The management of rhinoscleroma

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Introduction

In 1840 Bierkonesky described what he called an infective cutaneous cancer that affected the nose. Von Hebra (1870) described the same lesion but called it rhinoscleroma. Now it is believed to be an infection caused by a klebsiella rhinoscleromatis. Rhinoscleroma infection is worldwide but has a peculiar geographical distribution, as pointed out by Colledge (1936)—and which has been found true in Uganda where the following investigation was carried out. Previous workers in Uganda found a similar geographical picture (Kafero, 1951; Martin, 1967; and Roland, 1961). The treatment of this condition has been mostly confined to the use of Streptomycin and tetracycline but without any established knowledge of how much of these drugs was required to eradicate the infection (Roland, 1961). This infection, as we shall see is difficult to eradicate from a patient. Clinically, the condition looks like a tumour, as was thought by many in the past, but now we are more certain that it is caused by an organism called klebsiella rhinoscleromatis. This organism is responsible for the neoplasm-like manifestations of this disease that have caused so much diagnostic confusion.

The aims of this investigation, which was started in 1968, were:

1. To evaluate the role of antibiotics in the treatment of scleroma.
2. To establish what part surgery plays in the treatment of rhinoscleroma and its complications.

Materials and methods

Materials

A total of seventy-one patients was involved:

1. Seventy per cent from mainly south-west Uganda.
2. Thirty per cent from Rwanda, Burundi and Tanzania.

The patients were generally referred to Mulago Hospital Consultation Clinic, Kampala, from district hospitals of Uganda and the neighbouring Tanzania, Rwanda and Burundi, over a period of six years. They came from the south-west part of the country, mainly from the districts of Ankole and Kigezi. In Tanzania the patients referred to Mulago were from the north-west part which is bordering on Uganda's Kigezi and Ankole districts. In Rwanda, Burundi and Zaire, patients were limited to the highlands bordering on Uganda next to Kigezi District (Fig. 1).

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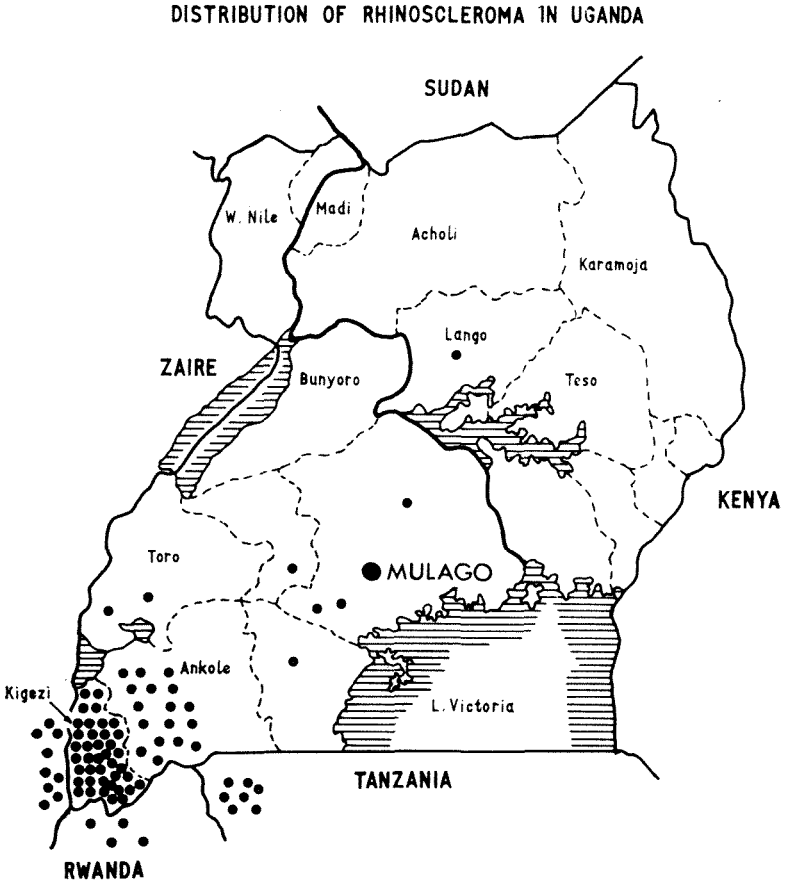


FIG. 1.

Shows the distribution of rhinoscleroma in Uganda as was seen in Mulago Hospital, Kampala in six years.

Methods

Each patient with a clinical diagnosis of scleroma was admitted to Mulago Hospital Department of Otolaryngology.

1. A complete otorhinolaryngological examination was made to assess the extent of the lesions and their distribution. The presence of lesions in the nose, oral cavity, pharynx, larynx or trachea was noted.

2. Two biopsies were taken. One was sent for culture and sensitivity to antibiotics, the second for histology. The discharge from the nose was taken on pus-swabs and sent for culture and sensitivity.

After establishing the diagnosis, the following routine was used. A choice of antibiotic was made from the following which were the ones to which sensitivity tests were carried out: streptomycin; tetracycline; erythocin; ampicillin; chloramphenicol; septrin; vibramycin (doxycycline).

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The choice of antibiotic for a particular patient was randomized. We printed twenty cards for each antibiotic. When a patient presented, a card was picked for him from a common pool. Then the patient was started on the antibiotic that was found on the card. It was then continued until a biopsy from the nose or nasopharynx yielded no growth of klebsiella rhinoscleromatis. These biopsies were taken at weekly intervals and sent for culture to see if organisms were still present or whether they had developed resistance to the drug in use. In some cases where the drug in use failed to alter the population of the organisms noticeably in eight weeks, it was discontinued.

3. *Surgical intervention*

(*a*) Nasal dilatation was used in this investigation in preference to excision or cauterization of the granulations in the nose. This change of method was motivated by previous experience with the latter method that was producing atrophic changes in the noses of patients (Ssali, oooo). Hagar's dilators were chosen because of their convenient sizes and their suitable curvatures. Removal of granulations or scar tissue was only carried out to find the stenosed nostril canal which in all cases were present although of 'pinhole' size in some. By using dilators it was possible to preserve the original mucous membranes which would otherwise have been removed if excision of granulations was carried out with a knife or cautery (Fig. 2). In cases where cautery or excisions was used, the patient developed atrophic rhinitis with subsequent foul-smelling discharge and crusting.

(*b*) Polyethylene tubes the size of normal nostrils were routinely inserted into each nostril and fixed in place using silk (Fig. 3).

These tubes remained in place for periods ranging from two to eight weeks. The longer the period the better was the result. Early cases needed a shorter period than the late cases. On the average the tubes stayed for twenty-four days. The healing was good with a well-epithelialized nostril devoid of crusts (Fig. 4). Crust formation, where it occurred, was minimal and often temporary as epithelium spread to cover all the nasal spaces. Polyethylene tubes when left in for twenty-eight to fifty-six days produced better results. A tendency for the nostrils to narrow was met with in a few cases but repeated dilation after the scar is fully matured and contracted prevented further stenosis in most of the cases. The biggest draw-back the tubes have is the discomfort they give to the patient.

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FIG. 2.
Advanced case of rhinoscleroma before treatment.

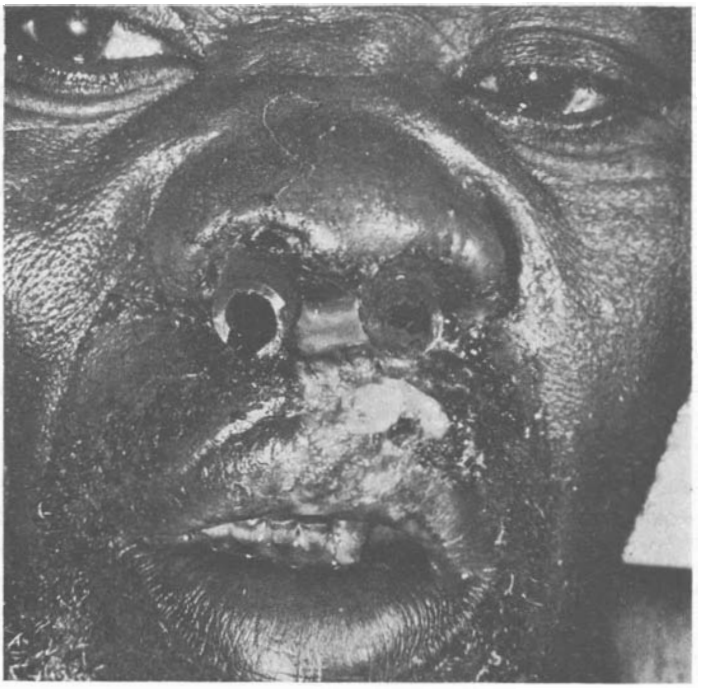


FIG. 3.
Same patient after dilatation and intubation, lip granulations removed.

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Results

Cultures

Biopsies were used for histology and cultures. Cultures from tissues on blood agar yielded growth of klebsiella rhinoscleromatis in 98 per cent of all cases that were clinically diagnosed as rhinoscleroma. Pus swabs from noses were less reliable. Pus swabs were positive in 60 per cent of biopsy-proven scleroma cases. It will be appreciated that biopsies were taken as the reliable means of ascertaining the presence or absence of organisms in the body tissues. These were taken weekly during the time the patient was maintained on antibiotics until two consecutive cultures were found negative. The table overleaf shows the various drugs used and their effectiveness against klebsiella rhinoscleromatis.

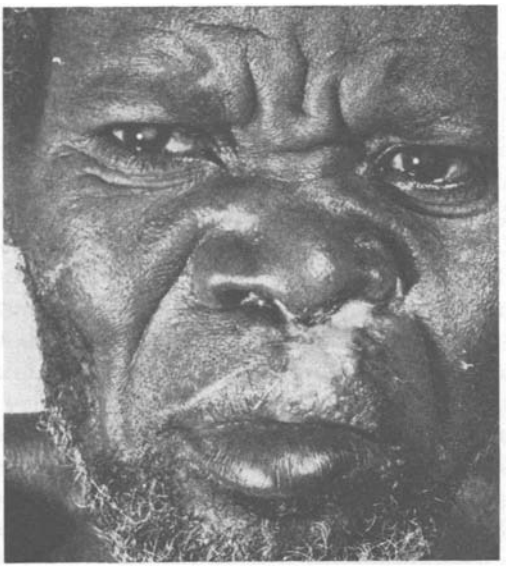


FIG. 4.
Eight weeks after treatment.

Up to now we have not yet found recurrence in any of the patients that have had two negative biopsies following a full antibiotic treatment course. Problems of irradication of organisms have been described by Hara *et al.* (1947) in a study of seven cases in one family. The success rate of the antibiotic regimen increased with a longer duration of treatment, save in cases where chloramphenicol and erythocin were used.

The best results were obtained using the following drugs, in order of activity, in vivo: ampicillin; septrin (bactrim, trimethoprim and sulphamethoxasole); vibramycin; streptomycin; tetracycline.

It was also established that a combination of two drugs produces speedier eradication of the infection especially if they are both bactericidal.

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TABLE I.

SHOWS ANTIBIOTICS USED AND THE RATE OF SUCCESS

Preparation	No. of patients	Dosage/day	Success rate	Mean duration in days
Streptomycin	13	0·5 g.	6	42
	7	1·0 g.	5	36
Ampicillin	12	2·0 g.	10	28
	10	1·0 g.	8	34
Septrin / Bactrim	6	8 tablets	5	28
Erythrocin	8	1·0 g.	Nil	32
Tetracycline	9	1·0 g.	7	72
	6	2·0 g.	6	56
Chloramphenicol	6	1·0 g.	Nil	42
Vibramycin	8	200 mg.	5	28

Discussion

Rhinoscleroma has a worldwide distribution but with greater incidence in central parts of Asia, South America and Africa. It is found in south-eastern parts of Europe and Russia and is common in Egypt. In East Africa the disease has been found to be particularly common in the south-eastern parts of Uganda, namely, Kigezi and Ankole districts (Fig. 1).

The frequency of scleroma in Kigezi, Ankole, Rwanda and north-west Tanzania remains up to now an unsolved mystery. Other areas of Uganda at similar high altitude as in Sebei do not have it. The question of a vector has been considered but not proved. Theoretically, a fly could transfer the klebsiella rhinoscleromatis organism from person to person by the mere fact that it feeds on mucus found on dirty noses. A fly could theoretically contaminate its feet and mouth parts while feeding on nasal secretions of an infected person, then transfer the organisms to another person by settling on his nose. The fact that this infection is prevalent in societies of relatively poor hygiene all over the world lends support to this means of transmission. In this same endemic area was found by the author a high incidence of atrophic rhinitis, attributed to a klebsiella ozaena (Ssali, 1973). Both these conditions are attributed to klebsiella organisms that are biochemically different. They both became more scarce as the educational and hygienic standards of a region increased.

Treatment with antibiotics showed that the drug should preferably be bactericidal rather than bacteriostatic. This is shown by the effectiveness of ampicillin, streptomycin and septrin. The drug must be given for a much longer period than usual otherwise the disease may recur. Four to six weeks of treatment was the average duration of antibiotic therapy.

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This investigation again showed that the usual therapeutic dose may have to be doubled if a speedy recovery is to be achieved. Some drugs like streptomycin which have ototoxic effects may necessitate periodic audiometry to be done on the patient. In vitro klebsiella rhinoscleromatis was found to be sensitive to the following drugs: ampicillin, streptomycin, septrin, tetracycline, erythrocin and chlorophenenicol but in vivo the picture was different. Only bactericidal drugs that could kill the organism in its intracellular habitat were the ones which effectively and rapidly eradicated the disease. A bacteriostatic drug which mostly prevents multiplication of the organism depends on the action of macrophages. The macrophages in this case are the Mikulicz cells already inhabited by the organisms and therefore incapable of destroying them. This showed that the body has no effective immune mechanism to combat the disease.

Basically, this disease is chronic and progressive, causing severe disability to the individual. Clinically, it consists of three recognized stages, namely:

1. *The atrophic stage*—this consists of changes in the nasal mucosa, similar to that of atrophic rhinitis, with crust formation and foul-smelling discharge. The columnar ciliated epithelium changes to squamous type.

2. *The stage of granulation formation*—here we find masses of granulations without ulceration. This may occur in the nose, pharynx, palate, larynx, trachea and bronchi.

3. The third stage consists of fibrosis and stenosis. This seals off the nostrils completely. At times the fibrosis spreads to the nasopharynx, and the trachea (Fig. 5).

Histologically, rhinoscleroma is a chronic granulomatous infection with characteristic fibrosis, eosin-staining, Russell bodies and vacuolated Mikulicz cells in which are found Friedlander's bacillus, as referred to here as klebsiella rhinoscleromatis. It is normally the disabilities caused by the third stage that make a patient seek specialist medical treatment. The patient wants to breathe through the nose again and swallow food properly. Unless the problem is approached systematically the patient gets no benefit either from antibiotics or surgery. If the antibiotic course is not long enough, organisms re-establish the disabilities within a relatively short time. Chemotherapy must be accompanied by surgical interventions to re-establish the air-way without creating atrophic changes which may by themselves be as bad if not worse than the original disease. Where the disease involved the pharynx, extensive excision of scar tissue failed to produce permanent improvement because the stenosis recurred invariably (Fig. 5). The trachea was successfully dilated and the air-way improved using gum-elastic bougies and a bronchoscope. The nasolacrimal ducts were found involved in two of the patients causing epiphora. Three of the cases in this series had dyspnoea resulting from stenosis of the larynx, trachea or a main bronchus. In these cases, ampicillin was used. Although the nasal

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lesions yielded no more growth of organisms after four weeks of antibiotic therapy, an additional course was given for seven days to ensure complete eradication. The trachea in each case was dilated using bougies and split laryngoscope.

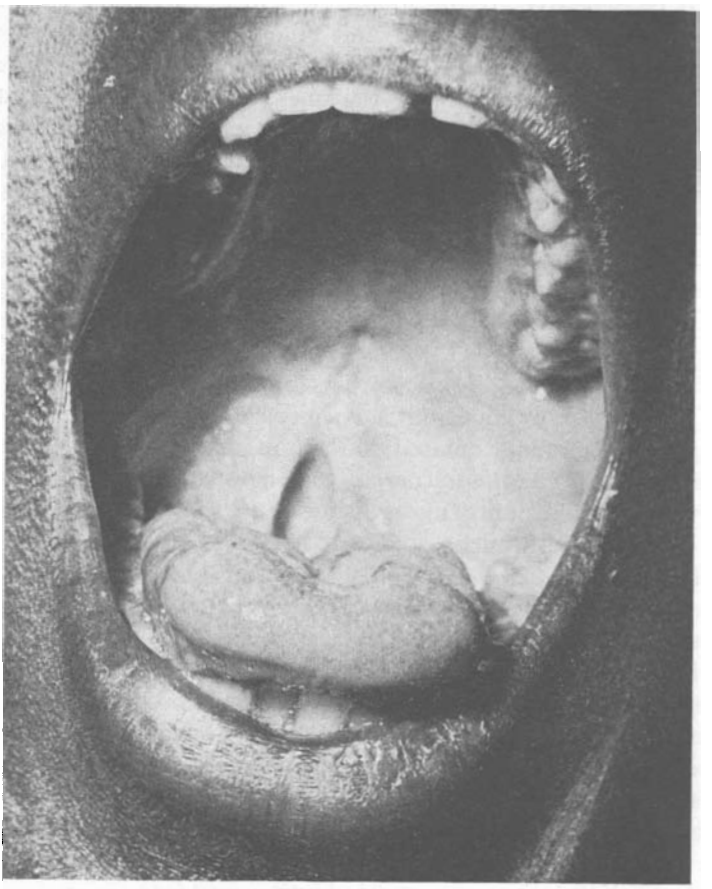


FIG. 5.
A case of advanced pharyngeal stenosis from scleroma. That small opening served for swallowing and breathing.

The left main bronchus was in all the three cases the one involved by scleroma causing obstruction by granulations. These were either picked up endoscopically using biopsy forceps or the bronchus just dilated with bougies. In each case improvement was satisfactory and X-ray of chest showed better aeration of the lung within two weeks following the first dilatation.

The hard palate was perforated in two of our cases causing difficulty in swallowing, because of regurgitation through the nose. In one case repair

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was impossible because of the fibrosis and the granular nature of the mucosa. A dental plate was used to close the hole. The second one had a mucosal flap used to close the defect.

Summary

This investigation showed that the organism klebsiella rhinoscleromatis was present consistently in our cases. It was found to be difficult to eradicate completely although seemingly sensitive to common antibiotics in vitro. A prolonged treatment with bactericidal antibiotics is therefore necessary to eradicate it. Nasal dilatation rather than excision of granulations is required to restore nearly-normal physiological nasal function. Excision or cautery of granulations inside the nose is discouraged so as to avoid atropic charges.

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