SCLEROMA IN DELHI AREA*

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Rhinoscleroma which is more recently termed as Scleroma was first described by Von Hebra in 1870. It is a chronic granulomatous condition afflicting the nose and the upper respiratory tract. This may or may not be associated with the concomitent involvement of the skin around the nose and the upper lip. This may result into unsightly, ugly and sometimes mutilating deformity of face. The exact incidence of the involvement of the skin has however not been worked out so far. The granulomatous involvement of the skin has been reported even at sites far away from the region of the nose and the mouth by Somani and Mehta (1964).

Although Von Frisch in 1882 was the first to have isolated the causative organism, the Frisch bacillus (Gram negative rods), yet the aetiogical role of this organism is very much mooted. However, with the experimental production of this disease in animals by Stephen and Smith (1961) the causative role of this organism is now on more sure footing.

The disease affects the poor and the unhygienic population who live under grossly dirty conditions. It is a disease of low contagion and occurs in endemic form in Poland, Hungry, South West Russia, Central America, Indonesia and Egypt. Stray cases however have been reported from all over the world.

Wahiet al (1958) had published a series of 85 cases from India (Lucknow, Indore and Jaipur). Kakar et al (1964–65) published a series of 20 cases from Delhi. Subsequently Behi and Bedi (1966) have reported two cases with the involvement of skin. Bedi (1967) have also reported a case of Rhinoscleroma with the unusual involvement of the skin and mucous membrane and the perforation of the palate. Kakar and Sood (1967) have reported rhinoscleroma "An unusual presentation". All this goes to show that the disease is not uncommon in Delhi area.

Cases of this disease are liable to present as diagnostic problem in the Department of E. N. T., Dermatology and sometimes in the Department of Surgery. It was with this aim in view that a collaborated study was undertaken so as to understand the variegated clinical patterns of this disease.

This series comprises a study of 30 cases seen in the Irwin Hospital during the last 4 years.

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AGE INCIDENCE

CHART I

Showing the relationship of age with the incidence of the Rhinoscleroma.

Age	No. of Cases	Percentage.
Below 20 years	2	6.6%
21–40 years	17	56.8%
41–50 years	5	16.6%
Above 51 years	6	20%

The disease occurs more commonly in young adults. The highest incidence is seen between the age 20–40 years, though a fairly good number of cases are seen in the elderly age group. The age range reported earlier is 8–68 years by Guevedo (1949) and 10–70 years by Wahi at al (1958).

CHART II

SEX INCIDENCE

Showing the relationship of sex with incidence of Rhinoscleroma.

Sex	No. of Cases	Percentage
Female	17	56.6%
Male	13	43.4%
		Ratio Female to Male–3 : 2

The sex incidence observed in our series, Female 56.6% aud male 43.4 to give ratio approx. is 3:2. Wahi et al (1958) have however found that males predominate ever females ratio being 3:2. Martin and shaw (1961) have reported male and female ratio 4:1.

CHART III

ECONOMIC STATUS

Showing relationship of economic status with the incidence of Rhinoscleroma.

Class	No of Cases	Percentage.
Rich.	0	0
Middle class	2	6.7%
Poor	28	93.3%

The disease occurs mainly (93.3%) in the poor strata of society only 2 patient belonged to middle class.

CHART IV

OCCUPATION

Showing the relationship of occupation with the incidence of Rhinoscleroma.

Occupation	No. of cases	Percentage
House wife	6	20%
Clerk	2	6.7%
Labourer	10	38.3%
Farmer	12	40%

The majority of the patients belonged our to labour class comprising farmers, labourers and the housewives.

These observations lead a strong support to the view that this is a disease of low socio economic status who live in dirty unhygienic home surroundings.

CHART V

INITIAL SYMPTOM

Symptoms	No. of cases	Percentage.
Nasal obstruction	16	53.3%
Skin involvement (Lip and Nose)	7	23.3%
Hoarseness	3	10%
Epiphora	2	6.6%
Epistaxis and Nasal discharge	1	3.3%
Regurgitation	1	3.3%

This chart depicts the initial presenting symptoms. The chief presenting symptom is usually the nasal obstruction 53.3% the next being the involvement of the skin (23.3%) and thereafter various other symptoms as are outlined above.

It is however worth pointing out at this stage that the educational and the social status of these patients is so low that the earlier symptoms are usually missed and they often land up in late stage when there is often gross deformity.

CHART VI

REGION INVOLVEMENT

Symptoms pertaining to	No. of cases	Percentage.
Nose	30	100%
Skin	7	23.3%
Larynx	6	20%
Palate and uvala	4	13.3%
Oro-nasopharyux	2	6.6%
Lachaymal sac	2	6.6%
Maxilla	1	3.3%
Orbit	1	3.3%

This chart shows the relative incidence of the various regions affected so as to show the exact involvement of the mucous membrane and the skin.

Nose is involved in all cases without exception thereby the importance of commonly labelling it as Rhinoscleroma.

Next is the involvement of skin (23.3%) thereby underlying the importance from dermatological aspect. Even the involvemet of skin far away from the nose is also reported Somani and Mehta (1964). There can also be involvement of the orbit, Mortada (1963); Kakar and Sood (1967).

CLINICAL CASES

It is worthwhile to mention the salient features of a few typical cases so as to demonstrate morphological features of the skin and mucous membrane involvement in scleroma which are of great interest from dermatological aspect.

Case No. 1. (Photograph 1). 50 years female showing characteristic hard induration of the skin of the nose and the upper lip for the last 2 years with concomitant involvement of the mucous memberane of the nose and the palate. Even with pressre the skin did not yield. Biopsy showed the presence of plasma cells, Mikulicz cells and Russel bodies.

Case No. II. (Photograph 2).—47 years female patient showing the induration of the nose and upper lip as well as the tumification of the skin for the last 3 years. On examination of the mucous membrane of the nose and the palate, typical granulomatous involvement was detected. Histopathology showed well defined picture of Rhinoscleroma.

Case No. III (Photograph 3.)—52 years female showing the big tumour formation in the region of nose and upper lip. There is considerable involvement of the nostrils and almost complete blockage, examination by posterior rhinoscopy however confirmed the involvement of the mucous membrane of the nose. Biopsy showed well developed pathology.

Case No. IV (Photograph 4).—45 years Male patient showing the typical mass formation and complete blockage of the nostrils. Patient had been suffering for about 4 years. Biopsy confirmed the diagnosis. These patients usually ignore the earlier symptoms and seek medical advice only when the disfigurement of face occurs and blockage of the nostrils is marked.

Case No. V (Photograph 5).—32 years male patient who was having the involvement of the nostrils and the granulomatous involvement of the skin of the nose and the upper lip for the last 5 years.

Patient was having complete blockage of both the nostrils for the last two years. He belonged to very backward strata of society and the hygienic standard was extremely poor. Biopsy was typical.

Case No. VI (Photograph 6).—48 years male patient who was having the involvement of the mucous membrane of the nose, gums and the palate for the last 2

years. Apart from the presence of extensive granulomatous infiltration, there was perforation of the soft palate. Biopsy confirmed the diagnosis of scleroma.

UNUSUAL CLINICAL FEATURES

Many patients with involvement of the skin around the nose and upper lip showed ulcerative lesions with crusting. There was great fragility and easy friability of the part. Some of the lesions were easily bleeding on touch.

These features which, were met with in a number of our patients have not been commonly seen and reported, therefore deserve special mention in proper appraisal of clinical morphology of scleroma with skin and mucous membrane affection.

HISTOPATHOLOGY

The histopathology of these patients was highly characteristic which showed the presence of plasma cells, Mikulicz cells and Russel bodies in most of the cases. However in later stage fibrosis was fairly well marked feature in addition to the above picture. It is However being contemplated to try experimental production of lesions and study the extent and nature of skin involvement on histopathological studies, which may throw further light on pathogenesis of this condition.

However if due consideration is given to the natural history of the disease, detailed examination of the nose, throat and the upper respiratory tract, the diagnosis can be made on clinical grounds with a reasonable degree of certainty.

This can be further confirmed by tissue smear and biopsy and culture examination as well as experimental studies.

DISCUSSION

Study of 30 cases in a short span of 4 years from Delhi area goes to prove that the disease is not uncommon in this part of the country. The clinical cases have been met with at various stages of involvement from the early stage of hardening or induration of the skin to late manifestations like gross multilation and destruction of the part affected. Apart from the involvement of the mucous membrane of the nose which is present in almost all the cases (100%) the skin is involved in 23.3% of all the cases. It is our desire to focus attention on this aspect of the disease which has hither to not received adequate notice. It is important to note that the involvement of the skin may be first and the presenting symptom of the patient–who may come with the involvement of the upper lip and the skin around the nose and the symptomless involvement of the mucous membrane of the nose may be detected subsequently on examination. Unless and until one is familiar with the clinical morphological features of this disease and diagnosis is liable to be missed and patient may shuttle from one department to another.

However with more knowledge now available on the possible aetiology of the disease, Stephen and Smith (1961) and the report of granulomatous involvement of the skin even at sites far away from the nose Somani and Mehta–(1964); it is all the more essential for Dermatologists to know all the facts of this disease otherwise it is likely to be misdiagnosed.

It is not the purpose of this paper to discuss the treatment and prognosis of this disease which have already been discussed elsewhere, Kakar et al (1964-65. The treatment chiefly consists of Streptomycin, X-ray therapy and broad spectrum antibiotics. Considerable improvement can be achieved if the patient presents at early stage. In the later stages the prognosis however is unsatisfactory.

SUMMARY

Scleroma in Delhi is not uncommon. Thirty cases were studied in a span of four years. Six cases are presented. Skin was found to be involved in 23.3% of the cases. The treatment consists of streptomycin and broad spectrum antibiotics. Improvement is significant if cases are detected at early stage of the disease.

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