Unmasking Hansen's disease through an ophthalmologist's eye

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Key words: Corneal hypoesthesia, Hansen's disease, Iris pearls, Lagophthalmos, Madarosis

Leprosy or Hansen's disease (HD) is a chronic granulomatous inflammatory disease caused by intracellular, rod-shaped, acid-fast bacillus, Mycobacterium leprae (ML). The ML commonly affects cooler parts of the body like the skin, peripheral nerves, earlobes, mucosa of upper respiratory tract, testis, and eyes.

Within the eye, it usually affects the anterior segment, which has a relatively lower temperature and rarely involves the posterior pole. Ocular damage in leprosy occurs in four ways: (1) direct bacterial invasion, (2) facial and trigeminal nerve involvement, (3) hypersensitivity reactions, and (4) secondary infection.[1] Ocular manifestations include lagophthalmos, ectropion, entropion, trichiasis, dacryoadenitis, dacryocystitis, episcleritis, scleritis, punctate/avascular/interstitial keratitis, corneal anaesthesia/nerve beading/ulceration/opacity, iridocyclitis, iris atrophy, and nerve paralysis.[2]

A 56-year-old male presented to us with complaint of bilateral gradual painless progressive diminution of vision. He was covering his forehead with a cloth and was wearing a mask for protection from coronavirus. He was on oral methyl prednisolone 8 mg/day since 1 year for allergic skin disorder. On examination his best-corrected visual acuity was 1/60 and 2/60 in the right eye (RE) and left eye (LE) resp. Examination revealed papules and plaques on upper/lower lid skin with loss of lateral two-third eye lashes in both eyes (BE). There was lagophthalmos in LE with a lid gap of ~4 mm on gentle closure, no gap on forced closure, and a good bell's phenomenon [Fig. 1b]. BE had superior corneal pannus [Fig. 2], decreased corneal sensations, absent exposure keratitis, and a quiet anterior chamber (AC). The iris had a normal pattern, absent posterior synechiae (PS) with multiple tiny round creamy white opaque spheres known as iris pearls (IP) scattered all over its surface bilaterally [Fig. 3]. Round, small sluggishly reacting to light pupils (SSRLP) having a poor dilatation with mydriatics were present in BE. Nuclear sclerosis grade 2 with posterior subcapsular cataract and peripheral cortical cataract was present bilaterally with IP over anterior lens capsule in RE [Fig. 3a]. On fundus examination, only faint red glow was seen. On removing the cloth and mask, loss of lateral two-third eyebrows, erythrematous patch on cheeks, and a depressed nasal bridge were appreciable [Fig. 1a, b].

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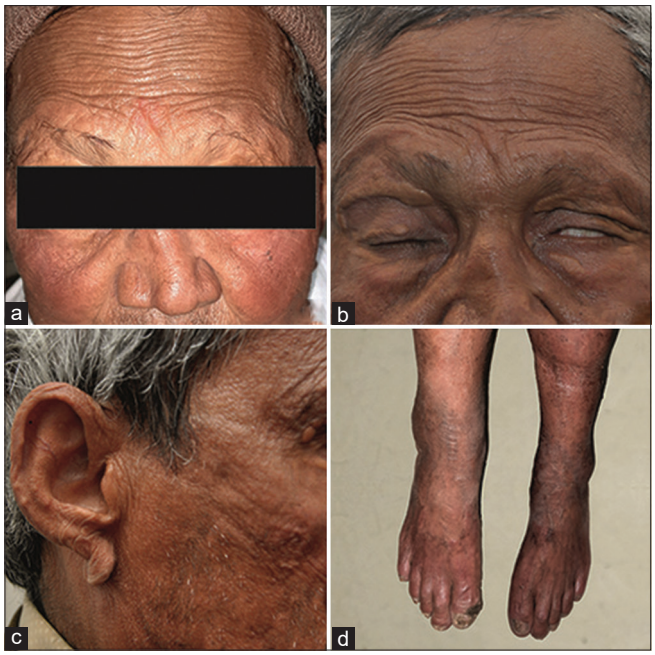

Figure 1: a: Photograph of face showing bilateral erythrematous patch on cheeks and a depressed nasal bridge. b: Photograph of face showing madarosis, papules, and plaques on upper/lower lid skin in BE with a lagophthalmos in LE. c: Photograph of face showing ear nodules. d: Photograph showing hyperpigmentation of skin of legs with involvement of toe

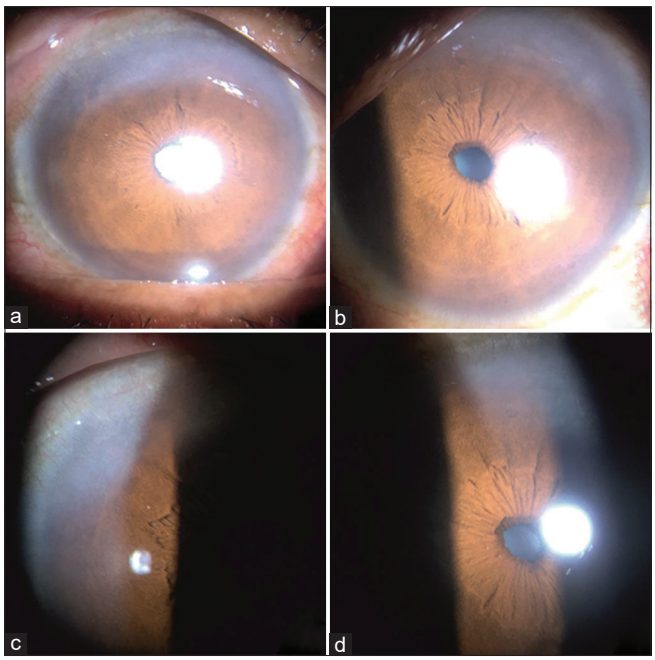

Figure 2: Slit-lamp photograph showing superior corneal pannus. a: Slit-lamp photograph (diffuse illumination) of RE. b Slit-lamp photograph (diffuse illumination) of LE. c: Slit-lamp photograph (slit illumination) of RE. d: Slit-lamp photograph (slit illumination) of LE

Intraocular pressure (IOP) was 08 mm Hg in BE. Gonioscopy revealed open angles up to the scleral spur with multiple IP scattered in the angles in BE [Fig. 4a-d]. A pattern resembling a necklace or beads of a rosary was seen in the superior angle in RE [Fig. 4a]. BE nasolacrimal duct syringing was patent.

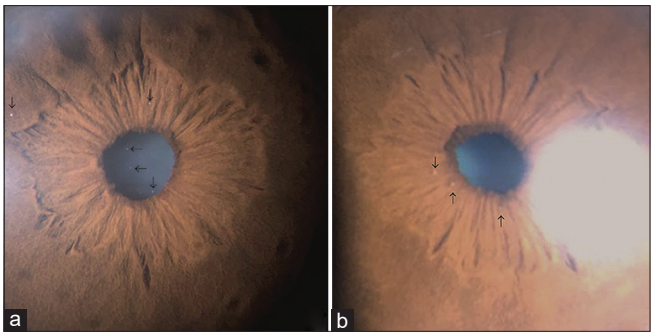

Figure 3: Slit-lamp photograph (diffuse illumination) showing iris pearls (IP) (black arrows). a: Slit-lamp photograph of RE showing IP on the anterior iris surface and anterior lens capsule. b: Slit-lamp photograph of LE showing IP on the anterior iris surface

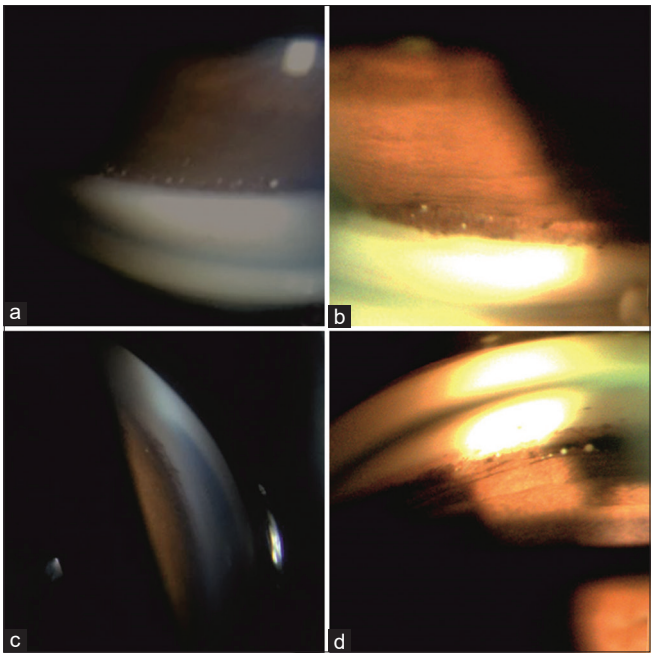

Figure 4: Goniophotographs showing Iris pearl(s).(a) Inferior goniophotograph showing superior angle of RE. (b) Inferior goniophotograph showing superior angle of LE. (c) Nasal goniophotograph showing temporal angle of RE. (d) Superior goniophotograph showing inferior angle of LE

The patient was diagnosed as HD and was referred to a dermatologist. On systemic examination, multiple macules and hypoesthetic skin lesions [Fig. 1d], ear nodules [Fig. 1c], and thickened ulnar nerves were present. A positive slit skin smear (SSS) for acid fast bacilli (AFB) confirmed lepromatous leprosy (LL) and multidrug therapy for HD was started. Phacoemulsification (PE) with intraocular lens implantation with iris hooks was advised for RE followed by LE. AC paracentesis (ACP) performed before PE in RE was AFB negative. Post op fundus examination was normal in RE. For the LE, blinking exercises, lubricant eye drops with ointment at night time were prescribed.

Discussion

Loss of hair follicles of eyebrows and eyelashes (madarosis) is a common sign of HD. It usually starts temporally and progresses nasally as was in our case. Another sign, corneal hypoesthesia

was present bilaterally with lagophthalmos and good bells phenomenon (LE) but without any corneal exposure and hence was managed conservatively.

IP are the diagnostic uveal manifestation of HD and pathognomonic of LL. They have been recognized in none among 250 patients,[3] 1/890 patients,[4] 2.02%,[1] and 4.8%[5] of leprosy patients in various studies. They are usually discovered mainly at the pupil portion around the collarette, in a pattern described as resembling a necklace or the beads of a rosary.[1] However, such a pattern was instead seen in the superior angle of RE on gonioscopy [Fig. 4a], which to the best of our knowledge is the first ever to be reported. They may occur in a clinically uninflamed eye[6] as seen in our case too.

In HD, pupils may be miotic, irregular, distorted, eccentric, with or without PS, and may have a sluggish or absent reaction.[2] Our case had small, round pupils without any PS with a diminished reaction.

Lewallen *et al*.[7] reported low IOP in a Hansen's population as compared to controls. Low IOP is caused by a reduced aqueous production due to destruction and late atrophy of the ciliary body.[8] Our case too had an IOP in single digits despite being on oral steroids.

Confirmation of the systemic disease is by the detection of bacilli on SSS or skin tissue biopsy. The diagnosis of lepromatous uveitis (LU) through skin, aqueous humor, and iris biopsy has been reported by Messmer EM *et al*.[9] Michelson JB *et al*.[6] and Campos *et al*.[10] also reported the diagnosis of ML through ACP in bilateral LU. The diagnosis in our patient too was ultimately established by an AFB positive SSS, while the RE ACP was AFB negative.

HD is not frequently encountered by ophthalmologists in their clinics nowadays. A delay in diagnosis leads to more complications and sometimes blindness. Hence, ophthalmologists should be aware and have a strong index of suspicion of HD in the presence of above ophthalmic features, including unexplained SSRLP/low IOP along with skin lesions/anesthesia, neurological involvement or nasal stuffiness.

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Conflicts of interest

There are no conflicts of interest.

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