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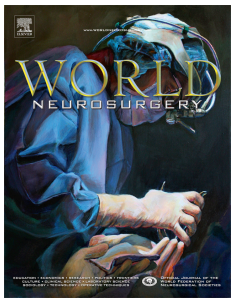

A retrospective analysis of vision impairing tumors among 467 patients with neurofibromatosis type 2

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Abstract

Background: Vision is extremely important for patients with hearing loss due to neurofibromatosis type 2 (NF2). Tumors adjacent to the anterior visual pathway can potentially impair the vision. To date, only a few case reports and small-series studies have been reported.

Objective: To evaluate the clinical features of tumors adjacent to the anterior visual pathway in a large series of NF2 patients.

Methods: Seventy-three patients with potentially vision-impairing tumors were carefully screened from among 467 NF2 patients.

Results: Among the 73 patients, 31 had intraorbital tumors, 21 had suprasellar meningiomas, and 21 had medial sphenoid ridge meningiomas. Of the 31 patients with intraorbital tumors, 17 had optic nerve sheath meningiomas (ONSMs), 9 had intraorbital schwannomas, 3 had spheno-orbital meningiomas (SOMs), 1 had an anterior cranial fossa-orbital meningioma, and 1 had a cranio-orbital schwannoma. To the date of the last follow-up, 43 (58.9%) patients experienced visual loss. In most cases, hearing loss tended to occur earlier than visual loss. Six patients underwent early operations, and they recovered well without any further vision damage. Six other patients underwent operations after having no functional visual ability in the affected eyes, and their visual ability was not saved.

Conclusion: Tumors adjacent to the anterior visual pathway, although uncommon in NF2 patients, can cause progressive visual loss. Early surgical intervention seems to be the