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Case Report

Interesting Unusual Associations in three Leprosy cases

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Leprosy is a chronic granulomatous disease with significant morbidity and a considerable social stigma attached to it. Here we report 3 cases with unusual associations of leprosy with other disorders. First case was a 53 year old female, treated case of lepromatous leprosy with recurrent painless bilateral periorbital swelling of 3 years duration predominantly involving the upper eyelids followed by laxity and hyperpigmentation, clinically diagnosed as Blepharochalasis. Second case was a 40 year old man, treated Borderline Tuberculoid (BT) case presented with a slowly growing asymptomatic swelling with multiple discharging sinuses on right foot since 2 months was diagnosed clinically as mycetoma foot and was treated with Modified Welsh regimen with marked improvement. Third case was 20 year old boy, born healthy, consanguineous parents, presented with generalised asymptomatic light and dark coloured lesions since 8years of age and large hypopigmented hypoaesthetic lesions on back and buttocks since 4 months. A diagnosis of BT leprosy with Dyschromatosis universalis hereditaria (DUH) was made and was given multibacillary multidrug therapy.

Key words : Leprosy, Unusual Associations, Blepharochalasis, Mycetoma, Dyschromatosis Universalis Hereditaria (DUH)

Introduction

Leprosy (Hansen's disease) is a chronic infectious disease caused by *Mycobacterium leprae,* a microorganism that usually affects skin and nerves leading to disabilities. The principle clinical manifestations of the disease are anaesthetic skin lesions, peripheral neuropathy and palpable enlargement of peripheral nerves.

Although the disease is usually well controlled by multidrug therapy (MDT), the disease can be aggravated by acute inflammatory reaction episodes that may cause permanent nerve impairments and resulting tissue damage.

Here we report three rare disease associations in leprosy patients from a Tertiary Care Centre in Maharashtra.

Case 1

A 53-year old female presented with laxity of the skin of the upper eyelids for the past 3 years which first manifested as recurrent painless bilateral

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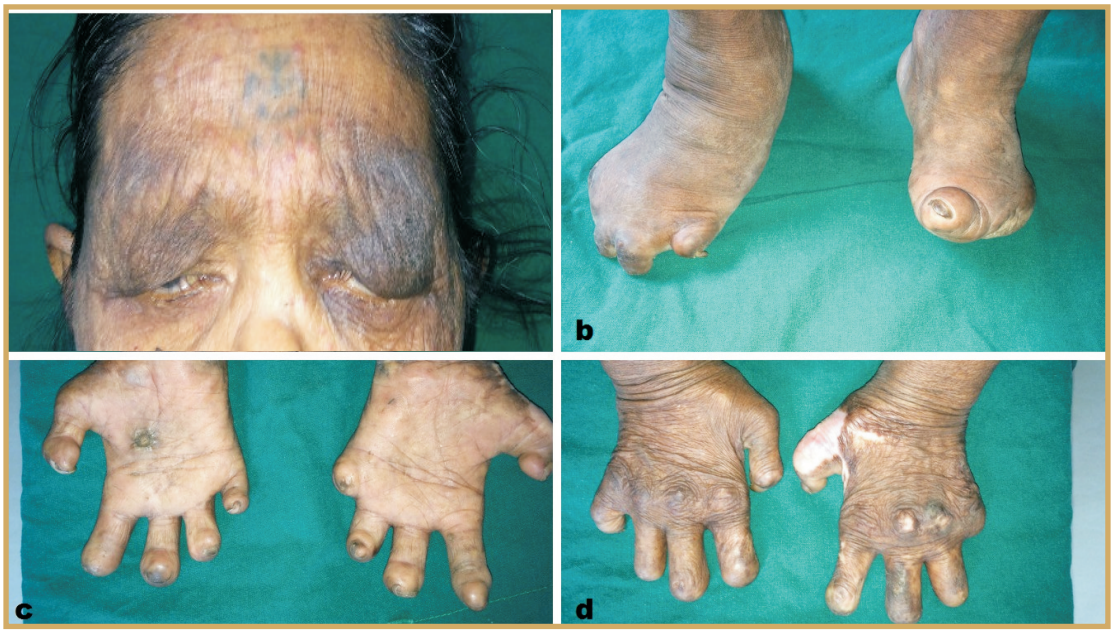

Fig. 1 : Case of leprosy with blepharochalasis

periorbital swelling mainly involving the upper eyelids followed by a gradual increase in the laxity of the skin at the same site. She was also a known case of lepromatous leprosy treated at the age of 5 years with Dapsone monotherapy taken for 3 years and had multiple disabilities. Local examination revealed lax wrinkled skin of the upper eyelids with hyper pigmentation. Lagopthalmos was present with interference of vision. Multiple localized well defined hypopigmented patches were seen over chest, back and face. The largest lesion measuring 12x6 cms and the smallest 2x2 cms. Madarosis and depressed nasal bridge (saddle nose deformity) were also present. There was resorption of finger and toes bilaterally of all the extremities. There was no swelling of the lips or the thyroid region. Her systemic examination was normal. Her haematological profile revealed low haemoglobin of 7.7gm%. Liver, renal and thyroid function tests, urine analysis, skull x-ray were all within normal limits. Slit skin smear was negative for AFB (acid fast bacilli). The patient denied consent for biopsy. The diagnosis of blepharochalasis was made based on the clinical presentation (Fig. 1). The patient even denied any corrective plastic surgical intervention for the same.

Discussion

Blepharochalasis is a rare degenerative disease of the skin of the eyelids, characterized clinically by unilateral or bilateral swelling followed by laxity, atrophy, wrinkling and pigmentary changes, predominantly of the upper eyelids (Burrows and Lovell 2004). The skin of the eyelids becomes so lax that it droops as redundant folds over the lid margins (Duke and Mac 1974). The term blepharochalasis was first coined by Fuchs in 1869, meaning eyelid relaxation in Greek (Collin et al 1979). It is also termed ptosis atonia, ptosis

adipose and dermatolysis palpebrum. The exact etiology of blepharochalasis is not known. Most of the cases are sporadic, but autosomal dominant inheritance has been noted in a few pedigrees (Burrows and Lovell 2004). Three stages are described in the evolution of blepharochalasis (Duke and Mac 1974). The first is the recurrent angioedema, while the second stage is characterized by discolored, flabby and lax skin, is called the stage of atonic ptosis. In the third stage, there is further relaxation of the tissues of the orbital septum, with prolapse of the orbital fat leading to interference of vision. This stage is called ptosis adipose. Systemic conditions associated with blepharochalasis are renal agenesis, vertebral abnormalities and congenital heart disease (Ghose et al 1984). Blepharochalasis may be associated with progressive enlargement of the upper lip due to enlargement of the labial salivary glands as well as thyroid swelling in Ascher's syndrome (Nicholos 1998). The only effective treatment is correction by plastic surgery after the disease has run its course; otherwise subsequent attacks of lid edema may interfere with the results (Nicholos 1998). Our patient was in second stage that is the stage of atonic ptosis.

Case 2

A 40 year old man presented with a slowly growing asymptomatic swelling with multiple discharging sinuses on right foot since 2 months (Fig. 2). He gave history of trauma to the same foot 4 years back in traffic road accident. Patient is a known case of lepromatous leprosy and took Dapsone 100 mg monotherapy for 5 years, 15 years prior to presentation.

Cutaneous examination showed ill defined swelling with multiple hyperpigmented nodules and discharging sinuses with serosanguinous discharge over the dorsal and posterior aspect of right foot. Systemic examination was unremarkable. Haematological and biochemical laboratory tests revealed diabetes mellitus. Serology for HIV was negative. Chest X-ray was normal. X-ray of the same foot showed osteoporotic changes in distal end of metatarsal, resorption of distal phalynx of 1st toe, bony deformity and cortical thickening in shaft of 2nd, 3rd, 4th metatarsals and cortical break. Biopsy revealed ill formed granulomas surrounded by foamy histiocytes with perivascular and periadnexal lymphocytic cell infiltrate in the dermis without evidence of actinomycotic or eumycotic granules. Zeihl-Neelson stain for actinomycosis was negative. Smears and cultures were negative for fungi, typical and atypical mycobacteria.

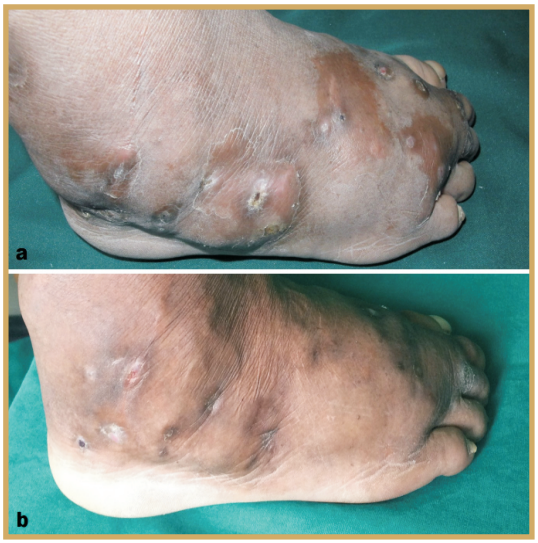

Fig. 2 : a) ill defined swelling with multiple hyperpigmented nodules and discharging sinuses over right foot (Before treatment) b) Decreased swelling and healing of sinuses after 3 cycles of Modified Welsh regimen.

The patient was started on 3 cycles of Injection Amikacin 250 mg twice a day for 21 days with 15

day intervals, and daily Cotrimoxazole (Sulfamethoxazole 800 mg and Trimethoprim 160 mg) twice a day and Rifampicin 450 mg, according to the Modified Welsh regimen, which was given as a trial on basis of clinical findings of myetoma. There was significant improvement in form of decreased swelling and healing of sinuses after 3 months.

Discussion

Mycetoma (often referred to as “Madura foot”) is a Greek term for “fungal tumor”. Dr John Gill provided the first formal description of mycetoma in 1842, in Madura (India), hence the name “Madura foot” (Welsh 1993). This infection results in granulomatous inflammatory response in the deep dermis and subcutaneous tissue, which can extend to the underlying bone. Mycetoma is characterized by the formation of grains containing aggregates of the causative organisms that may be discharged onto the skin surface through multiple sinuses. The grains are characteristic of etiologic agents, including a variety of bacteria (actinomycotic mycetoma) or fungi (eumycotic mycetoma). The foot is the most common site of infection, followed by the hand, trunk and scalp. Involvement is typically unilateral.

Talwar and Sehgal studied 60 clinically suspected cases of mycetoma. Of the 60 suspected cases, 20 were confirmed by culture and histopathological examination**.** The feet were found to be affected in 70% of these cases (Talwar and Sehgal 1979).

The combination of Amikacin with Cotrimoxazole is known as the Welsh regimen. Adding Rifampin to the Welsh regimen (Modified Welsh regimen) allows for remissions without recurrence (Damle et al 2008). Sixteen patients out of 18 patients of actinomycetoma completed the combination therapy, which lead to remission (Damle et al 2008).

Case 3

A 20 year old boy, born of healthy, consanguineous parents, presented with asymptomatic light and dark pigmented lesions all over the body since 8 years of age, the initial location of pigmented lesions was on the legs followed by progressive involvement of entire body sparing scalp, palms, soles, genitals and mucous membrane (Fig. 3). There was no history of photosensitivity, preceding dermatoses, drug intake, and exposure to chemicals or tars. There were no associated ophthalmological or auditory complaints. Developmental history was normal and no one in the family had similar complaints. Cutaneous examination revealed diffuse 1-2mm hyper and hypopigmented macules with few depigmented macules distributed symmetrically

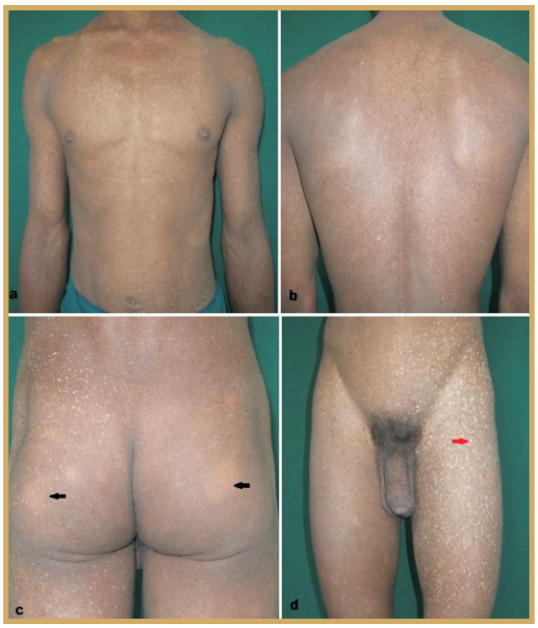

Fig. 3 : Case of leprosy with Dyschromatosis universalis hereditaria (black arrow - hypopigmented hypoaesthetic lesions of Hansen's disease; red arrow - hypopigmented lesions of DUH)

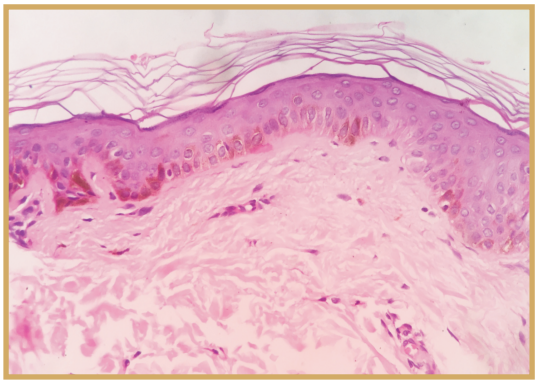

Fig. 4 : Skip areas of retained melanin with mild vacuolar changes in basal layer of epidermis seen in depigmented macules of DUH.

over the face, neck, trunk, bilateral hands and legs, sparing the axilla, palms, soles, genitals and mucous membrane. Biopsy of hypopigmented macules showed skip areas of retained melanin with mild vacuolar changes in basal layer of epidermis and dermis showing perivascular lymphocytic infiltrate and fibrocollagenous tissue (Fig. 4).

Additionally, he developed a few hypopigmented hypoaesthetic lesions on back and buttocks 6 months ago which was diagnosed as Leprosy with histopathological findings suggestive of Hansen's disease, however, slit skin smear for acid fast bacilli (AFB) was negative. Patient is currently on multibacillary multidrug therapy (MB-MDT) consisting of Rifampicin, Dapsone and Clofazimine for the same.

His systemic examination was normal. Haematological and biochemical laboratory tests were within normal limits. Ophthalmological and auditory examinations were also normal. The diagnosis of Hansen's disease with Dyschromatosis universalis hereditaria was made based on the clinical presentation and histological diagnosis.

Discussion

Dyschromatosis universalis hereditaria (DUH) is a rare disease with autosomal dominant (Wang et al 2005) and sometimes recessive inheritance (Bukhari et al 2006). It was first described in 1929 by Toyamo and then by Ichikawa and Hiraga in 1933 (Damle et al 2008). It is characterised by the presence of both hyper and hypopigmented, small, irregular macules uniformly distributed over the entire body, the pigmented macules vary in size and depth of color. The trunk and extremities are the dominant sites (Wang et al 2005). The face is involved rarely and the palms, soles and mucous membranes are usually spared. Most of the patients present within the first few years of life (upto 80% present before 6 yrs of age) (AlHawsawi et al 2002). Lesions of dyschromatosis universalis hereditarian need to be differentiated from xeroderma pigmentosum since both the disorders clinically show similar lesions while the latter has predominant lesions in photoexposed areas with atrophy and telangiectasia. Reticulate-acropigmentation of Dohi also shows similar lesions but are localised to dorsal aspects of extremities and rarely on face. The present case may have been misdiagnosed as leprosy and treatment for leprosy started. Both his earlier biopsy and skin smears were negative for AFB. Co-existence of both the diseases has not been reported, but this could be the first such case.

The pathogenesis of DUH has been postulated to be related to the interference with the neural-melanocytic interaction in early embryonic life in genetically susceptible individuals (AlHawsawi et al 2002). A defect in melanosome production and distribution in the epidermal melanin units with no significant alteration in the number of the melanocytes has also been suggested

(Al Hawsawi et al 2002). Extra cutaneous abnormalities reported in isolated cases of DUH include short stature and high-frequency deafness; abnormalities in erythrocytes, platelets and tryptophan metabolism; bilateral glaucoma and unilateral cataract and seizures. Generally it does not progress or worsen with age, once well established. These patients often suffer from depression because of cosmetic disfigurement so simultaneous counselling and psychiatric consultation can help them improve quality of life (AlHawsawi et al 2002).

To summarise, Blepharochalasis is a rare complication of leprosy which may be due to recurrent localised episodes of inflammation and reactions. Mycetoma is a reported association due to its similar demographic profile and anaesthetic foot of leprosy which is prone to trauma; DUH may not be directly associated with it and to the best of our knowledge DUH and Blepharochalasis with Hansen's disease are being reported for 1st time.

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