

SCLEROTIC HYPERPLASIA OF THE PHARYNX AND NASO-PHARYNX.

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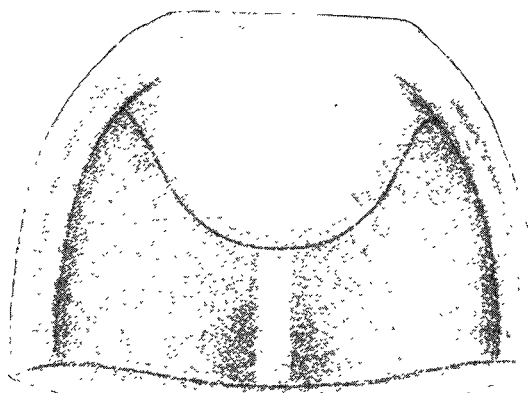
THE case upon which this paper is based is unique in my experience and in that of others, if one may judge from the unsuccessful result of an extensive literary search for references to a similar affection. I therefore believe myself justified in regarding the condition as a hitherto undescribed pathological entity which may not inappropriately be termed "sclerotic hyperplasia of the pharynx."

The patient was brought to me in August, 1899, by his medical adviser, Dr. R. T. Halliday, on account of enlargement of the uvula. He was then 34 years of age and was engaged in a shipbuilding yard. For about eight years he had been subject to slight attacks of sore-throat which had caused, however, no special inconvenience. Three years before I first saw him he began to feel something in the pharynx which he wished to expel; this sensation had gradually increased. Latterly choking fits had been excited by the "something" coming forward on the tongue or passing too far back, as was apt to occur while he was being shaved; he obtained relief in the one instance by pushing the offending body back with his finger, in the other by throwing forward his head. In either case if he did not at once succeed in getting it into its normal position he became panic-stricken and fell to the ground writhing. While in this state he lost control of himself and being a powerful man he was capable of doing great harm. He had, therefore, to avoid being in a crowd, and the barber and others had to be warned to stand clear of him at once should a choking fit set in. In addition, nasal respiration had been partially obstructed for a year, so that he was much troubled at night by the throat becoming dry. The patient was a tall, strong Highlandman who had had no serious illness and no venereal disease. His father and mother both lived to about the age of 70 years and left a family of 12 children, all of whom grew up to be adults.

On examination the pharynx presented a very striking appearance. The uvula was much larger than any I had ever seen and hung like a curtain, concealing the posterior pharyngeal wall. Its length was over one and a quarter inches, its width at the base was one inch, and in thickness it was also greatly increased. The shape and surface were in no way abnormal. The colour, however, was pale and the consistence was fibrous, so that the whole formed a firm unyielding mass.

On bringing the uvula forward on the dorsum of the tongue the posterior wall of the pharynx was found to present a peculiar aspect. A narrow tract of apparently normal tissue ran down the middle of the posterior wall. On

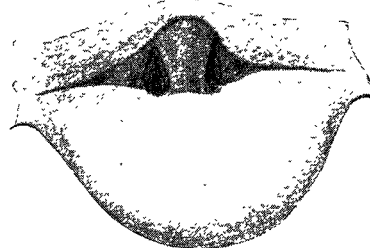
FIG. 1.



each side of this, extending outwards to the lateral wall and incorporating with it part of the right, and the whole of the left, posterior faucial pillar, was a broad prominent band which was thicker in its outer part. These bands passed upwards into the naso-pharynx and downwards to the upper end of the œsophagus. Their consistence was that of muscle and they could be pushed *en masse* towards the middle line. Their colour was a uniform

grey and their surface was perfectly smooth. Fig. 1 shows the pharyngeal aspect after uvulotomy. Examination of the naso-pharynx, which was possible only after uvulotomy, revealed marked changes (Fig. 2). The lumen of

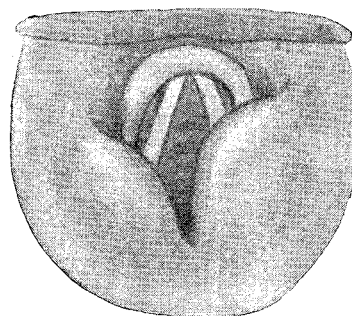
FIG. 2.



the cavity was reduced so as barely to admit a forefinger; this was the more striking in view of the patient's height (six feet two inches) and large build. The diminution in the space between the soft palate and postero-superior wall of the naso-pharynx was due partly to the continuation upwards over the latter of the bands above described and partly to thickening of the soft palate. The thickening on the roof, unlike adenoid vegetations, was least in the middle and most marked at the sides. Only a small portion of the septum and middle turbinates could be seen; the choanal arches and Eustachian orifices were concealed. The thickenings in the naso-pharynx, like those in the pharynx, were smooth, grey, and firm. By means of the laryngeal mirror the bands on the posterior wall of the pharynx were seen to pass downwards and to overhang the arytenoids, the sides of the epiglottis being in contact with the bands.

At a subsequent visit, in August, 1899, the greater part of the uvula was removed by means of the galvano-cautery snare without any untoward result. Since then the patient has experienced no discomfort and has considered himself cured. During the interval that has elapsed I have failed—except on the occasion noted below—to detect any change in the conditions described, and Fig. 1 and Fig. 2, which were made shortly after he came under my observation, illustrate the appearances at the present time. The occasion of the exception referred to was in October, 1900. After undue exposure he contracted a sore-throat which was more severe than any he had ever experienced. I saw him on the sixth day when the pain, which was confined to the left side, had begun to subside. There was then great swelling of the left band, especially about the level of the epiglottis; its surface was smooth and pale. Three weeks later the œdema had diminished considerably, so that a view of the larynx was obtainable; it was then found that the movement of the left arytenoid during phonation produced a clicking sound owing to its coming into contact with the overhanging tissues. Although five months have elapsed since this acute attack the left band continues rather thicker than the right and the clicking sound is still heard. Fig. 3 shows the appearances now

FIG. 3.



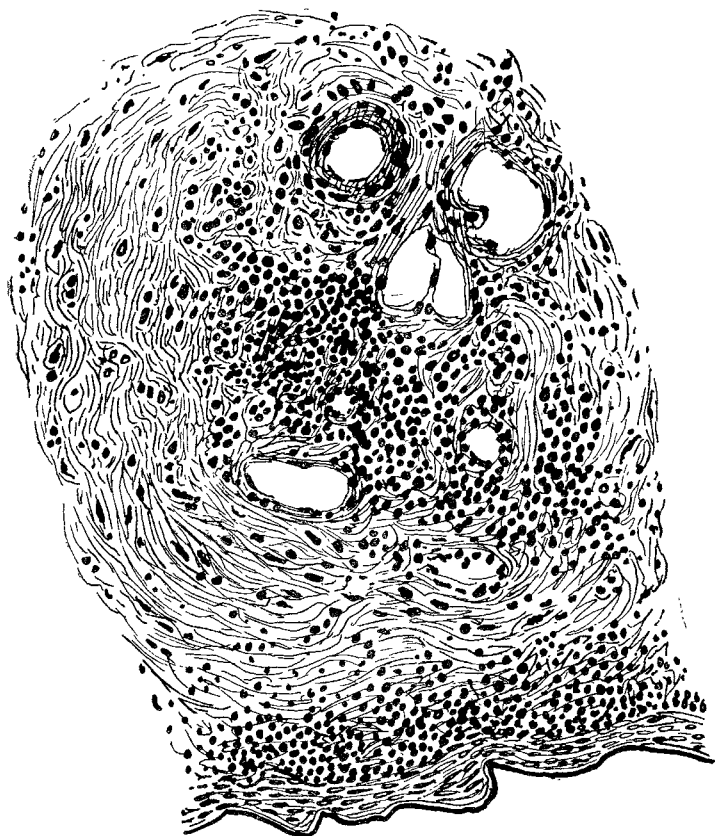
observed on making a laryngoscopic examination. The piece of uvula removed was preserved in absolute alcohol and the report on its minute structure by Dr. A. R. Ferguson, senior assistant to the Professor of Pathology in the University of Glasgow, follows:—

The piece of uvula submitted was incised longitudinally and paraffin sections were prepared from each portion. The section represents the entire cut surface so obtained.

Low power.—Almost the entire uvula is seen to consist of dense connective tissue of sparsely cellular character and fibrillated in appearance. This is disposed in denser whorls around the vessels, especially the arteries. Immediately surrounding the majority of the

vessels are dense areas of leucocyte infiltration. An irregularly distributed leucocyte infiltration is seen also running beneath the epithelial covering of the uvula. This latter is much compressed in appearance, the corneal layer especially being increased in thickness. The Malpighian layer shows no papilliform processes in any part

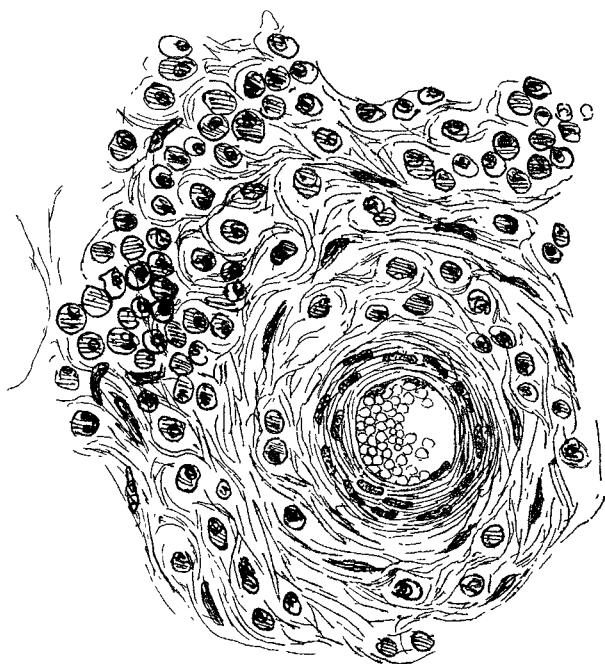
FIG. 4.



(Fig. 4). Occurring somewhat sparsely throughout the tissue are small groups of adipose vesicles, surrounded and in many cases diminished in size and separated by the dense connective tissue. Mucous gland tissue is abundantly present in the central portion of the uvula. The glandular tissue proper does not seem to have suffered to the same extent as the other portions in the prevailing fibrosis. The interstitial tissue of the glands, however, is densely infiltrated with leucocytes which are disposed in irregular areas. In the more central parts of the uvula the matrix of the connective tissue is more fibrous than at the periphery and the corpuscles correspondingly fewer in number. The vessels are small but comparatively numerous throughout; they are, without exception, surrounded by an irregular zone of leucocyte infiltration. A few striped muscle fibres, much interrupted and in great part completely isolated, are seen traversing that part of the tissue in which the glandular elements lie.

High power.—The epithelial cells of the surface are much altered by compression, having undergone a change comparable with that in the keratinous layer of the epidermis. The deeper layer shows no papillary processes, though the cells are well formed, their columnar character

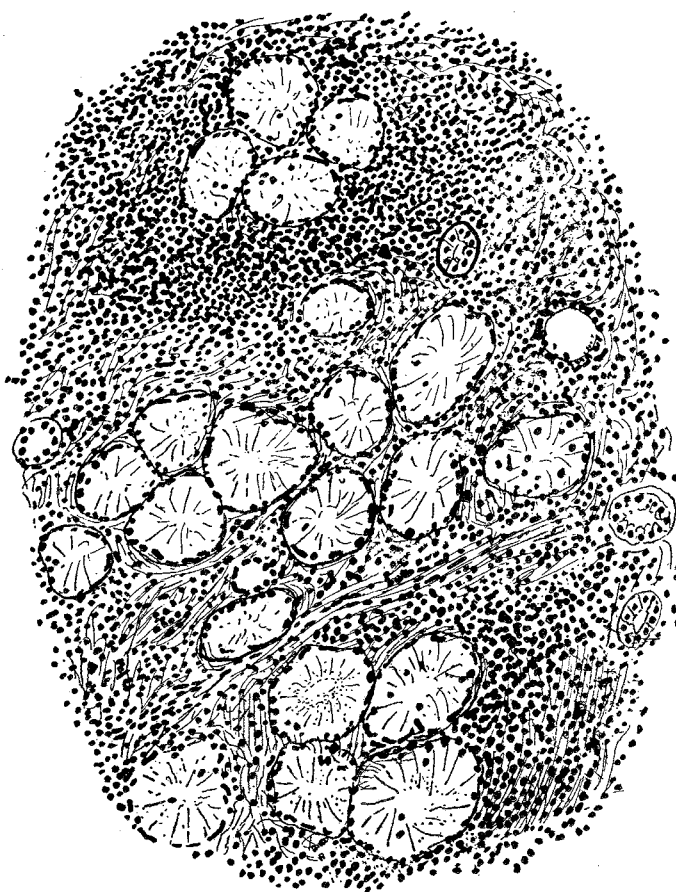
FIG. 5.



being pretty well preserved. The tissue beneath this layer is a well-formed connective tissue which is finely fibrillated, the fibrillae forming a very fine-meshed reticulum. The small vessels in this zone are surrounded by leucocytes. These latter are of the small mononuclear

type mainly, though many polymorphonuclear leucocytes are also seen. The other cells present are connective tissue corpuscles, many of which are observed in process of division. In this zone of the connective tissue many of the corpuscles have an irregular form with delicate processes; in the main, however, they are of the spindle-shaped type met with in young developing connective tissue (Fig. 5). The fibrillar reticulum can still be distinguished in the midst of these cellular areas. The degree of leucocyte infiltration is more pronounced in the interstitial tissue of the mucous glands than elsewhere. The mucigenous cells show no alteration in appearance, the majority being collapsed with very indefinite outline (Fig. 6).

FIG. 6.



A small portion of the affected tissue was removed from the patient under conditions which precluded its contamination with micro-organisms from any source, and this, after having been cut into small pieces with sterilised scissors, was introduced into a series of sloped glycerine agar tubes. These showed in the course of the second 24 hours of incubation at 37° C. a few small colonies resembling in general appearance, but somewhat larger than, those of streptococcus pyogenes. Film preparations from these colonies showed a short, beaded bacillus which, in forms staining deeply at the extremities, resembled a small diplococcus. Nothing in the least suggestive of bacillus rhinoscleromæ was obtained in the cultures.

Microscopical sections of the uvula were stained by various methods for the demonstration of organisms—e.g., carbol-thionine blue, Loeffler's alkaline methylene blue, Kühne's blue, Gram's method. These, carefully searched, showed no bacillus to be present in the depths of the affected tissue in any part. A few organisms, however, resembling that briefly described above as having been obtained from cultures, were seen between and immediately beneath the deeper cells of the Malpighian layer of the mucous membrane and were only found in this situation. These, it is accordingly inferred, represent but accidental invasions from the surface. The whole condition may thus be briefly summarised, from the point of view of its morbid histology, as a chronic hyperplasia of the interstitial tissue of the parts involved, of apparently progressive character, with no tendency towards degeneration, and (so far as can be demonstrated) unassociated with the presence of any specific micro-organism.

This case presents three striking features—namely, the greatly enlarged uvula, the thick band descending in each half of the posterior wall of the pharynx, and the thickening of the roof and floor of the naso-pharynx leading to a marked diminution of its lumen. In all the regions affected the morbid process is apparently the same, consisting in diffuse uniform thickening, which histologically proves to be a marked interstitial hyperplasia. The development of the disease has been very slow, and is probably still proceeding. In endeavouring to diagnose this condition several diseases have come under consideration.

1. *Rhinoscleroma.*—In this disease the naso-pharynx is the region most frequently involved and its aspect may come to resemble that presented by my patient. In rhinoscleroma, however, the reduction in the lumen of the naso-pharynx is due to cicatricial contraction and not as here to thickening of the tissues. Further, no description corresponding with the appearances of the uvula or posterior wall of the pharynx

in this case could be found. The fact, also, that rhinoscleroma has been observed in this country only in persons who have come from districts in which the disease is endemic renders it highly improbable that my patient's ailment is of this nature. Lastly, the absence of the specific bacillus and of Mikulicz's cells almost conclusively disproves rhinoscleroma.

2. *Tertiary syphilis*, owing to the various aspects it may assume in the throat, naturally suggests itself. Inquiry as to the patient's previous health yields no indication of his ever having contracted venereal disease; besides, this morbid process differs from tertiary syphilis in its perfect symmetry, in the absence of any tendency to ulceration, and in its being uninfluenced by iodide of potassium.

3. *Hereditary syphilis*.—Recently I have had under treatment a boy, aged seven years, the subject of late hereditary syphilis. In addition to laryngeal manifestations his palate and uvula presented a smooth uniform thickening which, although less marked, resembled that of the case under consideration. The greater part of the uvula was removed and Dr. Ferguson found, on examining it microscopically, that the enlargement was due to a diffuse fibroid hyperplasia which in its main features closely resembled the histological appearances presented in sections from the case the subject of this paper.

In consequence of the similarity of the histological appearances in the two uvulas it has been necessary to consider—(1) whether the man is the subject of late hereditary syphilis, and (2) whether the sclerotic hyperplasia in the child is independent of syphilis.

I have failed to discover any fact in support of the first question. The patient's parents spent their married life in the Highlands and had a family of 12 children, all of whom reached adult life, the physique of every member being above the average. The patient is the second youngest child and presents none of the stigmata of the diathesis. Owing to the want of corroborative evidence in the personal and family history of the patient, and because of the general dissimilarity of his pharyngeal manifestations to those observed in hereditary syphilis, I believe that this disease may be left out of account as an etiological factor.

The second question, as to whether the involvement of the pharynx in the boy is independent of syphilis, must be answered in the negative. That he suffered from hereditary syphilis when an infant is beyond doubt. It is also evident that the affection of his pharynx is of the same nature as that of his larynx and that the latter resembles the diffuse hyperplastic infiltration of the larynx which has been described in a few cases of hereditary syphilis. On these grounds the hyperplasia of the boy's uvula may be attributed to the diathesis.

While I have been unable to find a disease of which the sclerotic hyperplasia in the man is a manifestation, I would point out that an analogous condition probably exists in subglottic hypertrophic laryngitis. This affection is usually characterised by the presence of pinkish, smooth, firm, symmetrical folds beneath the vocal cords, which develop slowly, apparently in consequence of recurrent inflammatory attacks, during which they become more or less swollen. If we now turn to the case under discussion we find folds of thickened tissue in the pharynx presenting characters similar to those just mentioned; we get a clinical history of his having been subject to sore-throat for years, and of permanent symptoms due to hypertrophic changes having set in only at a comparatively recent date; examination during an inflammatory attack revealed marked swelling of part of the affected region; and, lastly, Dr. Ferguson informs me that A. Sokolowski's¹ account—which is probably the most detailed and thorough—of the histology of subglottic hypertrophic laryngitis might stand for that of the removed portion of the enlarged uvula, the sole difference being that in our case the deeper layer of the epithelium is not thrown into papillæ, but presents practically an even continuous surface, the columnar character of the deepest cells of the rete Malpighii, however, being maintained.

Considerable discussion has taken place as to the nature of subglottic hypertrophic laryngitis. In most text-books the causes mentioned are tuberculosis, syphilis, and rhinoscleroma, the last being probably the commonest. Of 100 cases of scleroma recently reported by A. Baurowicz² 34

were affected with subglottic hypertrophic laryngitis. If the frequency of subglottic hypertrophic laryngitis in scleroma be coupled with its great rarity apart from this disease it can be understood how certain observers who live where rhinoscleroma is prevalent—e.g., P. Pieniazek³ and Baurowicz of Cracow—and who consequently see subglottic hypertrophic laryngitis comparatively often, maintain that it is *always* a manifestation of scleroma. On the other hand, Sokolowski of Warsaw, who has also had an extensive experience of rhinoscleroma, denies this, and proves uncontestedly, as it seems to me, that subglottic hypertrophic laryngitis may develop independently of scleroma, tuberculosis, and syphilis; he thinks that the morbid process is of a specific nature, the etiological factor being as yet unknown, but he suggests that hereditary syphilis may play a part. A. Kuttner⁴ reports a case of subglottic hypertrophic laryngitis in which there were no grounds for suspecting the presence of any of the infective diseases mentioned; he maintains, therefore, that this laryngeal condition may appear as an affection *sui generis* which he proposes to term the genuine form of chondritis vocalis inferior hypertrophica. F. H. Bosworth⁵ also states that there can be no question of a simple idiopathic inflammatory process in the subglottic region giving rise to marked hypertrophy, but he thinks that in most instances a diathetic condition is present. There are thus good reasons for according to subglottic hypertrophic laryngitis independent rank amongst the diseases of the larynx.

The facts brought forward in this paper, if correctly interpreted, prove (1) that the pharynx and naso-pharynx may be the seat of a sclerotic hyperplasia unconnected with syphilis, rhinoscleroma, or other known infective disease; (2) that a similar morbid process may manifest itself beneath the vocal cords as subglottic hypertrophic laryngitis; and (3) that in the hyperplastic variety of hereditary syphilis the histological appearances closely resemble those of the above described sclerotic hyperplasia.

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ON THE EXISTENCE OF IMMUNITY AFTER ENTERIC FEVER.

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THERE seems to be a very general impression that an attack of enteric fever confers some degree of immunity upon the patient against further attacks of the same disease. The number of patients whom I found suffering from enteric fever in South Africa who had previously contracted the disease in India or elsewhere has made me feel that this conclusion is not based upon very good evidence.

A nursing sister of my acquaintance had two attacks of enteric fever in England during her nursing career, and she contracted the disease for the third time whilst doing duty with the forces in South Africa. An officer had suffered from enteric fever in India, and he also contracted malaria in that country. During the campaign he again became a victim and his illness was followed by three distinct relapses. These relapses did not occur in the usual way during the week or so after the temperature became normal, but came on when the patient was on an improved diet and beginning to get about—i.e., from three to five weeks after the temperature had fallen. This was, of course, altogether an unusual case and may have been complicated by his malarial diathesis. The following two cases are interesting as showing that the supposed immunity produced by very severe attacks of enteric fever was not sufficient to protect the patients for more than six months from a recurrence of their illnesses.

CASE 1.—A civil surgeon, aged 30 years, previously in good health, was admitted to No. 7 General Hospital at Estcourt in May, 1900, with symptoms of enteric fever. The attack was a most virulent one, the patient suffering from

Chondritis vocalis inferior hypertrophica, Archiv für Laryngologie, Band vii., S. 349.

³ Heymann's Handbuch der Laryngologie, Band i., S. 1305.

⁴ Chondritis vocalis inferior hypertrophica, Archiv für Laryngologie, Band v., S. 275.

⁵ A Treatise on Diseases of the Nose and Throat, vol. ii., p. 529.

¹ Ein Beitrag zur Pathologie und Therapie der chronischen hypertrophischen Kehlkopfentzündung, Archiv für Laryngologie, Band ii., S. 68; also Band iv., S. 239.

² Das Sklerom auf Grund der Beobachtung von 100 Fällen, Archiv für Laryngologie, Band x., S. 396; Zur Ätiologie der sogenannten