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LUPUS OF THE THROAT AND NOSE.¹

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NOTWITHSTANDING the valuable contributions of Chiari and Riehl, of Lefferts, of Homolle, and of Lennox Browne, on the subject of lupus of the throat and nose, there are still many questions regarding this disease which have not yet received a satisfactory solution. How is it related to lupus of the skin? By what means is infection carried? Does it ever attack primarily the mucous membranes, and is it then capable of being diagnosed? What is the prognosis as to laryngeal stenosis resulting, and as to the danger of a pulmonary or a general tuberculosis being developed? And, lastly, what are the best methods of treatment we possess for arresting its progress?

These are the questions I desire to bring before this Society, in the hope that our discussion of them may help to establish some points at present involved in more or less obscurity.

First, as regards etiology. Lupus of the throat and nose may originate in three ways: (1) By direct extension of the external disease along the mucous membranes; (2) as a secondary deposit at a more or less distant part; (3) by a primary deposit in the mucous membrane, without any external disease whatever. The first condition we see every day in cases of lupus attacking the alæ of the nose at the junction of the skin and mucous membrane, and subsequently destroying the cartilaginous septum and the anterior ends of the lower turbinates. In many of these cases, I believe, the disease really originates in the mucous membrane, but does not come under our observation till it has spread considerably, and the exact point of origin cannot be determined. The preference of lupus for attacking this part is no doubt due to its liability to abrasion during nasal catarrh. Another way in which lupus spreads to the pharynx

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is by way of the gums and buccal mucous membrane. This, however, is not so commonly met with as the extension along the nasal passages.

The occurrence of lupus as a secondary deposit in the larynx or pharynx is by no means so rare as was at one time supposed.

The following statistics, referring to cases in which the larynx was involved, show how frequently this condition will be met with if all cases of external lupus be examined :—

Holm	...	in 90 cases found 6 per cent.
Chiari	... „ 68 „ „	8·8 „
Browne	... „ 25 „ „	14 „
Haslund	... „ 109 „ „	9 „
Marty	... „ 89 „ „	9·1 „

Putting the above results together, we find that, in 381 cases of lupus of the external parts, there was secondary deposit in the larynx in 8·9 per cent.

By adding to the above those cases in which the pharynx or palate were alone involved, the percentage becomes much higher. Lennox Browne sets it down at 20 per cent., and that figure agrees with my own observations of 30 cases examined recently.

In view of the statistics I have quoted, I cannot see the foundation for Schroetter's conjecture that the larynx will not be found to be involved in more than 3 per cent. of all cases of lupus.

By what means does the secondary deposit take place? We may regard it as now fairly established that lupus is a localised tuberculosis in which the tubercle bacillus is present in small numbers. Infection may then take place by way of the lymphatics, or directly by the implanting of the bacillus on an abraded surface.

As the lymphatic glands are almost never enlarged in true lupus, I am inclined to regard the secondary deposit as due to direct infection. In support of this view it is to be noted that all cases of secondary lupus in the throat or nose have followed on lupus of the face, and never lupus of the trunk or extremities. It is easy to imagine how secretions could be conveyed to the mucous membranes in such cases. Again, the parts most liable to infection—the uvula and epiglottis—are also those most liable to injury.

That lupus may also originate primarily in the mucous membrane of the pharynx or larynx is now established beyond all doubt. Quite a large number of such cases are on record by competent observers. Of these the most important are the cases of Chiari and Orwin, in which the subsequent appearance of the disease on the skin confirmed the diagnosis.

Are we able to diagnose with certainty cases of primary lupus when there is no confirmatory evidence on the skin?

I think we are; especially if we have the opportunity of watching the progress of the case for some time. Practically the difficulty of diagnosis is limited to distinguishing lupus from hereditary and tertiary syphilis. In lupus there are two general conditions of the mucous membranes of the larynx and pharynx which strike one in the majority of cases, anæmia and anæsthesia.

The anæmia is most pronounced in the larynx, and to a less degree in the pharynx.

It is not so extreme nor so general as in laryngeal phthisis, but forms a marked contrast to the general congestion observed in syphilis. The anæsthesia is a very constant and characteristic symptom; but in one of my cases, where tracheotomy had been performed, hyperæsthesia was so great that a strong solution of cocaine had to be applied before the larynx could be examined. Though this symptom of anæsthesia is dwelt upon by most observers as being present in lupus, I should mention that Ramon de la Sota, speaking from a seemingly large experience, states that he has not observed any change in the sensitiveness of the mucous membrane.

The early changes in lupus of the throat are best observed in the uvula and epiglottis. The uvula becomes thickened and nodular, the colour of the mucous membrane remaining normal or becoming slightly heightened. After a time superficial ulceration of a sluggish character, and with little deposit on its surface, commences on one of the nodules, and spreads slowly and quite painlessly till the whole uvula is destroyed, or heals at one part only to start afresh in another.

On the epiglottis I have watched a similar process. It becomes thickened and irregular along its free border with slightly raised pale or greyish-red eminences. These we may observe to become white at their apices, a slough forming, which on separating leaves a small ulcer, with sloping edges and greyish-yellow base, but without any surrounding hyperæmia or infiltration of its margin.

These ulcers are slow of healing, and as one cicatrises another nodule breaks down or is absorbed without ulcerating. In this way a worm-eaten appearance is given to the edge of the epiglottis, which is very characteristic of the disease.

As the process goes on the epiglottis becomes paler till it comes to have a dead-white colour, and to the probe is stiff, fibrous, and resistant. Sometimes its free border is eaten into irregularly, so as to leave in it a triangular deficiency, or the whole border may be destroyed, as if cut across. If we are able to watch a case progressing as I have described, I think it is so entirely different from the active destruction which takes place in syphilis as to prevent any mistake in diagnosis. The laryngoscopic appearances, in fact, are far more allied, as Lennox Browne remarks, to tubercular disease than to syphilis, and yet the absence of cough, pain, emaciation, fever, etc., make the differential diagnosis from laryngeal phthisis very easy.

With regard to the painlessness of the ulceration, though this is the rule, there are exceptions to it. In one of Leffert's cases there was marked dysphagia, and in one of my own, where ulceration of the pharynx and epiglottis had proceeded painlessly, there was a considerable degree of pain on swallowing, while an ulcer on the summit of the right arytenoid remained unhealed.

In the vast majority of cases, however, the painlessness of the ulceration in lupus is a very striking feature.

In syphilis we often find extensive ulceration with comparatively little

pain, but never in my experience is it so completely absent as in lupus, where the patient seldom knows of the existence of the disease until it interferes with voice production.

The cicatrices of lupus are in no way characteristic of the disease, and from them alone I do not think a diagnosis can be made with certainty. The recurrence of ulceration in them, however, as Chiari first pointed out, is pathognomonic of lupus.

Lennox Browne has advanced, as distinguishing syphilis from lupus of the soft palate, that the latter always attacks the buccal surface, and the former the nasal.

In one of my cases, however, along with lupus of the nose, there was on the upper surface of the soft palate a mass of nodular, or warty-looking tissue, which blocked the choanæ, and hid more than half of the septum. Of course, this may have been adenoid tissue, as there was no ulceration present ; but beyond the fact that there were no vegetations on the vault, and that the whole pharynx was atrophic, is to be noted that an exactly similar condition is described by Chiari as occurring in one of his cases.

The limitation of lupus to the supra-glottic region has been suggested as distinguishing it from syphilis, but no great reliance must be placed upon this, for the invasion of the true cords is only a question of time and the severity of the case.

Do not think they possess any immunity from attacks of lupus, but that as the disease always begins in the epiglottis or ary-epiglottic folds, and only extends very slowly, the true cords are not involved till the disease has lasted for some time. When the disease extends to the interior of the larynx, it possesses the same characters as in the epiglottis. Irregular nodules are seen on the arytenoids, vocal bands and true cords, which undergo the same changes as we have already described.

The presence or absence of necrosis of bone has been termed "the crucial test" to differentiate syphilis from lupus, but many cases are on record in which necrosis of bone has occurred in undoubted lupus. I shall describe one such case later, which came under my own observation, in which lupus of the nose led to partial destruction of the nasal bones. In these cases, however, one must not forget the possibility of the syphilitic and tubercular taints being combined in the same patient.

Stenosis of the larynx only rarely results from lupus, and may be due to blocking of the larynx by lupoid tissue, to fixation of the vocal cords, or to the contractions resulting from cicatrisation. It is much less frequently met with than in syphilis, in fact it is remarkable how seldom tracheotomy is required in lupus. Schroetter, with his very large experience, says he has never seen a case requiring it, and the number of recorded cases is very small. Acute œdema may also give rise to dangerous laryngeal obstruction, but this must be extremely rare, as I have only met with one recorded case. (Dr. C. H. Paul quoted by Marty).

I need hardly refer to the value of anti-syphilitic treatment in aiding diagnosis in a doubtful case. Such treatment will always exercise an unfavourable influence on lupus, unless there be also present a specific taint. At the same time we must not forget that sometimes a syphilitic lesion does not yield to specific treatment.

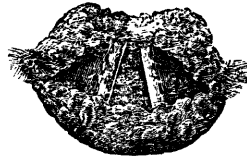
Is there a danger of lupus of the throat ending in pulmonary or general tuberculosis? This is a question to which little or no attention has been given in this country, and yet it is of the greatest importance in determining the prognosis of a case of lupus. The experiments of Koch, Cornil, and Leloir, Max Schüller, and others, have shown that general tuberculosis follows upon inoculations with the lupus bacillus. Numerous clinical observations, also, show how lupus may develop around fistulæ connected with tuberculosis joints or glands. Again, the coincidence of tubercular affections with external lupus is very striking in the light of recent statistics. Out of four hundred and eight lupus patients, no less than 67·9 per cent. were found to suffer from some other tubercular affection (Baumgarten, *Jahrsbericht über Mikro-organismen*, 1886). In thirty-eight patients, Besnier found pulmonary phthisis in eight, and Prof. Leloir, of Lille, on examining seventeen lupus patients detected signs of bacillary phthisis in no less than ten. Certainly there is no such frequent coincidence of the two diseases in this country.

I am not aware of any observations showing the liability of lupus of the mucous membranes to result in pulmonary or general tuberculosis. It is a subject which still requires to be worked out. French writers, however, seem to be much more impressed with the danger of such an issue than we are, and regard pulmonary phthisis as a common termination of lupus of the larynx. Recently I had an opportunity of observing a case in which general tuberculosis followed upon lupus of the face and tongue, which I shall relate at the conclusion of this paper.

With regard to treatment I have nothing new to bring before you. The methods of treatment are the same for lupus of the throat as of the skin. Scraping, scarifications, and the electric cautery are most efficacious in the severer forms, but the milder escharotics, nitrate of silver, chromic and lactic acids, are sufficient in a large proportion of cases to arrest the process, at least for a time. Mandl's solution of iodine has a favourable influence in the nodular thickenings of the larynx, but still better is scarification, with rubbing in of lactic acid, as in laryngeal tuberculosis. In some cases where ulceration is present, especially if accompanied with pain, insufflations of iodol or iodoform gives the best results. Tracheotomy, in the few cases in which it has been necessary, has always had a favourable influence on the progress of the disease. Dilatation, by means of Schroetter's bougies, was sufficient in a case related by Ganghofner to avert tracheotomy. In conclusion, I will describe briefly a few cases which have come under my observation recently, and which have been referred to in the foregoing paper:—

CASE I.—E. W., aged nineteen, came under my care six months ago, complaining of hoarseness and slight pain on swallowing. She was a healthy-looking and well-nourished girl, though of pale complexion. Symptoms had been present for two or three months before I saw her. On examination, the mucous membrane of the pharynx and larynx was seen to be abnormally pale, and so insensitive that the larynx could be probed without causing reflex disturbance. The uvula and soft palate were intact. The whole of the posterior wall of the pharynx was occupied by small cicatrices with projections of granulation-like tissue here and there, but there was no active ulceration except in the naso-pharynx, where there

was a superficial ulcer on the posterior wall about the size of a threepenny piece. The epiglottis was thickened—stiff, nodular along its free border—with small cicatrices between the nodules, of pale colour, and appeared about half its normal size. There was no ulceration to be detected at that time. Within the larynx there was thickening of a nodular character, involving the ary-epiglottic folds—the false cords—and the inter-arytenoid space. The vocal cords were normal in appearance, but there approximation was prevented by the inter-arytenoid thickening. There was no lupus of the skin. The teeth were exceptionally healthy, and there were no other evidences of hereditary syphilis. Mr. Reginald Harrison, who sent the case to me, had come to the conclusion that the case was not syphilitic, and the most careful inquiries into the family history failed to show any evidence of hereditary taint. From the cases of lupus of the larynx I had seen previously I diagnosed this as such, and the subsequent history of the case abundantly justified this opinion.



During the past six months I have watched this case, and seen several of the nodules along the edge of the epiglottis break down, forming shallow, sluggish ulcers, which healed slowly, leaving small cicatrices. One also found on the summit of the right arytenoid, about the size of a split-pea, and while it remained unhealed gave rise to considerable pain on swallowing. Iodol and morphia insufflations were most effective while ulceration was present, and the nodular condition of the larynx improved under brushings with Mandl's solution, the voice becoming much clearer. This patient is still under my care, as she returns every few weeks with fresh ulceration of the epiglottis.

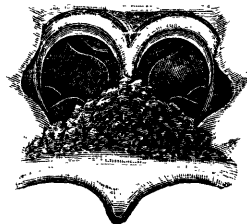
Of course, this patient had the benefit of the doubt, and was treated with iodide and mercury for some weeks. This treatment, she declared, made the pain on swallowing worse, and certainly did not cause healing of the ulceration.

The lungs, I should mention, are quite healthy, and there are no bacilli in the sputum.

CASE II.—T. R., aged eleven, had had lupus of the nose for two years, with destruction of the cartilaginous septum and anterior ends of the lower turbinates. Examination of pharynx showed great thickening of the uvula with nodules along its right border and tip of a pinkish, grey colour, and a small superficial ulcer, with grey base, on its anterior surface. There was no pain. The mucous membrane was highly insensitive and decidedly anæmic.



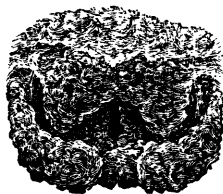
CASE III.—C. W., aged eleven, had lupus of the nose of some years duration; pharynx anæmic; very atrophic; and with reduced sensitiveness. On the posterior surface of the soft palate was a mass of granulation-like tissue, the colour of normal mucous membrane, which blocked the channel and hid the greater part of the septum. There was no ulceration.



CASE IV.—M. R., aged eight years, had suffered from lupus of the nose for over a year when she came under my notice. There was then a large ulcer on the bridge of the nose, exposing the nasal bones which were necrosed. The ulcer was surrounded by lupus

nodules. There was no history of syphilis nor any evidences of hereditary taint. The child was very delicate and ill-developed. The diagnosis of lupus was confirmed by a surgeon of large experience, who treated the case by scraping.

CASE V.—A. P., aged twelve. Lupus of the face for past two years. A year ago voice became husky and breathing slowly became more and more embarrassed. In January of this year breathing was so bad that the patient was admitted to a children's infirmary and tracheotomy performed. Examination showed absence of the uvula, and the whole arch of the soft palate together with the tonsils and base of tongue, was cicatricial. The epiglottis was greatly thickened and nodular—of a pale pink colour, with a considerable deficiency in the middle of its free border. The arytenoids and inter-arytenoid space and false cords were greatly thickened and studded with irregular nodules which entirely hid the true cords.



CASE VI.—S. M., aged seven years. Had suffered from lupus of the face—both cheeks—for 16 months, when she was admitted to the Children's Hospital three months ago. She then had an ulcer on the tip of the tongue, deep, irregular, and quite painless. The parents were quite unaware of this ulcer being present till it was discovered by accident on asking the child to show her tongue. The ulcer was scraped and examined for bacilli, but none were found. Six weeks after admission the child died from acute miliary tuberculosis.

REFERENCES.

- RAMON DE LA SOTA.—*New York Medical Journal*, July 10, 1886.
HOMOLLE.—*Des Scrofulides de la Muqueuse Bucco-pharyngienne*. Paris, 1875.
CHIACI AND RIEHL.—*Lupus Vulgaris Laryngis*. *Vierteljahres sch. für Derm. und Syph.* IX. Jahrg., Heft. 4. 1882.
LEFFERTS.—*American Journ. of Med. Sciences*. Vol. LXXV. April, 1878.
TÜRCK.—*Klinik der Krankheiten des Kehlkopfes und des Luftröhre*. Page 425.
LENNOX BROWNE.—*The Throat and its Diseases*. Second Edition. London, 1887.
ALEXANDER HASLUND.—*Zur Statistik des Lupus Laryngis, in Viertel. f. dermat. und syphil.* 1883. Page 471.
SCHROETTER.—*Laryngologische Mittheilungen*. 1875. Page 84.
„ *Vorlesungen über die Krankheiten des Kehlkopfes*. Wein, 1888. Page 166.
VON ZIEMSEN.—*Cyc. of Medicine*. Vol. VII., p. 848.
MORELL MACKENZIE.—*Manual of Diseases of the Throat*. Vol. I. 1880.
MARTY.—*Le Lupus du Larynx*. Paris, 1888.
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