

## Leprosy of the Larynx: A Case Report

Mansoureh Momen Heravi and Alireza Sharif\*

Department of Infectious Disease, School of Medicine, Kashan University of Medical Sciences, Kashan, Iran

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**Abstract-** Leprosy is an ancient deforming disease caused by *Mycobacterium leprae*, which is still poorly understood and often feared by the general public and even by some in the health care professions. Fortunately, the outlook for patients has dramatically improved over the last three decades with the introduction of multi-drug treatment and management strategies that have somewhat diminished the stigma of this diagnosis. We report a rare case of leprosy of larynx. A 45 year old man presented with complaints cough, dyspnea and hoarseness since many years ago. Because of demonstration of acid fast bacilli in smear of his sputum, the diagnosis of tuberculosis was made and anti tuberculosis treatment was initiated. But he developed fever and his symptoms exacerbated. In examination there was a tender erythematous nodule on right supraclavicular region, loss of eyebrows and lashes and disseminated hyper and hypo pigmented cutaneous lesions on abdomen, thorax, back with normal chest x ray. Laryngoscopy to rule out laryngeal tuberculosis was done and granulomatous lesion was seen. Laryngeal and skin biopsy was performed which numerous acid fast bacilli, macrophages and foamy cells suggestive of lepromatous leprosy were demonstrated in both specimens. Treatment was started on multi bacillary regime of WHO multidrug therapy. In conclusion, this report highlights the importance of systemic involvement in lepromatous leprosy especially when the initial presentation is laryngitis or respiratory symptoms. Laryngeal leprosy may mistaken with tuberculosis laryngitis due to respiratory problems and existence of acid fast bacilli in respiratory secretions.

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**Key words:** Leprosy; laryngitis; larynx; *Mycobacterium leprae*

### Introduction

Leprosy is an ancient deforming disease caused by *Mycobacterium leprae*, which is still poorly understood and often feared by the general public and even by some in the health care professions. Fortunately, the outlook for patients has dramatically improved over the last three decades with the introduction of multi-drug treatment and management strategies that have somewhat diminished the stigma of this diagnosis (1,2). An estimated 1.34 million cases of leprosy occur worldwide (3). The prevalence of leprosy varies markedly from country to country. However, the overwhelming majority of cases are found in developing countries, and 92 percent of the cases are detected in just 11 countries led by India and Brazil (4). Nonetheless, with increasing international travel, cases of leprosy may present anywhere (5). *Mycobacterium leprae* is a unique organism with capacity to invade not only Schwann cells but also other parenchymal tissues such as testis, lymph node, larynx, liver, bone and muscle (6). There are no recent reports on la-

ryngeal involvement in leprosy, and younger physicians might not be aware of the possible seriousness of the pathology of the larynx in leprosy patients. Several older publications call attention to the pathology of the larynx in leprosy, and all textbooks on leprosy consulted mention that leprosy can involve the larynx (7). Though the stage is all set for the final push of leprosy from the world, our knowledge and concepts of some aspects of this stigmatizing disease still need refinement (8). We report a case of leprosy of larynx with initial presentation of respiratory complaints suggestive tuberculosis.

### Case Report

A 50 year old man was presented with hoarseness, dyspnea, dysphagia and cough which had started many years ago but the severity of them had increased in recent months. Smear of sputum was revealed acid fast bacilli and anti-tuberculosis treatment was initiated, but 2 weeks later he developed fever and dyspnea was exacerbated so he was admitted with complaints hoarse-

\*Corresponding Author: Alireza Sharif

Department of Infectious Disease, School of Medicine, Kashan University of Medical Sciences, Kashan, Iran  
Tel: +98 361 5550210, 913 1613373, Fax: +98 361 5516090, E-mail: sharif\_a@kaums.ac.ir

ness, sweating, weight loss, anorexia, non productive cough. On physical examination there was deformity of anatomical structure of oropharynx, a tender erythematous nodule on right supraclavicular region, loss of eyebrows and lashes and disseminated hyper and hypopigmented cutaneous lesions on abdomen, thorax, back. His hands were deformed as claw hand. There was tenar and interosseous atrophy, flexion contracture of proximal interphalangeal joint (PIP), disability for abduction and adduction of fingers. There was no hypoesthesia in skin lesions. Evaluation of skin lesions and palpation of peripheral nerves for enlargement and/or tenderness no revealed abnormal findings. There was no sensory loss. Other systemic examinations were within normal limits. There was no history of contact with leprosy patients. The vital signs at admission was: T:37.3°C BP:110/70 mm/Hg, PR:80 /min, RR:20/min.

After admission corticosteroid was added to anti tuberculosis treatment

Laboratory findings, including a complete blood count, erythrocyte sedimentation rate (ESR) were normal. C reactive protein (CRP) was positive(3+). There was 3 times increase in SGOT and SGPT. Urine analysis showed mild pyuria, hematuria and proteinuria. Purified protein derivative 5 IU intradermal injection (PPD test) disclosed a 16 mm skin induration.

The chest X-ray no revealed abnormal findings. With suspicion of endobronchial tuberculosis ENT consultation and subsequently laryngoscopy was performed. Glottic and supraglottic swelling, erosion, blunting and granulomatous tissue were seen on laryngoscopy. Barium swallow was done due to dysphasia which started from one year ago and it revealed a diverticula's in pharynx. Regarding to skin lesions and loss of eyebrows and lashes, dermatology consultation and skin biopsy were performed. Skin biopsy showed numerous granulomas containing lymphocytes, histiocytes, fibroblasts and a few multinucleated giant cells in dermis. With suspicion of leprosy, HE & Ziehl-Neelsen staining of skin specimen was performed which exhibited collection of macrophages, a few multinucleated giant cells, sparse lymphocytes and some foamy cells in dermis and numerous acid fast bacilli in macrophages. Specimens of skin and larynx were sent to research center of leprosy for final confirmation. Numerous acid fast bacilli, macrophages and foamy cells suggestive of lepromatous leprosy were demonstrated in both specimens. Also at this time the culture of sputum for mycobacterium tuberculosis was reported negative and CT scan of chest was normal. So diagnosis of lepromatous leprosy was confirmed. So laryngeal involvement was due to lepro-

sy. Antituberculosis treatment was stopped and started on multibacillary regime of WHO multidrug therapy with Dapsone (100 mg daily), Rifampin (600 mg once-monthly) and clofazimine (50 mg daily and 300 mg once monthly). Ophthalmologic and neurologic consultation was performed which showed no abnormal findings. On a later review he was found to be having secondary infertility. He had three children but he couldn't have further children from 15 years ago. Ultrasonography of testis showed atrophy of both testis. During hospitalization after dose reduction of corticosteroid he developed some nodular erythematous lesions in forearm and swelling of wrist which disappeared with increasing dose of corticosteroid.

## Discussion

*Mycobacterium leprae* infection usually presents with cutaneous and neurological involvement. However, initial clinical manifestation in muscle, lymph node, larynx, liver, spleen, bone marrow, bone and testis (proven by biopsy) has been infrequently reported (6). Leprosy is longstanding disease so laryngeal lesions developed gradually and may be asymptomatic in some cases while others have been found to have variety of clinical manifestation in form of congestion, infiltration, nodulation, thickening and fibrosis. Anti-leprosy chemotherapy is equally effective on these lesions. A screening survey of laryngeal pathology in such cases should be done periodically (9). Most lepromatous patients have leprosy in the larynx, especially the epiglottis (7). It is emphasized that in areas where there is significant incidence of leprosy, laryngeal involvement may be expected even in the absence of local clinical manifestations and be got confirmed by histological examination and demonstration of acid-fast bacilli. Further, histological appearance may not be very characteristic at the outset (10). With earlier diagnosis and MDT, it has become rare to see a complete airway obstruction, but the possibility exists and all leprosy workers should be aware of this life-threatening event. However, with timely diagnosis and proper treatment, most patients will not reach the stage of a complete life threatening obstruction of the upper airway. With reductions in treatment duration, (single dose or 6 months) we might again see more serious laryngitis in leprosy patients. Early diagnosis of leprosy is still not universal. In highly endemic areas like Brazil and Paraguay, advanced cases of lepromatous leprosy are still seen (7). Fleury *et al.* reported eight patients with larynx involvement in leprosy. Seven were classified as lepromatous and one was borderline leprosy in

reversal reaction. One patient required an emergency tracheostomy and one had an almost complete upper airway obstruction (11). Malik *et al.* reported five instances of lepromatous leprosy involving lesions of the larynx (12). Yunus reported two cases of leprosy, one of which was admitted as a case of acute tonsillitis and was referred only later to the ENT surgeons with hoarseness after a diagnosis had been made by biopsy of the lymph node; the other was referred with a possible facial paralysis (13). In this patient claw hand indicated involvement of the ulnar and median nerve. Nerve damage occurs later in the disease toward the lepromatous end of the spectrum, where sensory loss generally involves the distal extremities first. Nerves commonly involved include the ulnar and median (claw hand), the common peroneal (foot drop), the posterior tibial (claw toes and plantar sensitivity), facial, radial cutaneous, and great auricular (5).

After dose reduction of corticosteroid this patient developed some nodular erythematous lesions which disappeared with increasing dose of corticosteroid. Nodular skin lesions may be suggestive of erythema nodosum that usually develops after initiating treatment of leprosy. Erythema nodosum leprosum (ENL or Type 2 reaction) is associated with BL and LL disease. This is thought to be an immune complex disorder and is more common during treatment but may also occur before or after the completion of therapy (5).

Hyper pigmentation was developed 3 months later after onset of treatment which was due to major side effect of clofazimine. Accumulation of drug in the cells of the leprosy granuloma causes pigmentation which clears after discontinuing treatment.

The incubation period for leprosy is uniquely long among bacterial diseases, a minimum of 2 to 3 years, averaging 5 to 7 years, and can be as long as 40 years or more (14), this patient had migrated to Iran from 20 years ago and he had no travel to Afghanistan during this time and also his symptoms had been started from many years ago, so probably he had acquired mycobacterium leprae from Afghanistan. In conclusion, this report highlights the importance of systemic involvement in lepromatous leprosy especially when the initial pres-

entation is laryngitis or respiratory symptoms. Laryngeal leprosy may mistake with tuberculosis laryngitis due to respiratory problems and existence of acid fast bacilli in respiratory secretions.

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