Two cases of scleroma in Iran

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SYNOPSIS The distribution of rhinoscleroma in Asia is reviewed and two cases from Iran are added to a region in which the disease seems not to have been previously reported. The pathogenesis of the disease is reviewed.

The clinical appearance of scleroma is usually characteristic and the disease is easy to diagnose (Fig. 1). von Frisch described the causative organism in 1882. Studies with fluorescent and immunofluorescent methods confirmed the exclusive role of *B. rhinoscleromatis* in the specific pathological picture (Hoffman, Correa, and Luian, 1963). Koch's postulates were, however, not completely fulfilled until recently (Hoffman, 1968). Although rabbits and mice could be infected experimentally, the lesions produced were not specific and self curative. Only 40% of a given litter showed abscess formation at the injection site after 15 to 20 days. The lesions disappeared spontaneously after 30 to 90 days. Multiple animal passage increased the virulence of the bacillus only slightly.

Recent experiments demonstrated that continuous and repeated injection of bacilli at the same site produces a granulation tissue indistinguishable from the characteristic lesion in the human nose (Hoffman, 1968). Equally good results can be achieved by a single injection of bacilli together with sterile mucus, which in some way prohibits phagocytosis of the organism and leads to specific pathological alterations.

These findings explain the low infectivity in the human subject and the restriction of the disease to the upper respiratory tract. Infection is favoured by close association with a diseased person for very long periods. *Bacillus rhinoscleromatis* has been isolated from the nasal mucosa of apparently healthy individuals. In all these cases a member of the family or some other close associate was infected. Manifest scleroma followed very frequently. Whole tribal groups and large families suffered infection (Oomen and Kirschner 1938; Aldoin, 1957). The disease affects the rural poor and 'unwashed' (Aldoin, 1957). Risk for both sexes is equal, although due

to socioeconomic factors the incidence from different countries shows moderate variations. Only 5% of all cases occur amongst children: the majority are found in people between 15 and 35 years of age, which is a reflection on the short life expectancy in the countries frequently affected.

Geographical Distribution

The disease occurs under all climatic conditions. It has been described from Siberia, southern Switzerland, to the high Andes. Most cases are from moderate climatic zones and the tropics. Wahi and Misra (1964) describe a scleroma belt on the Eurasian continent, extending from Yugoslavia, Bulgaria, and Rumania, on the northern and southern coast of the Black Sea to the Caucasus and the Central Asiatic Soviet Republics, from there to India and the Indonesian archipelago. No cases had been recorded from Iran and Afghanistan, which was attributed to the high altitude of large parts of these countries (Wahi and Misra, 1964).

The main European focus is Galicia (Poland and Russia). A high incidence of scleroma is reported from Egypt, spreading throughout Africa. Most of the Central and South American cases are from Guatemala (Quevedo, 1949). Australia is practically free of scleroma.

Asian Cases

The number of case reports depends not only on the frequency of the disease, but also on the standard of case recording and general public health statistics. Except for the presence of the disease in a given country no valid conclusions can be drawn from the literature.

Two cases from Turkey and three from Iraq are recorded. Numerous cases were collected in Kazakhstan, Uzbekistan, Turkmenistan, and

Tadjikistan (Barilyak, 1962; Derepa, 1965). Barilyak confirmed the diagnosis with impression smears and nasal scrapings in 94% of 646 cases.

Many case reports have appeared since 1889 from the Indian subcontinent. Wahi and Misra (1964) collected 85 cases from the literature before 1941 and added 30 of their own. Another 50 cases have been reported since.

Common environmental and socioeconomic factors in some tribes and communities of Indonesia produce foci of endemic scleroma. A series of 53 families with numerous cases has been recorded (Oomen, 1938). The disease is also found in the Philippines, Japan, and China.

Case Reports

CASE 1

A 28-year-old woman from Fassa (1,200 m above sea level) had been bitten by a donkey three years before admission. She complained of slowly progressing nasal obstruction and frequent epistaxes following the nasal trauma. On admission both nostrils were blocked by granulation tissue, which extended onto the upper lip. The nose was dilated and flattened in a typical fashion. Biopsy showed that the granulation tissue consisted of lymphocytes, plasma cells with Russell bodies, and characteristically distended histiocytes containing clumps of bacilli (Mikulicz cells). Bacteriological studies confirmed the presence of *Klebsiella rhinoscleromatis*. The patient received 2,000 r superficial radiotherapy and was seen six months later in an essentially unchanged condition.

CASE 2

A 30-year-old woman from Behbahan (altitude 800 m) developed slowly progressive nasal obstruction in the course of a year. Her physician removed a 'polyp' which very soon recurred. On admission

Fig. 1a and b Case 2: note the distended and enlarged turbinate through the dilated nostrils. The nasal canal is completely obstructed.

both nostrils were occluded by a soft mass of granulation tissue with a firm base in the gingivo-buccal sulcus. The base was distended in the midline (Fig. 1). Microscopic examination showed the typical granulation tissue with Mikulicz cells. The patient received a full course of streptomycin after local resection of some of the granulation tissue to permit nasal drainage and easier breathing. The postoperative course was satisfactory.

Discussion

Scleroma is almost exclusively a disease of the respiratory passages, since their mucus supports the growth of the bacillus (Hoffman, 1968). Sixty per cent of all nasal scleromata have eventually laryngeal, 28% nasopharyngeal, 16% pharyngeal, and 9% tracheal and bronchial complications and involvement. The progressively expansive growth of the untreated granulation tissue may produce pressure atrophy and destruction of bone, occasionally

	<i>Kl. Scleromatis</i>	<i>Kl. Ozaenae</i>	<i>Kl. Friedlaender</i>	Strain from Case 2
Capsule	+	+	+	+
Motility	-	-	-	-
IMVIC reaction	-	+	-	-
Dextrose	-	-	-	-
Sucrose	(+)	(+)	± or (+)	A
Gelatin	+	-	+	-
Oxidase test	-	-	-	-
Lactose	-	-	-	A
Maltose	-	-	-	A
Dulcitol	-	-	-	-
Rhamnose	-	-	-	A

Table Bacteriological studies in case 2¹

Slightly sensitive to chloromycetin.
Resistant to kanamycin, tetracycline, streptomycin, gantrisin, neomycin, penbritin, and orbenin.

Fig. 2 Granulation tissue consisting of plasma cells and fibroblasts. The enlarged histiocytes with clear cytoplasm are prominent throughout the lesion (Mikulicz cells). *H and E* × 120.

extending into the orbit or reaching the meninges through the ethmoid or sphenoid bones. Involvement of the skin of the cheek, neck, forehead, and back has been reported. Nasal lesions may grow through the Eustachian tube into the antrum. Rare cases of the oesophagus, tonsils, bowel, and cervical lymph nodes are recorded.

Clinical diagnosis is usually not difficult. It should be confirmed by bacteriological determination of the *Klebsiella*, from nasal smears, needle aspiration of the lesion and cytological determination of Mikulicz cells, or by biopsy.

Streptomycin and aureomycin are the recommended therapeutic agents. Resistant strains have been described. Treatment must be prolonged,

especially in all those cases in which reinfection from a carrier is to be expected (Shvartsberg, Kozitskaia, Derkach, and Zaitseva, 1952; Durska, 1957). Local resection before antibiotic therapy is helpful, since the penetration of the therapeutic agent into the often large obstructive mass is limited. Surgical treatment alone is not successful.

Peculiar historical and epidemiological parallels between leprosy and scleroma exist, although the two diseases are clinically dissimilar. Hansen's bacillus was known for a long time and Koch's postulates were only recently fulfilled. Infectivity of both organisms is low. Close association with lepers for prolonged periods of time is necessary for infection. There is no explanation why some contacts contract the disease and others do not.

The bacilli of both diseases collect in specific histiocytes, which permit pathological diagnosis. Virchow's leprosy cell with lepra globi in leprosy, and Mikulicz's cell in scleroma. Both diseases require prolonged therapy, which has become available only relatively recently.

The tissue affinity of *Mycobacterium leprae* and *Klebsiella rhinoscleromatis* differs specifically. *M. leprae* infects primarily the ectoderm, including the neuroectoderm, and relatively late all other organs. *B. rhinoscleromatis* grows only in association with nasal and respiratory mucus, which provides protection against host defence. Antibiotic therapy must therefore be prolonged to prevent internal reinfection.

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