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Original Research Article

Rhinoscleroma - A Clinical Study and Review of Literature

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ABSTRACT

Introduction: Rhinoscleroma is a chronic, progressive, and infectious granulomatous disease of the respiratory tract mucosa. There is often a delay in diagnosis due to unfamiliarity with the disease and also because culture is not always positive. Klebsiella rhinoscleromatis is the causative agent of this infection and the Mikulicz cell is specific to the lesion being a large macrophage with clear cytoplasm containing the bacilli. It is endemic in temperate and tropical zone countries.
Materials and Methods: In this study 28 cases of histologically diagnosed rhinoscleroma cases are being studied on the basis of their age distribution, common symptoms, sex ratio and response to treatment.
Results: In the present study, most common age group in Rhinoscleroma was in 3rd decade (32.14%) followed by 4th (28.57%) and 2nd (21.42%) decade. The median age group of Rhinoscleroma in our current study was 30 years and male to female ratio was 1:1. The most common symptom in rhinoscleroma in our study was nasal obstruction
Conclusion: In the present study of Rhinoscleroma, patients presented with varied nasal symptoms like nasal obstruction, discharge, nasal bleed, mass and deformity in absence of more generalized symptoms. Clinical diagnosis is often difficult and has to be relied on Histopathological examination of biopsy specimen and may require repeated biopsies. Many a times they may be overlooked in clinical practice.

Keywords: Rhinoscleroma, Klebsiella rhinoscleromatis, Mikulicz cell

INTRODUCTION

Rhinoscleroma is a chronic progressive granulomatous disease predominantly affecting the upper respiratory tract caused by Klebsiella rhinoscleromatis. [1] It is endemic in many tropical and temperate zone countries especially Eastern Europe, North and Central Africa, Southern Asia and the Middle East, as well as parts of South and Central America. Some authors have pointed out the predisposing factors of Rhinoscleroma as low living and bad cultural standards, combined with poor hygienic condition. [2]

Pathological diagnosis of Rhinoscleroma is made when tissue examination shows a granuloma with Mikulicz cells, hyaline bodies and Von Frisch bacilli. The plasma cells,

lymphocytes and Russel bodies are characteristic components of the subepithelial infiltrate. [3] The diagnosis of the disease is made on the basis of clinical presentation, bacteriological examination as well as the histopathological findings. [4,5]

The treatment of Rhinoscleroma is primarily medical using different antibacterial agents with variable results and increasing numbers of recurrences. Radiation therapy is indicated in resistant cases under special conditions. Surgery is reserved for critical situations such as airway obstruction or cosmetic reconstruction.

MATERIALS AND METHODS

The present study was carried out on both the outpatients and inpatients of Department of ENT. Only those patients presenting with chronic symptoms of nasal obstruction, epistaxis, nasal discharge, crushing, deformity, showing nodules, ulceration and atrophic changes on clinical examination and histopathological diagnosis of Rhinoscleroma were selected for this study. Clearance from institutional ethical committee was obtained prior to conducting study. The consent was taken from all the 28 cases. Mean age of patients in the study was 31.32 years with standard deviation of 11.29(31.32±11.29 years). A detailed history was taken and a thorough systemic and ENT examination with special emphasis on nodules, polyps, ulceration in the nasal cavity. Based on the clinical signs and investigation a diagnosis was arrived and appropriate medical or surgical or both modalities of treatment were carried out. The patients were followed up for a period of 3-6 months and special emphasis was given for the recurrence of the disease and recovery of the patient.

RESULTS

In the present study, most common age group in Rhinoscleroma was in 3rd decade (32.14%) followed by 4th(28.57%) and 2nd(21.42%) decade (Table.1). It is in comparison with the studies of Badraway (1966) where the patient in age group of 3rd and 4thdecade comprised 50% each. In a study conducted by U.Zafar et al the peak age of presentation of Rhinoscleroma was in 4thdecade. [6] In a study conducted by Magnina C in Peru, most common age group of patients was in 3rddecade (50%) which correlates with the current study. [7]

The median age group of Rhinoscleroma in our current study was 30 years and is comparable to French national retrospective study on rhinoscleroma carried out by de Pontual et al where the median patient age at diagnosis was 35.7 years. [8] But another study conducted in Gulf region by Abalkhail A the median age of the patients was 24 years. [9] Another study conducted by Hart CA, Rao SK also showed a peak incidence in age group of 2nd and 3rddecade. [10]

In our study the male to female ratio in rhinoscleroma was 1:1 (14 each) (Table.2) whereas other studies conducted by Hart CA, RaoSK, Magnina C et al, Abalkhail A et alshowed a female predominance of 1.3:1, 3:1, and 2.5:1 respectively. In our study the decrease in female patients of rhinoscleroma may be due to ignorance regarding the disease, lack of education, awareness and non-utilization of hospital services by the women.

Table.1: Age distribution

SL No.	Age group	No. of cases	Percentage
1	0-10	0	0
2	11-20	6	21.42
3	21-30	9	32.14
4	31-40	8	28.57
5	41-50	4	14.28
6	51-60	1	03.57
7	61 and above	0	0

Table.2: sex distribution

SL No.	sex	No of Cases	Percentage
1	Male	14	50
2	Female	14	50

The most common symptom in rhinoscleroma in our study was nasal obstruction which co-relates with other studies of U-Zafar et al, Hart CA, Rao SK. Abalkhail A et al. Oropharyngeal involvement was seen in eight patients (28.57%), (Fig.1) nasopharyngeal involvement in four (14.28%), laryngeal in two patients (7%). vestibular stenosis in the form of partial / complete in seventeen (60%) and Choanal stenosis in two patients (7%). Study conducted by Magnina C showed oropharyngeal involvement in 25%, laryngeal in 37.5% (Fig.2) and choanal in 50%. [7] Study conducted by Abalkhail A showed laryngeal involvement in 44% and nasopharyngeal involvement in 24%. [9] Comparing these studies laryngeal involvement in our current study was less and may be due to early presentation of the patients to the hospital and early diagnosis and starting of adequate treatment.

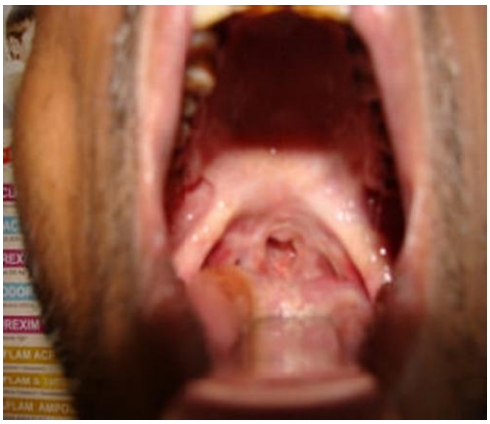

Fig.1: Oropharyngeal involvement in rhinoscleroma

All 28 patients of Rhinoscleroma were treated medically with combination of T.Rifampicin (450mg OD) and T. Ciprofloxacin (500mg OD) or T.Rifampicin (450mg OD) and T.Doxycycline (100mg OD) for a period of 6-12 weeks according to their clinical cure. It was assessed by improvement in symptoms and regression of granulomatous masses.

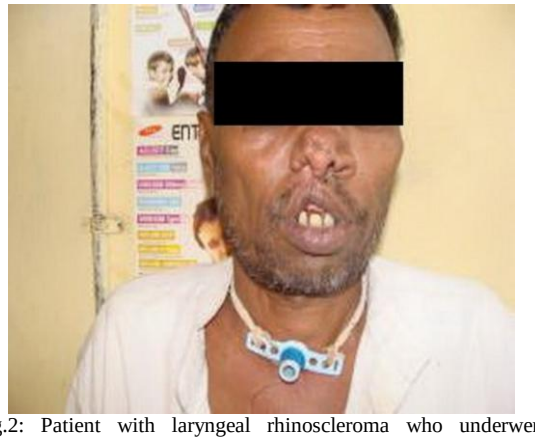

Fig.2: Patient with laryngeal rhinoscleroma who underwent tracheostomy for narrowed upper airway

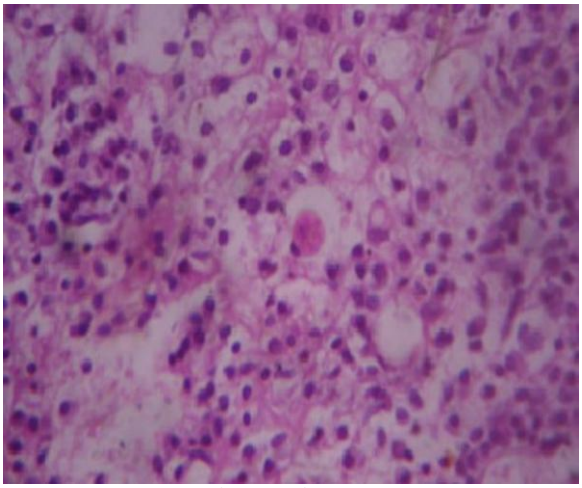

Fig.3: Histopathological features of Rhinoscleroma

Surgical modality along with medical treatment was used in 6 patients of the 28 patients, where surgical debridement of the granulomatous masses and cicatricial tissue was done and recanalization was done using polythene tubes. These tubes were left in-situ for 6-8 weeks and then removed and examined. Since none of the patients had any complaints of dysphagia, no definite treatment was given for the oropharyngeal involvement and all cases were treated conservatively. Out of 22 patients who have completed the course of medical treatment,

all the patients have recovered completely giving a 100% results with Ciprofloxacin and Rifampacin or Rifampicin with Doxycycline. No recurrence was seen. Rests of the 6 patients are still continuing the treatment till date. This is in par with the other studies and case reports to be an effective and affordable option. [11-13] Out of the 6 patients who underwent surgery, 4 have completed treatment with removal of polythene tubes and none of the patients have recurrences and is comparable to a case report by Vadher DJ. [14] Two of our patients are still continuing the treatment. Only 1 case of recurrent Rhinoscleroma was seen, who underwent surgery twice in duration of 20 years and has been operated (recanalization) again and is on treatment. Out of the 2 cases of laryngeal scleroma one was operated for stridor (emergency tracheostomy) during the follow up period.

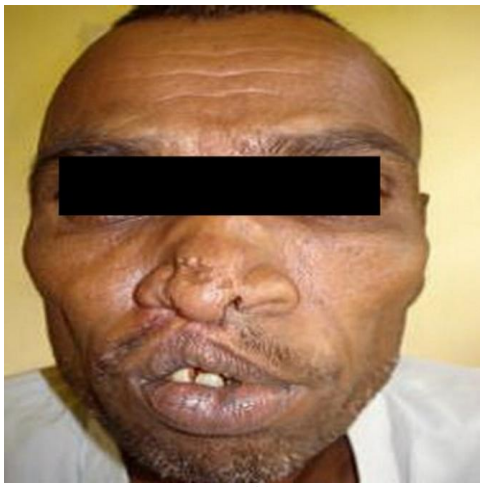

Fig.4: Typical presentation of advanced Rhinoscleroma

DISCUSSION

Epidemiology

i) Incidence: No racial predilection exists, but females are slightly more affected than males (about 1.3 to one) and patients are commonly afflicted in the second and third decades of life.

ii) Geographical distribution: Rhinoscleroma is more common in developing countries and rural areas and is endemic in Africa, South-East Asia, Mexico, central and South America and Central and Eastern Europe. [15] In India it is endemic in Madhya Pradesh, Uttar Pradesh, and Rajasthan. [16] Southern highlands province of Karnataka a belt north of Vindhya Mountains and also scatteredin some villages quite away from these mountains. [17]

iii) Socio-economic: There appears to be an association between low socioeconomic status, poor hygiene, poor nutrition, crowded living condition and the development of rhinoscleroma. [15]

iv) Transmission of disease: Transmission is proposed to occur via direct inhalation or inoculation by respiratory droplets but only after prolonged contact. Familial incidence observed further provides clue to transmission.

Bacteriology:

The infectious agent Klebsiella rhinoscleromatis (of the enterobacteriacea family) was first described by Von Frisch in 1882. This gram negative, non-motile, encapsulated facultative, glucose fermenting, intra cellular diplococcus, is hosted by humans alone. It is widely accepted that the mucopolysaccharide in the capsule of K. pneumonia protects the bacteria by effectively inactivating macrophages so they cannot phagocytize bacteria. Klebsiella rhinoscleromatis, because of its close biochemical relationship is proposed to share this defense mechanism. Ineffective phagocytosis of the organisms by macrophages results in characteristic Mickulicz cells, large macrophages with clear cytoplasm and intracellular bacilli. This ineffective phagocytosis may be responsible for the chronicity and granuloma formation seen with this disease. [15]

Host factor: Host factors may play a role in development of the disease. Cellular immunity is impaired in affected patients,

but humoral immunity remains intact. [15] Studies in patients with rhinoscleroma have shown alteration in the ratio of CD4 – positive lymphocytes (Helper T cells) to CD8 - positive lymphocytes (cytotoxic T cells) with a marked increase in the latter type. Patients have also shown impaired response of their CD4 lymphocytes to interleukin-2 and also a diminished proliferative response to concavalin A, T-lymphocyte mitogen. Iron deficiency has been proposed as a risk factor for rhinoscleroma and may contribute to the site predilection of nasal vestibule and subglottic region by altering epithelial regeneration and causing squamous metaplasia. This may explain its association with poor nutrition and why menstruating women and pregnant women have more severe course. [15]

Pathology:

According to the work of Canalis and Zamboni the majority of events leading to chronic infection occur in the subepithelium. First K. Rhinoscleromatis invades the sub-epithelium, multiplies and incites capillary proliferation. Next many polymorphonuclear cells enter into the subepithelium in response. They are able to phagocytize the bacteria but die at an accelerated rate without completing digestion of the organism. Finally macrophages (histiocytes) enter the area and phagocytose the decaying polymorphonuclear cells and klebsiella organism. Their phagosomesundergo massive dilation and at this point they become the characteristic Mikuliczcells. These macrophages are effective in lysing decaying PMN but are unable to destroy the Klebsiella organism. Eventually these macrophages rupture and release both active and inactivated organism into interstitium and cycle continues. Infection and subsequent mass lesions occur in areas of transition between squamous and ciliated respiratory epithelium such as nasal vestibule and subglottic area. [18]

Microscopy:

Mickulicz first described the large vacuolated histiocytes which now bears his name. These cells measure 100-200 m have eccentric nuclei, and contain the Klebsiella organism within their cytoplasm. Though these cells typify rhinoscleroma, their presence is not necessarily pathognomonic. Other disease such as Lepromatousleprosy, Glanders and Granuloma inguinale may demonstrate large vacuolated cells with intracellular organism. [19] Plasmacytes often show a notable degree of activity in which the accumulation and coalescence of globules of secretory products present in a characteristic appearance commonly known as “Cornil or Mott cells” (Fig.3). Such cells are encountered in variety of inflammatory lesions and are in no way specific for scleroma. The cell may degenerate and be cast off leaving a naked, eosinophilic hyaline mass known as Russell body. [20] Other cells associated with rhinoscleroma are Unna body, Morula cells. [21]

Clinical features:

Kouwenaar (1956) pointed out that the clinical picture varies according to the geographical location. The enormous swelling and distortion known as ‘Hebra Nose’ is common in Indonesia but seen in other locations, whilst laryngeal lesions are said to be more common in Europe and America. Spread to pharynx, larynx, lacrimal duct, upper lip and paranasal sinus may occur. In the granulomatous phase, masses of inflammatory tissue may cause pressure erosion on bone. Extension into the orbit and the cranial cavity may be encountered. [20]

Classically there are three clinical and histological stages of rhinoscleroma:

- Catarrhal - atrophic
- Granulomatous

- Sclerotic

The catarrhal atrophic stage may last weeks to months and may begin with anon-specific rhinorrhoea. This may evolve to a foul smelling, purulent rhinorrhea with crusting and nasal obstruction. Histologically squamous metaplasia of the epithelium is seen, with underlying infiltrates of PMN cells and some granulation tissue. The granulomatous or hypertrophic stage is usually the point at which the clinical and histological findings are most easily recognized. Patients present with bluish red or polypoid, anterior nasal masses which are non-tender, rubbery and maybe prone to bleed. These masses most often affect the anterior- inferior septum, but sometimes the maxillary antrum may be involved and act as reservoir for infection. Nasal deformity is not uncommon as destruction of nasal cartilage occurs to form a classic "Hebra nose" (Fig.4). The destructive process of the disease may leave the patient with anosmia, anaesthesia of soft palate, enlargement of the uvula, dysphonia or various degree of airway obstruction.

Histologically there is pseudo epitheliomatous hyperplasia, Mickulicz cells, monocytes, lymphocytes, Russell bodies and macrophages. Sometimes the inflammatory infiltrate is found to be angiocentric and causes a vasculitis like picture with hyalinization of the vessels. Finally in the sclerotic stage, former masses are replaced by extensive scaring, deformity and stenosis. Few Mickulicz cells or Russell bodies are formed. Patients may present at any of three stages and with host of non-specific complaints, including nasal obstruction, rhinorrhoea, epistaxis, dysphagia, nasal deformity, stridor, dysphonic and anosmia. On examination the diagnosis of rhinoscleroma should be considered if the patient comes from an endemic area and has nasal lesions involving the septum with relative sparing of the sinuses. [15]

Diagnosis:

1) Brush Biopsy specimen of nasal or respiratory tract mass or an incisional biopsy from an early accessible lesion sent for histopathology.

2) Biopsy specimen for bacterial culture on blood or Mac Conkey's Agar. A positive culture of K-rhinoscleromatis is diagnostic but only 50-60 percent of patients are culture positive.

3) The bacteria may be seen using Periodic Acid-Schiff, Giemsa, Gram orWarthin-Starry Silver Stains.

4) An immunoperoxidase technique for the K-capsular antigen has been shown to increase the specificity of histological findings in culture negative cases. [15]

5) Complement fixation test: It may be helpful test in patients of early stages, if biopsy is not adequate or in late cases when biopsy may not show the classical features of Mickulicz cells. Though conversion to a negative reaction has been correlated with regression of lesions, authors feel that the test is merely adjunctive evidence in diagnosis rather than confirmatory. [19]

6) Imaging studies are somewhat helpful in determining the extent of disease but should not be relied upon for diagnosis. On CT scans, rhinoscleroma appears as a homogenous, nonenhancing mass with distinct margins. Occasional bony or cartilaginous erosion may be seen, but adjacent fascial planes are not usually invaded. Magnetic resonance imaging may show masses obstructing the osteomeatal complexes and also show high signal intensity on T1 and T2weighted images in hypertrophic stage. [15]

Dawlatty (1991) described the "palatal sign" from lateral skull X-ray of advanced cases of rhinoscleroma causing thickening of the soft palate at its attachment with hard palate and tapering towards its free edge. This V

shaped soft palate is a sensitive and specific radiologic finding.

Treatment:

Medical: Streptomycin was initially widely accepted as a drug of choice. Tetracycline soon became preferred antibiotic with its potential for oral administration and avoidance of vestibulotoxic side effects. Use of tetracycline was limited by prolonged course of therapy and its contraindication in pregnant women and children. Ciprofloxacin, Trimethoprim-Sulfamethazazole, topical and systemic rifampicin and topical 2% acriflavine have also been used with some success. A shorter course (4-12 weeks) of ciprofloxacin 500 mg twice daily, along with nasal lavage twice daily has been stated in a number of case reports to be effective and affordable option. [15] Increased compliance, due to shorter duration of treatment, twice daily dosing and a low-side effect profile, makes ciprofloxacin an appealing drug of choice. Moreover ciprofloxacin achieves superior tissue penetration and is concentrated within macrophage. [15] Treatment for a minimum of 4-6 weeks and are continued until two consecutive cultures from biopsy material are proven negative.

Initial surgical debridement prior to chemotherapy is reported to be useful in granulomatous stage. Recent reports have shown good results with oral therapy with rifampic in with Trimethoprim Sulfamethaxazole combination and ciprofloxacin. [22] Local application of 2% acriflavine for a period of eight weeks has been notedto be efficacious and non toxic. [23] Locally applied rifampicin has been used with success. [24] Irradiation to a total dose of 3000-3500 Gy over three weeks destroys scleroma but with current antimicrobials is hardly used. [25] Response of drug therapy depends on stage of disease, site of involvement, disease extent and whether it is primary or recurrent. [26]

Surgical:

There are several indications for surgery in rhinoscleroma, including relief of airway obstruction and reconstruction of cicatricial defects. Reconstructive surgery is needed for nasal or lower airway stenosis from scarring or imperforation. [15] Surgery to re-establish the nasal airway by discrete removal of granulation and dilatation of airways combined with insertion of polythene tubes for 6-8 weeks appears to be the most appropriate mode of treatment. Lastly after complete eradication of the disease, plastic surgery of the nose may be carried out to give good cosmetic appearance. Recurrence is common in patients with rhinoscleroma and they should be followed regularly with nasal endoscopy and cytology. In addition, a prolonged course of antibiotic therapy, lasting weeks to months, may stave off recurrences.

CONCLUSION

In the present study of Rhinoscleroma, patients presented with varied nasal symptoms like nasal obstruction, discharge, nasal bleed, mass and deformity in absence of more generalized symptoms. Clinical diagnosis is often difficult and has to be relied on Histopathological examination of biopsy specimen and may require repeated biopsies. Many a times they may be overlooked in clinical practice. Management of these patients is challenging due to varied presentation and lack of definite protocols for treatment. Early medical treatment will decrease the deformity rates in these patients. Sometimes medical and surgical modalities have to be combined together according to clinical situation to reduce the morbidities.

Awareness regarding the disease process and health education should be provided to people regarding maintenance of hygienic conditions and utilization of health

facilities. Emergence of newer pharmacotherapeutic agents have reduced the disease burden to a large extent and newer surgical interventions with LASER, rhinoplasty will further open up a new chapter in the cosmetic reconstruction. Due to the chronicity of these diseases a long term follow-up is required to assess the recurrence of these diseases.

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