HEREDITARY COMBINED NEURINOMAS AND MENINGIOMAS

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(Received for publication March 1, 1955)

VON RECKLINGHAUSEN'S disease, in the form of multiple neurofibromatosis affecting not only peripheral nerves but cranial and spinal nerve roots, has frequently been demonstrated to show marked hereditary tendencies. Even when this disease, unaccompanied by widespread neurinomatosis, manifests itself only in the form of bilateral acoustic nerve tumors, it has been shown to be hereditary. In the remarkable report of a family by Gardner and Frazier,2 they traced the bilateral deafness as a true Mendelian dominant through five generations comprising 217 persons.

Cushing and Eisenhardt,1 in their classical monograph on meningiomas, have brought together in a most exhaustive manner the reports from the literature of cases of multiple meningiomas, and combined neurinomas and meningiomas occurring in the same patient. It is, indeed, with one of the two examples presented by these authors in Chapter V of their book, entitled "Combined Neurinomas and Meningiomas," that we are dealing in the present report. Stanley G., the subject of Case I, Serial No. 311, in Chapter V of their monograph, was referred to the care of one of us by Dr. Cushing upon his retirement from active neurosurgical practice. The present paper is a continuation of the remarkable history of this patient, and that of his only child. For the sake of completeness, we are taking the liberty of repeating the earlier history of Stanley G., as reported by Cushing and Eisenhardt, and will then report further upon our own experiences with this unique father and daughter.

CASE REPORTS

"Case I. Serial No. 311. Multiple psammomeningiomas involving cerebral dura, roentgenologically verified, and symptomatically associated with bilateral auditory disturbances presumably due to bilateral acoustic neurinomas. Suboccipital exploration with surgical verification of multiple tumors supposedly meningiomas in the right lateral recess adjacent to the porus acusticus. Recovery with fairly stationary condition after 5 years."

"July 17, 1932. Admission of Stanley G., a Jewish medical student, 24 years of age, complaining of headaches and impaired hearing. The family history revealed a tendency toward familial psychoneurosis."

"Past History.—For a period of at least ten or twelve years the patient had

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noticed a tendency to flushing and sweating of the right half of the body. A large lipoma was removed from the suboccipital region in 1922. For five years, progressive impairment of hearing and for four years inequality of pupils had been observed. An advancing right-sided deafness with tinnitus had been present for two years and tinnitus referred to the left ear for a year and a half. Some impairment of vision also had been noted for two and a half years; and latterly he had experienced colour hallucinations on stooping. Unsteadiness in gait with a tendency to deviate to the right and backward with attacks of weakness and trembling had been of recent occurrence. Despite a negative Wassermann he had received a thorough antiluetic treatment.

"Physical Examination."—This disclosed: (1) a left relative hyposmia; (2) bilateral choked discs of 2 diopters; (3) anisocoria, right pupil wider than left; (4) nystagmus poorly sustained; (5) hypeaesthesia of the right trigeminal skin field; (6) slight palsy of the right face; (7) impaired hearing in the right ear with sluggish vestibular response to caloric test (normal reaction on left); (8) a definite right-sided tendency to flushing and hyperhidrosis; (9) positive Romberg with falling backward and to the right; (10) slight sensory loss over right arm and leg; (11) increased deep reflexes on the left; (12) a smooth cranial hyperostosis palpable in the left frontal bone with enlargement of the temporal artery.

"X-ray films showed unmistakable evidence of multiple psammomatous meningiomas bilaterally situated and involving the cranial vault. They were chiefly parasagittal in situation, more particularly on the left side in correspondence with the palpable hyperostosis.

"Diagnosis."—It was clinically apparent that there was a growth in the right cerebellopontile angle, probably an acoustic tumor associated with bilateral parasagittal meningiomas over the cerebrum. Ventriculograms disclosed moderate dilatation of the lateral ventricles with slight depression of both frontal horns.

"Operation."—July 25, 1932. Novocaine. Suboccipital exploration. The usual bilateral exploration was made. A tense cerebellum was disclosed and on exploring the right recess normal-appearing 9th, 10th, and 11th, and 7th and 8th nerves were seen. Closely adjacent to the porus acusticus were two tiny tumors supposedly meningiomas about 1 cm. in diameter, apparently attached to the dura. The first proved on supravital examination to be a psammoma; the other was not examined. It was thought to be another meningioma (c.f. infra). The operation was then concluded.

"Subsequent Notes."—Convalescence from this procedure was protracted and stormy due to extreme vertigo and vomiting on the slightest change of position. In view of these distressing symptoms it was thought best to postpone an attack upon the meningiomas involving the forebrain. On August 20, 1932 he was discharged.

"In spite of his handicap and increasing deafness, he courageously resumed his medical course and succeeded in graduating in June 1936. He meanwhile had married and become a parent. On October 17, 1936 a thorough otological examination by Dr. Page Northington showed a total loss of vestibular response and air conduction on the right and partial loss in the left ear. In a report April 12, 1937, he stated that he was busy as an interne; that his general health was excellent, his mind active, his domestic life happy, and he was not in the least sorry for himself.

"Histological Note."—The first and smaller of the two tumors as stated was a psammomeningioma. When we came to cut the other, it proved to be a transitional type of neurinoma showing in addition to pali-