XIV.

SYPHILIS OF THE EAR.*

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Syphilis of the ear is an old knowledge, recognized in the early history of this disease, but the epoch-making discoveries of Schaudinn, in 1905, of the spirocheta pallida as the etiologic agent of this disease; the cultivation of the spirocheta pallida by Noguchi; the complement fixation reaction of the blood after Wassermann; Noguchi's luetin cutaneous test; the determination of an increase of globulin after Nonne; also the lymphocytosis and Wassermann of the spinal fluid; Noguchi's recent observations on the presence of the spirocheta pallida in certain cerebral diseases, like general paresis, and, finally, the Ehrlich-Hata discovery of arsenic as a therapeutic remedy, have added new substance to the study and are all of the greatest aid to the clinical diagnosis as well as being means of control in the treatment of syphilis.

It is not supposed that we will be satisfied with even our present advanced understanding of this disorder, but with characteristic persistence our profession will prosecute the study of the many still hazy features of this disease. My presentation of this subject will necessarily be limited to certain features, and only in a manner of résumé will touch upon the various characters, leaving to the discussion an elaboration of the undeveloped features.

It is elementary knowledge that all the tissues of the ear may be affected by this disease—skin, periosteum, bone, cartilage, mucous membrane, nerves and vessels—and that the various stages of the disease may even attack the different parts of the ear. The disease may be present in the acquired or inherited form. As a matter of convenience, I will appor-

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tion my discussion of the subject to the external ear, middle ear, inner ear and intracranial region.

EXTERNAL EAR.

As a matter of fact, our observations of syphilitic lesions about the external ear vary considerably, owing, perhaps, in some cases to the class of material available, and again to a neglect in recognizing the probabilities of this disease in all of its protean types in this particular region. At my clinic at the Chicago Post-Graduate Hospital I have learned from the great number of cases seen there to recognize many that otherwise might have passed as of no particular importance or attributed to some other condition.

The chancre or ulcus durum is hard and infiltrated, usually single and umbilicated; the spirocheta pallida may be found on the slide, and the neighboring lymph glands are enlarged and sensitive. It is acquired from a bite, kiss, finger, tooth-pick, pen, pencil, towel, instrument, etc. It may occur upon any part of the external ear, but most commonly about the external meatus.

Lues of the drum occurs as a papule or minute gumma. These readily disappear under proper treatment, but otherwise usually soften and perforate into the middle ear cavity, creating a suppurative condition there. (Kirchner, Politzer, Gruber, Triquet.)

The secondaries are, as a rule, in the form of condylomas situated at the posterior auricular attachment, especially in young people; but occasionally, when found about the entrance of the meatus, they resemble granulations or polypi. The maculopapular eruption has been observed in the canal as well as on the drum.

The tertiaries form an interesting class of external ear lues. According to Gruber, Kirchner and Moss and Steinbrügge, there is a type that is due to a periostitis of the bony canal—not to be confused with a gumma—which by the pressure exerted causes considerable pain, even on movement of the lower jaw, as in eating. Some of the exostoses one sees causing narrowing or occlusion of the canal are the result of a former active luetic periostitis.

Occasionally gumma of the mastoid are seen. Bruehl[1] recently reported such a case. These gumma may disappear

under treatment, or they may disintegrate and create considerable destruction. When they soften in the canal they appear as an ulcer covered by a yellow grayish secretion.

MIDDLE EAR.

If one investigated thoroughly into the etiology of all middle ear cases presenting at a large clinic, the frequency with which lues appears would create no little astonishment. It is true, the association does not necessarily determine the cause in all such middle ear cases, but the frequency of association, the symptomatology as brought out from the past history, the old landmarks still present of former active processes, all tend to strengthen one's convictions of the commonness of lues as a cause of middle ear disease.

There is no description that will characterize a middle ear lues clinically. We are today, in this respect, not much changed from the position taken by Lowenberg forty-five years ago, when he said they, at that time, knew practically nothing about it. We, as they, still have to build up our evidence from a careful scrutiny of the parts in search of tiny gummas, periosteal thickening and ulcerations, and also from the history, neighboring lesions, laboratory tests, the presence of sequestrums with tell-tale odor, and finally by the response to treatment. But from the new knowledge learned from Schaudinn, Wassermann and Noguchi, a more accurate understanding of diagnosis in middle ear cases will surely result.

An endarteritis of the mucous membrane, and a periostitis of the bony walls, aside from gumma, constitute perhaps most of the pathology in this region. When the periosteal thickening following the periostitis involves the inner or labyrinth wall, ankylosis of the stapes may result. Habermann called this luetic sclerosis. A few years ago (1903) I published a paper on osteosclerosis of the mastoid,[2] which report is in accord with what Moss and Steinbrügge speak of as sclerosing mastoiditis the result of luetic periostitis. These were cases that presented an obscure otalgia, that proved later to be due to luetic periostitis, both of the middle ear walls and mastoid cells, and this without other evidences of ear trouble.

Lues of the tube in the primary form usually occurs at the faucial end, the result of dirty instrumentation, like a cathe-

ter. As secondaries it appears as an erythema or as pearl-like plaques (Deprès) at the faucial mouth. Tertiaries may also occur.

Perhaps some of the strictures of the tube that we frequently meet with are the result of healed ulcerations of luetic origin.

INNER EAR.

Inner ear affections are labyrinthine or retrolabyrinthine. In the acquired form of inner ear lues the disease usually appears at the end of the secondaries, although it has been noted as early as one week after recognition of the chancre, and again in some cases delayed for years.

The ear symptoms are like those of any other nerve deafness, with or without the vestibular symptoms.

The deafness is rapid in its progress and soon becomes quite profound. There is no preponderance of lost hearing in either the upper or lower end, but the entire scale, as a rule, is affected, with occasional islands of hearing for a little while. It has been my experience to find that only one ear may be involved, although it is but a matter of a short time before the other likewise exhibits similar changes.

The inherited form of inner ear lues may appear at any time of life, from infancy to middle age and past, but the far greater number are observed first between the eighth and sixteenth years. The combination of interstitial keratitis, pegged incisors and deafness at this age is familiarly known as the hereditary triad of lues. But this arrangement of symptoms is not invariable. I have quite often seen the teeth normal, while again I have seen the cases when their hearing was normal. Sometimes the deafness is from middle ear changes, with no labyrinth involvement. But the majority of my cases when first seen had marked nerve deafness, even unto deaf-mutism.

The deafness comes on quite suddenly; in fact, often over night or after some prolonged exposure or exertion. Occasionally it may appear rather suddenly and progress with only fair rapidity. It is usually bilateral and, according to Fraser,[3] frequently associated with ozena.

Where Ménière's symptom complex occurs, it has, according to Grey, this striking peculiarity: that when due to lues

there is but one such attack, while if due to other causes there is the likelihood of similar attacks.

The pathology, as presented to us by Politzer, Moss and Steinbrügge, consists of a round cell infiltration and hyperplasia of connective tissue substance, especially of the periosteum. There may be a serous labyrinthitis following a severe hyperemia, and even pus; new bone formation; chronic endarteritis, and hemorrhage into the fibers of the cochlear nerve, leading to atrophy, particularly in the basal coil and of the cells of the spinal ganglion. Gumma may be found in the petrous bone. Periosteal thickening causing pressure in the internal auditory canal may result in paralysis of both the seventh and eighth nerves. According to Rosenstein, a chronic basal meningitis about the entrance of the auditory meatus is much the commonest cause of these symptoms.

It is my belief, based upon the experience I have had with the use of salvarsan, that lues of the ear responds favorably to the action of this remedy, in both the acquired and inherited form, provided treatment is commenced within the first few weeks, better if in the first week, of its manifestation, and also if pushed energetically and supplemented by the use of mercury and K. I. The socalled neurorecedives, that have attracted so much attention lately, have but very seldom occurred in my cases, and in no case was there lasting involvement of the nerves. I am of the opinion of Haike and Wechselmann that the superior activity of salvarsan over mercury in penetrating the nerve structure and exciting the hibernating spirochetes to "move on" may account for these nerve symptoms, which would most likely occur later on if left to other forms of treatment, because all other forms of treatment seem not to reach these nerve stations. Perhaps in some cases where the nerve manifestation appears within a few days following the salvarsan, obliterating the canals and ankylosing the stapes, the condition is simply a Herxheimer phenomenon, but I cannot attribute it to any selected toxic action of the drug.

INTRACRANIAL AFFECTION.

The intracranial affection may be located most anywhere within the skull, but, according to the symptoms produced, it is convenient to speak of a lesion of the cortex, of the mid-

brain or nuclei, of the cerebellar pontine angle, and of the cerebellum. In the latter case disturbed equilibrium is present, similar to that of vestibular origin.

The pathology of lues in this territory is meningitis, endarteritis and gumma, named in their order of frequency.

As the fibers of the cochlear nerves cross at various places along the route from their nuclear origin near the fourth ventricle to the cortex of the temporal lobe, a lesion located in the auditory area of one temporal lobe seldom creates actual disturbance of hearing in either ear (excepting sensory aphasia, that may be present and mistaken for real deafness). Bilateral lesions have been known to cause complete deafness.[4] Deafness in both ears may also result from a lesion that encroaches upon the cochlear nerves where the fibers of each are situated close together, as in the ponsmedullary region. This is known as a midbrain or nuclear deafness, and is not to be confused with the cortical deafness just referred to. In the midbrain variety of the disease various symptoms, as a rule, are present that designate the seat of the lesion, such as ophthalmoplegia, dysarthria, ataxia, apoplectic attacks, bulbar paralysis, headaches. In fact, all kinds of neighborhood symptoms may be present, depending upon the extent and rapidity of the process, and particularly if acute in character. Large lesions and chronic cases are less likely to exhibit symptoms.

A lesion of the cerebellopontine angle gives rise to a one-sided deafness which appears early and usually has associated with it a facial palsy, vertigo, nystagmus, glossopharyngeal paralysis, etc.

In the diagnosis one may consider a rapid onset, profound deafness, or, at least, of severe degree, slight or absence of tinnitus (Siebenmann), associated protean manifestations of cerebrocerebellar characters, normal drum and open tubes, as strongly presumptive of intracranial lues. As additional evidence we have a facial palsy that may improve under treatment; also such other direct evidence of the disease as pegged teeth and scarred eyes, the blood and spinal fluid reaction to the Wassermann test, and the skin reaction to Noguchi's test.

In the differentiation one must particularly keep in mind the possibility of leukemia, pernicious anemia, diabetes, rheumatism, digestive disturbances, arteriosclerosis, aneurism of

the basilar artery, neoplasm, traumatism, toxic neuritis, occupational causes, hysteria and malingering.

The following case of intracranial lues presents several interesting features, chief of which were profound deafness with diabetic symptoms:

Male, forty-one years, otherwise healthy, digestion good; eyes, urine, temperature and blood pressure normal. Excessive user of tobacco. Gradual impairment of hearing with mild tinnitus in left ear for two years. Sudden and profound loss of hearing in both ears on return from a long and tedious drive in the country. No vertigo, nausea, vomiting or tinnitus. On examination, he heard no voice at all, nor any tuning fork, either by air or bone. No spontaneous nystagmus, but both ears reacted normally both to the caloric and turning tests. Drums normal; tubes open. Wassermann strongly positive. Increasing doses of iodid of potassium and mercury up to 160 grains, and one-fifth of a grain respectively were taken daily for two weeks. Then six-tenths gram salvarsan intramuscularly. The latter was repeated four weeks later, with a continuation of the iodids and mercury. Between seven and nine weeks from commencement of the treatment symptoms of diabetes with other fourth ventricle disturbances began to appear, terminating in coma and death.

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