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A Brief Discussion of Neurofibromatosis Type 2 and Sporadic Vestibular Schwannomas

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DESCRIPTION

Treatment of vestibular schwannomas in patients with neurofibromatosis 2 balances growth control with hearing preservation, with the primary goal of preserving the patient's quality of life. The risk of functional deterioration after surgical resection of these lesions is higher than in sporadic cases. Stereotactic radiosurgery is a less invasive procedure that provides comparable, if not better, results than resection. Previous research into the effectiveness of stereotactic radiosurgery for vestibular schwannomas in patients with NF2 used Gamma Knife systems. The effectiveness of linear accelerator-delivered treatment has not been specifically addressed. Although model studies suggest that Gamma Knife is superior for lesion compliance, clinical studies with sporadic vestibular schwannomas show comparable results. In four patients with vestibular schwannoma, our experience with LINAC radiosurgery in NF2 shows good long-term growth control. Neurofibromatosis type 2 (NF2) is caused by a mutation in the NF2 gene and affects 1 in every 33-40,000 live births. The disease is inherited in an autosomal dominant manner, although spontaneous mutations account for approximately 50% of cases. While bilateral vestibular schwannomas are clinically pathognomonic for NF2, other tumours such as meningiomas and schwannomas are common along the neuro-axis. There are three clinical variants of NF2. Wishart's type presents earlier and patients often have multiple intracranial and spinal tumours. Gardner's type manifests itself later in life, has a less severe course and a significantly lower incidence of intracranial tumours. Finally, late mutations in the NF2 gene during embryogenesis cause cellular mosaicism, and these patients may not have bilateral vestibular schwannomas. The most common symptom of NF2 is hearing loss. Therapeutic recommendations focus on quality of life with an emphasis on hearing preservation or auditory rehabilitation. Approximately 75% of vestibular schwannomas will grow at an average annual growth rate of 3.2 mm/year. Compression of local structures by tumour growth can compromise facial and cochlear nerve function, as well as cause ataxia and hydrocephalus. Although surgical resection of vestibular schwannomas can reduce the tumour mass effect, there is a greater risk of iatrogenic hearing loss or facial weakness than with a sporadic lesion. Although some advocate early intervention to preserve hearing, others prefer a "wait and see" approach, reserving intervention until hearing loss occurs or tumour growth is evident.

CONCLUSION

SRS is the application of externally generated ionizing radiation to a stereotactically defined target with the aim of eradicating or inactivating the lesion. Unlike surgical resection, the goal of SRS is to control tumour growth rather than remove the tumour mass. Although SRS may not be suitable for large vestibular schwannomas, it shows better rates of normal facial movement and hearing preservation than microsurgical resection for small and medium-sized sporadic tumours. When used to treat vestibular schwannomas in patients with NF2, tumour control and hearing preservation rates, as well as facial nerve palsy complication rates, are comparable to surgical resection. We present our experience in the treatment of vestibular schwannomas in four patients with NF2 with the LINAC system equipped with circular collimators and compare its effectiveness with that of the gamma knife. Published experience with SRS in NF2 comes from institutions that use the gamma knife for treatment delivery, and similar studies using linear accelerator delivery systems are rare.

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CONFLICT OF INTEREST

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