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RHINOSCLEROMA (SCLEROMA)

REPORT OF A CASE IN SOUTH AFRICA

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Rhinoscleroma (syns.: scleroma, 'scrofulous lupus of the nostrils') is a specific chronic granuloma localized in the rhinopharyngeal tract and involving the tissues of the nose, sinuses, pharynx, larynx, trachea and bronchi. Although the disease is now stated to be world-wide,[1] so far as the authors are aware this is the first report of a case in South Africa.

The literature has been reviewed in 1946,[1] 1949,[2] and 1955.[3] The disease was first described by the Viennese dermatologist von Hebra in 1870, but it had undoubtedly existed in Eastern Europe for many years before that time.[2] Important milestones in the history of the condition are the description of the characteristic Mikulicz cell in 1876 and the isolation of the *Bacillus rhinoscleromatis* by von Frisch and Pellizari in 1882. In 1932 the Second International Congress of Otorhinolaryngology had collected 2,361 cases for discussion.

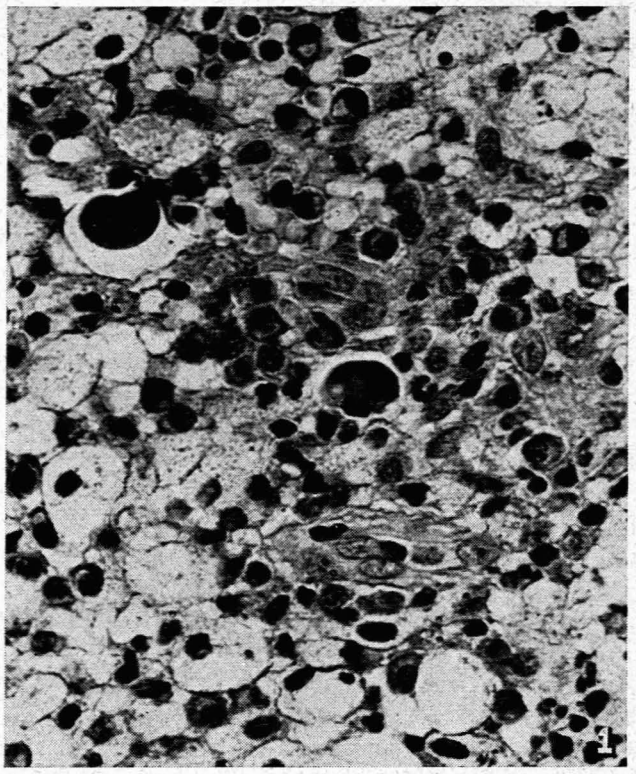
Incidence. There are now few countries in the world where cases have not been reported, but in many of them it is very uncommon in native-born inhabitants; e.g., in the USA up to 1949, 102 cases had been reported, only 13 of which were not in immigrants.[2] The disease appears to be particularly common in Central and Eastern Europe, Central America (El Salvador and Guatemala)[1] and in Indonesia.[4]

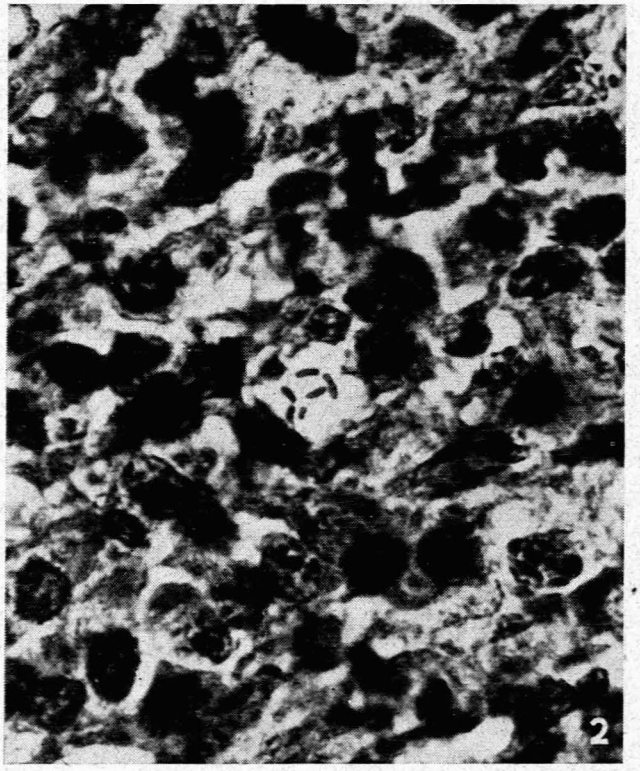
Etiology: Although the specific etiology is disputed, most authorities accept the *Klebsiella rhinoscleromatis* (Frisch bacillus) as the causative agent. This bacillus is a member of the Friedlander group and is biochemically[5] and immunologically[6] a distinct species. It can be isolated from cases with regularity and occurs as an intracellular inhabitant in the lesions. Cases of the disease exhibit positive intracutaneous skin tests to its antigens, and complement fixation and agglutination reactions have further confirmed its specificity.[5] Moreover, beneficial results with antibiotic treatment have been reported.[7–10] Levine [11] failed to find biochemically typical Klebsiella organisms in over 500 cultures from persons with or without disease of the ear, nose and throat other than rhinoscleroma. However attempts to reproduce the disease by various types of inoculation into humans has failed[1] although very similar histological lesions have been produced in white mice.[11] Thus Koch's postulates have not been fulfilled and some workers are concentrating on a possible virus etiology.[1] The disease occurs at all ages and in both sexes. It is most frequently seen in early adult life. Several reports of multiple cases in families have been reported. Its incidence is greatest at the poorer socio-economic levels.

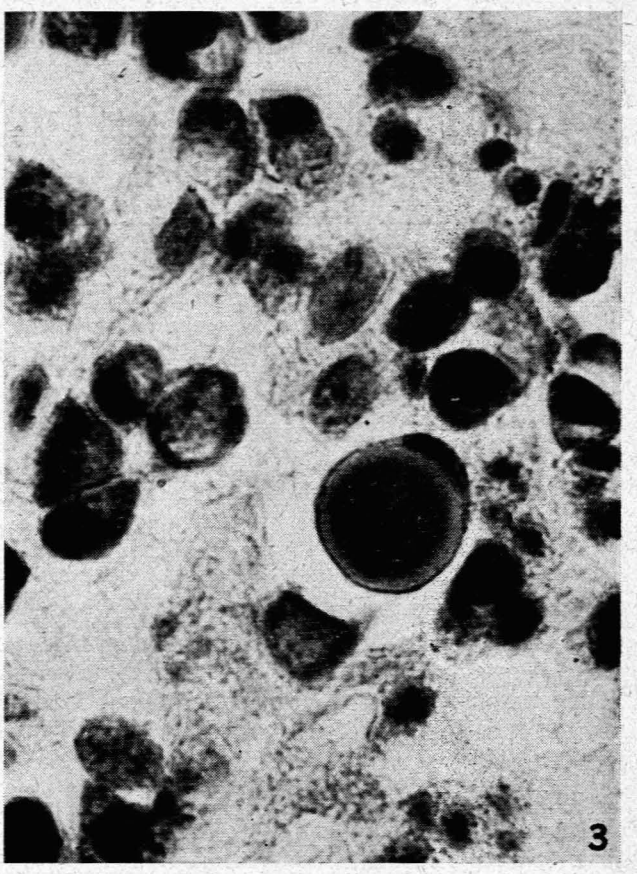
Transmission. Transmission experiments have failed and the exact mode of infection remains unknown. Probably long exposure is necessary.[3,4]

Pathology. The conditon begins slowly and insidiously, usually in the nasal septum or alae and spreads by peripheral extension to the nasopharynx, sinuses, larynx, trachea and bronchi, and less commonly to lips, tongue, uvula, soft palate, orbit, lacrymal passages, eustachian tubes and tympanic cavity. The condition is a submucosally infiltrating granuloma with pathognomonic histological features. A variety of clinical appearances are produced, e.g. atrophic rhinitis, localized nodules, polypi, and diffuse infiltrations. Untreated it pursues a chronic relentless course producing gruesome facial deformities. Bony and cartilaginous structures are not involved. Progressive fibrosis and cicatrization in the healing phase may lead to further deformities. The course may extend over a period of many years.

Histopathology. In very early cases there may be only massive plasma-cell infiltration with the characteristic bacilli lying free in the tissue spaces. Shortly







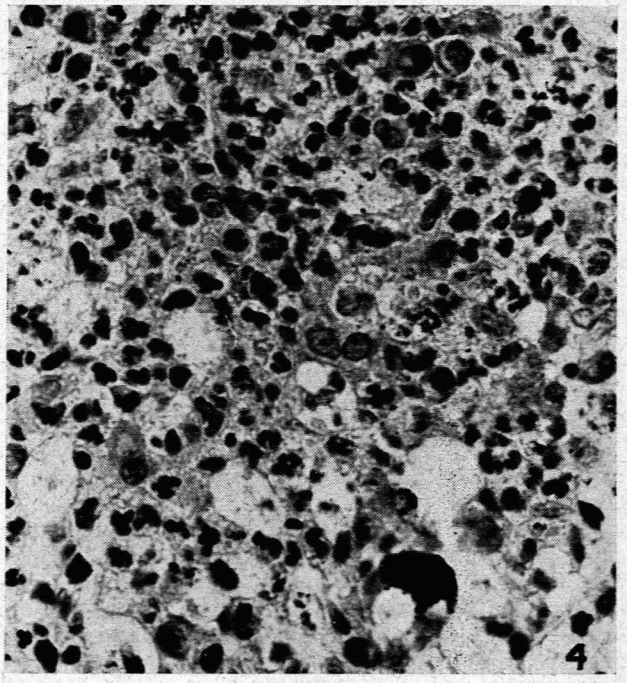


Fig. 1. Rhinoscleromatous granuloma. Stained H. and E. × 600. The section shows sheets of typical Mikulicz cells separated by a fibrous trabecula which is infiltrated by plasma cells. Two typical Russell bodies are present in the centre of the field.

Fig. 2. Rhinoscleromatous granuloma. Warthin-Starry technique × 1500. The section shows the typical rod-like bacilli lying within a Mikulicz cell.

Fig. 3. Rhinoscleromatous granuloma. Giemsa stain × 1500. The section shows a Russell body in an intracellular position.

Fig. 4. Rhinoscleromatous granuloma. Stained H. and E. × 400. The section shows a micro-abscess.

thereafter the Mikulicz cell appears, and in the active phase these cells are found in masses and sheets separated by fibrous trabeculae heavily infiltrated with plasma cells and a few lymphocytes (Fig. 1). The Mikulicz cell is probably a tissue histiocyte. They are large pale cells (50 μ) with small intensely pyknotic nucleus and foamy vacuolated cytoplasm. With the PAS stain, thin condensed cytoplasmic Schiff-positive partitions radiate out from the nucleus towards the cell boundary, but the intervening cytoplasm is unstained. Frozen sections stained with Oil Red O show no neutral fat. With bacterial stains, or with the Giemsa stain, many of the Mikulicz cells are seen to contain the characteristic rod-like bacilli (Fig. 2). These are argyrophilic and can be demonstrated in large numbers, both intracellular and extracellular, by the Warthin-Starry technique.

Russell bodies (rounded or oval hyaline masses, strongly PAS positive) are found in considerable numbers lying free and within plasma cells (Fig. 3). The fibrous trabeculae contain many dilated vessels, some of which show endarteritis obliterans. An additional feature is the presence of occasional microabscesses (Fig. 4). The overlying mucosa may show nonspecific pseudoepitheliomatous hyperplasia. As the lesion ages, progressive vascular sclerosis and fibrosis replace the cellular elements until cicatrization is produced.

Taken as a whole, the histopathological picture in the active stage is very characteristic. Although the Mikulicz cell itself can be seen in other conditions, e.g. simple or allergic nasal polypi, and other infections of the nasal mucosa (Friedmann[3]), we regard the Mikulicz cell containing typical bacilli as pathognomonic.

Clinical Picture. As the primary site is almost always in the nose, the disease often begins with the symptoms and signs of rhinitis, with ozoena, nasal discharge and epistaxis. Sense of smell may be preserved, for the olfactory cleft is not early involved. Later, progressive painless nasal obstruction is the main complaint. Intranasal inspection may show nodules, polypi, or diffuse infiltrations. Invasion of other structures produces symptoms due largely to mechanical obstruction and loss of function: viz. *larynx*—hoarseness, coughing, aphonia; *pharynx*—dysphagia; *tongue*—difficulty in speech; *middle ear*—deafness.

No pyrexia or constitutional symptoms are produced until late, when anorexia, weight loss and cachexia appear.

Diagnosis. A definite diagnosis may be established by biopsy, but this should be confirmed by cultural methods. Complement—fixation reactions, and agglutination and skin reactions are not to our knowledge available as yet in South Africa.

Prognosis. This used to be very poor, because spontaneous cure was rare and induced cures almost as uncommon. Progressive spread of the condition led to respiratory obstruction and death from intercurrent infection. New hope now awaits sufferers with planned surgery and antibiotic treatment. The organism has been proved sensitive to streptomycin, aureomycin, terramycin and chloromycetin, and apparent cures are being reported.[7–10]

REPORT OF CASE

J.P., an Indian boy aged 13 years, was born in South Africa, but was taken to India by his family when he was 2 years old and lived in a village in Bombay from 1945 to 1947. He now lives in Boksburg North and there are no other cases in the family of 2 adults and 9 children. His main complaint was blockage of the left nostril, and symptoms of this had been present for 2 years. During the last 2 months he had suffered frequent episodes of epistaxis and excessive sneezing.

On examination of the left nostril a large haemorrhagic polyp of the inferior turbinate, $\frac{1}{2}$ inch in diameter, was found. Similar smaller polypi were found on the posterior part of the right inferior turbinate. His general condition was excellent and no facial deformity was present. He was apyrexial. The larynx was normal. The polypi were removed surgically and sent to us for histological examination.

Histology. The characteristic histological features of rhinoscleroma were observed (Figs. 1-4) and the diagnosis was made on histopathological grounds.

Bacteriological examination was made a few days after operation. In the meantime, the patient had been treated with crysticillin. Swabs taken from both nostrils resulted in the isolation in pure culture of a bacillus with the characteristic morphological and biochemical features of *K. rhinoscleromatis*. The bacteriological studies were undertaken by Dr. V. Bokkenheuser of the South African Institute for Medical Research and we are indebted to him for the following detailed report:

> The bacteriological features of the organism from the case (J.P.) of rhinoscleroma are as follows:
>
> Primary culture on blood agar yielded an almost pure culture of a Gram-negative non-motile bacillus. The colonies were large and mucoid (Fig. 5) but capsules could only be demonstrated by intraperitoneal injection of the organism suspended in mucin into mice (Fig. 6).
>
> The culture fermented the following carbohydrates with production of acid only; glucose, mannitol, saccharose, maltose, salicin, adenite, xylose, sorbitol (after 6 days in-

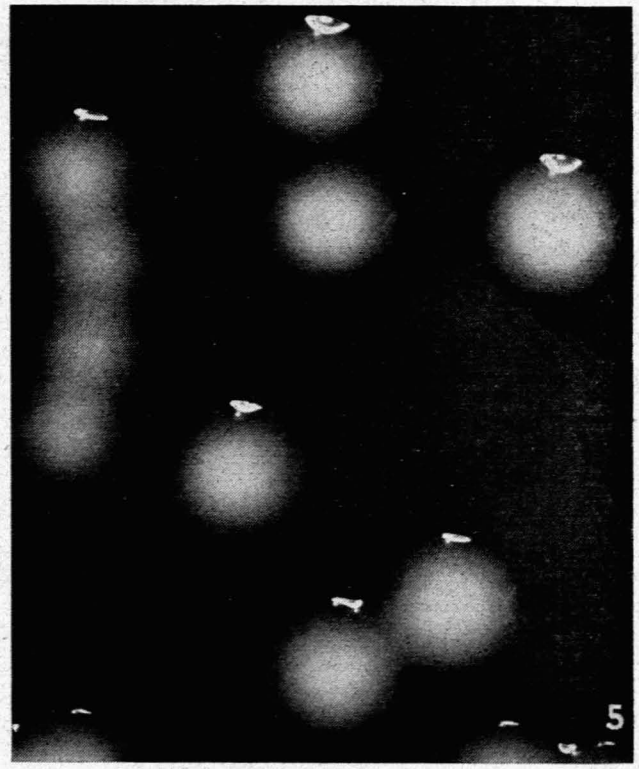


Fig. 5. Klebsiella rhinoscleromatis. The plate shows the mucoid colonies on blood agar.

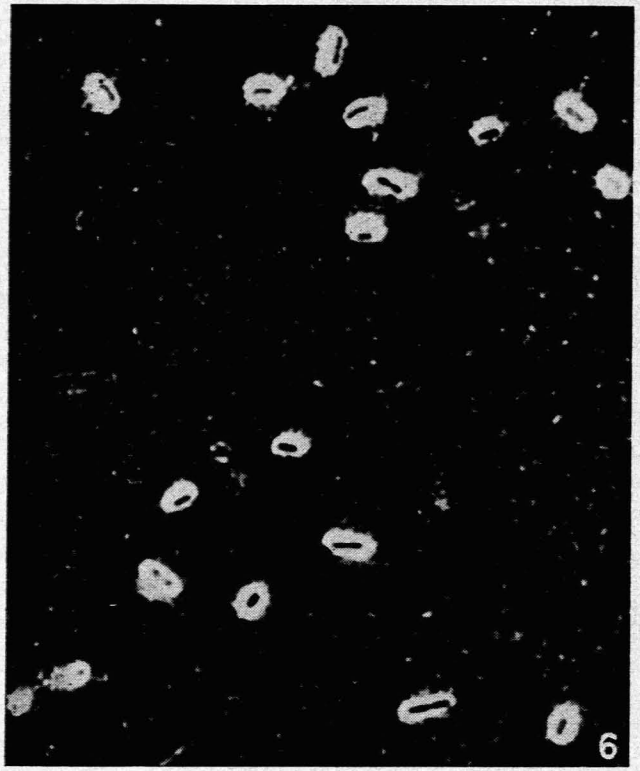


Fig. 6. *Klebsiella rhinoscleromatis.* Indian-ink preparation. The plate shows the encapsulated organisms.

cubation at 37°C), rhamnose, laevulose, dextrin, arabinose, glycogen and glycerol (after 10 days of incubation). Lactose and dulcite were not fermented. The H_2S, indol, urea citrate and Voges-Proskauer tests were negative.

Gelatin was not digested. The methyl red and nitrate tests were positive.

The organism has thus all the characteristics of the klebsiella organisms described in cases of rhinoscleroma (Kauffmann[14]).

A culture of the organism was sent to the Statens Serum Instituut, Copenhagen, for typing of the capsular antigen. Dr. Ida Orskav reported as follows: 'Biochemically the strain is designated as *Klebs. rhinoscleromatis*, and serologically it belongs to capsule type 3.'

Sensitivity tests showed the following spectrum of sensitivity: penicillin negative, streptomycin 2, sulphatriad negative, aureomycin 2, chloromycetin 2, terramycin 2, erythromycin 3, achromycin negative.

In view of this bacteriological report, further treatment with streptomycin and erythromycin has been advised.

DISCUSSION

The diagnosis of rhinoscleroma in this case has been established beyond doubt. From the history it appears that it must be regarded as an imported case; infection probably took place in India during the child's residence there in 1945–47. It is well known that the condition is prevalent in India and hitherto has been unknown in South Africa. The incubation period and evolution of the disease extended over a period of 7-9 years before the first symptoms were produced. A further 2 years elapsed before treatment was sought—a good illustration of the slow progress and chronicity of the disease. Further extensive antibiotic treatment to sterilize the site of infection is now urgently indicated.

SUMMARY

1. The first case in South Africa of rhinoscleroma is reported.

2. A brief review of the literature of the condition is presented.

3. Diagnosis in well established cases can be confidently made on histological grounds.

We wish to thank Dr. V. Bokkenheuser of the South African Institute for Medical Research for his expert assistance in the bacteriology, Dr. Ida Orskov of the Statens Serum Instituut, Copenhagen, for typing the capsular antigen, Dr. T. Pienaar for his clinical assistance, Mr. M. Ulrich for the photography, and Mr. D. Lloyd for his unstinted technical assistance.

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