Case Reports and Technical Note

Familial Bilateral Acoustic Neuroma
Affecting 14 Members from Four Generations

Case Report

PETER D. MOYES, M.D., F.R.C.S.(C)

Department of Neurosurgery, University of British Columbia, Vancouver, British Columbia

In 1882, von Recklinghausen stated that “multiple neuromas cannot be considered as a purely acquired disease,” and since that time a familial or inherited tendency has been demonstrated in some cases of neurofibromatosis. In 1900, Thomson found 10 such cases in the available literature. In 1920, Feiling and Ward published a paper on a familial form of acoustic tumor affecting three generations. In 1930, Gardner and Frazier published a clinical study and field survey of a family of five generations with bilateral deafness in 38 members; a follow-up report 10 years later added a further generation to this formidable history. Cushing was the first to describe the characteristic course and symptomatology of auditory and labyrinthine disturbances, headache, cerebellar ataxia, and increase in intracranial pressure.

The family that we are about to describe follows the pattern exactly except for the fact that failure of vision has not been demonstrated in any of the affected members. Our investigation, begun in March, 1965, was instigated by a singularly unusual family history supplied by the patient whose record follows.

Case Report

A 22-year-old man was referred to this office by his family doctor following investigation by an otolaryngologist who had detected minimal nerve deafness on the left side without any apparent vestibular disturbance. The patient had been prompted to see his physician because of his alarming family history, although he had no symptoms himself.

Family History. His paternal grandfather and paternal great-aunt had both died as a result of brain tumors with the presenting symptoms of bilateral deafness, ataxia, and paralysis; his father had died following an operation for the removal of the second acoustic neuroma; one paternal uncle had died following a similar operation; another paternal uncle had died before surgery; and a third paternal uncle had died in his 20's from a similar condition, without surgery. Since the start of our investigation, two more paternal uncles have been told they have bilateral acoustic neuromas but have so far refused surgery. One female cousin has died following removal of a second tumor, and her sister and brother have both had surgery. Our patient's sister has been investigated because of hearing loss and vertigo but a tumor was not discovered at operation. Two young children of a cousin, aged 6 and 3, have been investigated for tumor and another cousin's 15-year-old son is reported to be having surgery later in the year. The patient's maternal uncle had an acoustic neuroma removed at operation. Details of this family will be given later.

Examination. The tomograms carried out previously on our patient indicated slight enlargement of the internal auditory meatus on the left. Despite full neurological examination, we could find no other neurological abnormality nor any involvement of the fifth or seventh cranial nerves. The patient had red hair and freckles, but no cafe-au-lait spots and no evidence of peripheral neuromas. An air study, carried out after his admission to the Vancouver General Hospital, revealed a mass about 1 cm in diameter in the region of the left internal auditory meatus. The cerebrospinal fluid protein was 20 mg%.

Operation. On April 12, 1965, the tumor was removed completely via a suboccipital craniectomy. The postoperative course was
uneventful except that the patient developed a left facial palsy despite great efforts to avert this. He is being kept under careful observation but so far has shown no evidence of a tumor on the other side.

**Family History**

The history of four generations was obtained through the following means: by questioning the mother of our patient who supplied some information regarding members of her husband's family; by writing to the hospitals and doctors who treated various members of her husband's family; by writing to the hospitals and doctors who treated various members of this family (particularly Drs. William and Howard House of Los Angeles who operated on three members and examined five others); by seeking the information in birth and death registries; and by interviewing 10 members of the family (Fig. 1).

**First Generation: Male WU.** The family's first generation begins with this man who was a farmer in Dade County, Missouri. He died in Fresno in 1933; his autopsy reads "cerebellopontine brain tumor" as the cause of death, with bronchopneumonia as the other contributory cause. He was described by his wife, who is still living, as being pleasant and kind when he first married her; he was teaching school and she was his pupil. However, 20 years and 10 children later, he had become bad-tempered, cruel to his children, and had been deaf for an unknown period of time. Interviewing was difficult since his wife refused to talk about him or his family except to complain of his cruelty. She left him in 1927 with most of her 10 children and moved to Fresno. She took her husband back into the family in 1932 as a "boarder" because he was unable to care for himself any longer. He was apparently completely deaf and unable to walk, dress, or feed himself. He died in 1933 at the age of 59. He seemed to bear none of the von Recklinghausen stigmata according to his relatives who remember him.

**First Generation: Female RU.** This woman, the wife of WU, was his first cousin; she and her husband had the same paternal grandfather. At 82 years of age, she is in full possession of her faculties, but has bumps on

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**Fig. 1.** Genealogical chart showing the occurrence of bilateral acoustic neuroma in four generations of the family and also in a relative by marriage.