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Scleroma of the upper respiratory passages: a CT study

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Abstract

Scleroma is a chronic specific granulomatous disease endemic in Egypt. The nasal cavities were all affected in the 37 patients examined. The paranasal sinuses and nasopharynx were involved in 17 patients, while the larynx and upper trachea were affected in five patients, four of whom were females. Twenty-three patients were in the granulomatous stage; the rest of the patients were in the atrophic and fibrotic stages.

The main nasal and nasopharyngeal CT findings were soft tissue masses of variable sizes. The lesions were characteristically homogeneous, non-enhancing and had distinct edge definition; adjacent fascial planes were not invaded. The subglottic area was involved in laryngeal and tracheal scleroma. The lesions were mainly in the form of concentric irregular narrowing of the airway. In the trachea, crypt-like irregularities were diagnostic of scleroma. Intra-orbital, intra-cranial and infratemporal parapharyngeal scleromatous masses were detected in one patient.

Introduction

Scleroma is a chronic specific granulomatous disease showing an affinity for the mucosa of the upper respiratory tract (Shehata *et al.*, 1975).

In 1870, the disease was first described by Hebra, and in 1876, Von Mikulicz described the histology and characteristic foam cells which bear his name. In 1882, Von Frisch discovered the *Bacillus scleromatis* (Hoffman and Harkin, 1973). Hebra described the disease as Rhinoscleroma. In 1932, the International Congress of Otorhinolaryngology changed the name to Scleroma emphasizing that the disease attacks not only the nose but also the pharynx and other parts of the respiratory system (Sinha *et al.*, 1969).

Scleroma was found to arise at a junction line where two types of epithelium join. It starts at the line where the stratified squamous epithelium of the vestibule joins with the columnar ciliated respiratory epithelium of the nose; pharyngoscleroma arises at the transition line between the respiratory epithelium and the stratified squamous epithelium of the pharynx. Laryngoscleroma starts below the vocal cords where the stratified

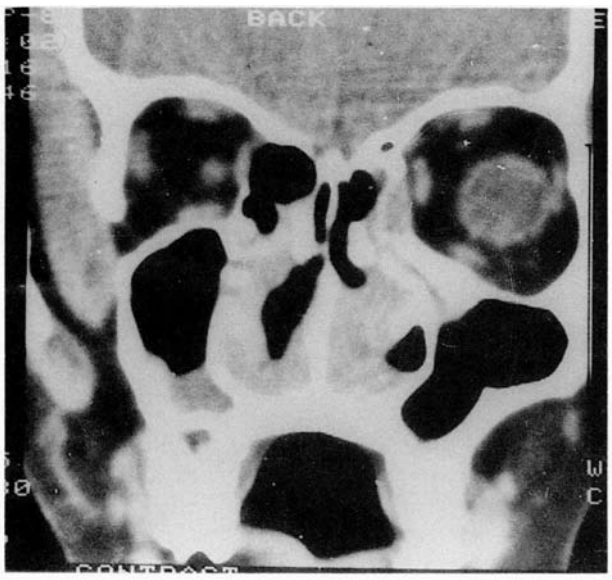

Fig. 1

Coronal CT scan: Bulky soft tissue mass filling and expanding the nasal cavity. Bony interruption of the medial wall of the left maxillary sinus with scleromatous tissue in its lower part.

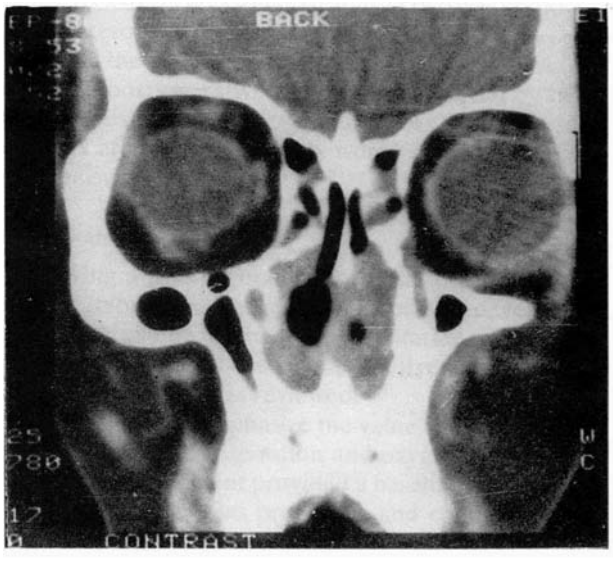

Fig. 2

Coronal CT scan marked nasal involvement by granulomatous mass with destruction of the cartilaginous part of the nasal septum. Anterior ethmoidal air cells are partially involved.

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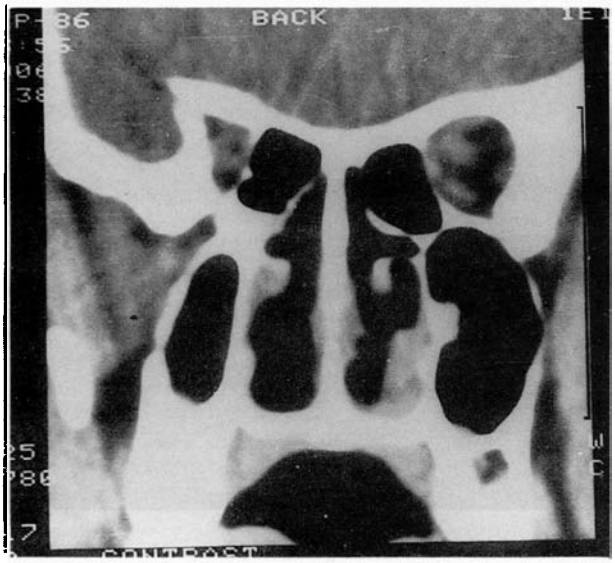

FIG. 3

Coronal CT scan: Nasal scleroma with atrophy of the middle turbinates and totally eroded inferior ones.

squamous epithelium of the cords merges with the columnar ciliated epithelium in the subglottic area (Sinha *et al.*, 1969). Unusual sites include the Eustachian tube, middle ear cavity, nasolacrimal duct and intracranial extension.

The clinical picture of the disease shows four stages: the catarrhal stage, characterized by prolonged purulent rhinorrhea; the atrophic stage with mucosal changes and crust formation; the granulomatous stage in which small firm granulomatous nodules may be found anywhere in the nose, pharynx, larynx or bronchi; the sclerotic or fibrotic stage where dense fibrotic narrowing takes place (Badrawy *et al.*, 1974; Taha *et al.*, 1975).

The aim of this work is to study the CT scan findings in cases of scleroma and to identify the degree of extension of the disease.

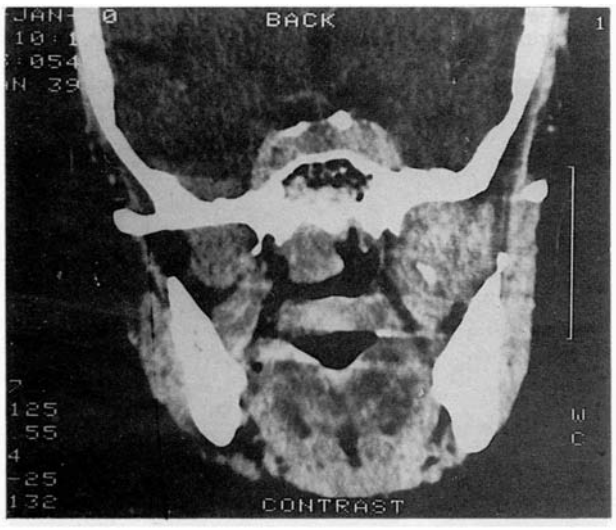

FIG. 5

Irregular nodular thickening of the mucosal lining of floor of sphenoid sinus—nasopharyngeal projecting scleromatous mass with right parapharyngeal involvement.

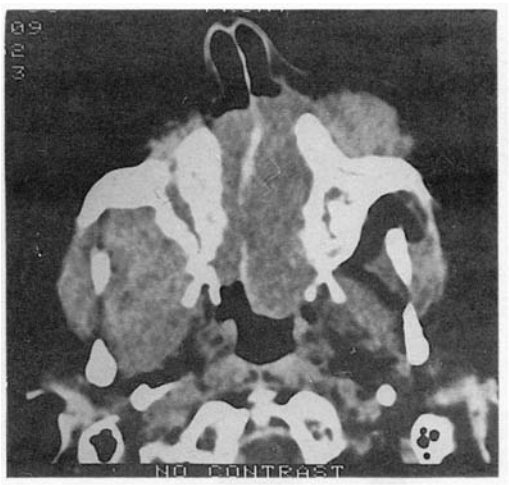

FIG. 4

Total bilateral nasal masses—extending into left side of nasopharynx, opacification of the maxillary antra with reactively thickened sclerozed bony walls.

Material and methods

This study was carried out on 37 biopsied and proven scleroma patients, attending the ENT Department of Alexandria Main University Hospital and referred to the Radiology Department for CT scanning.

Every patient was subjected to:

(1) a complete clinical otorhinolaryngological examination.

(2) biopsy and pathological examination from the nasal lesions.

(3) CT scan of the nose, paranasal sinuses, pharynx, larynx and trachea using the Somatom DR2 scanner 256 × 256 matrix axial. Scans of 4 mm with contiguous cuts were obtained, and coronal scans for the nose, paranasal sinuses and nasopharynx. Scans were performed before and following IV contrast injection of urografin 76 per cent (0.1 g sodium diatrizoate and 0.66 g meglumine diatrozoate per 1 ml urografin). The administered dose was two ampoules 40 ml.

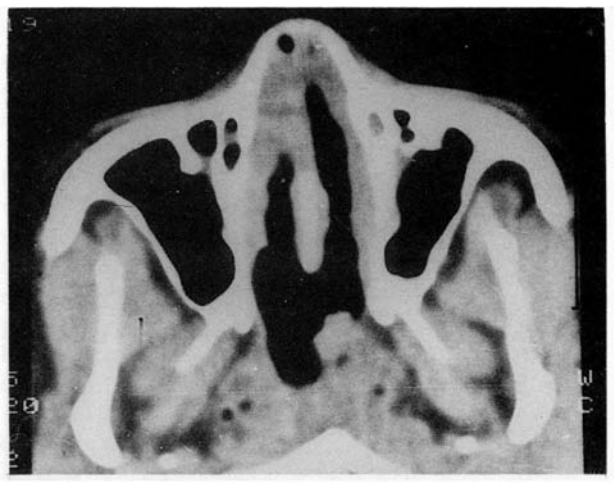

FIG. 6

Axial CT scan: Homogeneous non-enhancing mass occupies the left side of the nasopharynx encroaching on the left Eustachian tube orifice.

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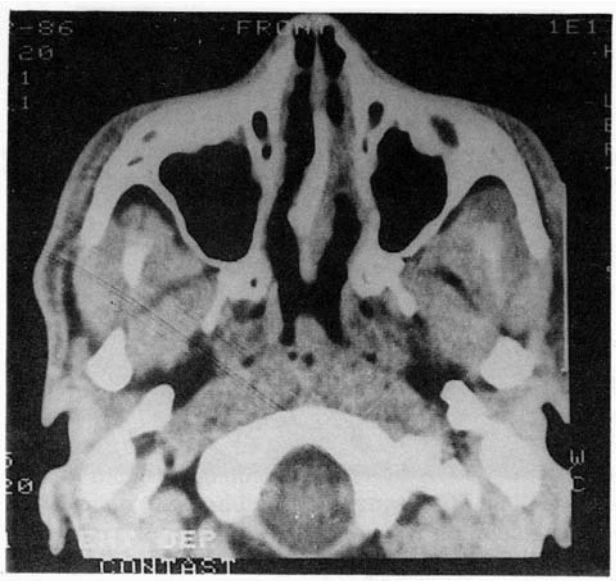

FIG. 7

Axial CT scan: Atrophic nasal changes. A central nodule is projecting anteriorly from the posterior pharyngeal wall.

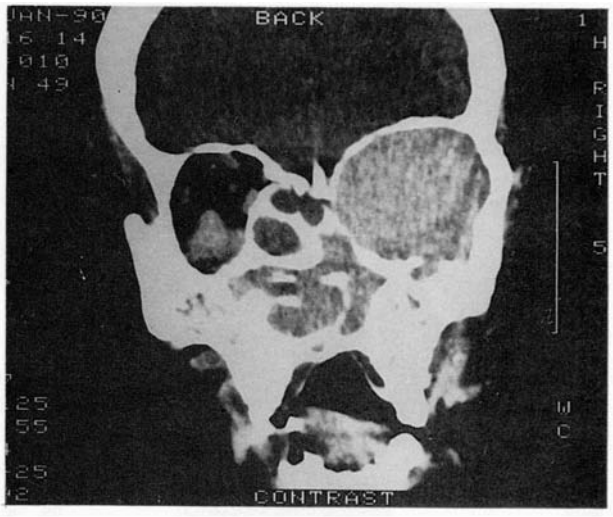

FIG. 8

Coronal CT scan: Bilateral nasal masses, bilateral ethmoidal involvement partly by obstruction and partly occupied by granulomatous mass-extensive; right orbital involvement and early left orbital mass.

Results

1. *Age*

The 37 patients examined ranged in age between 13 and 60 years, with highest incidence in the second and third decades of life (85 per cent of cases).

2. *Sex*

In scleroma of the lower part of the upper respiratory tract 'larynx and trachea', females were more commonly affected than males with a female to male ratio of 4:1, while males presented a higher incidence in scleroma of the pharynx and paranasal sinuses (male to female ratio 2:1).

3. Forty per cent of our cases were living in Alexandria, while 60 per cent came from rural areas. A positive family history was found in three of our cases.

4. Patients were classified according to the clinical stages of the disease manifested in the nose into three stages: 23 were in the granulomatous stage, nine in the atrophic stage, while the fibrotic stage was detected in five patients. None of our cases presented in the catarrhal stage.

CT Findings

1. The nose was affected in all 37 patients examined. Nasal lesions varied from small mucosal nodules in three patients to bulky soft tissue masses expanding the nasal cavity. The lesions were bilateral causing narrowing of the nasal air way reaching to complete obstruction in four cases where masses were bridging between the lateral nasal walls and septum (Figs. 1 & 4). Lesions were characteristically homogeneous, non-enhancing and with distinct edge definition.

The cartilaginous part of the nasal septum was destroyed in one patient (Fig. 2). The turbinates were always affected; one or more of either or both sides showed destruction, deformity, atrophy or complete absence of both bony and soft tissue components of nasal turbinates (Fig. 3).

2. Paranasal sinuses were affected in 17 patients. Maxil-

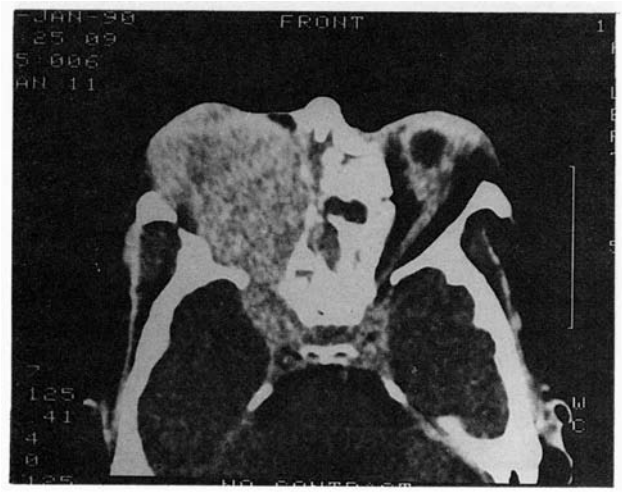

FIG. 9

Axial CT scan: Same patient right intracranial extension through the widened right superior orbital fissure.

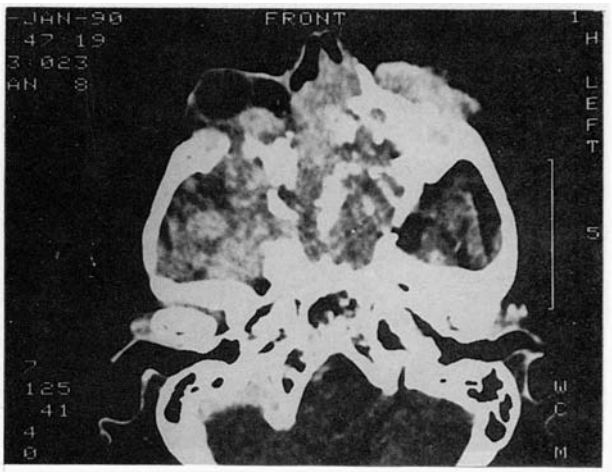

FIG. 10

Axial CT scan at a lower level. Same patient with marked right infra-temporal involvement. Bones are interrupted and sclerotic.

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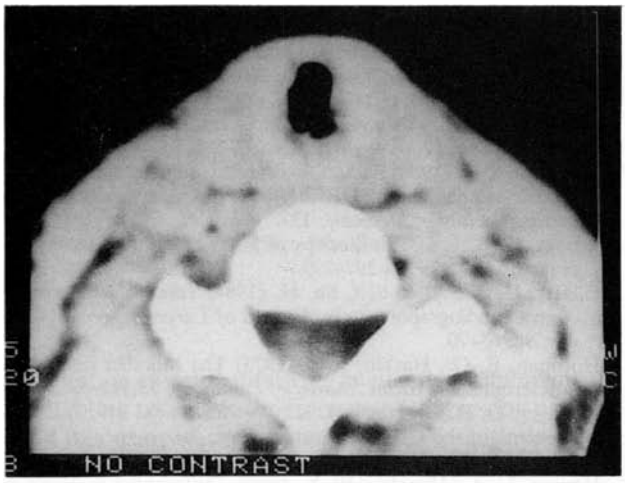

FIG. 11

Axial CT scan: Irregular narrowing of the subglottic airway with posterior web formation.

FIG. 12

Axial CT scan: Concentric narrowing of upper tracheal airway with small crypt or sinus like irregularities characteristic of tracheal scleroma.

lary sinus involvement was in the form of total opacification with thickened and sclerosed bony walls in one patient (Fig. 4). Nodular mucosal thickening with small interruptions of the medial bony walls (Fig. 1), or diffuse mucosal masses variably occupying one or both antra in the remaining patients.

The ethmoidal sinuses were involved in 13 patients; their CT scans revealed normally aerated air cells intervening between the involved opacified cells (Figs. 4 & 5). The opaque ethmoid air cells were partly occupied by scleromatous masses while some cells were obstructed with retained secretions (Fig. 8). Granulomatous masses in the sphenoid and frontal sinuses were detected in two patients (Fig. 5).

3. Nasopharyngeal lesions were found in 17 of the patients examined, the nasopharyngeal lumen was deformed and irregularly reduced in size by projecting bulky homogeneous soft tissue masses which were non-enhancing and had distinct edges (Figs. 4–7). Hypertrophied granulomatous masses were creeping around the cartilaginous part of the Eustachian tube (Fig. 6).

4. The pterygoid plates and muscles and the para-pharyngeal spaces as well as the pterygomaxillary and the infratemporal fossae were preserved in all but one patient, where massive bilateral intra-orbital and early intra-cranial extension was detected; limiting fat and fascial planes were preserved. In this patient the scleromatous tissue was denser compared to adjacent structures (Figs. 8–10).

5. The larynx including the subglottic and upper tracheal regions was involved in five patients, four of whom were females.

Laryngeal involvement was non-specific with irregular thickening of the epiglottis and aryepiglottic folds; smooth swelling of the false cord and upper part of arytenoids were found in one patient.

In all the five patients the subglottic area was affected in the form of irregular concentric narrowing with web formation in one case (Fig. 11).

The upper tracheal involvement characteristically showed small crypt-like irregularities (Fig. 12). The lesions were always mucosal and did not infiltrate deep to the cricoid or other laryngeal or tracheal cartilages.

Discussion

Scleroma is easily diagnosed clinically in endemic areas. The problem arises whenever a patient is presenting in non-endemic areas, where differential diagnosis between scleroma and other granulomatous or neoplastic lesions arise. Using CT scanning the extent of scleroma involvement can be assessed accurately.

In Egypt, the disease mostly affects the poorer classes living under unhygenic conditions (Dogheim, 1980). Males were commonly affected by scleroma of the nose and paranasal sinuses, while the subglottic and tracheal involvement were commoner in females. Acuna (1973) found a higher scleroma female incidence.

All our scleroma patients initially presented with findings in the nasal cavity; difficulties in diagnosis arise whenever scleroma lesions are found elsewhere in the upper respiratory passages without nasal involvement.

In the nose, paranasal sinuses and nasopharynx, scleroma lesions were in the form of small mucosal nodules or bulky masses, of soft tissue attenuation value; in either condition the lesions were homogeneous, non-enhancing and had distinct edge definition. Multiplicity of lesions and nasal involvement were characteristic. Som (1985) described scleroma lesions as being minimally enhancing. Badrawy (1966) reported solitary antral involvement in one case.

Bone and cartilage involvement in scleroma is problematic; Som (1985) described destruction of the nasal vault, Shum *et al.* (1962) reported erosion in the medial wall of the maxillary antra, whilst Badrawy *et al.* (1974) said that bone can be affected by scleroma in much the same way as benign tumours, this takes the form of displacement, pressure atrophy or complete absorption of bone. Unger *et al.* (1984) found that certain common inflammatory-like diseases may also destroy bone.

In the present work expansion of the nasal bones was common with bulky nasal masses; destruction of the turbinates and septum was found in some patients. Interruption of the medial wall of the maxillary sinus was seen in one patient; reactive sclerotic osseous changes were common.

In the nasopharynx despite the projecting masses and

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deformities of the nasopharyngeal airway the pharyngobasilar fascia was not involved; the parapharyngeal spaces were preserved even in the presence of muscle involvement. These characteristic features differentiate scleroma from aggressive malignant tumours, threatening fungal infection, post-operative oedema and haemorrhage. The Eustachian tube orifices may be occluded by granulomatous tissue. Similar finding were described by Steinberg and Clarck (1983), Silver *et al.* (1983) and Hoover and Hanafee (1983).

In the present study the main brunt of laryngoscleroma affected the subglottic area in the form of concentric irregular narrowing. Posterior web formation was found in one patient; comparable findings were described by Holinger *et al.* (1977). The subglottic involvement was always associated with variable lengths of tracheal involvement in the form of narrowing and stenosis. In the trachea the concentric narrowing characteristically showed crypt-like irregularities; this confirms similar findings described in one of our previous publications using positive contrast laryngotracheogram in scleroma patients. These crypts may represent the dilated orifices of goblet cells lining the fibrotic tracheal walls; this differentiates scleroma from other causes of tracheal stenosis (Gaafar, 1983; Gaafar and Abou Seif, 1984).

In one of our patients bilateral intraorbital, mainly extra-conal masses were detected, which were sizeable on the right side. This was associated with intracranial extension through the widened superior orbital fissure to the right parasellar region. This same patient was the only one showing parapharyngeal and infratemporal soft tissue involvement of denser attenuation value. She was a female aged 60 years and she had a therapeutic course of radiation orthovoltage to the maxillary antra and nasal cavities (35 Gy in four weeks) 20 years earlier. This may explain the relatively aggressive nature of her lesion; however, in spite of the extensive involvement adjacent fascial planes, the involved structures were respected. No malignant change could be detected on repeated biopsy examination; her right eye was totally blind.

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