non-specific chorioretinal manifestations, such as proliferation of RPE, hypopigmented patches, choroiditis, retinal nodules, chorioretinal refractile bodies, and retinal vascular sheathing. Serous retinal detachment,

uveitis-related macular edema, and optic neuritis have also been reported as posterior segment manifestations of leprosy.[5–8] In this series, none of the aforementioned lesions were encountered. The rarity of leprosy-related posterior segment findings could be attributed to the difficulty in performing posterior segment evaluation due to anterior segment structure opacities or inadequate posterior segment evaluation in community-based studies.[3,9]

Posterior segment pathologies were not specifically related to leprosy. Peri-papillary atrophy was found in two patients (case no. 1, 2, and 7 in Table 2), but the poor visual acuity was most likely associated with aphakia. Drusen was encountered in four eyes (case no. 2, 5, 6, and 8 in Table 2), which was thought to be age-related. We found bilateral extensive chorioretinal atrophy with RPE clumping in one patient (Figure 2). This could be a sign of atypical retinitis pigmentosa or other hereditary chorioretinal dystrophy; however, it might also be related to past chronic inflammation, associated with leprosy. Diffuse RPE atrophy as a manifestation of leprosy-associated uveitis has been reported, which can be caused by direct bacterial invasion on the ocular structures and the formation of immune-complex hypersensitivity reaction and T cells-mediated immune response.[7] This presentation has similarities with chorioretinal atrophy and perivenous pigment clumping found in pigmented paravenous retinochoroidal atrophy (PPRCA). One of the proposed etiologies of PPRCA is inflammation, such as tuberculosis, rubeola, and syphilis.[10,11]

Unfortunately, further investigation was impossible during the screening. We only performed posterior segment examination if the patients' visual acuity was 6/18 or worse due to time constraint and resources limitation. Singkawang is a small city, located 151 kilometers away from the Pontianak, the capital city of West Kalimantan, where adequate ophthalmological service is still scarce. Therefore, patients with visual pathologies were referred to facilities with better resources. Furthermore, as posterior segment involvement in leprosy may only affect the peripheral retina, vision may be spared. Hence patients may not always complain of visual impairment. However, as peripheral lesions may be present in leprosy patients, posterior segment evaluation should be performed in all leprosy patients.

Conclusion

Even though specific leprosy-related posterior segment abnormalities were not found in this study, there were few posterior segment abnormalities that are commonly found in normal population. However we found one bilateral case with lesions probably caused by ocular inflammation in the past, which may be related to leprosy. Therefore, we recommend that every leprosy patient should undergo comprehensive ocular examination, including posterior segment evaluation.

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Disclosure

No financial disclosures, conflicts, or proprietary interests to report. This manuscript has been read and approved by all the authors. -

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