

Our Experience with Rhinolaryngoscleroma & its atypical presentations in Egypt

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Background: Respiratory scleroma is a common nasal and laryngeal granuloma in some developing countries, with increase incidence in developed countries.

Although diagnosis is not difficult in endemic areas, atypical cases represent a great challenge in diagnosis and managements.

Methods: We reviewed medical records of respiratory scleroma cases in two hospitals, and included eleven cases in whom the presentation were uncommon and atypical.

Results: We reported 11 cases of atypical presentation of rhinolaryngoscleroma; One case of isolated tracheal granulations, two cases of isolated nasopharyngeal involvement, two cases of supraglottic swellings, two cases of bilateral septal swelling, one case of bilateral maxillary sinus masses, one case of extensive scleroma with intra orbital & intracranial extension, two cases of unilateral nasal masses.

Conclusion: Scleroma should be put in differential diagnosis of any sinonasal, pharyngeal, laryngeal and even tracheal lesions.

Keywords: Rhinoscleroma, atypical presentation, Egypt.

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Introduction

Scleroma is a chronic specific inflammation of the upper respiratory tract. It is caused by gram negative diplobacilli called *Klebsiella rhino scleromatis* or *Frisch bacillus*. [1]

The disease affects upper airway mostly the nose, oropharynx, nasopharynx, larynx and trachea; less commonly the nasal sinuses and other areas of upper air way. It is endemic in some rural low socioeconomic areas in Central America, North Africa like Egypt and Sudan. It is considered as one of the differential diagnosis of upper air way granulomas, atrophic inflammation and post inflammatory scarring. [2]

The history of scleroma started about 150 years ago. It was first described as a clinical entity by Von Hebra in 1870, who called it nasal leprosy, hard nose. Six years later, Mikulicz studied the microscopic histology. During the same year, Gerhard gave the first comprehensive description of laryngoscleroma. In 1882, Von Frisch discovered the *Klebsiella* organism and suspected it to be the etiologic agent. However, it was not until 1900 that Gerber characterized the lesion as a chronic inflammatory process and described its histologic appearance. [3-5]

Young and middle aged females are more commonly affected. It is not known to be contagious or familial as a rule, but Italian study reported 15 cases in one Italian family, and Another American study reported 7 cases in a single American family. [6]

Most cases were diagnosed in endemic areas, and recently in some non-endemic areas, explained by the increased migration over the world. Typical presentations are overlapping stages; Catarrhal stage, atrophic stage (sometimes called ozaena), Granulomatous (proliferative

or nodular) and sclerotic (cicatrical or fibrotic). During the catarrhal stage there are foul smelling purulent nasal discharges and nasal obstruction; atrophy and crusting of the nasal mucosa or hyperemia and exudates in the respiratory tract mucosa. [5,7,8]

In the granulomatous stage there are epistaxis, nasal deformity, hoarseness, anosmia and anesthesia of the soft palate; bluish red and rubbery granulomatous lesions which evolves into a pale hard granulomatous mass. [5,7]

Sclerotic stage symptoms are similar to the previous stage; the granulomatous lesions are surrounded by dense fibrotic tissue. Most patients are diagnosed in the granulomatous stage, because they are more symptomatic and other organs besides the nose may be involved. [5,7]

Typically, It affects the nasal cavity and larynx as primary areas, also less common affect naso and oropharynx, and rarely affect skin, paranasal sinuses, nasolacrimal sac, skull base middle ear, oral cavity and trachea. [5,7]

At present, rhinoscleroma is a diagnostic and therapeutic challenge due to its chronic course and wide differential diagnosis like neoplastic and other granulomatous lesions, presence of other atypical presentations and atypical affected areas. [8]

Aim of Study

To describe new patterns of atypical presentations of cases with respiratory scleroma in Egypt.

Patients and Methods

This is a retrospective case series study done in two institutes; Faculty of medicine in Cairo University, and Fayoum University.

After reviewing the medical records in the two institutes during four years from 2013 to 2017, and reviewing all cases diagnosed histopathologically as scleroma, we included eleven cases with atypical presentation of respiratory scleroma. We excluded cases in whom the diagnosis were strongly expected due to typical presentation of respiratory scleroma like subglottic masses or stenosis, bilateral anterior nasal masses and nodules at nasal vestibules.

Case Scenario 1 (Isolated Tracheal Granulations)

Case One: 36 years old house wife presented with mild biphasic stridor of one month duration, and muffled voice for three months. No histories of trauma or ICU admission. Laryngoscopic examination shows sub glottis granulations. CT scan showed concentric tracheal stenosis about 4cm length at tracheal ring 3, 4, 5, and large goiter compressing trachea. Examination was done with direct laryngoscopy and bronchoscopy under general anaesthesia shows mid tracheal stenosis with reddish smooth mucosal covering polypi. Patient was scheduled for total thyroidectomy and tracheal biopsy and dilatation. His respiration improved markedly, and biopsy shows characteristics of Scleroma.

Case Scenario 2 (Supraglottic swelling) " 2 Cases"

Case Two: A 33 years old housewife, presented with hoarseness of voice and expectoration for 3 months, and recently developed mild attacks of choking and stridor for 2 weeks duration. There was no haemoptysis or dysphagia or pain. Examination shows single right sided large pedunculated reddish supraglottic polyp arises from the ventricle, with smooth covering intact mucosa with NO ulceration or mamillation, with intact mobile both vocal cords. And free subglottic area and nasal cavity fig;1. She had microlaryngoscopic surgery (MLS) with complete removal of the polyp without need for tracheostomy. Her respiration became quiet, and her voice improves, and biopsy proved scleroma.

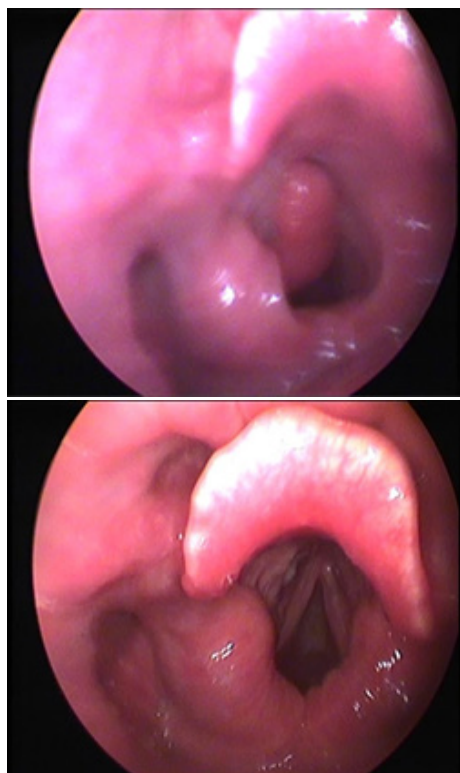


Fig 1. Preoperative left supraglottic smooth mass & post op. view.

Case Three: A 45 female patient, non smoker, presented to outpatient clinic with hoarseness of voice for 2 months, with no choking, stridor, haemoptysis, dysphagia or pain. Examination showed bilateral multiple sessile greyish glistening supraglottic polypi arises from the ventricles, with smooth covering intact mucosa. Both vocal cords were mobile. Subglottic area was free. Nasal cavity was free. She had microlaryngoscopic surgery (MLS) and biopsy from those polypi which proved to be scleroma. And she received rifampicine and improved upon.

Case Scenario 3 Isolated nasopharyngeal scleroma Two cases.

Case Four: Forty years old female patient complaining of bilateral alternating partial nasal obstruction with nasal tone of voice (rhinolalia aperta) for six months . There is no past history of nasal discharge or crustations with occasionally regurgitation of fluids from the nose.

An examination by anterior rhinoscopy was free. Nasal endoscopy shows stenotic narrowing of posterior choana with fibrous bands extending from the roof of nasopharynx to the soft palat (**Fig 2**). Oropharyngeal examination shows absent uvula. CT of paranasal sinuses shows narrow posterior choana in axial cuts. Patient was candidate for endoscopic dilatation of the choana and stenting and biopsy revealed scleroma.

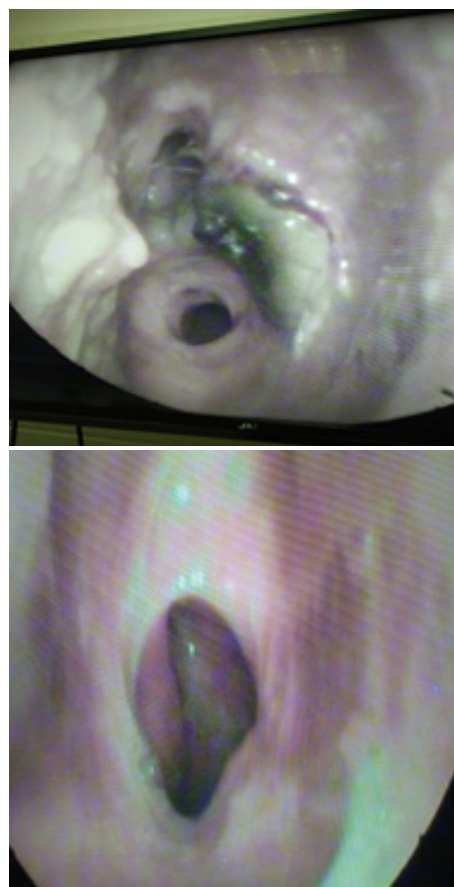


Fig 2. Endoscopic view of the narrowed choana.

Case Five: Sixty years old male patient complained bilateral nasal obstruction with nasal tone of voice (rhinolalia closa) and snoring for 3 months. Examination by anterior rhinoscopy was free. Nasal endoscopy shows large irregular reddish nasopharyngeal swelling with intact overlying mucosa of. Ear examination shows bilateral SOM. CT of paranasal sinuses shows large soft tissue irregular mass filling the nasopharynx

without bone erosion (**Fig 3**). Patient was candidate for endoscopic biopsy and debulking of the mass. It was proved to be scleroma.

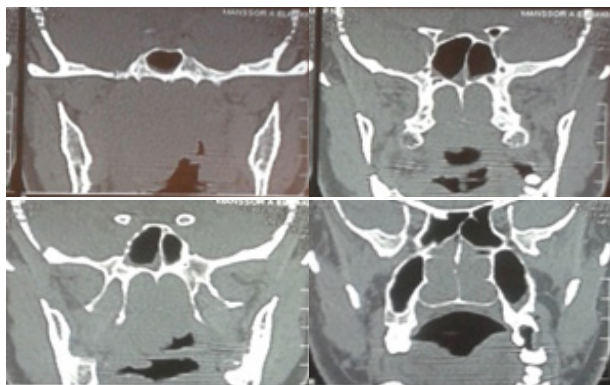


Fig 3. Pre op CT scan large nasopharyngeal opacity.

Case Scenario 4 (Bilateral Maxillary swellings)

Case Six: A 45 years old female presented with bilateral persistent greenish mucopurulent nasal discharge resistant to medical treatment for 4 months. Endoscopic examination was free apart from congested pulging left uncinete process and concha bullosa. CT scan shows bilateral homogenous maxillary opacity (**Fig 4**). Endoscopic sinus surgery was done bilateral middle meatal antrostomy. The maxilla was filled with polypi. Biopsy was surprisouly scleroma.

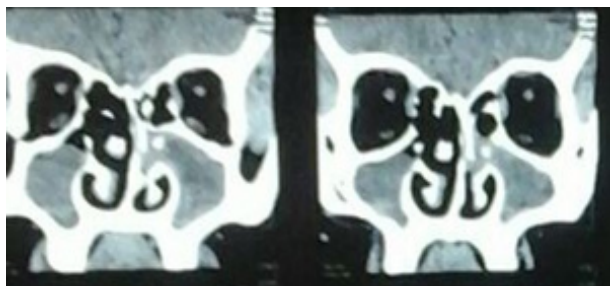


Fig 4. CT scan bilateral maxillary opacity.

Case Senario 5 Unilateral nasal mass Two cases

Case Seven: Thirty six years old female patient presented with left sided nasal obstruction and mucopurulent discharge for 2 months, , occasional epistaxis. Anterior rhinoscopy shows left sided grayish polypoidal irregular swelling. CT scan shows left sided nasal opacity with bone expansion with opacified all left paranasal sinuses fig; 5. Endoscopic excision biopsy was done, the mass was vascular and soft in consistency. Biopsy revealed features of scleroma.

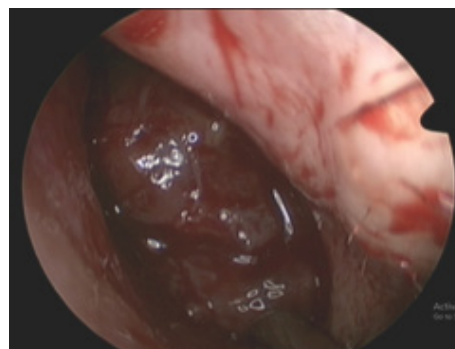


Fig 5. CT scan lt sided sinonasal opacity & Endoscopic view of left nasal mass.

Case Eight: 46 yrs old male patient presented with unilateral (Rt) nasal obstruction & discharge for 7 months. Examination showed unilateral nasal mass was detected completely filling the right nasal cavity & CT scan showed a unilateral nasal mass completely filling the Rt side of the nose with pan sinus opacity on the Rt side, biopsy was taken & proved to be Rhinoscleroma.

Case Scenario 6 Bilateral septal swellings Two cases

Case Nine: A 35 female patient presented with bilateral nasal obstruction for 2 months, associated with, occasional epistaxis. Anterior rhinoscopy shows bilateral smooth reddish swellings arises from nasal septum. CT scan showed bilateral opacity arising from the whole nasal septum, and pulging in to the nasopharynx and free paranasal sinuses, simulating septal hematoma (**Fig. 6**). Endoscopic biopsy and debulking was done. Biopsy revealed scleroma.

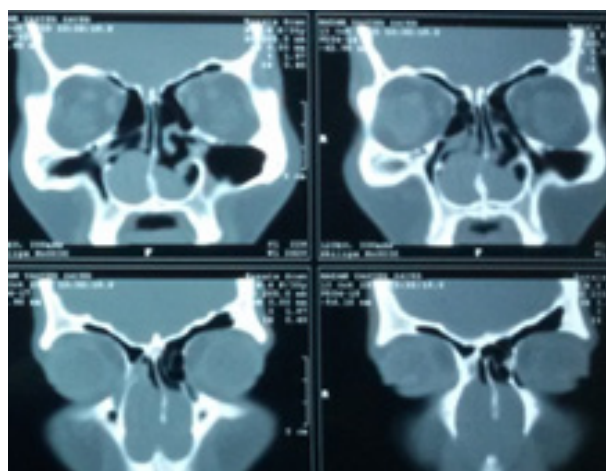


Fig 6. CT scan bilateral nasal opacity related to septum.

Case Ten: A 34 years old male patient presented by bilateral nasal obstruction, discharge & crustations for 8 months, examination showed bilateral septal swelling is occupying most of the anterior part of the septum , biopsy proved to be rhinoscleroma. Patient completely cured on rifampicine treatment.

Case Scenario 7 Extensive sinonasal with intracranial extension.

Case Eleven: A 37 years old male patient known to have respiratory scleroma presented with recurrent bilateral large nasal masses causing deformity and broadening, with bilateral proptosis. CT scan shows panopacity of sinonasal

area eroding lamina papyrea and sphenoid bone and skull base (**Fig. 7**). There were concurrent affection of the palate and whole larynx even supraglottic area and epiglottis. Endoscopic biopsy and debulking was done and laryngeal dilatation and tracheostomy.

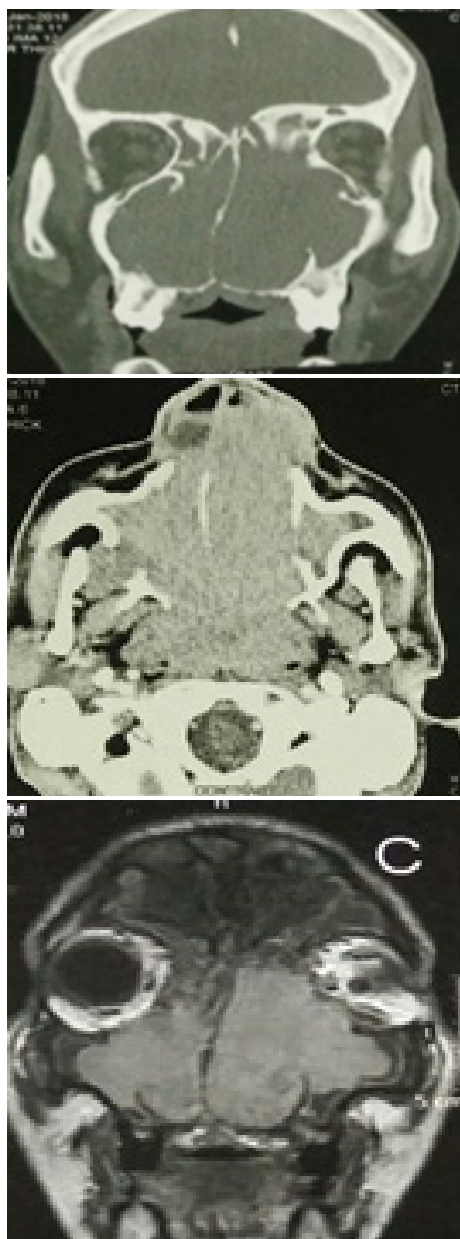


Fig 7. CT scan and MRI shows extension of the mass.

Discussion

Humans are the only known host of scleroma. Infection occurs by droplet infections with long contact period. The disease is endemic in some areas of the world, with high incidence in low socioeconomic, overcrowded and rural areas with malnutrition. [9] Respiratory scleroma is extremely rare in developed countries, but nowadays with more ease travelling and transportation and with more immigration, it should be suspected in travelers from developing endemic countries. [10]

Classic presentation of nasal scleroma in its three stages is bilateral. Combined nasal affection with other regional involvement, was most frequently encountered with the

larynx in 65% followed by the trachea in 52%, and less commonly the palate was affected by fibrosis in 14.5% (7 out of 48 cases) while only 2% (1 out of 48 case) showed tonsillar affection. Being a rare disease in developed countries, atypical presentations were reported in few studies. [5,11,12]

Our study included eleven patients; seven females and only 4 males, Their age ranged from 33 to 60 years old with mean 41 years old, with higher incidence in females as proved by Hart & Rao (2000) [13] who explained this sex predominance due to increased incidence of iron deficiency anemia and poor nutrition in child bearing ages. The youngest age reported in literatures was a newborn infant, with middle ear affection, while the oldest age was a 68 years old man with paranasal sinuses affection. [5] Familial predominance was reported by some studies. [6]

In our study there was one patient presented with isolated tracheal stenosis and granulations. Isolated bronchial scleroma was detected by Acuna 1973 [14] in one case. Many studies reported simultaneous affection of trachea and or bronchi with rhinoscleroma trachea in 12%, and bronchi in 2% to 7%. [15]

Egyptian study reported trachea affection in 17 patients out of 56 patients with respiratory scleroma. [16] Soni, 1994 [17] found four cases (20%) of 20 scleroma patients suffering from tracheal lesions and only one case with bronchial involvement.

A study from the United States showed that only 2 out of 22 cases with rhinoscleroma had tracheal involvement limited to the first two tracheal ring. [5] In Mexico, tracheal and bronchial scleroma has been reported in 2 percent of cases with RS. [18]

In our study, we reported two cases presented with laryngeal supraglottic swelling, both were associated with RS. In literatures, There was one case from El Salvador with supraglottic, Glottic multiple polypoid-like lesions which proved to be scleroma. [19]

In this study, Isolated nasopharyngeal scleroma was reported in 2 cases, this is in accordance with Schwartz RA, Goriniene E, (2008) [11] who reported nasopharyngeal involvement in 18%–43% of cases, also the nasopharynx was involved in 13 patients out of 56 patients (23%) but not isolated i.e.; associated with nasal disease. [16]

In this study we reported a case of Bilateral Maxillary swellings with nearly free nasal cavity, (Evrard et al., 1998) [20] reported an unusual case of sphenoidal sinuses rhinoscleroma. Involvement of the paranasal sinuses with Rhinoscleroma was previously reported by DiBartolomeo JR (1976) also. [18]

Two cases in this study presented with a unilateral nasal mass completely filling the nasal cavity with pansinus opacification on CTscan. This agrees with (Edgardo Bonacina et al., 2012) who presented a 51-year-old Egyptian immigrant in Italy, with left nasal cavity and ethmoidal mass causing inferior turbinate destruction. [21] However, (Fawaz S et al., 2011) reported that absolute unilateral affection is rare. [12]

In this study 2 cases presented with bilateral septal swelling which is considered atypical & a rare finding to arise only from the septum sparing the lateral nasal wall & paranasal sinuses. Cases of rhinoscleroma with intraorbital extension are very rare, only three cases exist in otolaryngological literature, and none has been reported in ophthalmological

journal. [22]

Intracranial extension is very rare but was reported by (Bahri et al., 1972), who found that scleroma, may invade the brain through the cribriform plate and produce a tumor-like lesion at the base of the brain. [23]

In this study we reported a case of intra cranial & intra orbital extension of Rhinoscleroma in a male patient 39 years old with highly extensive lesion.

Other rare atypical presentations were reported by other authors; Egyptian study of 88 patients reported 16 cases (18%) with atypical presentation or complication two cases of them presented with Pott's Puffy like tumor, whereas intracranial involvement occurred in one patient, intraorbital extension in three patients, cheek swelling in one patient, Diagnosis was confirmed with electron microscope. [12]

Other presentations of RS reported in the literature include septicemia followed by fatal septic shock and disseminated KR infection. [24]

Cutaneous involvement is very rare to occur, a study in a skin centre in Singapore reported four patients presented with only cutaneous involvement. [25] And another study in Senegal reported skin involvement. [26]

Conclusion

Rhinoscleroma should be put in differential diagnosis of any sinonasal, pharyngeal, laryngeal and even tracheal lesions especially in endemic areas. In developed countries Physicians should be more familiar with respiratory because of the increased incidence in travelers and immigrants.

Compliance with ethical standards.

Funding

No funding was received.

Conflict of interest

The authors declare that they have no conflict of interest.

Ethical approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed consent Informed consent was obtained from all individual participants included in the study.

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