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Rhinoscleroma in Western Kenya: Report of Three Cases with Characteristic Features

Kan TORIYAMA[1], Fukumu UZUTA[1], Masachika SENBA[1], Hideyo ITAKURA[1] and Noah O. KAMIDIGO[2]

[1]*Department of Pathology, Institute of Tropical Medicine, Nagasaki University. Sakamoto−machi 12−4, Nagasaki 852, Japan.*
[2]*Provincial Pathologist, Rift Valley Province, Nakuru, Kenya*

Abstract: We report three cases of rhinoscleroma in western Kenya with their characteristic histological features. During the period of seven years from 1979 to 1985, 23,721 surgical pathological specimens were examined in western Kenya. Forty−eight cases were diagnosed clinically as tumor or tumorous lesion in the nasal cavity. Three of them were identified as rhinoscleroma histologically. Rhinoscleroma showed characteristic appearances. There was moderate infiltration of foamy−shaped histiocytes (Mikulicz's cells), plasma cells with Russell's bodies and lymphocytes beneath the nasal mucosa. Rod−shaped bacilli were phagocitized by Mikulicz's cells and were demonstrated clearly by Warthin−Starry stain. Rhinoscleroma is a relatively rare disease in this area. All patients were adults and lived in the tropical areas with relatively plentiful rainfalls. The most common tumor or tumorous lesion in the nasal cavity in western Kenya was histologically squamous cell carcinoma, followed by rhinosporidiosis, undifferentiated malignant tumor and malignant melanoma. We have not seen any rhinoscleroma which showed malignant transformation or was complicated by other malignant tumors.

Key words: Rhinoscleroma, *Klebsiella rhinoscleromatis*, granuloma, nasal tumor, western Kenya

INTRODUCTION

Rhinoscleroma is a chronic granulomatous disease which is caused by Gram negative, diplobacillus, *Klebsiella rhinoscleromatis*. Von Hebra and Kaposi (1870) first described the disease as one of sarcomas in Austria. Von Frisch (1882) reported that the disease was a granuloma which was caused by specific diplobacilli species. Nowadays, *K.*

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rhinoscleromatis is generally accepted as the causative agent(Levine, 1951). The disease usually affects the nasal mucosa, gradually involves the upper respiratory tracts and finally results gross deformity of the nose or destruction of the respiratory passages. Histologically, the lesion is characterized by the specific infiltration of the two types of cells, namely, foamy-shaped Mikulicz's cells and Russell's bodies. The possible way of infection of the disease is still unclear. Although the disease had been considered as a lesion of the nose alone, it has been found with increasing frequency in the larynx, trachea, bronchus, cervical lymph node and orbit(Jay *et al.*, 1985; Stiernberg *et al.*, 1985; Gaafar and Helmi, 1984; Strobel *et al.*, 1983; Agarwal *et al.*, 1981; Soni *et al.*, 1985; Badrawy and El-Schnnawy, 1974). The disease can occur in any generations but slightly more common in the second and third decades of life, and both sexes are equally affected(Jay *et al.*, 1985) The disease is likely to be found amongst peasants and the lowest class of worker at the social, economic and hygienic levels(Kerdel-Vegas *et al.*, 1963). Nowadays, the disease occurs sporadically all over the world and is endemic in Central and South America, Egypt, Indonesia, eastern Europe and Uganda(Kerdel-Vegas *et al.*, 1963; Shum *et al.*, 1982). No histological and epidemiological reports of the disease have been published from Kenya. In the present communication, we report three cases of rhinoscleroma in western Kenya with their characteristic features and discuss the association with malignant tumor of the nasal cavity.

Materials and Methods

During the period of seven years from 1979 to 1985, we examined a total of 23,721 surgical specimens in western Kenya and found out three cases of rhinoscleroma and 45 cases of tumors and tumorous lesions in the nasal cavity. These specimens were taken at the mission hospitals, district hospitals, private clinics and provincial hospitals in western Kenya(Rift Valley, Nyanza and Western Province)and brought to the histological department of the two hospitals, the Rift Valley Provincial General Hospital and Nyanza Provincial General Hospital. When the specimens were brought to the hospitals, the clinical data and general informations relevant to the desease were collected as completely as possible.

The paraffin sections were stained with hematoxylin-eosin(H.E.), periodic acid Schiff(P.A.S), silver impregnant, elastic van Gieson, Mallory's for collagen fibers, Gram and Warthin-Starry procedures.

Results

1. Prevalence of rhinoscleroma in western Kenya

In Table. 1, the site of lesion, age, sex, ethnic group and inhabitation of the patients were described. Out of 48 cases of tumor or tumorous lesion in the nasal cavity

Table. 1. Rhinoscleroma in Western Kenya (1979−1985)

Case	Site of lesion	Age	Sex	Ethnic group	District	Province
1	nasal cavity	Adult	Female	Luhya	Busia	Western
2	nasal cavity	75	Female	Kalenjin	Kericho	Rift Valley
3	nasal cavity	Adult	Male	Kalenjin	Kericho	Rift Valley

which were diagnosed clinically, three were diagnosed histopathologically as rhinoscleroma. We could not find out rhinoscleroma in any place other than the nasal cavity. Two of them were from the Kalenjin group in Rift Valley Province and one was from the Luhya group in Western Province. Rhinosleroma was a relatively rare disease in western Kenya.

2. Histology of rhinoscleroma

The nasal mucosa which covered the affected area showed moderate atrophic change (Photo. 1) and the peripheral mucosa of the lesion showed often moderate thickening with acanthosis and hyperkeratosis. Beneath the mucosa there was chronic inflammatory cell infiltrations. The most characteristic histological feature of the disease was the infiltration of Mikulicz's cells, plasma cells with Russell's bodies (Photo. 1 and 2). The Mikulecz's cell was a foamy vacuolated histiocyte that measured 100 to 200 microns in diameter and had a clear cytoplasm and eccentric nucleus (Photo. 3). In our cases, numerous number of rod−shaped bacilli were identified in the cytoplasm of Mikulicz's cells and extracellular tissues. These short rod−shaped bacilli were Gram negative and demonstrated clearly with Warthin−Starry stain and measured 2 to 5 microns in length (Photo. 3 and 4). The Russell's body could be recognized as a degenerated plasma cell and showed non−nucleated round structure. It was 10 to 40 microns in diameter and had a homogenous cytoplasm which was stained a bright red color with H.E. stain (Photo. 5).

3. The association with rhinoscleroma and malignant tumors in the nasal cavity

In Table. 2, histological diagnosis of tumors and tumorous lesions in the nasal cavity, including rhinoscleroma was described. Out of 23,721 surgical pathological specimens examined, 48 cases had been diagnosed clinically as tumor or tunorous lesion in the nasal cavity. According to our histopathological examination, the tumors or tumorous lesions in the nasal cavity in western Kenya were diagnosed as follows: squamous cell carcinoma, rhinosporidiosis, undifferentiated malignant tumor, malignant melanoma, benign tumor, rhinoscleroma and malignant lymphoma. Although 33 cases out of 48 tumors and other diseases in the nasal cavity were malignant neoplasms, we could not find out any rhinoscleroma which was associated with malignant tumors or showed a malignant transformation.

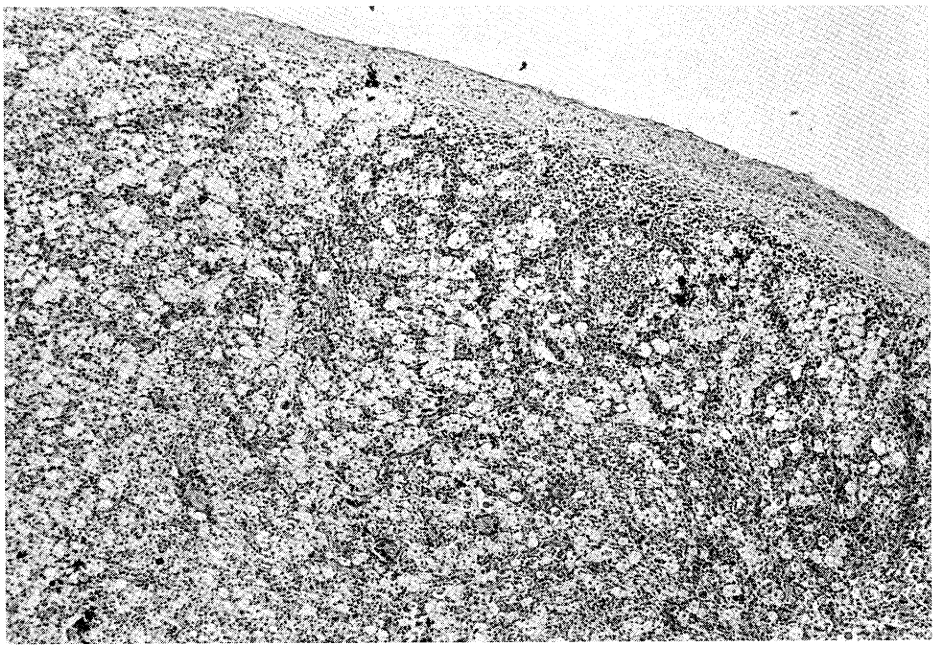

Photo. 1. Marked infiltration of Mikulicz's cells and plasma cells beneath the atrophic nasal mucosa (H.E., ×40).

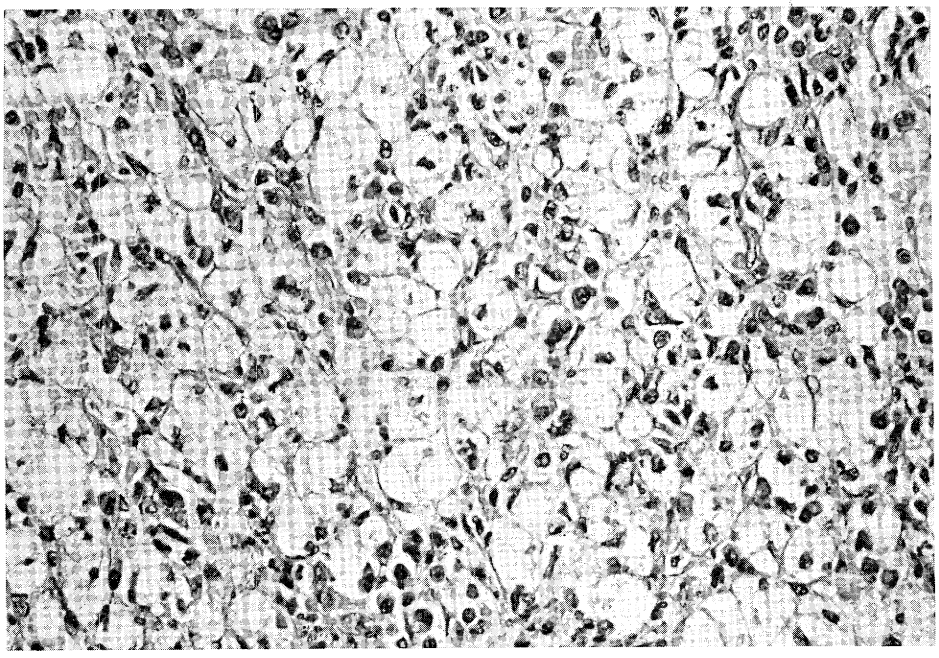

Photo. 2. Mikulicz's cells (H.E., ×200).

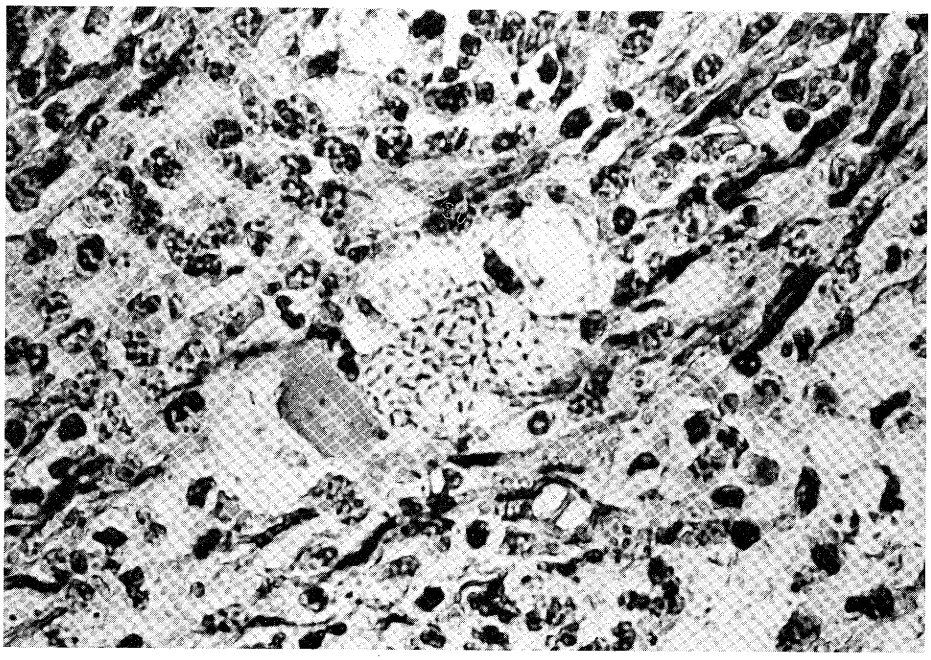

Photo. 3. Rod−shaped bacilli in Mikulicz's cells
(Warthin−Starry stain, ×400).

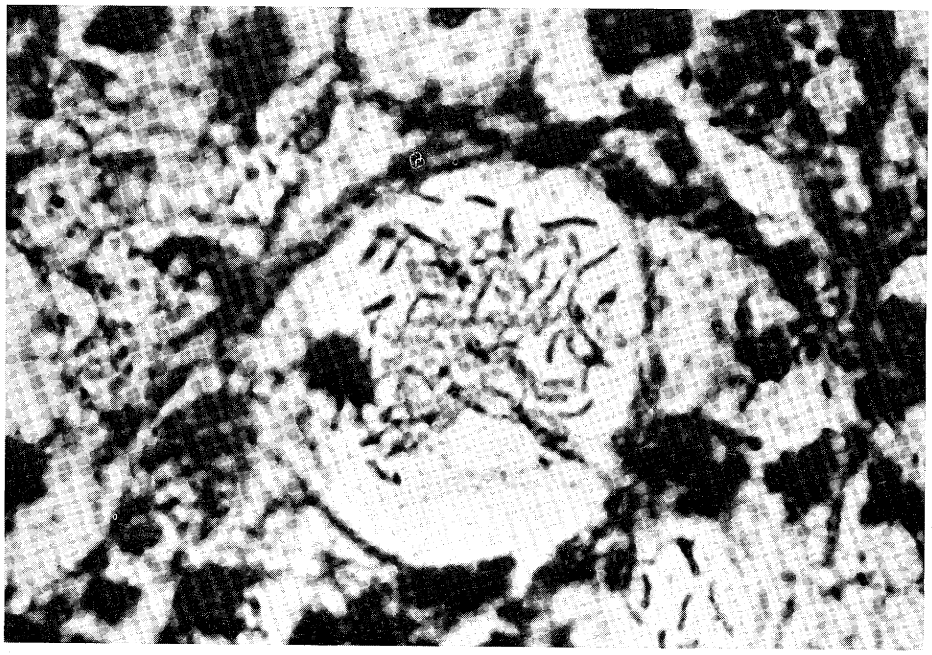

Photo. 4. Rod−shaped bacilli in Mikulicz's cells
(Warthin−Starry stain, ×1000).

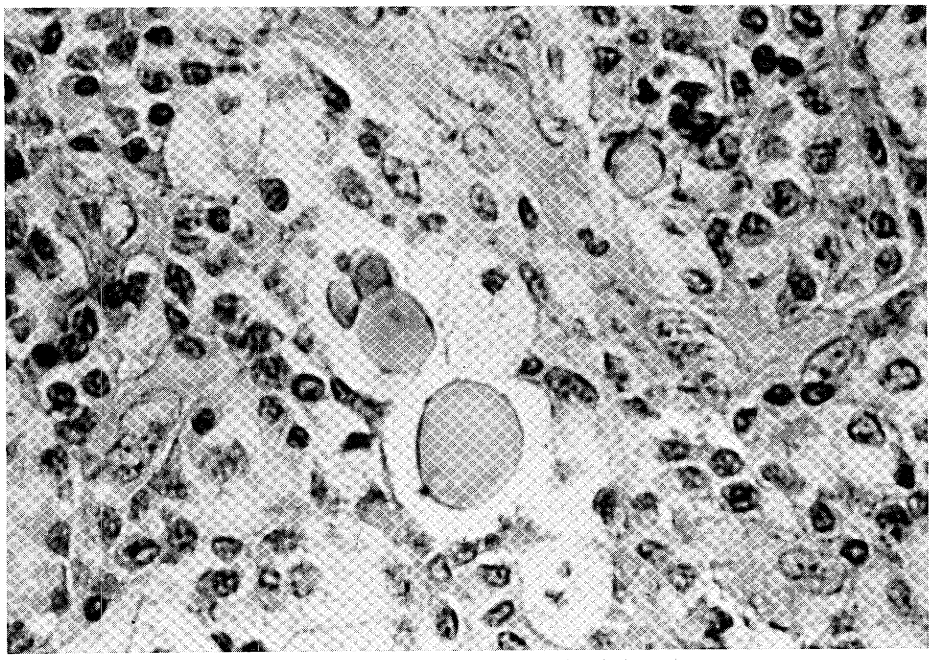

Photo. 5. Russell's bodies with plasma cells. (H.E., ×400).

Table 2. Tumorous lesions and tumors of the nasal cavity in Western Kenya (1979 to 1985)

Histological diagnosis	number
Carcinoma	22
Rhinosporidiosis	9
Undiffirentited malignant tumor	6
Malignant melanoma	3
Benign tumor	3
Rhinoscleroma	3
Malignant lymphoma	2
Total	48

DISCUSSIONS

Since 1951, few cases of rhinoscleromas have been reported from middle Africa except from Uganda, Rwanda, Burundi and Upper Volta (Table 3). Roland (1961) reproted two cases of the disease in Kenya. We identified three cases of rhinoscleroma among the total of 23,721 surgical pathological specimens examined in the period between 1979 and 1985 in western Kenya. All our cases were located exclusively in the nasal cavity alone. Although in the United States of America the disease is becoming one of the medical problems because of the recent influx of immigrants from endemic central America (Shum *et al.*, 1982), our study suggests that rhinoscleroma is a relatively rare disease in western Kenya. Sometimes the disease was associated with malignant tumors or showed a malignant transformation (Kerdel-Vegas *et al.*, 1963; Shum *et al.*, 1982). In our cases, however, there was no rhinoscleroma which was associated with malignant tumors or showed a malignant transformation. The clinical course of rhinoscleroma has been devided into three stages (Jay *et al.*, 1985). The first stage is the catarrhal stage which is characterized by purulent rhinorrhea lasting for several

Table 3. Reported cases of rhinoscleroma in Middle Africa since 1951

Country	No. of cases	Reference
Ghana	1+?	Vernoon & Jager (1969), Murpht (1981)
Ivory Coast	1	Bauduceau et al. (1984)
Kenya	5	Boland (1961), Toriyama et al. (1986)
Malawi	10	Liomba & Hutt (1980)
Mali	1	Penalba et al. (1983)
Mozambique	2	Azevedo (1971)
Nigeria	9	Roland (1961), Edward et al. (1977)
Rwanda-Burundi	34	Thjis (1956), Fain & Falaise (1957), Marneffe (1959)
Zaire	?	Claveau (19689)
Senegal	5	Reynaud (1956, 1966)
South Africa	1	Beynaud & Dorfman (1956)
Uganda	103	Kafero (1951), Roland (1961), Martin (1967), Ssali (1972)
Upper Volta	37	Claveau (1976)
Zaire (Eastern)	6	Liomba & Hutt (1980)
Zambia	3	Bhagwandeen (1977)
Zimbabwe	5	Carter (1966)

* modified the original data of Liomba and Hutt (1980)

weeks or months. The second stage is the granulomatous stage which is most commonly seen and shows soft granulomatous nodules in the respiratory tracts. After several years, the disease progresses to the third stage, sclerotic stage in which fibrosis of the affected area occurs. All our three cases showed the granulomatous stage, when the disease shows histologically most characteristic features of the infiltraion of Mikulicz's cells and Russell's bodies. Although there were numerous number of the bacilli in Mikulicz's cells and extracellular tissues, we did not culture the microbe for identification of organism. The presence of both Mikulicz's cells and Russell's bodies confirms the diagnosis of rhinoscleroma (Convit *et al.*, 1961). Although we reported three cases of rhinoscleroma in western Kenya and all of them showed the granulomatous stage, we might overlook the disease which showed the caterrhal or sclerotic stage. Further study is required to elucidate the epidemiology of rhinoscleroma in western Kenya and also the mode of infection of the desease.

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ケニア西部における rhinoscleroma と鼻腔内腫瘍

鳥山　寛，宇津田　含，千馬　正敬，板倉　英世（長崎大学熱帯医学研究所病理学部門）
NOAH. O. KAMIDIGO(ケニア共和国，リフトバレー州病理医)

Rhinoscleroma は *Klebsiella rhinoscleromatis* を原因菌とする，主として鼻腔粘膜に生じる慢性肉芽腫性疾患である．かつては欧米先進諸国にも存在したが，現在では熱帯，亜熱帯地方に局在しており特にエジプトを中心とする北アフリカ，エルサルバドルを中心とする中南米からの報告が多い．過去にケニアからの詳細な報告はなかったが，我々は1979年から1985年にわたってケニア西部，リフトバレー，ニャンザ，ウエスタン州の外科生検材料，総数23,721のうち48例の鼻腔内腫瘍様病変を検索し3例の rhinoscleroma を得た．患者は3例とも比較的，降水量の多い農村地帯に居住する成人であった．病理組織学的に rhinoscleroma は病変部におけるミクリッツ細胞及び形質細胞由来であるラッセル小体の浸潤を特徴としており組織球由来のミクリッツ細胞に貪食された *K.* rhinoscleromatis と思われる桿菌は Warthin−Starry 法で明瞭に染色された．腫瘍及び腫瘍様疾患としては扁平上皮癌，rhinosporidiosis，未分化悪性腫瘍，悪性黒色腫の順に多かったが rhinoscleroma と他の悪性腫瘍との合併，或いは rhinoscleroma の悪性変化はみられなかった．

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