Sinus involvement in cases of rhinoscleroma: a cause of antibiotic resistance and early recurrence after medical treatment

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Theory
To investigate the true incidence of sinus infection in cases of rhinoscleroma, and the possible role of reinfection from the sinuses to explain recurrence and resistant to treatment in cases of scleroma.

Materials and methods
Twenty-five patients with histologically proved granular rhinoscleroma were included in this cohort prospective study.

Results and conclusion
More than one-third of the cases of rhinoscleroma in our series were associated with histologically proved sinus lesions, and these sinus-positive lesions were associated with resistance to standard medical treatment and with early recurrence.

Keywords:
chronic sinusitis, granuloma, rhinoscleroma, rifampicine

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Introduction

Rhinoscleroma is a very rare, chronic specific granulomatous disease of the upper respiratory tract that is endemic in certain areas of the world like Egypt, Mexico, and central Asia and Europe [1].

Because of the rarity of the disease, many aspects of the disease still remain unknown; among these aspects is the high tendency of the disease to recur after medical treatment and the resistance of the organism to medical treatment [2].

The disease has always been described as a nasal disease affecting the mucocutaneous region of the nose [3], but recently, some data have emerged about the possibility of sinus involvement in cases of rhinoscleroma. All these reports were based on radiological and clinical evaluation or were presented as case reports [4,5].

In this work, we study the true incidence of the involvement of the ethmoidal sinuses in rhinoscleroma on the basis of histopathological analysis of specimens in a relatively large cohort of patients and postulate a theory of reinfection from the sinuses as a cause of early recurrence of the scleroma lesions and as a cause of resistance to standard medical treatment.

Patients and methods

Twenty-five patients with histopathologically proved granular rhinoscleroma (Figs 1–3) who were candidates for laser debulking of granulations were included in this study from May 2010 until July 2011.

The following patients were excluded:

(1) Patients who received any form of medical treatment in the last year.
(2) Patients with atrophic rhinoscleroma, as detection of a clinical cure in these patients is difficult.

All patients gave their consent to participate in this study and for the surgical procedures and the study design was approved by the local ethical committee.

Patients were subjected to the following:

(1) Complete ear, nose, and throat examination including endoscopic examination of the nose and larynx.
(2) Punch biopsy from nasal lesions as well as biopsy from anterior ethmoids after limited anterior ethmoidectomy, which was done under general anesthesia at the time of laser debulking.
Biopsies were divided into two parts: the first part was sent for histopathology for confirmation and documentation of the disease (Fig. 4) and the second part was sent for bacteriological culture and sensitivity to antibiotics.
(3) Computed tomograpic scan at the time of diagnosis (Fig. 5).

All patients received medical treatment after debulking in the form of rifampicin 150 mg twice daily for 3 months; after that, another punch biopsy was taken from the nose to confirm cure (Fig. 6), and if the patient was clinically and pathologically free, treatment was stopped but if not, it was continued for another 3 months, with frequent laboratory tests for liver and kidney functions.

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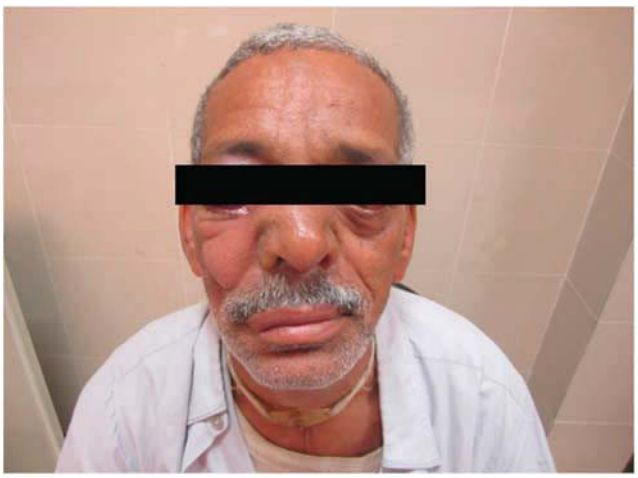
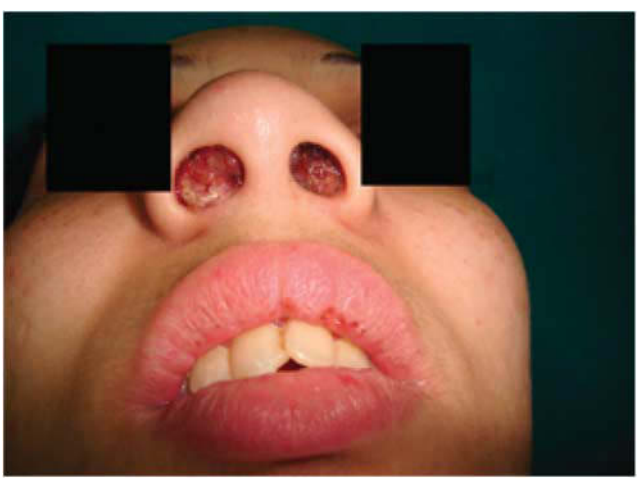
Figure 1



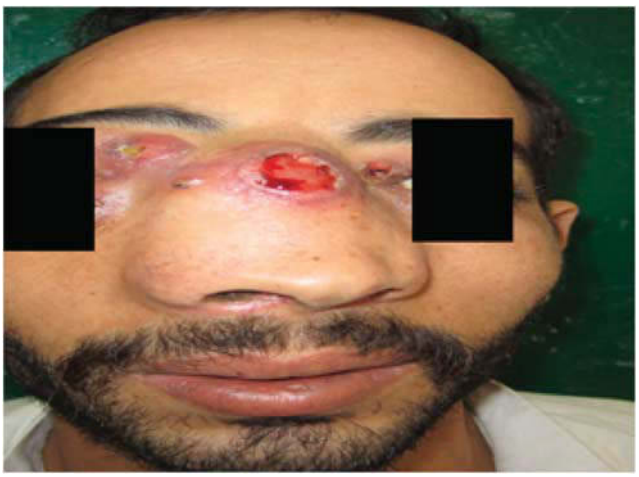
A case of granular rhinoscleroma with laryngeal involvement showing severe facial disfigurement.

Figure 2



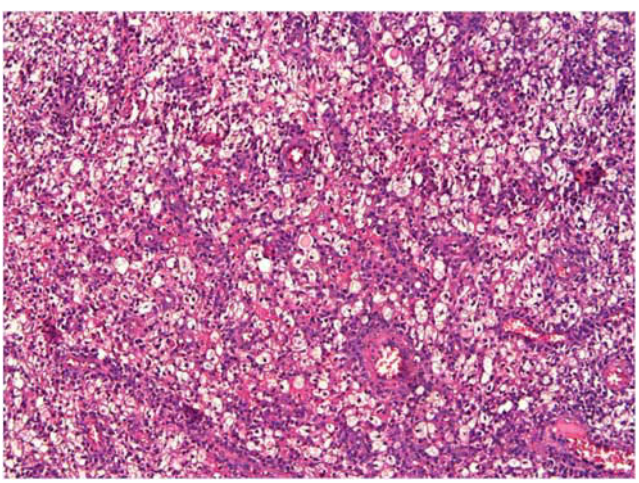
A case of granular rhinoscleroma showing totally occluding nasal granulations.

Figure 3



A case of extensive rhinoscleroma with severe facial disfigurement and facial ulceration.

Figure 4



Rinoscleroma, cellular phase, showing many Mikulicz cells with a foamy cytoplasm together with many plasma cells and a few Russel bodies, × 200.

Follow-up was conducted at 12 months after cure, and punch biopsies were taken from clinically suspicious cases.

Results

Twenty-five patients with histopathologically proved rhinoscleroma were included in this work; of these, two patients did not attend the follow-up visit after 1 year and were thus excluded from the study.

Out of these 23 patients, 14 were women (60%), whereas nine were men (40%). Twenty patients (86%) were between 18 and 49 years of age, whereas one patient was a 12-year-old boy and two patients were 51 and 56 years old, respectively.

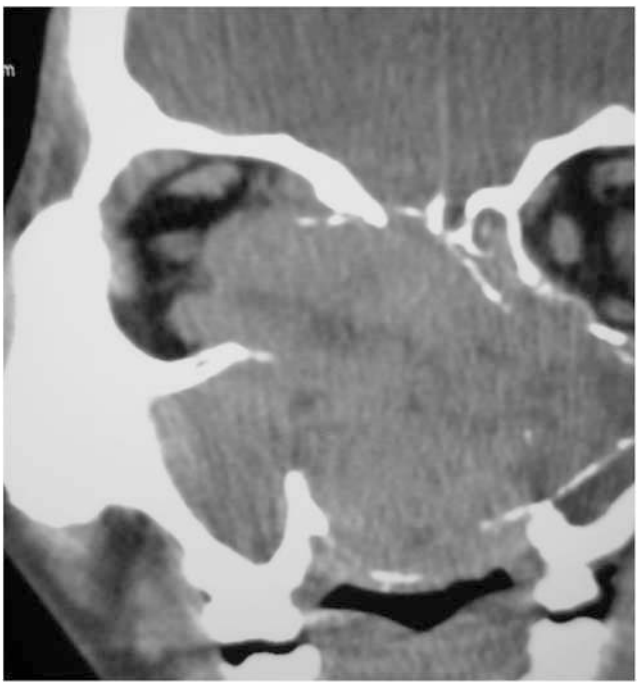
In all cases, histopathological examination was carried out to confirm the diagnosis and the classic picture of plasma cells, Mikulicz cells (large foamy macrophages containing coccobacilli), and degenerated uniform plasma cells (Russell bodies) was detected. In all 23 patients, granulomatous lesions were detected in the anterior part of the lateral nasal wall septum, whereas in 15 patients (62%), mixed atrophic and granulomatous lesions were observed in the nose.

In 12 out of our patients (52%), there were lesions in the nasopharynx and in 10 patients (43%), there were subglottic and tracheal lesions.

During anterior ethmoidectomy, there was normal ethmoidal mucosa in 20 patients (86%), whereas three patients (14%) showed variable degrees of ethmoidal granulations.

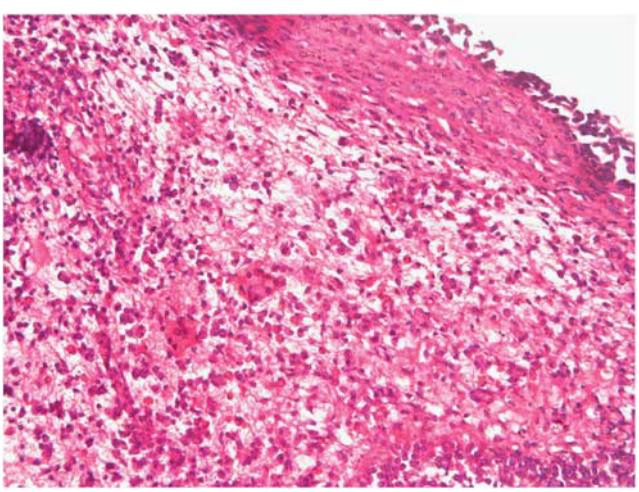
In our work, nine patients (39%) showed histologically positive lesions in the anterior ethmoids; among these,

Figure 5



Computed tomograpic scan of one of our advanced cases showing massive involvement of all sinuses.

Figure 6



Rhinoscleroma (post-treatment), showing only plasma cells within a fibrotic background; note the absence of Mikulicz cells, × 400.

three patients had ethmoidal granulations, whereas six patients showed normal sinus mucosa.

In this study, 19 patients (82%) were clinically and pathologically cured after 3 months of continued treatment, whereas four patients (18%) needed another 3-month treatment course; all four of these patients had sinus-positive lesions.

After the 1-year follow-up, seven patients (30%) showed recurrence of lesions as proved by histopathological biopsies from suspected lesions and all these patients had sinus-positive lesions.

The paucity of rhinoscleroma cases justifies the limited number of patients in our study, which did not allow a statistical study of the relation of sinus infection to the period of treatment and recurrence of cases.

Discussion

Rhinoscleroma is a very rare, chronic specific granuloma of the nose and respiratory tract that was first described by Von Hebra in the 1870s, but it was in 1882 that Von Frisch described the classic bacillus scleromatis [6].

The disease is endemic in certain areas in the world like Egypt, Mexico south East Asia, and central and Eastern Europe [1].

Owing to the rarity of the disease and the limited number of publications in the literature, many aspects of this disease are still unknown or at best unclear example.

(1) Mode of infection and transmission.
(2) Variable clinical presentations.
(3) Why it does not affect the lymphatic system like other granulomatous diseases.
(4) Presence of genetic predisposition.
(5) Lines of prevention and treatment.
(6) High incidence of bacterial resistance and high incidence of recurrence of lesions reaching 25% after 10 years [2].

Rhinoscleroma has always been described as a nasal lesion affecting the mucocutaneous junction of the nose but with computed tomography and endoscopes, scleroma has been found to infect the maxilla and ethmoids more than was typically expected [4,5].

In 1991, Abou-Seif *et al.* [7] reported sinus (maxillary and ethmoid) involvement in 37 rhinoscleroma patients in the form of tissue masses of variable sizes.

In this work, we have studied the incidence of sinus involvement in cases of rhinoscleroma and linked this to the incidence of recurrence and resistance for standard treatment in a trial to postulate a theory that reinfection from sinuses may be the cause of recurrence and resistance to the treatment of rhinoscleroma.

Analysis of our results led to the following conclusions:

(1) Our demographic data showed the disease to be slightly dominant in women compared with men, with the highest incidence in the third and fourth decades of life, which agrees with the well-known demographic distribution of the disease [2].
(2) The incidence of sinus involvement in our work was 39%, and there have been no similar studies in the literature to compare with our work, but Abou-Seif *et al*, in 1991, on the basis of a radiological study, only showed paranasal sinuses to be involved in 17 out of 37 cases [7].
(3) The incidence of recurrent cases in the 1-year follow-up after cure was 30%, which was slightly less than that obtained in the work of Gaafar and colleagues,

who reported a recurrence rate of 41% in patients after 1–3 years. All our recurrent cases were among the previously diagnosed sinus-positive cases. This strongly indicates the possibility of reinfection from sinuses as a cause of early recurrence in those cases.

(4) Sixteen percent of our group of patients were resistant to medical treatment and required a second course of treatment; again, all of these patients were among the sinus-positive cases, thus also indicating that reinfection from sinus lesions may be the cause of resistance to antibiotic treatment as it is well known that sinus perfusion is much less than nasal perfusion.

Conclusion

This is the first article in the literature to address the true incidence of rhinoscleroma in the sinuses on the basis of histopathological specimen examination apart from case reports.

More than one-third of the cases of nasal scleroma are associated with sinus infection of the disease and this may be associated with normal sinuses on endoscopic or clinical examination.

These sinus-positive cases are linked with early recurrence and resistance to medical treatment, highlighting the necessity to prolong treatment courses in these cases.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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