Neurofibromatosis type 2

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A 45-year-old woman developed a gradual onset of sensorineural hearing loss in the left ear in 1984. Head CT scan with contrast demonstrated bilateral cerebellopontine angle (CPA) enhancing masses. She underwent a resection of the left CPA tumor with pathology confirming schwannoma. Despite several resections, she had progressive hearing loss and she became deaf in 2003. Her brain MRI (figure) demonstrated bilateral vestibular schwannomas, which are sufficient for a definite diagnosis of neurofibromatosis type 2 (NF-2), according to the National Institutes of Health, Manchester, or National Neurofibromatosis Foundation diagnostic criteria.¹ In

addition, this patient also has multiple meningiomas, which are associated with NF-2.² The patient has been treated with imatinib mesylate (Gleevec) plus hydroxyurea in a clinical trial for her progressive meningiomas. At her last follow-up visit, she was neurologically and radiographically stable after 12 months of treatment.

References

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Figure. T1-weighted MRI revealed large gadolinium-enhancing masses in bilateral cerebellopontine angles (large white arrows) with extension into the internal auditory canals (small white arrows), characteristic of vestibular schwannomas. Several extra-axial enhancing masses were present in the posterior fossa and the right middle cranial fossa (black arrows), consistent with meningiomas.

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