Scleroma of the Nose and Pharynx

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Scleroma of the respiratory tract was first recognized just over a century ago. The disease is endemic in a number of North African, Central American and Latin American countries. However, it has been only rarely seen in the United States. The progression of scleroma takes place through three stages. The first stage of rhinitis has an early catarrhal phase which progresses to one of atrophic rhinitis. The second stage is seen as a proliferative granulomatous growth which obliterates the nasal fossae. The third cicatricial stage is usually accompanied by pronounced scarring and retraction of the tissues involved.

Because of the increased ease of international travel by both sea and air, more cases can be expected to occur in the United States. This paper is presented to alert physicians to suspect scleroma in any case of granulomatous disease of the respiratory tract.

Scleroma of the upper respiratory tract was first recognized over a century ago. It was described as a clinical entity by von Hebra in 1870, who called it nasal leprosy, hardnose and exuberant syphilis of the 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. Because the preponderance of cases presented with nasal involvement, it was often called rhinoscleroma.

At the Second International Congress of Otolaryngology held in Madrid in 1932, it was suggested that the term rhinoscleroma be changed to scleroma in order to emphasize that the disease may attack any part of the respiratory tract. Similarly, it was suggested that such terms as syphilis of the nose, nasal leprosy, leprosy of the Slavics, scleroma neonatorum and scleroma respiratory be discarded in preference to scleroma alone.

Scleroma is particularly a disease of the young and middleaged adults. There is a slightly higher incidence in females. Most patients afflicted with the disease seem to belong to the lower economic class and have poor nutritional status. The disease

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is not known to be contagious or familial as a rule, but Losageno reported 15 cases in one Italian family, and Hare and co-workers reported 7 cases in a single American family. In the endemic areas of the province of Lublin in Poland, there is a higher proportion of familial incidence of scleroma observed among children as compared with adults.6

Scleroma is well-known in North Africa, Central America and Latin America—with the heaviest concentration of the disease being seen in the Guatemala-El Salvador region. Quevedo’s Guatemalan series now consists of over 1,200 cases.7 In neighboring Mexico, scleroma is frequently seen.8 Scleroma is an extremely rare condition in the United States. The number of cases reported in native-born Americans is less than 60. With the increased ease of international travel by both air and sea in the last decade, one can expect the incidence of scleroma to increase in the United States.

Since scleroma may mimic various diseases of the respiratory tract throughout its stage of progression, this paper is presented to alert physicians to consider the possibility of scleroma in evaluating granulomatous disease of the respiratory tract.

Scleroma is a specific, chronic, progressive granulomatous disease of the respiratory tract. Because of the preponderance of cases with nasal involvement, earlier papers report the disease as rhinoscleroma. While most patients do, indeed, present with nasal involvement, the pharynx is involved in more than half the cases. In Mexico, laryngeal scleroma is evident in 15 percent of patients with respiratory scleroma. Tracheal and bronchial scleroma has been reported in 2 percent of such cases.8

Unusual cases of scleroma have been reported in the literature. The youngest age reported was in a newborn infant, with involvement of the middle ear. The oldest age reported was in a man of 68 years.4 Scleroma has been reported with involvement of the paranasal sinuses.9 Cervical lymph node involvement has been recorded.10 Scleroma has also been shown to undergo intracranial extension.11,12 Malignant changes have been reported in 5 percent of patients.13

Bacteriology

Klebsiella rhinoscleromatis is believed to be the causative agent of scleroma. K. rhinoscleromatis is a Gram-negative diplobacilli commonly known as the Frisch bacilli after its discoverer.1 K. rhinoscleromatis is considered to play an important role in the pathogenesis of the disease.

Klebsiella rhinoscleromatis grows poorly on the MacConkey’s agar. Colonies are thick, grayish-white, slimy and mucoid and tend to string out when touched with a loop (Figure 1). On Gram staining, Gram-negative rods are seen. India ink smears show the presence of capsules. Some recent reports suggest that Klebsiella rhinoscleromatis may be identical with Type C of Klebsiella pneumoniae.14

Cellular Pathology

Histologically, one observes connective tissue containing Mikulicz cells, Russell bodies, plasma cells, lymphocytes, polymorphonuclear leukocytes, fibroblasts and the Gram-negative diplo-
bacilli within the vacuolated portion of the Mikulicz cell.

The Mikulicz cell is considered to be a macrophage. It is seen as a large, foamy, mononuclear cell measuring up to 100 microns in diameter. It has a pale, reticular, ill-defined cytoplasm with an eccentric nucleus and central vacuoles. If special bacterial stains, such as Giesma or Warthin and Starry stains, are used, one will find many Frisch bacilli within the vacuolated cytoplasm of the Mikulicz cell (Figure 2).

The Russell bodies are elliptical formations measuring 20 to 40 microns in diameter. They are smaller than Mikulicz cells but still twice as large as a normal plasma cell. They have a homogeneous brilliant red refractile cytoplasm (Figure 2). The Mikulicz cell is a major histologic feature of scleroma. However, the plasma cell, which undergoes some hyaline degeneration to become the larger Russell body, is not specific. Russell bodies may also be seen in any inflammation in which plasma cells are a prominent element. Such is the case in leprosy, malignant bubo and venereal granulomatosus.

Cultures for K. rhinoscleromatis are most often positive during the granulomatous phase of the disease.

Nasal Scleroma

Nasal scleroma characteristically progresses through three stages. Stage I (rhinitis stage) begins with symptoms of acute catarrh that gradually progresses to symptoms compatible with atrophic rhinitis. At the onset of the disease, the patient may present with symptoms of acute rhinitis or a common cold. However, a feeling of malaise with hyperthermia and leukocytosis are conspicuously absent in scleroma. Later in Stage I, the early catarrhal symptoms become mucopurulent. There is a profuse, brownish discharge from the nose associated with pus formation within the nasal chambers. During this atrophic phase of Stage I, it is difficult to distinguish atrophic nasal mucosa of scleroma from that of ozaenae (atrophic rhinitis) or leprosy.

Stage II (granulomatous stage) is a proliferative stage in which nodular granulomatous outgrowths swell to fill the entire fossae of the nose. The nasal tip and the alae become enlarged and rigid. The lower nasal cartilages fan out and the soft tissue between them distends, producing an elephantiasis-like deformity. At this stage, scleroma may clinically mimic carcinoma of the nose. Most patients present to their physician during this stage of the disease. It should be noted that during this stage of the disease, the characteristic Mikulicz cells and Russell bodies are most often present. The Frisch bacilli are usually abundant during this stage and readily cultured.

In Stage III (cicatricial stage), gradual development of fibrosis occurs with progressive scarring of the area. Visible contraction and deformities of the affected parts are seen as the inflammatory mass is replaced by extensive dense scar.

Reports of Cases

Case 1. In February 1973 a 23-year-old Mexican-born man was seen because of nose bleeds. The patient's history indicated that catarrh with nasal congestion had been present throughout most of his life. Because of nasal obstruction, a tonsillectomy was carried out in Mexico in 1970. Continued progressive nasal obstruction necessitated surgical operation on the nasal septum in Mexico in 1971.

In the present illness, there was a history of progressive nasal obstruction with foul smelling, brownish discharge. On examination, ballooning of the nasal alae was noted. The nasal septum was extremely widened bilaterally and inferiorly. There was total nasal obstruction due to extremely hard, keloid-like tissue filling the nasal cavity bilaterally. On examination of the oral cavity, thick granulation-type tissue along the free border of the soft palate and lateral pharyngeal bands was seen (Figure 3). Dense scarring of the soft palate had completely eliminated the
In July 1974, because of persistence of disease and recurrence of symptoms, the patient returned for examination. Recurrent bilateral nasal masses were noted with thick crusting along the nasal mucosa, and a brown, foul-smelling discharge was seen in the nasopharynx. The nasal fossae were stenotic with ingrowth of thick, firm scar tissue. Polytomography of the nasal fossae showed intact bony margins of the nose and paranasal sinuses. The patient was taken to the operating room, where repeat biopsy confirmed the persistence of nasal scleroma. Disc sensitivities were carried out, and tetracycline proved to be the drug of choice. Administration of doxycycline hyclate (100 mg twice a day for six weeks) was begun.

Results of follow-up examinations in July 1975 showed excellent resolution of the scar tissue with a patent nasopahrynx. The lateral pharyngeal band granulation tissue had regressed completely, and the foul-smelling discharge had disappeared.

CASE 2. A 17-year-old Mexican-born girl was seen in March 1963 because of difficulty breathing through her nose. The patient gave a three-year history of chronic nasal obstruction with catarrh. In 1962 nasal surgical procedures had been carried out in Mexico to relieve pronounced obstruction. On examination, protruding granulomatous tissue was noted which completely filled both nostrils and which bled when touched. Large granulomatous lateral pharyngeal bands were seen on examination of the throat. Findings on x-ray studies of paranasal sinus and chest were normal. Biopsy was carried out and the finding reported as “possible leprosy.” The patient was brought to surgery for additional surgical excision of the proliferative mass. The pathology report identified the mass as scleroma, and Giemsa stains indicated the presence of Gram-negative diplobacilli (Figure 4). Administration of chlortetracycline hydrochloride was begun. Throughout the antibiotic therapy, nasal discharge and crusting continued to be present.

The patient was seen again in December 1963 with recurrence of large granulomatous tissue along the lateral pharyngeal bands and total nasal obstruction. The excessive tissue was treated with electrocautery and administration of chlortetracycline hydrochloride was begun again. The patient returned every six months because of recurrent disease and had to be taken to the operating room several times. In May 1967 the granu-
loration tissue had replaced both the right and left lateral pharyngeal bands. The uvula was no longer distinct but had been replaced by tense scar tissue along the free stenotic border of the soft palate. Repeat culture and disc sensitivities were carried out and chlortetracycline was shown to be the treatment of choice (Figure 5). However, the patient returned pregnant at this time and refused to accept further antibiotic therapy. She was not seen again.

**Therapy**

Quevedo reports that streptomycin administered for 21 days every three months over an average period of five years appears to give the most favorable response. Streptomycin is effective given in large doses when scleroma is limited to the nose but not when the lower respiratory tract is involved. In such patients, the addition of tetracycline usually renders a good result. Treatment is started with 0.5 or 1 gram of streptomycin each day for two full weeks, depending on the tolerance of the patient to the drug. After one or two months, 1 gram of tetracycline is given every day. It is necessary to repeat this treatment periodically. Administration of corticosteroids is helpful in the granulomatous stage of the disease, during postoperative recovery and in patients with complete stenosis of the nose. Findings on sensitivity tests of Frisch bacilli most often have shown tetracycline to be the treatment of choice. Disease limited to the nasal area may be effectively treated by partial rhinectomy in conjunction with tetracycline therapy. In all cases, identification of the organism should be achieved and antibiotic sensitivities obtained before beginning therapy.

**REFERENCES**


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