RHINOSCLEROMA

REPORT OF CASE WITH EXTENSION TO THE INTESTINES

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Rhinoscleroma in its pathological and symptomatological aspects has of recent years closely attracted the attention of the medical world.

Recognition of its increase in frequency was made by the International Congress of Otorhinolaryngology, held at Copenhagen in 1928, on which occasion localities heretofore free from this disease reported occurrence of cases. It was thereupon decided to make a wide investigation, the results of which should be announced in a congress which proposed to assemble in Spain in 1931 during the month of September—all of which plans were carried out.

We are aware that rhinoscleroma has been known for years in Central America, because more than forty years ago Alvarez was able to study it in the Republics of Guatemala and Salvador, countries which contain a large percentage of indigenous population. The situation is somewhat different in Costa Rica, where, as Peña Chavarría and Nauk [1] noted in 1929 (the date of the publication of some papers on the clinical and pathological aspects of rhinoscleroma), the course of four years' local hospital practice had yielded only four cases. Nevertheless, this disease seems to be on the increase in Costa Rica, as during the last few months the writers have had the opportunity of studying four other cases. This increase seems to have been general, apart from the Continent of the Americas, as Machulko-Horbatzewitsch [2] has noticed it in Ukranie.

Of the last four cases studied by us, we have been able to make an adequate necropsy on two, as the patients died from complicating diseases. During one autopsy we found a rhinoscleromatic lesion so curiously localized that we believed it to be worthy of special mention—hence this paper. Nowhere else have we found mention of intestinal rhinoscleroma; indeed, Askanazi [3] declares that "Rhinoscleromatic metastasis is not observed in any other organs than those of the respiratory

tract.'' He also applies the same statement regarding the bacillus of Frisch.

The following are the particulars regarding the patient in whom we minutely studied Frisch's Disease:

T. G. F. Day laborer, aged 19. Half-caste of Nicaraguan origin. With a few absences, this patient has been hospitalized here more or less permanently for three years. On account of complete laryngeal obstruction, aphonia, and attacks of suffocation, we performed a tracheotomy in 1928, placing a cannula, which the patient himself daily removed and cleaned. The youth told us that the disease had begun at the age of eight years, and he associated its beginning with a traumatism caused by a blow. One morning, while cleaning the tube, he became asphyxiated and died shortly afterwards. As we observed at autopsy, the rhinoscleromatic lesions were laryngeal, but had not attacked the soft tissues of the nose and cheek.

Autopsy: No. 1360. 1932. Body of a youth well proportioned and well nourished. In the nostrils and contiguous skin, no enlargement, hardness, or rhinoscleromatic lesion is observed. There is no marked narrowing of the nasal cavities. The mucous membrane is smooth, gray, and a little thickened. In the nasal septum one observes some retracted scars. The mucous membranes of the pharynx, of the bucal cavity, of the tongue, and of the faucial isthmus, have no pathologic lesions. The epiglottis, the arytenoepiglottic and the pharyngoepiglottic folds, are very much thickened and hardened, and are covered with a smooth mucosa, light gray in color. The wall of the larynx is thickened, hard and rigid, as are also the true and false vocal cords. Below the vocal cords, especially on the anterior wall, one sees some very retracted lesions. As an effect of the sclerosis, the trachea, which has a diameter of about 1 cm. is transformed into a rigid tube, the wall of which measures 0.7 to 1.2 cm. in thickness. On section one perceives numerous segments of cartilage and bone. The surface is, in parts, smooth and light gray in color, and in some places is covered with dark red matter, which in its turn covers superficial ulcers. In the upper part of the trachea one sees the opening of a tracheotomy, diameter 1 cm. with coarse, roughened edges. The same type of lesion, though less severe, is found in the principal bronchi, and in the major intrapulmonary bronchi. The superior cervical lymphatic glands are

not enlarged. In the lungs, especially in the upper parts, one finds extensive tuberculous lesions in the form of small, round, irregular nodules, partly caseated in the centre. The lymphatic tracheobronchial and lower cervical lymph glands are large, hard, and sprinkled with little nodules, yellowish-gray in color. In the spleen and liver we find numerous tubercles. The spleen is slightly enlarged. On section, the malphigian follicles are clearly visible. In the cut surface of the liver (1555 gms.) one distinguishes the outline of the lobules. Very little blood exudes. In the right suprarenal capsule and in both kidneys one sees extensive caseated lesions. There are no tuberculous lesions in the intestine or in the mesenteric lymph glands.

Commencing in the ileocecal valve, the cecum and the ascending colon are covered with numerous button-like nodules, whose diameter measures approximately one half cm. The mucosa covering the nodes shows no changes; the lesions decrease in number until they gradually lose themselves in the hepatic flexure of the colon.

A histopathologic examination reveals to us the nature of this curious intestinal lesion. The tissue of the sub-mucous membrane examined with slight magnification, shows a proliferation clearly composed of numerous large vacuolated cells with relatively small pycnotic nuclei, Mikulicz cells. Under the mucosa there are large foci formed exclusively of these cells, in the middle of which many plasma cells stand out prominently mixed with small lymphocytes. There are also many polymorphonuclear leukocytes, a large number of them eosinophilic, situated particularly beneath the epithelium. Joined to the Mikulicz cells one sees some hyaline cells full of droplets, which by their precise formation appear as a rosette around the nucleus.

The muscular coat of the intestine is not infiltrated; the *muscularis mucosae* is fairly well preserved, even though here and there are noted tiny gaps where the mucosa is lacking. That which remains is infiltrated with cells. The glands are more separated one from another than is normal and there are in them fewer Mikulicz cells, and more leukocytes than in the submucous membrane. Included in the tissue of the rhinoscleromatic granulation we meet isolated lymphatic nodules which often show germinal centres. By means of the microscope we can clearly see encapsulated bacilli in some of the

Mikulicz cells, especially in those situated below the mucosa. In the mesenteric lymph glands no rhinoscleromatic lesions are seen.

The tissue of the epiglottis shows an infiltration formed of plasma cells and lymphocytes. There are a few Mikulicz cells, but hyaline cells exist in a slightly larger quantity. The epithelium and cartilage show no alteration. The epithelium of the trachea is transformed into a thick stratified squamous layer that is keratinized. The epithelial intrapapillary prolongations are quite lengthened and have penetrated into the tracheal wall. In the lower portion of the trachea one finds some superficial ulcers. The wall of the trachea is greatly thickened and infiltrated with numerous plasma cells. Below the epithelium are foci formed almost exclusively of Mikulicz cells. The connective tissue is thick, forming broad cords which divide the masses into separate nodules. The smooth musculature is in part hypertrophied. One sees destruction in nearly all the elastic fibres—indeed, some parts consist only of remnants, shrunken, and deeply stained.

The cartilaginous rings in some parts have become triangular due to a conical proliferation of their internal aspect. The cartilaginous capsules are large, irregular, and contain several nuclei, especially in the region of the proliferation. The peripheric portions of the cartilage are generally calcified. In many parts one finds areas of ossification with the formation of bone medulla that is rich in cells. At times one sees two cartilaginous rings bridged together by bone and cartilage. In other parts of the trachea these rings are partially destroyed, thus causing the rest of the cartilage to assume an irregular, indented form. In the interior covering of the wall, numerous islands of cartilage and bone are seen. The newly formed bone is generally surrounded by a fringe of osteoblasts, and in some spots one sees osteoclasts in a large number, situated in bone cavities.

In the large bronchi one finds similar lesions, tho' less pronounced. Mikulicz cells are here much more numerous than in the trachea, and are also situated in the deeper layers. As the bronchial walls are much more delicate than those of the trachea, the proliferations of cartilage and bone reach up to the epithelium and force it into the lumen. In the small bronchi we see no rhinoscleromatic lesions.

The cervical lymph glands show extensive tuberculous lesions, but none of rhinoscleromatic origin.

The result of this autopsy proves a fact which we consider important for the pathogenic study of rhinoscleroma, and this is that the lympathic tissue and especially the lymphatic nodes of the intestine sustain little damage so far as we have determined; neither do we find rhinoscleromatic lesions in the mesenteric and cervical lymphatic glands though in relation to this last statement, we must take into consideration that possibly they may have been veiled by existing tuberculous lesions.

Even though other workers have encountered the bacillus of Frisch in lymphatic nodules, we do not imagine multiplication takes place in such tissue, for did it do so, it would produce anatomical lesions which we have not been able to find, neither which have been discovered by other investigators.

This relative immunity of the lymphatic tissue possibly explains the rarity of the rhinoscleromatic intestinal lesions. One can easily imagine, in the case under discussion, that the intestinal infection was caused by the deglutition of the Frisch bacillus—a method of infection analogous to that of intestinal tuberculosis. Nevertheless, in contrast to the bacillus of Koch, which chiefly affects the lymphatic tissue of the intestines in which the first tuberculous changes develop, the Frisch bacillus apparently does not enter the lymphatic nodes of the intestine. Only thus, in such a chronic infection as rhinoscleroma, can the rarity of intestinal lesions be explained. The localization of rhinoscleroma in the cecum may be explained by the fact that excrement rests there for a longer period of time. Whether the Frisch bacillus is able to penetrate intact intestinal mucosa, or whether small imperfections in it are necessary for infection, has not been determined up to the present. In this particular case at least, we discovered no anterior lesions.

There is no agreement, up to the present, in those who have studied the pathologic problems of rhinoscleroma to prove those elements which have given origin to Mikulicz cells. Many authors think they come from plasma cells. In our opinion one cannot solve this problem by simple morphological deductions, because in spite of the fact that the Mikulicz cells and plasma cells are always united and intermingled, and that there is an inverse relationship in their

frequency, it is also an evident fact that the nuclei of the Mikulicz cells are always pycnotic and one does not recognize in them the typical structure of the nucleus of plasma cells. We and other authors consider the most reasonable belief to be that the hyaline cell is the result of the transformation of the Mikulicz cell. In the first, one sees almost always the pycnotic form of the nucleus, the vacuolated protoplasm, and in addition, the size coincides with that of the Mikulicz cells. In relation to the number of Mikulicz cells that one finds in rhinoscleromatic tissue, we have discovered that the more cells there are, the more acute and progressive is the spread of the rhinoscleromatic lesions. In the case described, we found a very scanty sprinkling of Mikulicz cells in the epiglottis; they were more numerous in the trachea, and the numbers were considerable in the bronchi. In the intestine, on the contrary, the majority of cells in the infected tissue were those of Mikulicz. This difference corresponds with the age of the lesions. Probably, Frisch's bacillus or its toxins, have some influence on the development of the Mikulicz cells. Thus it is indicated, and indeed we have been able to confirm it as a fact, that Frisch bacilli are practically always found within the Mikulicz cells, and therefore the possibility arises that the proportion of Mikulicz cells depends on the number or virulence of the bacillus Frisch which infects the tissue.

It appears as if there were a definite relationship between the number of Mikulicz cells in the tissues and clinical symptoms.

R. H. aged 11, came to the Hospital for the first time in August, 1928. He told us that the trouble had begun at the age of four years in consequence of a neglected coryza, which produced a slight respiratory difficulty. A biopsy was made on the date of admittance to the Hospital. In the section only a few cells of the Mikulicz type were found. The infiltration was composed almost entirely of plasma cells. This stage of the disease was, we believe, stationary, for in the four subsequent years the progress was little advanced. In 1932 the patient again entered hospital, as his condition had become more severe during those last few months. A second biopsy showed that the majority of the infiltrated cells were those of Mikulicz, intermingled with a few plasma cells. This

period of the disease, we believe, was a period of exacerbation.

At the present time (August 1932) the patient is fifteen years old and is slightly anemic. On examination of the head we note congestion of the conjunctiva, caused by circulatory difficulties arising from the contracting scleromatic tissues, which have caused occlusion of the lacrimal ducts. As may be seen by the accompanying illustration, there is malformation of the nostrils, one of which, the left, is completely closed, and the right partially so. The mucosa of the palate is congested, but so far no rhinoscleromatic invasion has taken place, and in the case of this patient (contrary to what was found in the former case under discussion) the lesion has extended to the exterior, localizing itself in the soft tissue of the nose. As an etiological point of importance we wish to note that this patient has two brothers, both attacked by scleroma.

The third case under observation was also a great help to us. The patient, a boy of nine years, who had suffered from rhinoscleroma for about one year, died of an intercurrent illness—lobar pneumonia. The progress of the disease ran a slow course, but had extended all over the interior, invading the nasal mucosa, the epiglottis, the larynx and the upper third of the trachea. Histologically, we found a very dense infiltration of plasma cells. In a few sections only did we find a scanty sprinkling of Mikulicz cells.

Nevertheless, in clinically doubtful cases in which histologically no definite picture is found, one must resort to a bacteriological examination, conducted in the following manner and recommended by us on the grounds of experience: Disinfect the skin or mucosa thoroughly, and make a small incision in a manner that leaves the lips of the wound well separated. For the cultures one extracts samples from the bottom of the wound without touching the sides, thus avoiding contamination with other bacteria. Generally one obtains in this manner a pure culture of Frisch's bacillus even in the first inoculated tube.

SUMMARY

Three cases of rhinoscleroma are presented, autopsy studies of two being described.

Intestinal lesions with Mikulicz cells and the *Bacillus rhinoscleromatis* were found in one. The number of Mikulicz

cells in any given lesion varies in direct relation to the activity of the disease, so that these cells may not be found in tissue examined during a period of quiescence. In clinically suspicious cases not presenting Mikulicz cells on biopsy, samples should be taken for culture of the bacillus of rhinoscleroma; technique for this procedure is described.

The occurrence of rhinoscleroma in three members of the same family is considered worthy of note.

From general observation, rhinoscleroma seems to be on the increase in certain parts of the world.