Multiple bilateral cranial nerve schwannomas in a patient with neurofibromatosis type 2

Sir,

Neurofibromatosis type 2 (NF2) or multiple inherited schwannomas, meningiomas, and ependymomas syndrome is an autosomal dominant disease characterized by the development of bilateral vestibular schwannomas. We present a rare case of NF2 with multiple cranial nerve schwannomas involving the 3rd, 5th, 6th, 7th, 8th, 11th, and 12th cranial nerves. The patient, in addition, had anterior third right parasagittal meningioma and C2–C4 and C7 neurofibromas. A patient of NF2 with so many bilateral cranial nerve schwannomas has never been described in the literature.

A 20-year-old female presented with decreased hearing in both ears and decreased vision in her left eye for 2 years. Neurological examination revealed a visual acuity in the left eye of 6/60. There was bilateral sensorineural hearing loss. Magnetic resonance imaging revealed multiple extra axial lobulated masses along the course of bilateral 3rd, 5th, 6th, 8th, 11th, and 12th cranial nerves (Figures 1 and 2). Symmetrical masses were seen along the course of both trigeminal nerves involving their cisternal segments (Figure 1c). Symmetrical lesions were also seen causing widening of both internal auditory canals with cisternal extension. Lobulated masses were also seen along the course of 9th, 10th, and 11th nerves in the jugular foramen. The lesions were showing relatively homogenous signal characteristics, appearing isointense to gray matter on both T1- and T2-fluid attenuation inversion recovery images. There was no significant intralesional hemorrhage or necrosis seen in any of these masses.

Surgical treatment of patients with NF2 is complex. Management in such patients consists of removal of only symptomatic lesions with regular surveillance for the other lesions. Bilateral vestibular schwannoma may complicate the decision of surgery. The goal of surgery in such cases is to decompress the brain stem and to prolong the period of cranial nerve function. Another alternative is to wait until the affected ear becomes deaf. Larger tumors may require partial

Figure 1: (a) Magnetic resonance imaging constructive interference in steady state images axial sections showing bilateral 3rd nerve schwannoma, (b) magnetic resonance imaging constructive interference in steady state images axial sections showing right 6th nerve schwannoma, (c) postcontrast magnetic resonance imaging constructive interference images axial sections showing bilateral 5th and 8th nerve schwannoma, (d and e) postcontrast magnetic resonance imaging constructive interference images axial sections showing intraspinal and extraspinal components of 11th nerve schwannoma, (f) postcontrast magnetic resonance imaging constructive interference images axial sections showing 12th nerve schwannoma.

Figure 2: Postcontrast magnetic resonance imaging images axial sections showing various schwannomas.
removal with decompression performed when brain stem compression develops.

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**Conflicts of interest**
There are no conflicts of interest.

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