FULMINANT DISFIGURING REFRACTORY RHINOSCLEROMA - AN ENCOUNTER

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Abstract

Rhinoscleroma is a rare chronic granulomatous disease of the upper respiratory tract caused by bacilli Klebsiella rhinoscleromatis, primarily involving nose, pharynx, oropharynx, larynx and adjacent structures. It can be effectively treated with long-term specific antibiotics. However, progressive and diffuse sclerotic deformity of the nose can occur and result in severe stenosis, leading to disfiguration and debilitation. We discuss a case of a 32 year old male with progressive bilateral nose block since 25 years. Patient had visible external nasal deformity of flattened broadened dorsum with bilateral nasal stenosis, despite having undergone two nasal surgeries along with conservative treatment. He was subjected to recanalization of bilateral nasal cavities along with specific medical treatment. Surgical intervention is a good option in treating extensive rhinoscleroma with symptomatic obstruction, along with prolonged antibiotic therapy and follow-up to help prevent relapses.

KEYWORDS – Rhinoscleroma, Recanalization, Nasal stenosis, Nasal stent

INTRODUCTION

Rhinoscleroma is a rare disease of the upper respiratory tract caused by gram-negative bacillus Klebsiella rhinoscleromatis, which primarily involves nose, nasopharynx, oropharynx, larynx and adjacent structures. Progressive diffuse sclerotic deformity of the nose can occur resulting in severe stenosis, disfiguration and debilitation.

Historically, the disease was first diagnosed in 1870 by Von Hebra. He also coined the term rhinoscleroma (rhino = nose, scleroma = hard). Frisch in 1877 discovered the bacillus Klebsiella rhinoscleromatis, which was named after him. 1

The nasal vestibule is located caudal to the nasal valve area and just posterior to the external nares. Although the valve is the narrowest part of the nasal cavity, significant stenosis of the vestibule can cause debilitating nasal obstruction. Collapse of nasal valve may or may not be present. 2

Ciprofloxacin, a fluoroquinolone, is an antibiotic with excellent tissue penetration and a broad antibacterial spectrum of action. Adverse effects are comparatively few and include gastrointestinal symptoms in 3 to 6% of patients and uncommon central nervous system and allergic symptoms. Long term compliance is improved due to twice daily dosage. 3

From a bacterio-pathological point of view some similarity was noticed between tubercle bacillus and Frisch bacillus. Both cause chronic specific granulomatous lesions and both respond well to streptomycin therapy. 4 On that basis, rifampicin has been recently used both systematically and locally (nasal instillation or infiltration) with success for the treatment of scleroma (Gamea 1988). 5
Acriflavine is a mixture of 2,8 diaminoacridine and 2,8 diamino-10 methyl-acridinium chloride (Albert, 1951). In addition to its use as a dye, it has an active antibacterial action against most gram-positive and negative organisms. Acriflavine has also been reported to produce in vitro a lethal action on K.rhinoscleromatis bacilli cultured on nutrient agar plates (Rizk, 1977).

In most cases, treatment involves prolonged antibiotic therapy and aesthetic surgical reconstruction when necessary.

CASE REPORT

We would like to report a case of a 32 year old male with progressive bilateral nose block, with discharge and crusting since 25 years. Patient had visible external nasal deformity of flattened broadened dorsum with saddle deformity and bilateral extensive nasal stenosis, despite having undergone two nasal surgeries along with conservative treatment. Palatal arches were pulled up, multiple palatal adhesions and reduced movements of the soft palate were also observed, thus compromising nasopharyngeal isthmus patency. Bilateral tympanic membranes were retracted and dull. Rest of ENT examination was normal.

Routine investigations were done and the patient was subjected to surgery that included recanalization of the nasal vestibule, soft palate release and placement of stent, and removal of stent, along with aquasol (Vitamin A) and acriflavine nasal drops.

During surgery, cicatrized tissue adherent to the lateral wall of the right nasal cavity and septum was released. Biopsy was taken. Septal perforation was noted intra-operatively. Soft tissue was released at the floor to make it more concave. Lateral wall was pushed laterally to release the adhesions and to create the turbinates. Valve area was created. Cicatrized tissue over olfactory area also was released. The same procedure was repeated on the left side. Patient was started on oral medications with Rifampicin 450 mg and Ciprofloxacin 500 mg.
Second stage surgery was done after three weeks. Soft palate was released. Stent was placed in both nasal cavities and sutured across the septum. Stent was removed after three weeks.

Patient had an uneventful recovery and came for follow up every three weeks.
FIGURES

Fig 1 - Photograph showing preoperative nasal deformity
Fig 2 - Photograph showing palatal adhesions
Fig 3 - Photograph showing nasal vestibular stenosis
Fig 4 - Photograph showing septal perforation
Fig 5 - Photograph showing placement of nasal stent
Fig 6 - Photograph showing removal of nasal stent
Fig 7 - Photograph showing widened nasal vestibule on second look surgery

DISCUSSION

Rhinoscleroma is a chronic inflammatory disease characterized by granulomas in the nose and the upper airways, and essentially has three stages:

1. Exudative or catarrhal-atrophic stage - which resembles atrophic rhinitis, examination and biopsy at this stage may be non-specific.

2. Proliferative or granulomatous stage - characterized by lymphocytic infiltration and plasma cells, with painless, non-ulcerative nodule formation; may also be characterized by extranasal spread to trachea, larynx, bronchi, sinuses, bone, cartilage, ear, orbit and intracranial spread.

3. Cicatricial or fibrotic stage - occurs due to protracted inflammation and healing, presenting as stenosis of nares.\(^2,3\)

The disease usually starts with clinical features of atrophic rhinitis with crusting, except affecting smell perception. However, as this patient had extensive cicatrisation, he complained of anosmia as well. As the disease progresses, granulomas develop which cause nasal obstruction and ultimately cicatrization. Many a times these stages do occur concurrently.

Specific diagnosis is made by bacterial isolation by culture on blood or MacConkey agar and by identification of histopathological features and bacilli in biopsies using periodic acid Schiff (PAS), Giemsa and Warthin-Starry stains. These stains combined with immunoperoxidase staining using Klebsiella capsular type 3 antiserum increase accuracy and specificity of both histological and bacteriological diagnoses.\(^6\) Biopsy, which shows infiltration of submucosa with plasma cells, lymphocytes, eosinophils, Mikulicz cells (foamy macrophages with numerous intracellular gram-negative rods) and Russell bodies (degenerating plasma bodies). The latter two are diagnostic but only in active stage. The first two stages are usually managed conservatively, while the third stage requires surgery.\(^2,3\)

Antibiotics including streptomycin, chloromycetin, tetracycline, cephalosporsins, ceforizanide, clofazimine, have been tried. Other drugs include acriflavin, mitomycin C, ciprofloxacin.\(^3\)

Surgical treatment described includes excision of stenotic segment\(^6\) followed by transposition of soft tissues, split and full thickness skin grafts, composite chondro-cutaneous grafts taken from the ear, intraoral tunneling of the oral mucosa, and local nasolabial flaps.\(^2\)
In this patient, biopsy showed plasma cells, lymphocytes, foamy macrophages, and numerous congested blood vessels. The patient gave history of lack of relief with homeopathic treatment. Surgery was done, which included recanalization of the nasal cavities, wherein cicatrized tissue was excised, and nasal cavity was expanded by pushing lateral wall laterally and excising soft tissue from the floor. Patient was started on rifampicin, ciprofloxacin and acriflavine. A second stage surgery was done which included soft palate release and placement of nasal stent, which was removed after a few days. Patient is now relieved of nasal obstruction and is able to perceive smell very well.

CONCLUSION

Surgery is the preferred modality of treating cicatricial stage of rhinoscleroma, when conservative method fails to control the disease. Post-operative medical supplements help in faster resolution. Patient co-operation and education is must for the proper management of this condition and the correction of external deformity is always undertaken only after the disease is burnt out with confirmation with biopsies.

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REFERENCES


