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DEAFNESS FOLLOWING MUMPS: THE POSSIBLE PATHOGENESIS AND INCIDENCE OF DEAFNESS

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Mumps is thought to be the most common cause of unilateral acquired sensorineural deafness in children. Mumps deafness is usually sudden in onset, profound or complete, and may be associated with vestibular symptoms. The authors' clinical survey of 55 patients with unilateral deafness which could reasonably be ascribed to mumps indicates that the hearing loss is exclusively unilateral, severe or total and permanent, and that approximately 45% of the patients experienced dysequilibrium of vestibular origin. An analysis of the present series of mumps deafness also suggests that the primary route of invasion of the virus is hematogenous, and thus the term "viral endolymphatic labyrinthitis" is proposed as the possible pathogenesis of the deafness, since both tympanogenic and meningogenic routes of viral invasion to the labyrinth can be excluded on the basis of the clinical and cerebrospinal fluid studies. This view of the pathogenesis, particularly that mumps meningitis is not associated with deafness, is supported by several reports including those of VUORI *et al.* (1962), AZIMI *et al.* (1969), LINDSAY (1973), NADOL (1978), *etc.* The incidence of deafness following mumps appears to be extremely low, approximately 1: 20,000, as estimated by EVERBERG (1957).

Mumps is thought to be the most common cause of unilateral acquired sensorineural deafness in children. It is also well known that mumps deafness is usually sudden in onset, profound or complete, and may be associated with vestibular symptoms. However, the mechanism of onset of the disease and inner ear pathology in connection with this mumps infection have yet to be elucidated because histopathologic reports on the human temporal bone are extremely limited in number (LINDSAY *et al*,. 1960; SMITH and GUSSEN, 1976). Furthermore, no definite agreement of opinion has been reached as to the incidence of this remarkably severe and irreversible inner ear deafness, though unilateral in the majority of the cases, among the patients infected with this virus. From the findings of detailed clinical studies on 55 fresh cases which the authors have encountered in the past 10 years and in which inner ear deafness can reasonably be ascribed to mumps, the authors discuss the possible pathogenesis and the incidence of deafness following mumps.

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Studies on Clinical Cases

Since 1974, a definite cause-effect relationship between deafness and mumps has been established by the clinical course, history of the illness and serological and/or virological examinations in a total of 55 cases. In clinical cases with deafness which is definitely shown to have developed with mumps, the age distribution, male-female ratio and presence or absence of vestibular symptoms are as presented in Table 1.

The hearing loss in these patients was exclusively unilateral, profound or total, and more than 45% of the patients developed dysequilibrium of vestibular origin. In age,

Table 1. Age and sex distribution of patients with mumps deafness.

Age (years)	No. of cases	Male: Female
– 5	19 (5*)	11: 8
6–10	19 (7*)	9: 10
11–20	6 (4*)	4: 2
21–30	6 (4*)	2: 4
31–	5 (5*)	1: 4
Total	55 (25*)	27: 28

Asterisk (*) indicates number of the patients with associated vestibular symptoms (25/55=45.45%).

the youngest was a boy aged 1 year and 6 months and oldest a 38-year-old woman. As for the age distribution, those aged under 10 years accounted for the majority with 38 cases (69.1%). The male-female ratio was 27: 28, there being little difference between the sexes. Generally, there is a tendency for the percentage of the accompanying vestibular symptoms to be higher in adults and lower in children. Viewed overall, these vestibular symptoms improve rapidly despite the hearing loss being severe, and compensatory remission appears to be obtained after the lapse of about 1 month even in the older patients. No relationship whatsoever is observed between the development of deafness and the severity of mumps infection.

Discussion

While not objecting to the opinion that inner ear deafness following mumps infections is due primarily to labyrinthitis caused by mumps virus, the authors wish to mention the route of invasion of this virus to the inner ear.

In discussing the mechanism of development of labyrinthitis, three routes are generally mentioned for the pathogen to reach the labyrinth: 1) infection *via* the middle ear (tympanogenic), 2) infection *via* the cerebrospinal fluid (meningogenic), and 3) hematogenous infection. In infection through the middle ear or tympanogenic route, viral otitis media develops and reaches the perilymphatic space *via* the inner ear fenestrae to present pathology of perilymphatic labyrinthitis. This route can be excluded since there are no clinical findings or history that suggest viral otitis media. As to the infection *via* the cerebrospinal fluid or meningogenic route, this route is premised on the presence of meningitis or meningoencephalitis which is considered to reach the perilymphatic space *via* the internal auditory meatus or the cochlear aqueduct to present perilymphatic labyrinthitis. According to the authors' observations on the clinical course and studies of the findings of the cerebrospinal fluid in seven fresh in-patients, however, findings suggestive of meningitis or meningoencephalitis were not obtained at all. Thus, this route of invasion could also be ruled out. In fact, that mumps meningitis is not associated with deafness as a sequela is also supported by many reports including those of VUORI *et al.* (1962), AZIMI *et al.* (1969) and more recently, NADOL (1978), although meningitis is one of the relatively common complications of mumps.

As to the mechanism of development or pathogenesis of deafness due to mumps infection, therefore, hematogenous infection is considered to be the most probable. In other words, the viremic state due to mumps reaches the inner ear hematogenously, causes an inflammatory change in the stria vascularis of the cochlea and then results in severe impairment of the endolymphatic system. Thus, the term "viral endolymphatic labyrinthitis" is proposed as the possible pathogenesis of the deafness. In fact, LINDSAY (1973), the first to report on the pathology of the human temporal bone in a patient with mumps deafness, also states that the susceptibility of the stria vascularis by the mumps virus during the viremia can be explained by its relatively slow circulation and the intraepithelial location of its capillary network.

In view of the clinical course in which the hearing loss is very severe and advances destructively and rapidly, the deafness can hardly be explained by perilymphatic labyrinthitis, while pathology of endolymphatic labyrin-

thitis considered to spread to the entire endolymphatic structure appears to be easy to understand theoretically.

That the incidence of accompanying vestibular symptoms is high at 45% or more may be due to the influence of endolymphatic labyrinthitis on the saccule, through which endolymph of the cochlea can anatomically pass directly *via* the ductus reuniens. Therefore, the tendency for the vestibular symptoms to be relatively less severe and to improve in a relatively short time compared with the severity of the hearing loss can easily be understood if one thinks the vestibular symptoms are due to the pathologic changes in only the saccule.

As to at what incidence this inner ear deafness develops in mumps patients, references mentioning this point are very limited in number. Some authors have made conjectures that are considerably incorrect or unreliable. A report of EVERBERG (1957) is the only one that gives relatively theoretical values as to the incidence of deafness due to mumps infection. According to his report, the incidence of deafness as a complication of mumps is estimated to be about 0.05 per thousand, that is, one out of 20,000 patients with mumps. Cases accumulated by the authors live in metropolitan Tokyo and adjacent areas where the population has increased sharply, and the population which serves as the base is large. So, it may be said that the numerical value estimated by EVERBERG (1957) is not unreasonably low but within a relatively rational range.

The reason why inner ear deafness develops unilaterally in an overwhelming number of cases has remained obscure up to the present. From the fact that the incidence of unilateral deafness is low at 1/20,000, it may be inferred that the incidence of bilateral deafness is the product of the incidence of unilateral deafness and therefore would be exceedingly lower.

Conclusion

On the basis of results of a clinical study of 55 cases of inner ear deafness due to mumps, the authors discussed the pathogenesis and incidence of the deafness.

Results obtained are summarized as follows: 1) In inner ear deafness found in mumps patients, the viremic state reaches the inner ear hematogenously and brings about an inflammatory change in the stria vascularis of the cochlea to present severe impairment of the endolymphatic system (viral endolymphatic labyrinthitis). 2) The incidence of deafness in mumps patients is extremely low, and about 0.005% given in the report of EVERBERG (1957) is considered a reasonable numerical value. There seems to be a relationship between this low incidence and the reason why unilateral deafness is found in the majority and bilateral deafness is exceedingly rare.

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