

Type-II Lepra Reaction and Granulomatous Uveitis – An Unusual Presentation

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Abstract

We describe a case of young male who presented with lepra reaction with multiple macula-papular rash over face, forehead, ear lobules with peripheral neuropathy (Ulnar nerve thickening) and bilateral granulomatous uveitis. Slit skin smear was negative but skin nodular biopsy showed multiple discrete deeper dermis noncaseating well formed epithelioid cell granulomas with multinucleated giant cells intermixed with lymphocytes. Fite Faraco stain was negative suggestive of lepra reaction. Patient responded well with topical steroid and cycloplegic drops with disappearance of iris nodules. Systemic steroid was given for 12 weeks with gradual tapering to control lepra reaction and to prevent further ocular morbidity. Since ocular involvement can be seen even after completion of anti-leprosy treatment, the need for screening and periodic eye examination of the patient should be emphasized, for early identification of potentially sight-threatening lesions which can be easily treated. An ophthalmologist and a trained leprologist should preferably be included in the treatment of Hansen disease with ocular manifestations.

Delhi J Ophthalmol 2020;30;63-66; Doi <http://dx.doi.org/10.7869/djo.530>

Keywords: Erythema Nodosum, Iridocyclitis, Lepromatous Leprosy, Leprosy, Uveitis

Introduction

Leprosy (Hansen disease) is a chronic granulomatous multi-organ inflammatory disease caused by intracellular acid-fast gram-positive bacillus, the *Mycobacterium leprae*. Leprosy predominantly affects the skin, peripheral nerves, and eyes. Up to 75% of individuals with leprosy have ocular involvement and 40% have ocular disability.¹ *Mycobacterium leprae* has a strong preference for low temperatures, hence are mainly found in the skin, nose, earlobes and peripheral nerves. Within the eye, the organism is found only in the anterior segment which has relatively lower temperature. The organism has not been observed in the posterior segment or the optic nerve.² The eye is affected via direct invasion or during lepra reaction. Ophthalmic manifestations of leprosy include lagophthalmos, corneal ulceration, acute or chronic iridocyclitis, and secondary cataract.³ Ocular complications may also occur indirectly through impairment of lid closure (VII nerve) and corneal anaesthesia (V nerve) and through damage to adnexal tissues.³ Most of the ocular complications may lead to visual impairment and blindness; therefore, early detection and appropriate treatment is essential. Typically, the systemic disease is confirmed by detection of bacilli on slit skin smear or skin tissue biopsy.⁴ Histologic findings include multiple bacilli with acid-fast or Fite-Faraco stain positive, along with iris pearls. In addition to histopathology, polymerase chain reaction (PCR) can also be used to diagnose leprosy.⁴ In typical type-II lepra reaction without active leprosy, there is erythematous maculo-papular rash along with negative slit skin smear and Fite Faraco stain. Skin nodular biopsy shows noncaseating epithelioid cell granulomas with multinucleated giant cells intermixed with lymphocytes.

Case Report

A 36-year-old male presented with pain, photophobia, redness and diminution of vision in both eyes for 10 days duration. He was treated in primary health centre with antibiotics and lubricating drops. On examination, his distant visual acuity was 6/24 in both eyes; intraocular pressure was 21mmHg in right eye and 22mmHg in the left eye. There were Koppe and Busacca nodules [Figure-1(a)], koppe nodule, synechia and pigment release [Figure-1(b)], segmental synechiae [Figure-1(c)], matton fat keratic precipitates [Figure-1(d)] and hypopyon [Figure-1(e)] formation in both the eyes. There were multiple erythematous maculo-papulo-nodular lesions were present in the forehead, face [Figure-2(a)], forearms, arms and back. Ear lobules (Pinna) were grossly erythematous and thickened [Figure-2(b)]. Ulnar nerves were thickened. Other features including lagophthalmos, exposure keratopathy, corneal ulceration, conjunctival or scleral leproma were absent. He had history of similar episode of ocular pain, redness, photophobia twice in the past and had a history of one-year treatment for Leprosy five years back. A diagnosis of granulomatous uveitis was made and the patient was started on topical prednisolone acetate 1% 1drop 1 hourly along with Homatropine 2% 1 drop thrice daily. Skin specialist opinion was taken and was diagnosed as a case of type 2 Lepra reaction and the patient was put on systemic prednisolone (1mg/kg/body weight) slowly tapered over 12 weeks. Slit skin smear from forehead, cheek and ear lobules was negative for acid-fast bacilli. Skin incision biopsy was taken from macula nodular lesion of the arm which showed multiple discrete deeper dermis non caseating well formed epithelioid cell granulomas with multinucleated giant cells intermixed with lymphocytes.

[Figure-3]. Fite Faraco stain was found negative suggesting a lepra reaction [Figure-3]. Mantoux test was negative with normal radiogram and contrast-enhanced computed tomography of the chest. On haematological investigation, angiotensin converting enzyme levels was normal [21U/L]. Serological test for syphilis, human immune deficiency virus

and viral markers were alnegative. Complete blood count and blood sugar estimation was normal. Systemic involvement and iridocyclitis gradually improved with disappearance of iris nodules [Figure-4]. Later on he developed complicated posterior subcapsular cataract [Figure-5(a)] with diminution of vision (6/60) in both eyes. Ultrasonography –B scan was performed before surgery and the retina appeared normal. He then underwent cataract surgery with posterior chamber intra ocular lens [PCIOL] with surgical peripheral iridectomy [Figure-5(b)]. Post operatively his visual acuity was 6/12 in both eyes improving to 6/9 with correction. Intraocular pressure was 17mmg in both eyes. Patient is kept under regular follow-up.

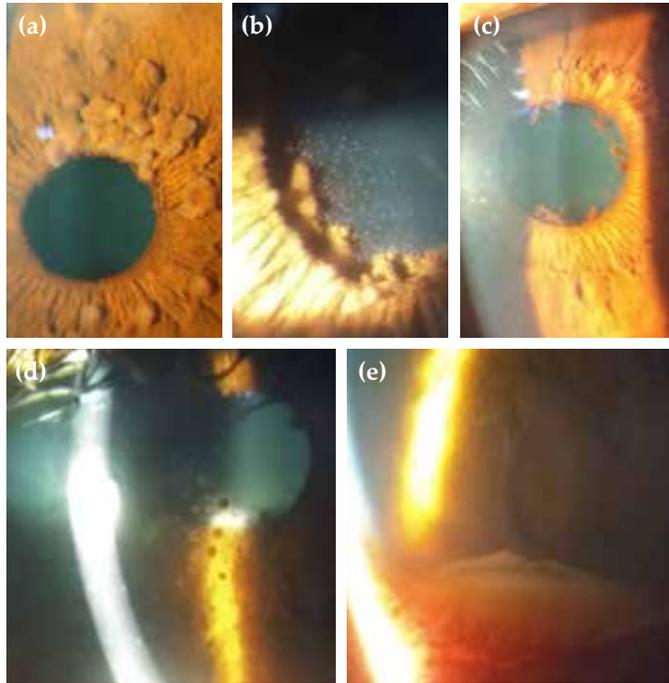


Figure 1: (a) koppe and bussaca nodule; (b) koppe nodule and pigment release; (c) segmental synechia; (d) mottan fat keratic precipitates; (e) hypopyon formation.

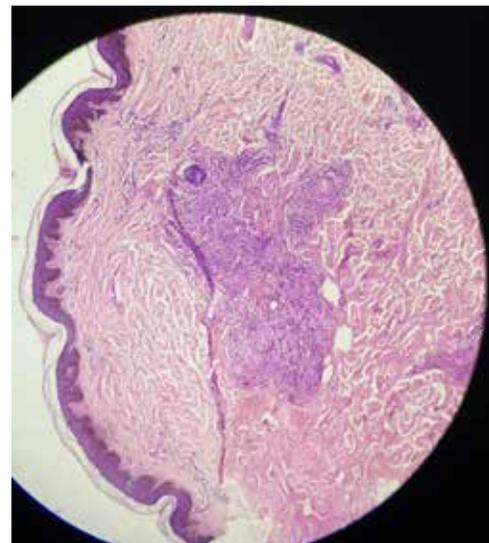


Figure 3: Nodular biopsy shows noncaseating well formed epitheloid cell granulomas with multinucleated giant cells intermixed with lymphocytes.

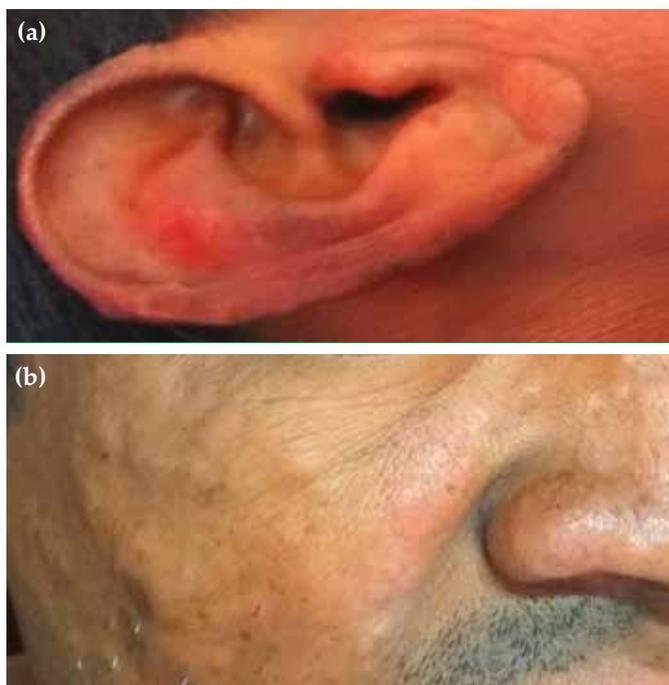


Figure 2: (a) macula papullo nodular lesion involving face; (b) Lepra reaction of pinna.



Figure 4: Disappearance of iris nodules after treatment.

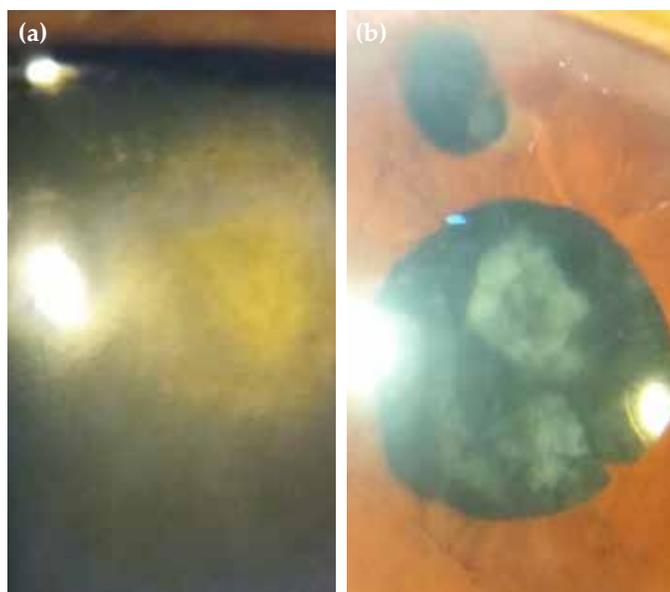


Figure 5: (a) Complicated posterior subcapsular cataract (PSC); (b) posterior chamber intra ocular lens and surgical peripheral iridectomy.

Discussion

Leprosy is a multi organ infectious disease affecting mainly the skin nerves and eyes. Skin lesions commonly are macules and plaques, rarely papules or nodules. Lesions vary from being hypopigmented with reduced sensations in tuberculoid leprosy to multiple confluent nodular lesions in lepromatous leprosy.^{1,2,3} Nerves are damaged in two locations, either peripheral nerve trunks near the fibro osseous tunnels or small dermal nerves in the skin lesions. Leprosy is also associated with type-1 and type-2 reactions.⁵ Type-1 reactions occur in patients with borderline leprosy and never with polar leprosy. This reaction manifests with signs of inflammation within macules, papules and plaques with appearance of new lesions and fever.⁵ Erythema Nodosum Leprosum (ENL) or type 2 reaction classically presents as tender erythematous nodules on the face, arms and legs. It occurs in lepromatous leprosy with skin infiltration and bacterial index of 4 or more.⁵

The present case was an outpatient case being treated for conjunctivitis elsewhere. Detailed ophthalmic evaluation was performed revealing features of granulomatous uveitis. This finding, along with systemic features and positive skin biopsy, confirmed the diagnosis of lepromatous leprosy with lepra reaction. Campos et al. reported the diagnosis of mycobacterium leprae through AC paracentesis in a case of bilateral iridocyclitis[6]. Lepromatous uveitis has also been diagnosed through skin, aqueous humor, and iris biopsy, as reported by Messmer et al.⁷

The reported frequency of iridocyclitis is 7%-24%.⁸ Iridocyclitis is a potentially blinding clinical manifestation of erythema nodosum leprosum (type II reaction), which results from antigen antibody reaction, mainly in multibacillary (lepromatous) leprosy.^{1,5} Acute inflammatory reaction

affecting the facial nerve, cornea and iris is characteristic of type II reaction, which may severely damage the eye, directly or indirectly.

Iridocyclitis is generally managed with topical steroids and cycloplegic drugs over a prolonged duration. Evidence strongly indicates that the most serious effects on body tissues consequent to infection of the skin, nerves, and eyes with Mycobacterium leprae are because of the immune response.⁵ In both lepromatous and non-lepromatous leprosy, adverse immunological reactions, either cell-mediated or humoral, may develop suddenly and have considerable severity, necessitating treatment with steroids or immunosuppressive drugs.⁹ The present case represents a clinical and histological demonstration of iridocyclitis, resulting from lepromatous leprosy, which could have progressed into blindness if left undetected or treated inappropriately. A close and long follow-up is required in these cases, as these patients are at risk of significant ocular morbidity, despite completing the multidrug therapy.⁹

Conclusion

It is recommended that an ophthalmologist and a trained leprologist, must be included in the treatment of Hansen disease with ocular manifestations. The risk of ocular complication increases with increased duration of disease and with lepra reactions. Since ocular involvement can be seen even after completion of anti-leprosy treatment, the need for screening and periodic eye examination of all the patients with leprosy should be emphasized, for early identification of potentially sight threatening lesions which can be easily treated.

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Cite This Article as: Das S, Pradeep B, Ojha P. Type-II Lepra Reaction and Granulomatous Uveitis – An Unusual Presentation.

Acknowledgments: Nil

Conflict of interest: None declared

Source of Funding: None

Date of Submission: 28 June 2019

Date of Acceptance: 6 October 2019

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