

Leprosy of the larynx

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Abstract

The result of a study conducted on 30 patients of lepromatous leprosy regarding laryngeal involvement is presented. The incidence of leprosy laryngitis is 36.6 per cent in this part of India in lepromatous leprosy patients. Leprosy is long standing 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.

Introduction

The introduction of modern chemotherapeutic agents has reduced the incidence of laryngeal leprosy in the most countries while it is still seen commonly in some of the developing countries such as India, Pakistan and Bangladesh.

WHO estimates that there are over 12 million cases of leprosy in the world, a third of whom are in India (Antia, 1988). Bacillary positive leprosy of long standing may produce a variety of clinical manifestations in the upper respiratory tract.

Mitsuda and Ogawa (1937) published a series of 150 necropsies out of which laryngeal stricture was found to be cause of death in 1.3 per cent of the cases. Yoshie (1956) observed a laryngeal lesion in 64.7 per cent of lepromatous leprosy (LL) cases. Pinkerton (1938) reported that approximately 40 per cent of moderately advanced cases present leprosy lesions of some form in the larynx. Barton (1974) while working in India, reported involvement of the larynx in 31 per cent of 29 patients with early lepromatous leprosy. Malik *et al.* (1975) described laryngeal biopsy findings in five cases of lepromatous leprosy. Gupta J. C. *et al.* (1980) reported histological findings from larynx in 30 lepromatous leprosy cases. Gupta G. P. *et al.* (1984) reported 10 cases of laryngeal leprosy in study of 44 cases of lepromatous leprosy. Since then a few more scattered cases of leprosy larynx have been reported in the literature (Popli *et al.*, 1990).

Material and method

Thirty patients with bacillary positive untreated lepromatous leprosy were taken for the present study. The ages ranged from 15 to 55 years; there were 19 males and 11 females. In each case a complete ENT examination was performed. Laryngeal involvement was assessed by indirect laryngoscopy. Direct laryngoscopy was performed in patients where the laryngeal view was obscured

by the shape and position of the epiglottis. Laryngeal findings were duly recorded, and punch biopsies from abnormal looking areas of the larynx were taken by direct laryngoscopy under local anaesthesia.

Observations

Laryngeal leprosy pathology was found in 11 patients (36.6 per cent) out of 30 cases of lepromatous leprosy.

The commonest laryngeal symptom was hoarseness (10 cases). It is varied from slight huskiness to almost complete aphonia. The patients also complained of cough (12 cases), throat pain (two cases), haemoptysis (two cases); one patient had breathlessness. All these laryngeal symptoms had lasted from one to six years.

The epiglottis was the most frequent site of involvement in the larynx and was found in all 11 cases of lepromatous leprosy. The next most common sites involved were the vocal cord (seven cases) and aryepiglottic fold (four cases). Each region of the larynx was found to have different types of lesions as shown in Table I. The false cords and pyriform fossae were not involved in any case in the present study. Mobility of the vocal cords was not impaired.

A laryngeal smear was taken in all laryngeal leprosy patients and was positive in three cases.

Almost all cases of leprosy laryngitis had advanced disease in the nasal cavity and smears were found positive in all cases for lepra bacilli.

A biopsy was performed in five cases with gross positive findings in the larynx which revealed the typical picture of lepromatous leprosy with positive AFB.

Discussion

There has been a tremendous decline in laryngeal leprosy among lepromatous leprosy patients with the discovery and use of new multidrug therapy and partly due to improvement in standard of living and socioeconomic

TABLE I
SHOWING THE DIFFERENT TYPES OF LESIONS AT DIFFERENT SITES IN
THE LARYNX

Anatomical sites	No. of cases
I Appearance of epiglottis	
Thickened	5
Nodular	2
Ulcerations	2
Destroyed and deformed	2
II Appearance of vocal cord	
Congested	3
Ulceration	2
Nodule	2
III Appearance of aryepiglottic fold	
Swollen	3
Oedematous associated under	1

status, especially in developed countries (Popli *et al.*, 1990). The survey carried out by Gupta G.P. *et al.* (1984), and Kumar *et al.* (1988) in India revealed that about 40 to 50 per cent of leprosy patients still have involvement of the laryngeal mucosa at the time of presentation. In the present series of 30 patients with lepromatous leprosy, laryngeal pathology was detected in 11 patients (37.6 per cent). These findings are similar to the incidence as reported by Pinkerton (1938), and Yoshie (1956) who studied cases about four or five decades earlier in developed countries.

Direct spread of the mucosal lesion from the nasal cavity to the larynx is the possible route; occasionally lymphatic spread has been described. In this study all the laryngeal leprosy patients had nasal lesions and were highly AFB positive, thus favouring the direct spread of leprosy lesion to the larynx.

There is no controversy regarding the fact that epiglottis is almost always involved in leprosy of the larynx. After passing the posterior channel, the inspired air flows over the free margin of the epiglottis, which protrudes into the air stream in a similar way to the inferior turbinate in the nose. The temperature of inspired air at this point is approximately 2°C cooler than the normal body temperature (Negus, 1958). It is generally accepted that *M. leprae* favour cooler parts of the body. Other sites of involvement in leprosy are the aryepiglottic fold, arytenoids, vocal cords, and ventricular bands.

The symptomatology of laryngeal leprosy varies from case to case and is not pathognomonic. The characteristic early symptom is the 'leprosy huskiness' a peculiar vocal quality that strikes the experienced observer as suggestive. The patients also present with a dry unproductive cough and sometimes a tickling sensation in throat. Dyspnoea is rare and less likely to require a tracheostomy in the present era.

Bacillary positive leprosy of long-standing may produce a variety of clinical manifestation in the larynx. The lesions generally develop gradually and are asymptomatic.

Various changes are seen in the epiglottis. Nodule formation occurs in the form of a granuloma. Small discrete nodules, pale in colour are initially seen on the free border but also extend on to the anterior and posterior surface. As the nodules enlarged and increase in number, they give the appearance of more generalized irregular infiltration.

Thickening of the fibrotic type of lesion is due to diffuse infiltration, which may have a marked inflammatory ele-

ment and then the epiglottis may appear thickened by two or three times. Punched out ulcers are formed over the mucosa of the arytenoids, corniculate cartilages and in the region of the false cords. The vocal cords are also affected but in the late stage various forms of pathology can occur. The nodules in the vocal cords are probably more common than is realized, but they are rarely seen because they occur late when the distortion and infiltration of the structure make them difficult to see. Such problems were encountered in the present study; as a result direct laryngoscopy was performed in these cases subsequently revealing the vocal cord pathology.

Congestion, swelling, infiltration, nodulation, ulceration, atrophy and scar formation with contractures and distortions are the successive steps in the disease in the larynx as in all the other diseased parts of the mucous membrane. This does not necessarily imply that apparently normal mucous membrane is free from lepromatous changes when examined histologically. Desikan and Job (1968) for example report a case in which an apparently normal vocal cord showed histological evidence, of a sub-mucosal granuloma.

The recurrent laryngeal nerve escapes in most cases (except during an acute reaction), while the superior laryngeal nerve is involved more often by deep infiltration of its branches.

The histology of leprosy in the larynx is similar whichever part is involved *e.g.* epiglottis, arytenoid *etc.* The lining epithelium was stratified squamous epithelium which at places was thinned out and ulcerated. The sub-epithelial clear zone was not apparent in the most of the tissues. A very narrow band of fibrous tissue with some fibroblasts separates the epithelium from the marked collection of histiocytes in the sub-epithelial region. This band of connective tissue with its rich network of capillaries supports the histiocytes. Some histiocytes exhibited vacuolization.

Lepra staining showed a parallel arrangement of lepra bacilli in the histiocytes (lepra cells); free bacilli in compact globular masses were also seen. The absence of a clear submucosal zone and the presence of acid fast bacilli in the epithelium allows shedding of a large number of *M. leprae* from the mucosa even when this is intact.

Systemic anti-leprosy chemotherapy rapidly modifies the laryngeal pathology by decreasing the bacterial count. Davey and Rees (1974) reported zero or near zero bacterial counts in nasal discharge after two months treatment with Dapsone.

Several drugs closely related to the sulphonamides (*e.g.* dapsone, danino-diphenyl sulphone, DDS) have been effectively used in the long-term treatment of leprosy. Both lepromatous and tubercular leprosy lesions can be suppressed by treatment extending over several years. It appears that 5 to 30 per cent of *M. leprae* organisms are resistant to dapsone. Consequently initial combined treatment with Rifampicin is advocated. Waters and Rees (1962) in a pilot study of Riminophenazine derivative B. 663 in low dosage (100 mg twice weekly) in eight cases of lepromatous leprosy found the rate of fall of morphologic index was similar to that obtained with sulphone therapy or with B.663 300 mg daily. The multidrug therapy (MDT) recommended for paucibacillary leprosy by the WHO (1982) consists of dapsone 100 mg daily for six months and Rifampicin 600 mg once a month for six months. The modified regimen for multi bacillary cases

comprises an initial intensive phase of 14 days during which supervised doses of rifampicin 600 mg, clofazimine 100 mg and dapsone 100 mg daily is followed by a continuation phase of treatment, for at least two years, with 600 mg rifampicin once a month and daily dosage of 100 mg dapsone and 50 mg of clofazimine. The cases in the present series were treated by this regime of multidrug therapy recommended by WHO.

Once the disease process is checked, the healing starts with variable degrees of fibrosis. The recent multidrug regime has revolutionized the leprosy lesions of the mucosa giving a speedy recovery. Treatment is continued until all signs of activity have been absent for three years. Thus it seems necessary that in places where there is a significant incidence of leprosy, laryngeal examination should be done as a screening procedure. Evidence of laryngeal pathology in an endemic area should always raise the suspicion of leprosy.

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