Letters to the Editor

Neurofibromatosis

As part of a team involved in the treatment of individuals with neurofibromatosis type 2 (NF-2, bilateral vestibular schwannomas), I was very interested in the article entitled "Acoustic Neuroma in an Adolescent Without Neurofibromatosis: Case Study," published in *JAAA* (Sells and Hurley, 1994). The authors describe a 15-year-old male with an acoustic neuroma but without associated NF. In the abstract, the authors state that "... an enhanced MRI 3 years post neuroma removal demonstrated no evidence of tumor regrowth and no evidence of other neoplasms." This MRI is not shown in the article; however, Figure 8 shows what appears to be, and what is labelled as, a post-op CT scan. I wonder if a gadolinium-enhanced MRI actually was done in the instance? As the authors note, it would be unusual for a 15 year old to exhibit a unilateral acoustic tumor in the absence of NF-2. Parry et al (1994) report a high incidence of spinal tumors (96%) and cataracts (84%) in early onset Wishart type NF-2, sometimes in the absence of multiple cranial tumors. Others also have reported similar spinal and ocular abnormalities (Pearson-Webb et al, 1986; Kaiser-Kupfer et al, 1989; Evans et al, 1992). In light of these results, NF-2 in this young man should not be ruled out without a complete spinal MRI and ophthalmological evaluation.

Sincerely,
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Evans D, Huson S, Donnai D, Neary W, Blair V, Newton V, Harris R. (1992). A clinical study of type 2 neurofibromatosis. *Q J Med* 84:603–618.

Kaiser-Kupfer M,Freidlin V, Datiles M, Edwards P, Sherman J, Parry D, McCain L, Elridge R. (1989). The association of posterior capsular lens opacities with bilateral acoustic neuromas in patients with neurofibromatosis type 2. *Arch Ophthalmol* 107:541–545.

Parry D, Eldridge R, Kaiser-Kupfer M, Bouzas E, Pikus A, Patronas N. (1994). Neuofibromatosis 2 (NF2): clinical characteristics of 63 affected individuals and clinical evidence for heterogeneity. *Am J Med Genet* 52:450–461.

Pearson-Webb M, Kaiser-Kupfer M, Eldridge R. (1986). Eye findings in bilateral acoustic (central) neurofibromatosis. Association with presenile lens opacities and cataracts but absence of Lisch nodules. *N Engl J Med* 315:1553–1554.

Sells JP, Hurley RM.(1994). Acoustic neuroma in an adolescent without neurofibromatosis: case study. *J Am Acad Audiol* 5:349–354.

Response to Otto

First, you are correct that Figure 8 and the accompanying text refer to the MRI mentioned in the abstract. The term "computerized tomography" was mistakenly substituted.

Your second concern is that Wishart NF-2 needs to be ruled out in this case. As NF-2 is an autosomal dominant genetic disorder with high penetrance, some characteristics of NF-2 would be present in a first-degree relative. Such is not the case here, unless this case represents a spontaneous genetic mutation that no one previously or presently supports.Further, a complete preoperative examination was carried out at the facility that performed the surgery. This included an ophthalmological examination and stem to stern imaging studies. No juvenile posterior subcapsular cataracts or additional tumors were detected at the time of the preoperative examination.

A recent record review showed that a 1-year postsurgery cervical spine imaging study and a 2-year postsurgery lumbosacral spine imaging study were both negative for additional tumors. Last, the most recent yearly ophthalmological examination demonstrated this young man to be free of cataracts.Thus, the case still represents an acoustic neuroma without associated neurofibromatosis.

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