CLINICAL MANIFESTATIONS OF RHINO SCLEROMA

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Von Habra (1870) on the basis of predominant sclerosing lesions in nose named the lesion "Rhinoscleroma"; later it was recognized that there was wide-spread involvement of the upper respiratory tract. At the International Conference of Otolaryngology in Madrid (1932) it was decided to name the disease "Scleroma" - The disease is uncommon. Wahi & Misra (1959) in India for instance collected only 85 cases over a 5 year period. Since then they have 93 more cases observed in the G.M. & Associated Hospitals, Lucknow, India. It is now realized that India has, perhaps, many autochthonous areas (Wahi & Misra 1963).

The disease is common in males at an average age of 23.8 years; the youngest observed was 16 and the oldest 75 years. The disease has a special affinity for primitive indigenous races. Contrary to Sivak (1942) our own cases have been observed in quite dry and desert areas. In agreement with Bellinoff (1933) the disease seems to be associated with poverty and is rare amongst "white-collar workers" and is non-existent among the intellectuals. Poor hygienic conditions, rural climate and ever crowding at home predispose to it. In agreement with Szmurlo (1933), Sivak (1942), Hara (1947) and Shaw and Martin (1961) we recognize four stages. The first is a non-specific prodromal stage of atrophic rhinitis predisposing to a 2nd stage of specific atrophic rhinitis. This later proceeds to a granulomatous stage and ultimately terminates in scleromatous scarring. Our observations are based on the clinical study of 178 patients.

Table 1

SITE OF LESIONS

	Location	Present series	Behindoff	Walkowrlschs
1	Nose	94.4 %	60 %	95 %
2	Upper lip	23.3 %	—	54 %
3	Pharynx	29.7 %	16 %	67 %
4	Nasopharynx	37.1 %	28 %	—
5	Palate	25.8 %	—	20 %
6	Larynx	20.2 %	60 %	22 %
7	Tongue	2.2 %	—	5 %
8	Lacrymal sac	3.3 %	—	6 %
9	Lower lid	3.9 %	—	—
10	Gums	5.0 %	—	—
11	Trachea & Bronchi	0.5 %	9 %	6 %
12	Oesophagial	0.5 %	—	—
13	Lymph.gland.	0.5 %	—	—

The nose appeared to be the actual site of origin of scleroma as was stated earlier by Shaw & Martin (1962) and the only site of lesion in 52.8 % of the cases. Only 4.4 % presented wide nasal fossa and in contrast to ozaena they had small, greyish-white crusts and the "stench" was usually not appreciable. In addition to these a further 15.7 % of cases of scleromatous rhinitis had granulomatous and fibrotic lesions in other parts of the body. The alae nasi in several of these cases had superficial fissures covered with scaly crusts. In all cases there were concentric gimlet like narrowing of the internal naris and posterior choanae. In 2.2 % of cases a complete stenosis of the posterior choanae by a scleromatous, insensitive mass was observed. In two cases the disease presented itself as atrophic rhinitis giving rise to advanced scleromatous lesions in pharynx and larynx. Such cases had also been observed by Shaw & Martin 1961).

Fortyeight and eighteen percent of these cases showed combined granulomatous and scleromateus scarring. This produced a wide range of apparent facial deformity from minimal thickening of the alae nasi to broadening of the nasal bridge and skeletal disfiguration caused by pushing out of the cartilages and nasal bones of the antero-lateral wall of the nose. Granulomatous lesions were seen projecting out the nostrils as hard, fleshy, red, nodular, insensitive masses covered with leathery mucosa. In 30.9 % of cases there was practically complete nasal obstruction resulting in rhinolalia clausa and mouth-breathing. Advanced lesions manifested themselves with a massively deformed nose, obstructed by an almost stony hard mass with loss of columellar mobility. Ulceration was observed as a late complication in 15.1 % in this series. Two cases were noteworthy. The first with recurrent epistaxis revealed on examination small symmetrical, pinkish, insensitive, ulcerated masses at the junction of the cartilagenous and bony septum. The second patient was observed to have an irregular, firm reddish, granular mass arising from the anterior end of the inferior turbinate. On biopsy both cases revealed scleroma and responded well to treatment with aureomycin. The posterior choanae showed varying degrees of stenosis in 19.7 % of the cases. Evidence of sinusitis was found in only 10.2 % of the cases.

The upper lip was involved in 17.9 % of the cases. In earlier stages it was found to begin as a subcutaneous infiltration in continuation of the nasal lesions. Later on the lip became thick, stony, hard, immobile and hideous. Superficial fissures and frank ulceration was noticed in 6.2 % of the cases. One case showed a huge chronic ulcer of 4 month duration which involved the whole upper lip and floor of the nasal vestibule. The surface of the ulcer was granular and in the centre there was a large perforation in the lip which exposed the roots of the incisors. In addition this patient had had bouts of dyspnoea with edema of the feet for the past year. On investigations his E.S.R. was found raised, he had a reversed A-G ratio and diffuse fibrosis was revealed by roentgenograph of the lungs. The diagnosis of mid line granuloma was considered in view of these findings and because of his failure to response to steroids and the antibiotics. Histologically, however, Mickuliczs cells were shown. Another case was a lady complaining of a gradual narrowing of the mouth. Both lips were hard and scleromatous, the angles adhering together leaving only a circular hole of ½ cm in diameter. The induration extended down to the chin. She had constant dyspnoea and dysphagia.

The nasopharynx was involved in 37.1 % of our cases ranging from atrophic nasopharyngitis to advanced scleromatous lesions. At time this is resulted in subtotal obliteration of the torus with fibrosis of the openings of the Eustachian tubes. This caused tinnitus and deafness.

The oropharynx was invaded in 29.7 % of cases. Almost all cases had a scleromatous contracture of the faucial pillars and posterior wall, narrowing the pharyngeal lumen and pulling the tongue upwards. In one case the pharynx was the primary site of the disease. It had advanced to the scleromatous stage without nasal or nasopharyngeal lesion.

The palate was involved in 25.8 % of cases. The sclerosed fixed and shrunken so called Gothic soft palate, was observed in 11.7 % cases with absent uvula and obliterated retropalatine isthmus. The hard palate was infiltrated and fibrosed in 10.2 % of cases. Ulceration of the palate responsible for smarting, burning and chillis was observed in 18 % cases. These superficial ulcers perhaps result from trauma during mastication.

Laryngoscleroma was first described by Gerhardt (1876); Watkin (1921) first reported a case of scleroma larynx and trachea. Later on 3 cases of primary scleroma were presented by El. Hakeem (1954). We have observed laryngeal involvement in 20.2 % of our cases. Two of these cases had primary scleroma of the larynx. The subglottic region is the commonest and most dangerous site for the disease. Diffuse involvement of the larynx was found in 13.8 % of our cases whereas in 6.2. % the lesion was restricted to the posterior and lateral walls. Vocal cord mobility was impaired in 7.3 % of cases. Dyspnoea sufficient to demand a tracheostomy was met within about half of the cases of laryngoscleroma. Isolated involvement of the aryepiglottic folds the epiglottis and the larynx was observed in only one case each. Two cases had isolated cordal lesion, which have never previously been reported. Interarytenoid edema simulating tuberculosis was seen in 2 cases. Atrophic laryngitis was the earliest clinical evidence of subglottic involvement. Later cases showed granulomatous infiltration with pinkish nodules and scarring with stenosis.

Cunning (1947) and Dixon (1947) described tracheal extension from subglottic lesion limited to the upper 5 rings. Morrison (1938) described simultaneous nasopharyngeal and tracheal lesions without laryngeal involvement. Clerf (1942) and Lucas and Negus (1942) have reported scleroma of the lower respiratory passages. During tracheostomy we have observed tracheal extension of laryngoscleroma in 6 cases. The lesion was localized to the posterior and lateral wall in 5 cases and in one case produced a dense annular diaphragm, opposite the 5th tracheal ring. A 70 years old patient consulted us because of bouts of severe dyspnoea occurring during the past year. He had noisy respiration after exertion. He had atrophic rhinitis and the larynx was normal. The roentgenograph was reported to reveal evidence of Rt. lower lobe bronchi-ectasis. A diagnostic bronchoscopy showed a granulomatous mass near the carina partially blocking the right main bronchus: a biopsy in this case was positive for scleroma.

Hara (1951) and Shaw and Martin (1961) described patients dying of meningitis from orbital scleroma. We have observed involvement of lacrhymal sac in 3.3. % of our cases and in two patients the involvement was bilateral and was associated with lacrimal fistulae.Sercer (1938) also reported such cases

and believed that extension occurred via the nasolacrimal duct or its lymphatics. In 5 cases the lower lid was infiltrated with production of ectropion. A note worthy patient consulted us with a history of gradually increasing bilateral swelling of the lids. Two years later she developed nasal obstruction and after a further two and a half she was observed to have advanced scleromatous lesions in the nose. The biopsies from both the nose and the lids were positive for scleroma. This case apparently had primary involvement of lid with later extension to nose via the nasolacrimal duct or its lymphatics. Szmurlo (1933) described orbital involvement as a rare secondary site of the disease but primary involvement of the lids and lacrimal sac has not been previously described.

Granulomatous lesions and ulceration of the gums was seen in 2.2 % of our cases. In contradiction to Roland (1961) we have not observed bony destruction of the alveolus.

The tongue was observed to be invaded in 6.2 % of our cases. In 3.9 % of our cases invasion of the tongue margin in continuation with palatoglossal folds was responsible for extra-articular trismus. In two cases the tongue was infiltrated and adherent to the post pharyngeal wall, leaving a passage less than one fourth of the normal size. In another patient the tongue was retracted upward and the scleromatous lesion of tongue, pillars and palate resulted in a narrowing of the palatoglossal isthmus to 1 cm in diameter causing dysphagia and dyspnoea.

Enlarged cervical lymph nodes were observed in 42.7 % of our cases, Szmurlo (1933) believed this is due to secondary infection. Sercer (1925) was able to culture the scleroma bacillus from the gland extract. We observed a case of scleroma of the pharynx having a single large lymphatic metastasis in the supraclavicular region which histologically revealed the presence of scleroma.

Deafness associated with tinnitus was found in 8.3 % of our cases. Ear discharge secondary to Eustachian tube obstruction and chronic suppuration of the middle ear was observed in 3.8 % of our cases. Conductive deafness was found in 6.6 % of cases and perceptive hearing loss in one case, perhaps due to streptomycin toxicity.

Remote skin manifestations mentioned by Somani (1964) have not been observed in our present study.

SUMMARY

Scleroma usually starts in the nose and may later spread to the nasopharynx, palate, pharynx, tongue, gums, lips, larynx, trachea and oesophagus. Four recognisable clinical stages in the natural history of the disease are: 1. the prodromal stage of nonspecific atrophic rhinitis; 2. the stage of specific atrophic rhinitis; 3. the granulomatous; and 4. the scleromatous stages. Metastasis to the cervical nodes is considered to be an evidence of lymphatic dissemination. Cases occur with primary lesions in the pharynx and larynx without any involvement of the nose. Scleroma may also start in the eyelids. Ulceration may be the first evidence of the disease. Lesions have been observed in two separate locations without any continuity. In advanced cases ugly facial deformity and disfigurement may occur.

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