Leprosy of the Nose Clinical Reassessment¹

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Leprosy, one of the most important tropical granulomatous diseases, caused by *bacillus leprae* has a special predilection for the mucous membrane, skin and peripheral nerves. Although leprosy is uncommon in Egypt, it is considered to be endemic in certain localities of this country, especially in upper Egypt. It is noteworthy that the oldest known descriptions of the disease can be traced to the time of the Exodus, 1411-1314 B.C. (⁴).

The nose, lined by mucous membrane, is one of the chief organs that suffers greatly from this disease. It is frequently deformed and disfigured and it contributes to the characteristic facies of leprotic patients. In spite of this important fact, the nose received little attention from the workers in this field, to the extent that Cochrane (⁵) reported that neither he nor others had thoroughly investigated early changes in the nose. It was therefore felt that a detailed clinical study of leprosy of the nose would fill a gap in leprosy research.

MATERIALS AND METHODS

Over a two year period (1971-1972), a group of 260 male leprotic patients from the Alexandria Leprosarium male hospital, were systemically examined with particular stress on the nose. These patients represented different age groups, clinical types and stages of morbidity (Table 1).

RESULTS

General clinical data. The collected data were analyzed, tabulated and classified into categories to give a clear detailed clinical picture of the disease in the nose. Young adults and children were the main victims of the disease. In addition, some elderly patients were seen with a long period of morbidity indicating that the disease does not apparently affect the life span. The study of the age at onset of the disease (Table 2) showed that the disease commonly attacked the younger population and, if we keep in mind the long incubation period,

Age groups	Lepromatous	Tuberculoid	Dimorphou	
1-10	4		_	
11-20	40	32		
21-30	68	24		
31-40	28	8	5	
41-50	16	6	3 ·	
51-60	12	4		
61-70	5		-	
71-80	3	2	_	
	176	76	8	

TABLE 1. Age groups and clinical types of disease.

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it becomes clear that the disease may be contracted in childhood and youth. In a high percentage (87.7%, 228 cases), the onset was before the age of thirty.

A nearly constant finding was the low social standard of living, the poor hygienic surroundings and the malnourishment among the majority of the studied cases.

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	Age groups in years								
Clinical types	Below 10	11-20	21-30	31-40	41-50	51-60	61-70	70	Total
Lepromatous	24	84	48	4	12		_	4	176
Tuberculoid	8	48	12	8			-	-	76
Dimorphous	-		4	4		-	-		8
	32	132	64	16	12	·	-	4	260

TABLE 2. Age of onset of the disease.

Type of disease	Patients examined	Nasal affection	Skin affection	Neural affection
Lepromatous	176	176	176	152
Tuberculoid	80	72	32	72
Dimorphous	4	4	4	4
	260	252	212	228

TABLE 3. Frequency of leprotic lesions.

The earliest initial leprotic manifestations were noted in the skin in 83, in the nose in 74 and in the peripheral nerves in 60 cases, and in more than one site in 43 cases.

Nasal involvement. In the studied series, 252 cases (97%) had nasal involvement while 8 cases only showed a clinically leprosy free nose. Those with nasal involvement were 176 lepromatous, 72 tuberculoid and 4 dimorphous cases (Table 3 and Figs. 1 and 2).

Considering that mucous membrane granulomata, ulceration, discharge and bleeding are signs of activity, there were 168 with active nasal lesions and 84 cases in the inactive stage with different degrees of scarring and atrophy.

Nasal symptoms. The group of symptoms given by any case were nearly the same and were of great significance in the assessment of the degree and stage of morbidity. In the order of diminishing frequency, they were as follows.

Epistexis. This was the most frequent and characteristic symptom. It was the initial symptom in 82 cases (31.5%) and the presenting symptom in 224 cases (86%). As the disease tended toward healing it diminished in frequency till it was absent in inactive long-standing cases. Epistexis was always scanty and self-arresting, it was commonly spontaneous and occasionally induced by nose-picking or crust removal. In some early cases with active extensive lesions, especial-

ly on the septum, epistexis was severe and frequent. Bleeding was commonly unilateral but occasionally bilateral. Right side bleeding was more frequent in incidence and amount.

Nasal obstruction. Although this was a common and probably also an early symptom, it did not attract the attention of many patients at the onset of the disease. However, as the diagnosis became established, it was present in 124 cases (47.7%). Obstruction was unilateral or bilateral, intermittent or continuous, according to the underlying lesions and the amount of accumulating discharge and crusts.

Nasal discharge. This was unilateral or bilateral, antenasal or postnasal, and was present in 121 cases (46%). In a few cases with early lesions, the discharge was always thin, mucoid and scanty. However, in cases with more advanced active lesions it was frequently thick, purulent, yellowish green, profuse, with a foul odor and occasionally tinged with blood. Due to the impaired function of the nasal mucosa, the greater amount of the nasal discharge usually remained inside the nasal cavity, often becoming dry and forming crusts. Only small amounts were expelled as antenasal or postnasal discharge. In late cases, with marked destruction of most of the intranasal structures, the discharge was almost entirely replaced by dry crusts that were seen adherent to the nasal



FIG. 1. Leprosy of the face, lepromatous type, involving mainly the nose.



FIG. 2. Tuberculoid leprosy of the face with multiple discolored patches.



FIG. 3. Collapse of the nasal ala, by the contraction of healing nasal lesions.

walls or filling the nasal cavity. In some further advanced cases where most of the intranasal structures were completely destroyed, there was neither discharge nor crusts but a wide roomy nasal cavity.

Crusts. These represented a common clinical finding. Although they were variable in amount, site, color and consistency, they were constantly found in all active stages of the disease.

The intranasal crusts consisted partly of desiccated nasal discharge and partly of sloughing tissues, clotted blood and external dust particles. Hence, they were always variable in color, shape and texture.

The frequent initial sites of these crusts were commonly on both sides of the anterior septum, and as the disease affected other sites they were found also adherent to walls of the nasal cavity. The adherent crusts on the ulcerated areas of nasal mucosa, cartilage and bone were not easily removed and forcible trial for their removal led to considerable bleeding. The crusts were commonly dark yellow, but occasionally they were brown, grey, black, green or showed a combination of colors. Admixture with blood gave the crusts a reddish coloration.

The amount of these crusts were proportionate to the extent and degree of intranasal lesions. In cases with small septal ulcers, the crusts were thin, scanty and of a lighter



FIG. 4. Falling nasal tip after loss of the columellae.

color. They were localized and presented only on the affected areas, hence no symptoms of nasal obstruction were present. As the disease affected wider areas of the nose, the amount of crusts increased to the extent that they filled the whole nasal cavity.

In consistency they were commonly soft, and according to the amount of nasal discharge, they were either very loose and soft or very dry, firm or hard. The loose and soft crusts had offensive odor, but this diminished gradually as they became more dry.

In the very late cases, where most of the intranasal structures had been destroyed and the disease was arrested, the nose displayed the picture of a very wide nasal cavity without crusts or discharge.

Nasal deformity. Up to 124 cases (47.7%) complained of different degrees of disfigurement of the nose. In some of them it was the main presenting symptom, in others it appeared later in the course of the disease. In the majority, the symptom was due to tissue destruction or fibrous tissue contraction, but in some it was merely due to the presence of a single or multiple lepromatous nodules on the overlying skin.



FIG. 5. Depressed and collapsed nose affecting its lower half.

Altered speech. Some patients (18 cases, 6.9%), noticed a gradual change in the tone of their voice with a sort of increased nasal intonation. These were those who had bilateral nasal obstruction or widened nasal cavity after extensive destruction.

Anosmia. Unexpectedly, this was the least frequent symptom, in spite of the different variable degrees of nasal involvement. Thirteen patients only had this symptom, but they regained the olfactory sense after nasal lavage with the exception of three cases (1.2%) who remained constantly anosmic.

A few other symptoms were rarely present, such as headache, nasal discomfort and epiphora, but they were very infrequent and not related to the nasal affection.

Nasal signs. Nasal deformity. A high percentage of nasal deformity was encountered in this survey (47.7%), an incidence which can be found only in a very few other diseases of the nose. All degrees and forms of disfigurement were detected, varying from mild alar collapse with columellar contraction and falling nasal tip through collapsing nasal bridge to a completely disfigured nose. In all cases, the lower half of the nose was the site of disfigurement (Figs. 3, 4, and 5).

When the skin over the alae, columellae or nasal vestibules, was the site of the lesion, as was frequently found, the deformity was due to fibrotic contraction of the affected part resulting in collapsing alae, short columellae, depressed tip or narrowed anterior nasal orifices which sometimes attained a state of complete atresia. When the amount of fibrosis was unequal on the two sides, irregular deformity or tilting of the tip, alae and columellae took place. In rare cases, there was partial or complete loss of the *alae nasi*, columellae or nasal vestibules (Fig. 6). Less frequently, the nasal septum was des-



FIG. 6. Irregular deformity of nasal alae, tip and columellae by the effect of contracting fibrous tissue.

troyed either alone or in association with other nasal cartilages and bone leading to depression and collapse of the nasal bridge and deformity of the lower half of the nose (Fig. 7). The degree and extent of the nasal deformity usually was corresponding and proportionate to the underlying intranasal destruction.

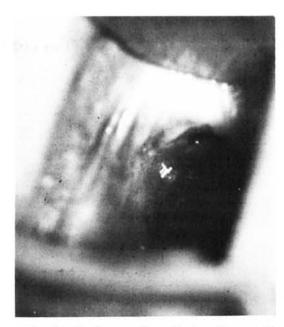
Swellings. Some of the patients with lepromatous leprosy had multiple nodules over the nose (17 cases, 6.5%). These were mainly present over the nasal alae, tip, and columellae, but others were seen on the lips, cheek and other parts of the face. The nod-



FIG. 7. Loss of the nasal septum, columellae and vestibule resulting in extensive lower nasal deformity.

ules were usually multiple and of different sizes. They were shiny, dark red or brown, firm, and well defined at the margins with disturbed sensation. The surface of some large nodules showed superficial irregular furrows giving the appearance of lobulation (Fig. 1).

Nasal vestibule. The lining of the nasal vestibule was involved in 100 cases (38,5%). It was found to have no relation to the site and degree of involvement of other intranasal structures and may even precede or follow the clinical course of the disease in the nose. Leprotic lesions were commonly seen beginning at the muco-cutaneous junction as small raised nodules that eventually ulcerated, acquired a covering of thin yellowish crusts and later healed by fibrosis. The lesions began posteriorly in the vestibule, extended anteriorly on the superior wall, and less frequently on the medial wall, but rarely on the lateral wall or the floor. The black thin hairs of the vestibule in the affected parts became discolored and brittle and fell out easily till the affected area became entirely hairless. As healing occurred by fibro-



F1G. 8. Healing nasal vestibule with smooth glazed fibrosis and loss of hairs.

sis, the healing areas appeared pale and glazed with diminished or absent sense of pain and touch (Fig. 8). Fibrosis of the vestibule alone rarely led to nasal deformity but when it was associated with lesions of the alae, columellae or the septum, the associated deformity was common and manifest. In a few instances, the cicatrizing fibrous tissue was so excessive that unilateral or



FIG. 9. Stenosed healing nasal vestibule by the effect of contracting fibrous tissue.

bilateral stenosis of the anterior nares was seen (Fig. 9). In a single case, a complete left nasal atresia was found.

Nasal cavity. Some characteristic findings were frequently observed in the nasal cavity and can be outlined as follows. In early cases the whole nasal cavity was apparently normal in width, shape and general appearance, the only abnormality being pallor and thickening of the mucous membrane of the septum and turbinates, with a moist appearance and increased mucoid discharge. The discharge was thin, translucent and tenacious and was commonly seen as thin threads on the anterior lower parts of the nasal cavity. The pain and touch sensations were occasionally diminished but the olfactory sense was always intact. All these patients complained of nasal obstruction and antenasal discharge (Fig. 10).

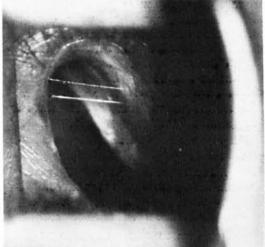


FIG. 10. Early nasal leprosy in the chronic catarrhal stage.

In all advanced cases, the common picture was that of ulceration, discharge and bleeding with obstruction of the nasal cavity by the accumulating thick purulent discharge, clotted blood and dry crusts. This appearance affected mainly the anterior lower portion of the nasal cavity. The more widespread the lesions, the more the amount of discharge and crusts and the more the degree of nasal obstruction.

Two types of late cases were seen. There were those in whom the disease was arrested in its earlier stages by long-term medical treatment. In such cases the nasal cavity had nearly the normal shape and dimensions, apart from small areas representing the sites of healed ulcers. These were in the form of pale, firm patches with absent sensation.

Secondly, there were those where the disease was so advanced that the nose was markedly deformed with extensive destruction of the intranasal structures leading to a picture comparable to dormant atrophic rhinitis.

In the later stages of the disease, in contrast to other granulomatous diseases, the healed nose tended to have a wider nasal cavity, with fibrosed retracted lateral walls. In such cases, the major parts of the septum and turbinates were absent, giving a wider nasal cavity. In only a few cases did the cicatrizing tissues and the collapsing nasal dorsum reduce the size and dimension of the cavity.

Nasal septum. Different lesions were frequently seen depending on the stage and type of lesions.

Superficial scattered ulcers on one or both sides of the anterior inferior cartilaginous septum, involving the mucosa (Fig. 11), were

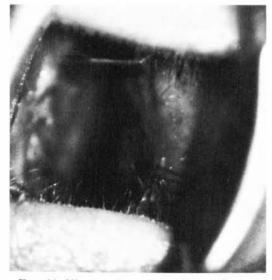


FIG. 11. Ulcers and crusts on the nasal septum in the granulo-ulcerative stage.

seen in early cases (124 cases, 47.7%). Ulcers were usually single, rarely multiple, always superficial, well defined, painless and covered by blood clots or thin crusts. The surrounding mucosa was congested and apparently healthy.

Healed ulcers were usually met with in

inactive cases. They were observed at the same anterior cartilaginous part of the septum, and appeared as pale, thin, dry and anesthetic patches and occasionally were covered by thin scaly crusts (53 cases, 20.4%).

Perforation of the nasal septum, mainly in the anterior cartilaginous part (Fig. 12), was

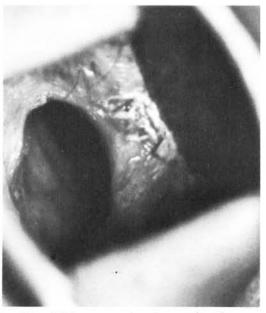


FIG. 12. Large nasal septum perforation.

the commonly encountered finding. This was seen in a larger group of patients (88 cases, 34%), and were always single. They were usually small having diameters of a few millimeters, but in more advanced cases, larger dimensions were seen. In a few cases, the bony septum was also partially destroyed and in a single case the whole septum except its superior olfactory part was completely lost (Figs. 7 and 13).

In the active cases, the edges of the perforation were ulcerating, bleeding and usually covered by crusts. In the healed, controlled cases, the septal perforation was arrested, and the fibrous tissue covered the raw edges and hence neither bleeding nor crusting was encountered.

Inferior turbinate prominence. In many cases, the inferior turbinate was free of lesions and healthy (68 cases, 26%), but in the majority (192 cases, 74%), there were one or more types of lesions related to the degree and state of affection.

In the very early cases, this turbinate ap-

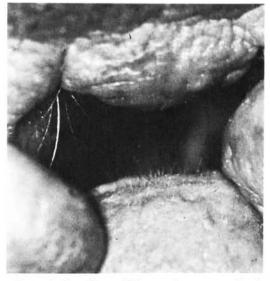


FIG. 13. Total loss of the nasal septum and columellae.

peared pale, resembling most other intranasal structures, and was covered by thin mucoid discharge. In the majority of the affected patients (128 cases, 49%), the inferior turbinates were smaller, paler and firm with evidence of sensory loss. This affected mainly the anterior half of the turbinate while the posterior part was usually healthy (Fig. 9).

Ulceration of the turbinate, mainly on its anterior part, was rarely met with. It occasionally extended backwards over the posterior half. Ulcers were either superficial or deep, involving the submucosa and bone. In all these cases there was excessive discharge, crusts and occasional bleeding.

Atrophy of the turbinate, partial or complete, was also met with, again frequently affecting the anterior rather than the posterior parts and the right side more than the left. There were 20 cases with bilateral inferior turbinate atrophy and another 28 cases with only right inferior turbinate atrophy.

Middle turbinate prominence. There were 148 cases (56.9%) with varying degrees of middle turbinate affection, denoting a less frequent involvement than seen on the inferior turbinate. Most of the lesions were seen on the anterior half. In early cases the lesions were pale and covered by thick mucus. More advanced instances showed ulcers and thick crusts while in healing cases the anterior end was small, firm and more pale with diminished sensations. In very advanced cases, there was partial or complete atrophy of the turbinate and the residual posterior parts showed firm fibrosis. In 16 cases this turbinate was completely absent.

Nasal floor. The most anterior part was less frequently affected, the middle part was found to have the same clinical picture as that of the septum and inferior turbinate, while the posterior part of the floor was rarely involved.

Olfactory area. This is the area occupying the mucosa of the upper third of the septum, the superior turbinate and the cribriform plate (¹²). Clinical examination did not reveal any lesion in this area. Even in cases with marked septal destruction and absence of inferior and middle turbinates, the upper third of the septum was found healthy and intact.

Examination for the sense of olfaction was assessed by the history and the ability to differentiate the smell of common odors. Only 13 cases with diminished or absent sense of smell were noted but, after cleansing their noses, we were left with only 3 cases with permanent anosmia (1.2%).

DISCUSSION

The nose was found to be involved in as many as 97% of leprosy patients, illustrating the marked tendency of the disease to affect this organ. The remaining 3% had healthy noses without any significant history of nasal signs or symptoms despite their long-standing affection by the disease. This gives support to Lagoudaky's (9, 10) postulation that the nose is not essentially the channel of contraction of the disease nor the first organ to suffer. In a recent study Beretti et al (1), of a series of 210 patients, found that 80% had upper respiratory lesions which diminished to 59.4% in the treated cases. The disease was generally contracted in childhood and youth. The age of onset and the duration of the disease showed no relation to the degree and extent of nasal affection, but the clinical type of the disease had an influence on the clinical picture of nasal involvement. Nasal lesions were of earlier onset and of shorter course in the lepromatous type and of longer course in the tuberculoid type while the dimorphous type assumed an intermediate position. Loss of thermal and touch sensations of the nasal mucosa occurred earlier in the tuberculoid than in the

lepromatous type, while loss of pain sensation was late in both types. Loss of the sense of smell was very rarely encountered.

The nasal mucosa was always the primary site of affection within the nose. Other nasal structures seemed to be involved later as the disease gradually progressed. The degree and the sequence of events in the intranasal involvement was found to follow a constant rule, which is reported now for the first time. This progression consisting of the constant antero-inferior nasal involvement, the less frequent posterior affection and the rare superior lesions, can perhaps be explained on the basis of the variability of temperature in the various parts of the nasal cavity. As the inspiratory air current passes through the nasal cavity, it becomes gradually warmer and when it reaches the posterior parts it has acquired a higher temperature. It can be stated that the inspired air practically never reaches the level of the olfactory area (12). Leprosy seems to affect the colder areas and its lesions become less severe as the local temperature rises. This may explain the sequence of events in the intranasal lesions and the frequent lower nasal deformity. As the incidence of nasal septum deviation is more frequent to the left (6), the right nasal cavity is frequently the wider and hence possibly cooler. Thus, we found more incidence of right nasal epistaxis, extensive leprotic lesions and more cases of right turbinate atrophy.

Brand (³) gave a similar explanation of the effect of local tissue temperature on the distribution of leprotic lesions and hence suggested a possible explanation for the absence of pulmonary parenchymal lesions in leprosy.

As the main clinical findings were tissue destruction, necrosis and excessive crust formation, the end result was a wide nasal cavity characteristic of secondary atrophic rhinitis and very rarely it ended with stenosis or atresia of the anterior nasal openings or cavities.

The collapse and deformity of the nose was due to the contracting fibrous tissue and also to the destruction of the supporting cartilaginous framework by direct leprous infection. In very few cases the bony nasal skeleton was also partially affected but it was very rarely collapsed.

Regarding the clinical classification of the

different forms of the disease, as reflecting involvement of the nose, we found that the modern scheme of classification as described by the Sixth International Congress of Leprosy (Madrid, 1953), in which leprosy was classified into type types and two forms, was followed by many workers (^{2, 5, 7, 8}).

Scott-Brown *et al* (11) described a method of clinical classification of nasal lesions into three stages: early with nasal catarrh, advanced with intranasal nodules, and late with skin and nerve involvement.

We find that all the previously described methods are inadequate for a detailed description of the clinical picture of nasal leprosy and have followed a modified new scheme, which gives clear and illustrative clinical information. From this the following three major, progressive stages are identified.

- 1. A chronic catarrhal stage, in which there is only pale moist mucosa with thin mucoid discharge. This was clinically apparent in 24 cases.
- 2. A granulo-ulcerative stage that was seen in cases with granulomatous nodules on the outer or inner surface of the nose, and those with ulceration of intranasal structures. These were the most frequent cases (180 cases), representing the commonest and most frequently seen clinical picture.
- Fibro-atrophic stage; seen in the very late healing cases having the final picture of nasal leprosy. There were 48 such patients, with intranasal atrophy, fibrosis and external nasal disfigurement.

SUMMARY

A detailed clinical study of leprosy of the nose is given for a large number of patients. The study covered all the clinical manifestations of early, advanced and late stages of the disease and laid stress on the incidence of the different symptoms and signs. It was possible to detect constant clinical findings regarding the usual picture of the affected nose. The rule of the different susceptibility of the intranasal tissues to the leprotic affection according to the degree of local tissue temperature was well illustrated and a modified method of clinical classification was used to facilitate the clinical staging of nasal leprosy.

RESUMEN

Se presenta un detallado estudio clínico sobre lepra de la nariz en un gran número de pacientes. El estudio cubre todas las manifestaciones clínicas de las etapas tempranas, avanzadas y tardías de la enfermedad y hace énfasis en los diferentes síntomas y signos. Fué posible encontrar características clínicas constantes con respecto al cuadro general de la nariz afectada. La norma de la variación en susceptibilidad a la enfermedad leprótica de los tejidos intranasales, de acuerdo con el grado de temperatura local del tejido, quedó bien ilustrada y se utilizó un método modificado de clasificación clínica para facilitar el enfoque clínico de la lepra nasal.

RÉSUMÉ

On a fourni une étude clinique détaillée de la lèpre du nez chez un nombre élevé de malades. Cette étude a couvert toutes les manifestations cliniques des stades précoces, avancés et tardifs de la maladie, mettant l'accent sur l'incidence des différents symptômes et des signes. Il a été possible de dégager des observations cliniques constantes en ce qui concerne l'image clinique habituelle du nez atteint par la lèpre. Le rôle d'une susceptibilité différentielle des tissus intranasaux à l'affection lépreuse, et ceci selon la température du tissu en cause, a été bien illustrée. Une méthode modifiée de classification clinique a été utilisée pour faciliter la gradation clinique de la lèpre nasale.

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