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A Contribution to the Study of Congenital Syphilis.

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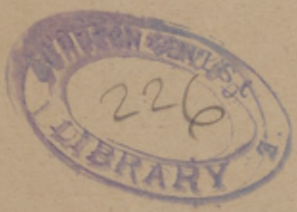

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A CONTRIBUTION TO THE STUDY OF CONGENITAL SYPHILIS.

By JOHN N. MACKENZIE, M. D., Baltimore.

I HAVE selected the case, whose essential features are given below, from my note-book, not only on account of its peculiar interest, but also to serve as the text for some remarks on the manifestations of congenital syphilis in the throat, and their behavior under the influence of acute disease.

Agnes C., aged eight, the child of syphilitic parents, was brought to the London Hospital for Diseases of the Throat and Chest in the latter part of September, 1879. Her history was as follows: Healthy at birth. When five weeks old, lost power in the right upper extremity. This lasted for five weeks, during which time she had no control over the limb. When eight weeks old, contracted a diarrhœa which lasted uninterruptedly for nine months. With the diarrhœa, round, irregular, elevated patches of a red color appeared on the back and buttocks, which were associated later on with an eruption around the anus and genitals. These lesions disappeared some months afterward with an attack of measles. When an infant, would cry out in the night, clinch her thumbs, and burrow her head in the pillow.

From her first to her third year, an interval of perfect health. At the age of three, inflammation, with a cloudy condition of the right eye, appeared, followed in two weeks by implication of the left. The eyes remained inflamed for two years. (The patient's letter from the Royal Ophthalmic Hospital states the condition to have been keratitis.) At the expiration of this time both upper eyelids became completely paralyzed, so that she had no power over them whatever. She could lift them up with her fingers, and frequently did so; but the muscular control of the lids was gone. There was no inflammation of the eyes beyond some remains of the keratitis at the time, and their exposure to light gave rise to no irritation. The mother noticed nothing wrong about the eyeballs during this period. With the ptosis the child complained of intense occipital cephalalgia; convulsive movements occurred during sleep, during which she would contract her brows and rub her head with her hands,

* Read before the American Laryngological Association, May 12, 1884.

leading the mother to suspect trouble with the brain. These symptoms lasted for over a month, at the end of which time, together with the paralytic condition of the lids, they completely disappeared. (During this period she was under intelligent medical supervision, but I am unable to discover what treatment was adopted.)

When they had passed away, the mother noticed for the first time that the right eyeball was turned inward toward the nose, and that she had become deaf in the right ear, from which there issued a slight but constant discharge. Shortly afterward a little white spot appeared on the left tonsil, which slowly ulcerated, the ulceration involving the whole of the back portion of the throat. At the same time she complained of great difficulty in deglutition and pain in the bones, the cervical glands became enlarged, she spoke in a nasal tone of voice, and had a profuse discharge from the nose. During the period of sore throat there was suppuration of the right lachrymal sac, which opened, and continued to discharge for eighteen months. The left sac then became inflamed, suppurated, and ruptured spontaneously, and the discharge has been gradually diminishing in quantity since. The ulceration of the throat healed slowly; would constantly break out afresh; in fact, it never completely cicatrized.

In August, 1879, the child became suddenly aphonic from exposure to cold, and, in the latter part of September, was brought to the hospital suffering from dysphagia and dyspnœa of several weeks' duration.

Condition on application at the Clinic.—Large head, prominent frontal eminences; cloudy corneæ, opacity of the right lens; right internal strabismus; marked flattening of the nasal bones and prominence of the lower jaw; the remains of a dacrocystitis of the left side. The pegged condition of the teeth is absent. Muco-purulent catarrh of nose and middle ear. On looking into the mouth, the whole of the hard palate is seen to be in a state of active ulceration, and there is necrosis with perforation of its central portion. The palatal destruction extends over the anterior arch of the alveolar ridge, involving the bone to a limited extent. The soft palate, uvula, and anterior faucial pillars are completely destroyed, and the posterior wall of the pharynx is the seat of a large, angry-looking ulcer which extends into the naso-pharynx as far upward as the view extends. The patient has to keep her handkerchief constantly to her mouth to absorb the discharge, which accumulates with great rapidity.

Laryngoscopic examination is not possible; but there is absolute aphonia, with dyspnœa, and the glands at the cornua of the hyoid bone are hard and swollen.

Liquids and solids, which are not finely divided, return through the nose, and the little patient complains of cough, pain in the head, and night sweats.

The ulceration continued to advance, in spite of tonic and anti-syphilitic treatment, until the 18th of October, when the patient was seized

with scarlet fever of a mild type. With the appearance of the exanthem the ulceration in the throat began at once to heal, and, when the stage of desquamation was reached, cicatrization was complete, the dyspnœa and dysphagia mitigated, and the voice restored.

Shortly after convalescence the ulceration became again active, and in the early part of the following December a severe, acute pneumonia supervened. The pulmonary inflammation, with which she was confined to the house for four weeks, seemed to exercise no appreciable effect upon the palato-pharyngeal ulceration, which only cicatrized some weeks after resolution had taken place.

The throat then presented the following appearances: The hard palate was covered with cicatrices, and there was a small point of incipient ulceration at its posterior portion. The palate was continuous behind with a dense mass of cicatricial tissue which completely filled the fauces and pharynx, and in which all trace of the original anatomy of these parts was lost. In its center a small, oval depression existed, which communicated with the parts below by an aperture, through which the tip of the little finger could not be passed. From this central sulcus, which represented the remains of the pharyngeal cavity, a small contracted passage led into the naso-pharynx which would just admit an ordinary probe. The child breathed entirely through her mouth, upon closing which, the respiration had a loud, roaring sound. By forcible expiration she could drive a faint column of air through the nose.

Before passing to the consideration of the subject to which I desire to call special attention, there are two features in the above case that deserve particular notice.

While deep ulceration of the pharynx (I refer to the *lower* pharynx) is not rare in congenital syphilis, such a degree of resulting stenosis is excessively uncommon. I know of but two similar cases. In the one,* a mere slit formed the only communication with the structures beneath, and in the other† the pharynx was so constricted as to scarcely admit the point of the little finger.

Of great interest, too, is *the paralysis of both upper eyelids.* The history of the case would point to a cerebral lesion as its probable cause, possibly a gummatous deposit in the course of the oculo-motorius nerve. The subsequent paralysis of the abducent and the symptoms referable to the organ of hearing on the same side may also be susceptible of a similar explanation. Any reasonable doubt as to the syphilitic nature of the paralysis would be dispelled by the easy exclusion of other causes and the curative effect of (presumedly anti-syphilitic) treatment. The case, then, adds one to the short list

* Catti, "Wiener med. Presse," No. 18, May 2, 1875.

† Levis, "Med. and Surg. Reporter," Philadelphia, May 3, 1879, p. 381.

of affections of the cranial nerves in hereditary syphilis observed by Graefe,* Barlow,† Dowse,‡ and Galezowski.*

In the "American Journal of the Medical Sciences" for October,‖ 1880, I called attention to the frequency with which the throat is involved in congenital syphilis, and gave a systematic description of the lesions which are found in the pharyngo-bronchial tract and œsophagus during the course of that disease. In opposition to the then generally received doctrine, I ventured to maintain, as the result of careful investigation of the subject, that, so far from being rare, as was generally supposed, laryngeal affections in congenital syphilis are among the most common and characteristic of its pathological phenomena, and that the invasion of the larynx may be looked for with the same confidence in the congenital as in the acquired form of the disease. Further experience and the study of cases recorded by others have only served to strengthen the positions taken in the paper referred to and the views there expressed, for which I must refer to the original.

Among other things, the following conclusions were reached in reference to deep, destructive ulceration of the oro-pharyngeal cavities: 1. That deep ulceration may invade the palate, pharynx, and naso-pharynx at any period of life from the first week up to the age of puberty. Thus, of thirty cases analyzed with reference to the period of invasion, fourteen occurred within the first year, and ten within the first six months of life, the remainder occurring at periods more or less advanced toward puberty. 2. When the eruption of inherited syphilis is apparently delayed until the latter period, that lesions of the palate and pharynx are found with a peculiar constancy, and often first attract attention to the existence of a diathesis of which they are the sole pathological expression. 3. That females are attacked more frequently than males. Thus, out of 69 cases of pharyngeal ulceration, 41 occurred in the former sex. 4. That ulceration may occur in any situation; but its most frequent seat is the palate, for which it exhibits the closest elective affinity. 5. That, when situated at the posterior portion of the hard palate, the tendency is to involve the soft palate and velum, and thence to invade the naso-pharynx and nose; while, situated more anteriorly, it seeks

* Virchow's "Archiv," Bd. lxxxiv, S. 269.

† "Trans. of the Pathological Society of London," 1877.

‡ "The Brain and its Diseases," vol. i, p. 76.

* "Recueil d'ophthalm.," Aug., 1879, p. 454.

‖ "Congenital Syphilis of the Throat; based on the Study of 150 Cases," N. clx, p. 321.

a more direct pathway to the latter, which is established by perforation of the bone. 6. That the next most common seats of ulceration in order of frequency are the fauces, nasopharynx, posterior pharyngeal wall, nasal fossa and septum, tongue, and gums. 7. That ulceration, especially that of the palate, shows a disposition to centrality of position, together with a special tendency to caries and necrosis of the bone, a fact probably explicable by the great vascularity of the periosteum and medullary membrane in youth. 8. That the tendency to necrosis exists at all periods of life, but especially in early youth, when it is more destructive and less amenable to treatment. 9. That while deep pharyngeal ulceration generally precedes or co-exists with similar affections of the larynx, the latter occurs too without evidence of pre-existing pharyngeal lesions. 10. That laryngeal ulceration does not commonly follow the pharyngeal destruction of so-called latent syphilis, those palato-pharyngeal ulcerations found in tardy congenital syphilis having little tendency to invasion of the larynx, but rather to implication of the nasal pharynx and nose. 11. That simultaneous or consecutive ulceration of the palate, pharynx, and nose seems to be characteristic of syphilis, or at least occurs more frequently in this than in any other disease.

I bring these facts again into prominence because they differ from commonly accepted views, and because they possess at least a certain value, by reason of the method by which they were obtained. I desire also to reiterate what was said in connection with the confusion of these lesions with so-called "scrofulous" ulceration. Without entering into a discussion of the subject, suffice it to say that there is no just ground for belief in an ulcerative scrofulide of the throat. It needs only the most superficial review of the writings of those who maintain its separate existence to show the utter confusion which prevails as the result of erroneous views, handed down among the traditions of an obsolete pathology.

It is obviously a point of great practical importance that this fact should be recognized, and especially in view of the rapidly destructive tendency of inherited syphilitic ulceration in the oro-pharyngeal cavities and larynx.

The throat ulceration of congenital syphilis not only exhibits a special tendency to rapid invasion of the deeper tissues; it often possesses an inherent virulence which places it apparently beyond the reach of therapeutic control. This is markedly true of the ulceration which occurs in the earlier years of life. Cases are now and then encountered where the ulcers stubbornly refuse to cicatrize, or do so sluggishly and imperfectly, healing at one point and becoming simul-

taneously active at others. *Under such circumstances, when remedial measures are apparently of little or no avail, they sometimes cicatrize, as if by magic, on the accession of an acute disease.* It is to this that I wish to direct particular attention.

The clinical study of the cases upon the analysis of which the report referred to was based disclosed certain striking facts in connection with the influence of some of the ordinary infectious diseases of childhood upon the progress of the inherited syphilitic affection. From the historical narratives furnished by this particular group of cases, it would appear (1) *that, while congenital syphilis affords no absolute protection against certain acute infectious diseases, its existence in the individual seems often, other things being equal, to mitigate their severity and exert a favorable influence on their course;* (2) *that certain acute diseases, accompanied by an exanthem, favor the dissipation, at least temporarily, of the throat and other manifestations of the disease;* (3) *that while at no period of the disease is the child exempt from these affections, they are more liable to be contracted during the period of latency—that curious interval of apparent health in congenital syphilis which Cazenave has poetically called the sleep of the virus.*

These remarks are limited to scarlet fever, measles, and chicken-pox, but they could doubtless be extended to embrace others of the exanthemata.

They do not apply, for obvious reasons, in the case of excessive virulence of the syphilitic cachexia or malignant epidemic influence of the intercurrent disease.

Of special interest is the effect produced by acute febrile disease upon the throat lesions of congenital syphilis. *Chronic inflammatory conditions and ulceration of the larynx, pharynx, and nasal passages are often influenced in a remarkable manner through the presence in the individual of an intercurrent febrile affection. This is, moreover, eminently true of those acute blood diseases with special tendency to local manifestations in the throat, such as scarlet fever, measles, diphtheria, etc. According to personal experience, scarlet fever and measles exert, as a rule, a favorable influence on the course of the throat affection, their supervention being of itself sufficient to cause its complete disappearance. The poisons of the two diseases in their circulation in these regions appear to be mutually destructive, and the throat escapes by virtue of such reciprocal antagonism.* The*

* It is quite possible that this may also be true of other mucous surfaces of the body.

cure here may be permanent, or relapses of the inflammatory or ulcerative process may follow the removal of the antagonistic influence of the intercurrent disease.

These remarks do not apply to diphtheria. When this affection supervenes during the existence of lesions in the throat, the patients rapidly succumb to the disease. The existence of syphilis in the child apparently increases the tendency to membranous formation; indeed, in some instances, apart from the presence of the diphtheritic process, there seems to be a *special tendency to fibrinous formation in the nose and retro-nasal space.*

The influence of acute disease upon the manifestations of constitutional syphilis is a subject which has received some attention at the hands of syphilographers, especially certain of the French school; but very little is known as yet, beyond the empirical fact, that the lesions of that disease, and especially the cutaneous syphilides, are often modified by the introduction into the blood of the virus of an intercurrent febrile affection. This modification may consist either in the permanent or temporary dissipation of existing syphilitic lesions, or in the exaggeration or intensification of the morbid process. Thus, for example, various syphilitic affections, such as skin eruptions, exostoses, etc., have been observed to disappear during the course of erysipelas,* acute rheumatism,† cholerine,‡ variola,# febrile furunculosis,‖ etc. Laségue△ has recorded a case of ulceration of the pharynx and tonsils which disappeared during an attack of erysipelas, while in a similar one observed by Martellière◊ a fatal result ensued from that disease. The dissipation of syphilitic eruptions has also occurred during pregnancy,‡ and as the result of vaccina-

* *Vide* Cazenave and Schedel, "Practical Synopsis of Cutan. Dis.," etc., Phila., 1829, p. 353; Rayer, "Traité des mal. de la peau," Paris, 1835; Lamarche, "De l'érysipèle salutaire," Thèse de Paris, 1856, and the excellent articles of Mauriac; "Étude clinique sur l'influence d'érysipelas dans la syphilis," Paris, 1873; published also in the "Gaz. des hôpitaux," Nouv. sér., 1873, pp. 305, 321, 346, 385, 410, 443, 466, 506, 546, 569, 594, 601. See, also, Bidenkap's case (cited by Bäumler, von Ziemssen's "Cyclopædia," Am. ed., vol. iii, p. 98, 1875).

† Rayer, *op. cit.*, p. 546 (Mauriac); see also Jourjon, "Infl. des mal. aiguës sur quelques manifestations cutan. de la syph.," Thèse de Paris, 1870.

‡ Cazenave, "Traité des syphilides," p. 593 (Mauriac).

Gore, "Lancet," Sept. 2, 1858.

‖ Diday (quoted by Mauriac, *l. c.*).

△ "Traité des angines," pp. 110–112.

◊ "Sur l'angine syphilitique" (cited by Mauriac).

‡ Gilbert, "Bull. de l'acad. de méd.," 1851, tom. xvii, p. 156.

tion,* and there is a case on record where the latter apparently exerted a curative influence in caries of the pharyngeal vault.†

The remarkable power of erysipelas over the cutaneous syphilides has suggested its artificial production as a therapeutic agent in these affections,‡ while their behavior under the operation of the vaccine virus led to the now almost forgotten practice of Lukonski.* It has, finally, even been proposed by an enthusiastic pupil of M. Hardy to inoculate the poison of small-pox in cases of syphilis which have resisted all other methods of treatment.‖

It is sufficiently evident, then, that a reciprocal antagonism exists between the poison of syphilis and that of a number of acute diseases. By what pathological law this is brought about is, in the present state of our knowledge of the mutual relations of disease, a matter of pure speculation. In the case whose history has been recited above it is interesting to contrast the curative effect of the exanthematous fever on the ulceration in the throat with the persistence of the latter during the acute pulmonary affection.

This remarkable influence of the febrile state upon syphilitic inflammation and ulceration of the nasal passages and throat is also, in a measure, true of simple inflammatory conditions of these cavities. It were foreign to the purpose of the present article to elaborate this latter and cognate subject, and I shall therefore simply offer for your consideration the fact that *simple catarrhal inflammation of these regions occasionally disappears completely, and is permanently cured during the course of an acute febrile disease.* Whether this occur as a phenomenon of so-called "substitution," or as the result of a profound impression made upon the nutrition of the parts by virtue of which abnormal secretion is arrested and the inflamed tract placed in a condition favorable to resolution, can only be determined by the accumulation of more exact scientific data concerning the reciprocal antagonism of pathological processes.

Without, then, attempting any special explanation or generalization, I present the foregoing observations from my clinical experience as a contribution to the study of an interesting but imperfectly understood subject.

* *Vide* "Revue méd.," 1861, tom. i, p. 157, Jeltzinzki.

‡ Jeltzinzki, *l. c.*, "Sur la cure radicale de la syphilis par la vaccination."

† Sabatier, "Propositions sur l'érysipèle considéré principalement comme moyen curatif dans les mal. cutanées," etc., Thèse de Paris, 1831.

* Jeltzinzki, *l. c.*

‖ Garrigue, "De l'influence des mal. aig. sur les diathèses," Thèse de Paris 1870.

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