Summary and Conclusion

Summary

Rhinoscleroma is a chronic granulomatous slowly progressive infection that affects the nose and other respiratory tract areas. The disease found predominantly in rural areas and is commoner where socio-economic conditions are Poor. The disease is more common in females in the child bearing age.

Rhinoscleroma has been widely reported from countries in the Middle-East, India, South-east Asia, Central and South America and Africa. In Egypt the disease is endemic in areas of Gharbia, Giza,Sharkia, Kaliobia, Menofea and Fayoum .

Rhinoscleroma is caused by Klebsiella pneumoniae subsp. rhinoscleromatis which is a gram-negative diplobacillus. Klebsiella rhinoscleromatis was identified as a causative agent of rhinoscleroma by Von Frisch in 1882.

Rhinoscleroma involves oral cavity in 18%, pharynx in 18-43%, Eustachian tube in 27%, larynx in 26%, trachea and bronchi in 10% of cases. The nose is involved in almost all cases, yet the reported rate of sinus involvement is low with maxillary sinus being involved in about 22% of cases.

An accurate history, a thorough physical examination and a high index of suspicion facilitate the diagnosis of rhinoscleroma. Endoscope

is essential tool for the diagnosis and take biopsy from the affected areas and areas of active disease

The biopsy is the most important in diagnosis of scleroma. The pathological findings of rhinoscleroma are characterized by three histomorphological STAGEs which may coexist. These stages are catarrhal (rhinitis), proliferative and fibrotic. In Proliferative STAGE features granulomatous inflammation predominantly in submucosal tissue and characteristically shows a mixed inflammatory cell infiltrate including plasma cells, these plasma cells are typically associated with prominent Russell's bodies and Mikulicz cells, these Mikulicz cells and Gram negative infiltration in the subepithelium is characteristic of rhinoscleroma but not pathognomonic. In catarrhal and sclerotic stage; Mikulicz cells are very rare if not absent and difficult to see the microorganisms making diagnosis more problematic.

A positive culture of Klebsiella rhinoscleromatis is diagnostic of rhinoscleroma but the limitation is that only 60% of the biopsy proven cases were positive for Klebsiella rhinoscleromatis.

Serological and immunochemical and PCR tests should be done to confirm the diagnosis. Complement fixation tests and agglutination tests can be used, but they are diagnostic only when the tests show positive results. Immunochemical examination performed with antibodies to the O2K3 antigen on Klebsiella rhinoscleromatis can be used to make a definitive diagnosis. Molecular assay based on PRA (PCR Restriction Assay) of rDNA16S gene from Klebsiella pneumoniae used for differentiation at the subspecies level. PCR in rhinoscleroma is possible but not routinely available.

Radiological investigation in rhinoscleroma may include several modalities. X-Ray show expansion. CT scanning can be used to assess accurately the extent of the disease. In MRI Rhinoscleroma showed mild to marked high signal intensity on both T1- and T2- weighted images. PET scan may be used in cases of scleroma there is increased metabolic activity in part affected.

In catarrhal stage antibiotic treatment is used as a single treatment to eradicate the infection but drug treatment combined with surgery in cases with granulomatous lesions or scarring stenosis may be a good choice for curing rhinoscleroma

The mainstay of treatment for rhinoscleroma is quinolones. Additional potentially efficacious antibiotics are cephalosporins, rifampin, the anti-leprotic agent clofazimine, tetracycline, streptomycin and trimethoprim-sulfamethoxazole (Cotrimoxazole).

Corticosteroid therapy in combination with antibiotics has given good results in some cases, also levamisole may be used as an immune-enhancing agent to improve the patient cellular immunity. Supportive measures also improve patient health.

Topical antibiotics such as acriflavine 2% or rifampicin have been used with significant results.

Surgery and laser ablation can be used only after complete cessation of disease activity has been achieved with medication Patients must be clinically and histologically free of disease as well as have negative tissue cultures; otherwise the risk of recurrence or dissemination is high.

After treatment induced remission, prolonged follow-up is essential to early detection of any relapse as the rate of relapse is high.

We suggest an Egyptian protocol including algorithm for diagnosis and treatment of rhinoscleroma.

Treatment can be summarized in the following points

1. Prophylaxis
2. Antibiotic for long period most preferred quinolone or cotrimoxazole in combination with rifampicin until negative biopsy
3. Levamisole regarded as an immune-enhancing agent
4. Local antibiotic; rifampicin or acriflavine
5. Supportive measures
6. Surgical treatment after negative biopsy
7. Prolonged follow up for early detection of relapse

Conclusion

Rhinoscleroma is a chronic disfiguring and debilitating disease. We have much to learn about its epidemiology and pathogenesis. We still have a long way from providing optimal therapy. Finally, relapses occur and prolonged follow-up is needed to spot early relapse.