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Leprosy in ENT

by

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In England and Wales 1,172 cases of leprosy have been notified since 1951, of which 815 have subsequently been removed from the register (Public Health Laboration Services, 1983). At the end of 1982, the number of patients on the central register of the Department of Health and Social Security was 357, whereas the numbers notified for the years 1980, 1981 and 1982 were 28, 23 and 27 respectively. In Scotland only one patient has been notified in the last three years and the number of patients under surveillance is 10.

All these patients had contracted the disease abroad. Most of the patients were Asians, either from the Indian subcontinent or Africans or Asian origin.

Usually it is the neurologist, dermatologist or physician who will diagnose leprosy. However, the patient may present to the ENT surgeon with a variety of complaints and leprosy should still be kept in mind for granulomatous lesions of the upper respiratory tract in immigrant populations of the United Kingdom.

Two cases of leprosy are here reported, one of which was admitted initially as a case of acute tonsillitis by the general physicians but was finally diagnosed to be suffering from lepromatous leprosy; the other was sent to the ENT department with a provisional diagnosis of facial paralysis.

Case No. 1

S.A., a 30-year-old Yemeni male who had lived in Riyadh for 11 years, was admitted to the medical ward with a history of fever, with night sweats and rash all over the body for the previous 10 days. This had been accompanied by vomiting. He did admit to having had a rash in the lower limbs for the previous 18 months and general ill health for the same period. There was no other relevant history. Examination was normal except for a temperature of 39°C. and a pulse of 100/min. He had cervical, axillary and inguinal lymphadenopathy, the lymph nodes being small, mobile and firm. There was a maculopapular rash on the upper and lower limbs. The liver was enlarged and was 2 cm. below the costal margin. It was not tender and the spleen was just palpable. Examination of the pharynx showed large tonsils, with pus in the left tonsiller crypts. A provisional diagnosis of acute tonsillitis and ? mycosis fungiodes was made and he was started on intramuscular penicillin. FBC showed him to have a haemoglobin of 12.2 g., WCC 18.5 k./c.mm. of which the neutrophils constituted 85 per cent and lymphocytes 11 per cent. ESR was raised at 118 mm./1 hr. U and E and LFT were normal. VDRL and HAA were negative. A blood culture was negative. A throat swab, MSU and sputum did not grow any infecting micro-organisms. A blood slide for the parasite did not show any evidence of malarial infection. B_{12} and folate were normal. However, ANA and RA Latex tests were positive. There was no evidence of any recent exposure to toxoplasma with the titres being less than 64. IgG was raised at 2,000/100 ml. and IgA was raised at 568 mg./100 ml. An ultrasound scan of the abdomen was carried out which confirmed an enlarged liver and spleen. As the temperature persisted and the patient did not improve, a diagnosis of infectious mononucleosis was considered but the monospot test was negative. After one week biopsy from the auxillary lymph node was carried out and blood was sent for viral studies, for CMV and Q fever; these proved to be negative. The lymph node biopsy, however, proved to be lepromatous leprosy. A repeat general examination was carried out when a maculopapular rash was found over the palate, tonsil and pharynx and there were palpable nerves all over the body, along with small-muscle wasting in the thenar muscle. He had a right conjunctivitis and an ophthalmological opinion was sought which showed him to have localised nodular congestion which proved to be a lepromatous nodule. He had also lost the outer half of the eyebrow. CNS examination was quite normal and there was no evidence of any loss of sensation.

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A nasal swab and nasal scrapings were taken and acid-fast bacilli were seen.

He was referred for an ENT opinion because of hoarseness, which he had had for about three months. On close questioning he admitted to having had a yellowish nasal discharge from the nose which was blood-stained at times. There was no nasal blockage. He did have occasional sore throats but had never had dysphagia or dyspnoea. Examination showed him to have a deflected nasal septum to the right with dry crusts on both sides of the nasal septum and on the inferior turbinates. The uvula was nodular and ulcerated and there was a macular rash on the soft palate (Fig. 1). Indirect laryngoscopy showed him to have a thickened left vocal cord only. X-rays of the sinuses were quite clear. Microlaryngoscopy was carried out and the thickened left vocal cord (Fig. 2) was stripped. Biopsies were also taken from the uvula, soft palate, nasal mucosa, turbinates and septum. All these proved histologically to be lepromatous leprosy (Figs 3 and 4). He was started on Dapsone and Rifampicin and when, 12 months later, nasal swabs were taken no AFB's were seen and the patient remains well, with occasional bleeding from the nose and a little crusting in the nose. There had been no definite evidence of atrophic rhinitis or any external nasal deformity.

Case No 2

RAA, a 26-year-old Indian male (Fig. 5) who had come to Saudi Arabia three years earlier, was referred to the ENT clinic with a provisional diagnosis of right facial paresis possibly due to chronic suppurative otitis media. He complained of loss of sensation on the right side of the face and eye for the previous four weeks along with pain in the face, head and eye. This had been gradual in onset and the patient was now in terrible pain. There was no nasal, pharyngeal, aural or ocular symptoms. He did not have any headaches, nausea or vomiting. He had not had any recent viral infection. However, he admitted to having pain in the left index finger for the previous couple of days. There were no other relevant history. He had had a hydrocoele operated upon four years earlier and had had hepatitis nine months earlier.

Examination of the ear, nose and throat was absolutely normal. He had a maculopapular rash on the right side of the face which roughly involved the distribution of the second and third divisions of the trigeminal nerve. Central nervous system examination was normal apart from a decreased right corneal reflex. Taking into account the severe pain, a provisional diagnosis of viral neuritis of the trigeminal nerve, possibly due to Herpes Zoster, was made and the patient had further investigations, including FBC, ESR, U and E, LFT, VDRL and virology studies. He had normal FBC with ESR of 1 mm./hr., normal U and E, liver function tests and had a negative VDRL. Virology studies were not diagnostic and a skull X-ray was normal. He was referred to a neurologist for a second opinion and, when seen by him a week later, the patient had developed a swelling of the left index finger and left big toe and the facial rash had increased to beyond distribution of the trigeminal nerve. He had by this time developed analgesia and anaesthesia of the area. On examination, another large macular rash was found on the left thigh. A dermatologist's opinion was sought, with leprosy in mind, and he found that the patient had thickened posterior auricular nerves as well. A diagnosis of 'tuberculoid leprosy in reaction' was made and this was confirmed by biopsies from the skin of the face and thigh. The swabs did not grow any fungi. The patient was also referred to an ophthalmologist and ocular involvement was excluded. A Lepromin test was performed and was found to be positive at 24 and 48 hours.

He was treated with Rifampicin and Dapsone, and Prednisolone was started two weeks later, as pain was not getting less and the lesions on the left foot had increased in size and new lesions had appeared on the left calf. He gradually improved and, when seen in the clinic about two months after the start of chemotherapy, he had improved a lot and his skin lesions were getting smaller.

Discussion

There are about 15,000,000 people suffering from leprosy in the world at the present moment. Mycobacterium leprae has probably been infecting man for centuries. It is quite possible that the swellings noted in the Ebers Papyrus (Barton, 1974*a*) (1552–1350 BC) (Ebbell, 1937) were lepromatous. It was introduced into the West by the Greek armies returning from India. It is endemic in the tropics and sub-tropics, although cases have also been reported from temperate zones as well. It is not endemic in the UK and it is present sporadically in Saudi Arabia, but cases are seen in immigrants as both countries have a high immigrant population. In Saudi Arabia most of the cases are seen in the south, in the area adjoining the Yemen.

Leprosy is an infectious disease which affects the peripheral nervous system primarily and also the skin, upper respiratory tract mucosa, reticuloendothelial system, eyes, bones and testes (Cochrane and Davey, 1964). It is caused by mycobacterium leprae which has not yet been

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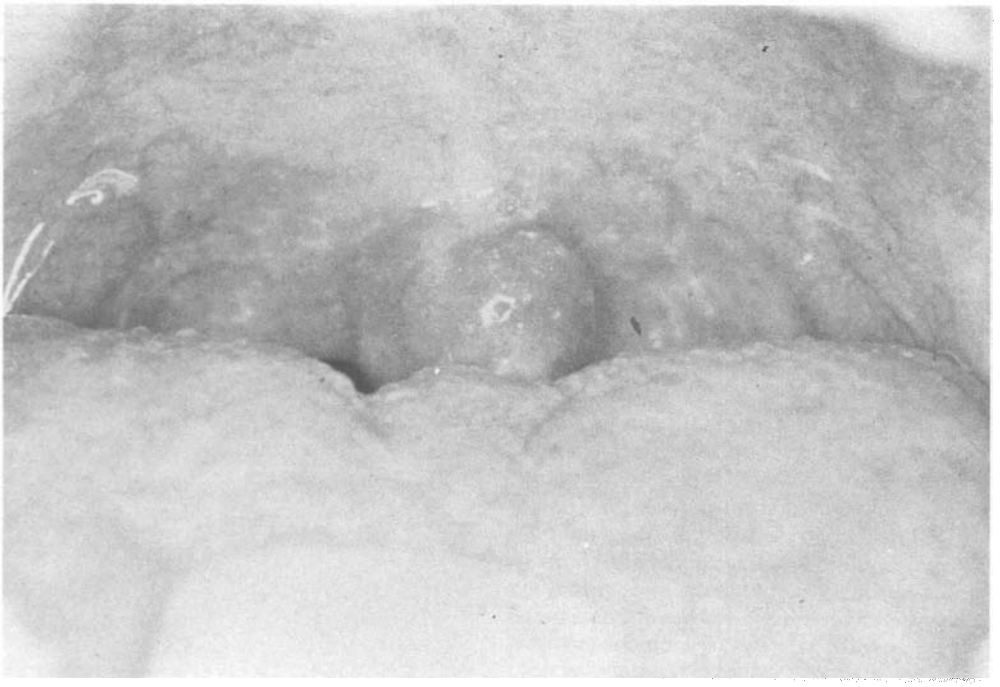


FIG. 1.
Palate showing macular rash

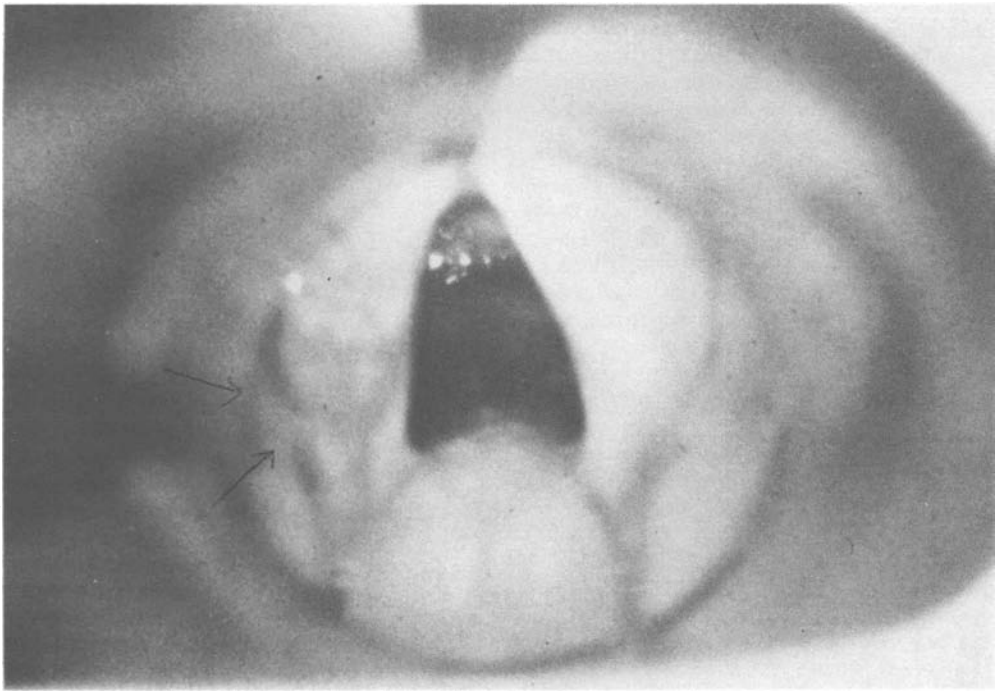


FIG. 2.
Larynx showing thickened left vocal cord

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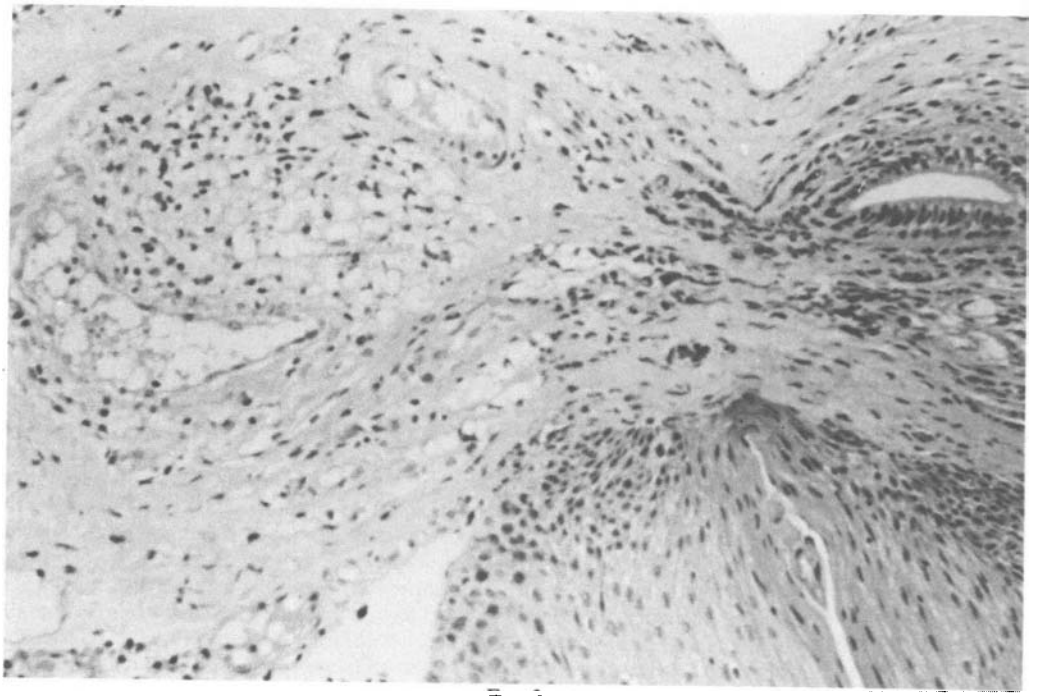


FIG. 3.
Lepromatous leprosy: Laryngeal biopsy showing groups of foamy histiocytes in the subepithelial connective trissue. HE×100

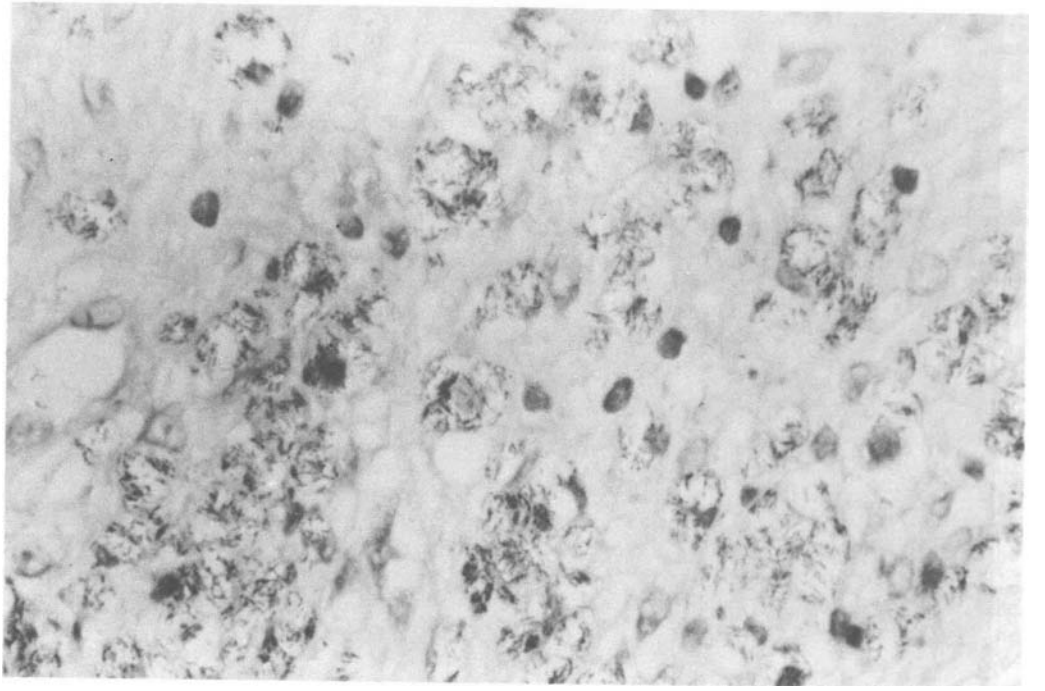


FIG. 4.
Lepromatous leprosy: Foamy histiocytes seen in Figure 3 are loaded with a lepra bacilli. Wade Fite×400

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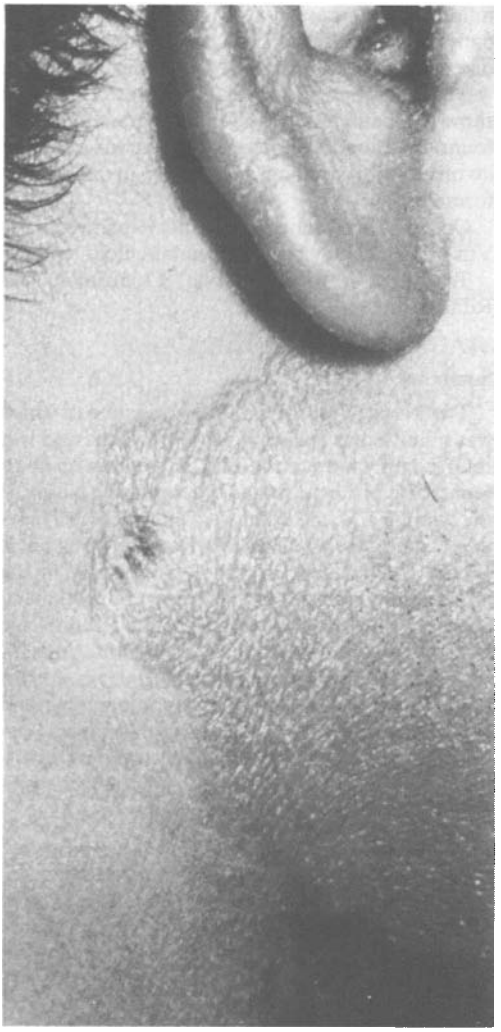


FIG. 5.
Lepromatous rash in the infra-auricular region

cultured but has been shown to produce a leprosy-like disease in mice and armadillos (Storrs, 1974).

Depending on the immune response of the patient, leprosy can be divided into three main forms:
—Lepromatous
—Tuberculoid
—Borderline
Tubercoloid leprosy involves nerves and skin and the patient presents with large anaesthetic macules and plaques, whereas lepromatous leprosy involves not only the skin and nerves but all tissues, only the CNS being spared. It is this type of leprosy which is of interest to the ENT surgeon, although borderline leprosy can also present to him. As 80 per cent of leprosy patients have upper respiratory tract involvement (Berett, 1970) and may involve the ENT surgeon, an attempt is made to review the clinical picture of leprosy from an ENT surgeon's point of view.

Nasal discharge was first described by Goldschmidt in 1891 to be rich in acid-fast bacilli. Sticker, 1897, at the First International Leprosy Congress, proved that mycobacterium leprae found in nasal discharge had actually come from the infected nasal mucosa. This seems to be the route of exit of the bacillus. The route of entry still remains obscure. Barton (1974b) has divided nasal involvement in leprosy into three stages:
(1) *Early Stage*, in which the mucous membrane is dry, pale and yellowish, and granulomas can be found scattered on the normal-looking mucosa. This thickened mucosa has been found to have resulted from submucosal infiltration and thickening. The mucosa does not shrink on application of cocaine and the dryness is due to damage to the parasympathetic nerves. These changes are found even before the skin changes are seen.
(2) *Intermediate Stage*, in which there is mucosal thickening producing nasal blockage. This leads to nose-picking and traumatic ulceration of the anterior septum. This stage is accompanied by nasal secretion which is mucoid, muco-purulent and blood-stained and contains large numbers of AFB.
(3) *Late Stage*, in which drying of the nasal discharge occurs, with the formulation of crusts and secondary invasion with opportunistic micro-organisms. Destruction of the septal cartilage and bone of the inferior turbinate occurs, leading to wide nasal airways which give the typical appearance of atrophic rhinitis. Occasionally adhesions will be seen.

Estimates of nasal involvement vary from 90–97 per cent (Shehata *et al.*, 1974). The mucous membrane is involved early and it is usually the anterior end of the inferior turbinate which is the most common site.

The classical triad of the lepromatous nose, i.e. saddle nose, septal perforation and atrophic rhinitis, is seen only in the late stages. This is the result of destruction and fibrosis and forms a typical picture which is quite different from that of other granulomas, such as syphilis or lupus. In the early stages, however, owing to columellar collapse, the nasal tip may droop and the alae may collapse. There may also be nodules in the nasal skin.

Other symptoms and signs include:
1. *Nasal obstruction:* 47.7 per cent of patients (Shehata *et al.*, 1974) will have nasal obstruction

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due to narrowing of the airway, due either to granuloma formation or to the stenosis.

2. *Nasal discharge:* 46 per cent of patients (Shehata *et al.*, 1974) have thin, mucoid discharge which later changes to purulent, yellowish-green, foul and blood-stained.

3. *Epistaxis* is the presenting symptom in about 86 per cent of the cases.

4. *Nasal deformity* occurs in 47.7 per cent of cases (Shehata *et al.*, 1974).

5. *Rhinolalia Aperta* is found in 6.9 per cent of cases.

6. *Anosmia:* 40 per cent of the patients (Barton, 1974b) will have some anosmia in lepromatous leprosy.

The tongue and tonsils can also be involved in leprosy (Prabhu, 1946) although the most common sites are the soft palate and the free edge of the epiglottis. Cases have been reported in which the granuloma has involved the pyriform fossa and larynx. This occurs usually after nasal involvement and after the diagnosis has already been made. Laryngeal involvement produces hoarseness due to infiltration of the mucous membrane and may even produce stridor. The whole of the larynx may be involved and in the later stages, when fibrosis occurs, there may be laryngeal stenosis producing stridor and eventually leading to death. This occurred in two cases out of 150 autopsies performed by Mitsuda and Ogawa (1937) on patients who had died of leprosy. Malik *et al.* (1975), while following 280 cases of hoarseness, found five patients to be suffering from lepromatous laryngitis due to diffuse infiltration. No nodular granuloma was found by him. However, Gupta *et al.* (1980) did show one patient out of 30 to have granuloma. He found that almost all parts of the larynx could be involved with swelling, thickening or granuloma formation.

Occasionally isolated cranial nerve palsies, of the Vth or VIIth, may be seen in tuberculoid leprosy.

Treatment is with Dapsone, Clofazimine and Rifampicin.

Summary

Two cases of leprosy are reported, 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. This is a rare diagnosis but it should still be kept in mind, particularly in the immigrant population of the United Kingdom and Saudi Arabia and particularly of the Saudis from the southern provinces.

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