

## Case Report

### A Case Report on Recurrent Lepra Reaction-Type 2

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#### Abstract :

Lepra Reactions (LR) comprises immunologically mediated inflammatory states that cause considerable morbidity. Type-2 LR occurs exclusively in patients with the lepromatous end of the leprosy spectrum (BL-LL). In 90% of the cases it follows during or after completion of chemotherapy, generally within 2 years. Here we report a case of type-2 LR. He is a twenty years old fisherman with history of hypopigmented skin lesion 3 years back and was treated as a case of lepromatous Leprosy (LL). After six months of completion of treatment he developed type-2 LR presenting with recurrent bouts of fever, polyarthritis, erythema nodosum, orchitis, mononeuritis multiplex and was treated in Faridpur Medical College Hospital (FMCH) on three occasions within last one and half years.

**Key words:** Leprosy, Erythema Nodosum, Lepra Reaction.

#### Introduction :

Leprosy, Hansen's disease (HD) is a chronic granulomatous disease caused by the bacteria, *Mycobacterium leprae* named after physician Gerhard Armauer Hansen<sup>1</sup>. Its clinical manifestation are largely confined to skin, peripheral nervous system, mucosa of the upper respiratory tract, eyes and testes<sup>2</sup>. Leprosy has affected humanity for over 4000 years and was well-recognized in the civilizations of ancient China, Egypt, and India<sup>3</sup>. In 2005, the World Health Organization (WHO) estimated that 1.5 million people were permanently disabled because of leprosy at that time<sup>4</sup>. Though the incidence of this disabling disease is decreasing throughout the world, still it is not uncommon in Bangladesh. About 70% of world's leprosy patient lives in India, with the disease endemic in Brazil, Indonesia, Mozambique, Madagascar, Tanzania and Nepal<sup>5,6</sup>.

The mechanism of transmission of leprosy is due to prolong close contact and usually by nasal droplet<sup>3</sup>. The clinical manifestations are determined by the degree of

patients cell mediated immunity (CMI) towards *M leprae*. High level of CMI with elimination of leprosy bacilli, produce tuberculoid leprosy, whereas absent CMI results in lepromatous leprosy<sup>6</sup>. Incubation period is 2-5 years for tuberculoid cases and 8-12 years for lepromatous cases<sup>6</sup>. Complications of leprosy are due to nerve damage, immunologic reactions and bacillary infiltration<sup>6</sup>.

There are two types of LR - (1) Type 1 Reaction or Reversal Reaction (2) Type 2, Reaction or Erythema Nodosum Leprosum (ENL)<sup>7</sup>. Patients having high load of leprosy bacilli as in Multi- bacillary/ infiltrative type of leprosy get type 2 reaction. When large numbers of leprosy bacilli are killed followed by release of their antigens, these antigens from the dead bacilli provoke an arthus type allergic reaction (Coombs and Gell type III hypersensitivity) producing antigen antibody immune complex reaction in the presence of complement system. Immune complexes are precipitated in the tissues (skin, eyes, joints, lymph nodes, kidneys, liver, spleen, bone marrow, endothelium and testes) as well as in the circulation<sup>7</sup>. It may occur in the early stages of treatment and even after completion of the treatment with multidrug therapy (MDT), because body takes a long time to clear the dead bacilli within the macrophages. Histologically ENL demonstrate a leukocytoclastic vasculitis<sup>8</sup>. Type 2 reaction commonly occurs within first two years after the start of leprosy treatment<sup>2</sup>. Therapy for type 2 reactions may include analgesics, such as acetylsalicylic acid or paracetamol and corticosteroids, such as oral prednisolone. Who do not respond to corticosteroids or in whom corticosteroids are contraindicated, clofazimine at high doses or thalidomide may be used under close medical supervision<sup>9</sup>.

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### Case Report :

Mr Mahiuddin a 20 years old fisherman was admitted for third time in FMCH with the features of recurrent bouts of erythema nodosum, erythematous skin lesion, loss of function of multiple peripheral nerves and polyarthritis involving both large and small joints for one and half years. He had a history of symmetrical hypopigmented, anaesthetic skin patch 3 years back without any feature of nerve lesion. He also had history of contact with leprosy patient. At that time he was diagnosed as a case of multibacillary leprosy and he took supervised treatment for 1 year with MDT. With the treatment his majority of skin lesions disappeared but he developed generalized reddish brown hyperpigmentation due to clofazimine. After 6 month of completion of treatment he developed polyarthritis associated with marked morning stiffness involving both large and small joints of limbs involving proximal interphalangeal, metacarpophalangeal, wrist, elbow, ankle and metatarsophalangeal joints bilaterally symmetrically. He also developed multiple erythema nodosum in different parts of the body especially lower limbs as well as some hypopigmented or erythematous macules and multiple leproma. He had associated paraesthesia of distal limbs, weakness and wasting of small muscle of the hand specially hypothenar muscle. He had thickening of multiple nerves (bilateral supraorbital, posterior auricular, anterior cervical, radial, ulnar, median, radial cutaneous, common peroneal, sural nerves). Among them some are also beaded and tender. Patient also developed orchitis, both testes are tender and left testis is soft and atrophic without gynaecomastia and loss of pubic hair. There were no features of uveitis or urethritis. It was accompanied by high grade remittent fever not associated with chills and rigor. Complete blood count revealed hemoglobin of 11 g/dl, total leukocyte count of 8,000/dl with neutrophil count of 72%. Erythrocyte sedimentation rate was 100 mm in the first hour. Renal function tests, urinalysis, chest X-ray, blood sugar, antinuclear antibody, rheumatoid factor, anti HCV antibody studies were within normal limit.

At that time he was treated with tab prednisolone 60 mg daily and pain, nodule, neurologic lesion and fever subsided. Patient presented further two times with same clinical features and was treated similarly with prednisolone. He developed cushingoid facies due to repeated use of prednisolone.



**Figure-1:** Comparison of facies between 1st and 3rd admission (cushingoid facies).



**Figure-2:** Raddish brown complexion of the face and backish infiltration of the ala of the nose.



**Figure-3:** Erythema nodosum in hand.



**Figure-5:** Leproma over back of the ear.



**Figure-4:** Residual bluish black hyperpigmentation of erythema nodosum in both legs.



**Figure-6:** Wasting of the small muscles of the hand



**Figure-8:** Leproma over chest.



**Figure-7:** Arthritis of the hand joints



**Figure-9:** Hypopigmented patch in back.

#### **Discussion :**

Leprosy mainly occur in poor socioeconomic background and the time of peak onset is second and third decade. Lepromatous form of leprosy is twice as common in male as in female<sup>2</sup>. Our patient is a fisherman and had a history of contact with leprosy who used to work with him.

Indian studies found prevalence of LR is almost 50% in those with LL and 9% in BL cases and the mean time to presentation with Erythema Nodosum Leprosum was 3.7 months after starting multi-drug therapy (MDT)<sup>10</sup>. Most patients develop during the treatment with MDT. There is a report from Bangladesh in which patient develop LR 16 years after completion of MDT<sup>11</sup>. In our patient LR develop after 6 month of completion of treatment.

Type-2 reaction may be the first presenting sign of the disease and usually last for few weeks to several months. In the beginning general symptoms like fever, headache and body ache appear before or along with the characteristic nodules that appear on the skin<sup>8</sup>. Type 2 reactions exhibit the typical signs of erythema nodosum and variable sized plaques appear in crops. Nodules blanch on pressure, usually multiple and tend to be distributed bilaterally and symmetrically. They appear preferentially on cooler parts of the skin (found on face and outer surface of limbs and less frequently on the trunk). They usually spare the warmer parts of the body like hairy scalp, axilla, groin and perineum<sup>8</sup>. In our patient we also found the same feature.

ENL differ clinically from other conditions like tuberculosis, streptococcal and viral infections and sarcoidosis, by the fact that the lesions in other cases persist for longer duration (other lesions last up to 7

days) and requires longer therapy, where as ENL lesions of lepra reactions do not last longer than 2 or 3 days<sup>8</sup>. In our patient each crop of EN persists on an average 2 days. In LL there may other form of skin lesion, like diffuse infiltration of face, madarosis, waxy shiny appearance of skin (varnished appearance). There may be nodular leproma and plaque like skin lesion<sup>8</sup>. Our patient had infiltration in the nose and multiple leproma in different parts of the body.

Arthritis with type I and II lepra reaction is typically acute onset, symmetric or asymmetric, oligo or polyarthritis and may resemble rheumatoid arthritis (RA) or seronegative spondyloarthropathy<sup>7</sup>. In our case, the symmetric polyarthritis could be misdiagnosed as RA and clinical consideration of ENL was difficult initially in the absence of cutaneous manifestations.

Nerves may also get affected in type 2 reactions. It may be thickened tender and may be beaded. The most commonly affected nerve is ulnar nerve at the elbow resulting in clawing of the hands<sup>2</sup>. In our case almost all nerves are thickened and tender and he had the feature of mononeuritis multiplex. Ocular tissue may get affected in type 2 reactions. It may lead to the development of iritis or iridocyclitis and impairment of vision<sup>2</sup>. Our patient does not have any ocular manifestations.

Often there is non pitting swelling of hands and feet. LR may lead to orchitis leading to testicular dysfunction and infertility<sup>2</sup>. In our patient there is pitting oedema of the legs and focal bluish black hyperpigmentation which developed after healing of EN. Though he had feature of orchitis he did not have any feature of hypogonadism. Hitherto we report a severe and steroid dependent case of ENL, occurred six months after successful treatment of LL, but he also had an unusual initial presentation of arthritis.

Type-2 lepra reaction should be treated with tab prednisolone 40-80 mg daily, reducing over 1-6 months<sup>5</sup>. Clofazimine may be useful for reducing or withdrawing corticosteroids from patients who have become dependent on corticosteroids. If despite two course of prednisolone the lesion persist or recur then tab thalidomide may be prescribed<sup>2</sup>. Even with the treatment of adequate dose of prednisolone the patient continued to develop lepra reaction. So we prescribed thalidomide 300 mg daily and hope he will do well with the treatment.

**Conclusion :**

Leprosy is a curable disease, but it is one of the important causes of disability in human being. The main cause of disability is the disease itself as well as LR. About 50% of the group of lepromatous spectrum develops type-2 LR<sup>2</sup>. So, early detection of the cases and proper treatment of this disease as well as LR should be the main aim of our treatment to get rid of this disabling disease.

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