A Rare Case of Multiple Intracranial, Intraspinal, and Peripheral Schwannoma with Intracranial Meningioma

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Bilateral vestibular schwannoma (VS) are found in 90 to 95% of patients with neurofibromatosis 2 (NF2). It is reported that more than 99% of VS cases in NF2 are benign, but they remain an important cause of mortality due to their location.¹ Schwannomas can develop along the course of the cranial, spinal, and peripheral nerves, differently than

vestibular. Often, it arises from the oculomotor, trigeminal, and facial nerves.² Bilateral trigeminal schwannoma with NF2 is rare.^{3,4} Intracranial meningiomas appear in 45 to 58% of patients with this disorder.⁵ We here present a case of NF2 with intracranial, intraspinal, and peripheral involvement.

Fig. 1 Contrast T1-weighted MRI images (axial, coronal, and sagittal) showing bilateral trigeminal schwannoma (right is larger than left) with bilateral multiple small schwannoma of seventh, eighth, and lower cranial nerves with velum interpositum meningioma (a–d). Postoperative axial T1-weighted contrast MRI showing excision of right-sided tumors with residual middle fossa component of right trigeminal schwannoma (e). Large subcutaneous nodule on radial aspect of left wrist (f) and old scar of previously operated schwannoma on left side of nape of neck (g). Excised tumors (eight in number) from right cerebellopontine angle region, largest was of trigeminal schwannoma (h).

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Fig. 2 Contrast MRI of spine showing scattered multiple schwannoma along the whole length of spinal cord (a–c).

A 63-year-old man presented with 32-year history of hearing impairment in both the ears, with a 20-year history of multiple subcutaneous nodules all over the body with imbalance while walking for 18 months. He also had nasal regurgitation of fluid for 6 months. On examination he had multiple subcutaneous skin swellings all over the body along with diminution of vision in both the eyes (6/60), with bilateral fifth, seventh, eighth, and lower cranial nerves involvement. Bilateral cerebellar signs were present.

On magnetic resonance imaging (MRI) of the brain and spine, there were bilateral trigeminal schwannoma (right > left) along with bilateral multiple small schwannoma of the seventh, eighth, and lower cranial nerves with velum interpositum meningioma (►Fig. 1). Whole-spine screening MRI showed multiple schwannoma along the length of the spinal cord (►Fig. 2).

Right-sided trigeminal schwannoma along with other right-sided small schwannoma were excised. Pathologic examination of tumors revealed schwannoma. Postoperative course was uneventful. Other lesions were managed conservatively and referred for gamma knife radiosurgery.

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Conflict of Interest

None.

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