Vestibular Schwannomas and Results of Radiosurgery

Schwannomas Vestibulares e Resultados com Radiocirurgia

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ABSTRACT

The present article aimed to describe some of the epidemiological aspects of vestibular schwannomas and available treatments emphasizing on stereotactic radiosurgery, a description of the method, its evolution over time and assessment of long-term results based on a literature review.

Key words: Radiosurgery; Stereotactic; Vestibular Schwannoma; Tumor control; Complications

INTRODUCTION

Vestibular Schwannomas are benign tumors due to excessive production of Schwann cells from the myelin sheath of one of the vestibular branches of the 8th cranial nerve. The tumor generally occupies the inner ear canal and progressively extends to the cerebellopontine angle, compressing the brainstem and other cranial nerves. In the United States, the estimated incidence is approximately 1 new case in 100,000, accounting for near 8% of all intracranial tumors. Sporadic vestibular schwannomas are unilateral, not considered hereditary and affect both sexes, with a slight predominance in women. They represent 75% of cerebellopontine angle tumors.

The natural history of vestibular schwannomas remains uncertain. There are few literature studies with long-term follow-up of a significant number of patients. The mean age of the patients at diagnosis is 45-50 years. According to published articles, the tumor growth rate is variable, on average 1mm/year.

A number of authors have suggested the presence of an extracanalicular tumor, young patients and tumor growth in the first year of radiological follow-up as predictive factors of tumor progression. However, these studies have demonstrated that irreversible hearing loss may occur even with no evidence of tumor growth in radiological examinations, contributing to the concern that conservative behavior represents a lost opportunity to preserve hearing.

Some authors demonstrated that 43% to 54% of tumors will grow in a 3-year observation period and that a small percentage (4% to 6%) will spontaneously regress.

In a series of 962 patients with 1000 vestibular schwannomas who underwent surgery the clinical presentation described hearing loss in 95%, imbalance in 61%, tinnitus in 63%, changes in facial sensitivity in 9% and facial paralysis in 6%. Mean hearing loss time until diagnosis is approximately 3-4 years.

Management options for patients with vestibular schwannoma

There are a number of therapeutic options for patients with vestibular schwannoma and several aspects should be assessed to make them more adequate and individualized for each patient. The options include partial or total microsurgical resection, radiosurgery, which can be the first choice, or performed after partial surgical resection and confirmed tumor growth progression, fractionated radiotherapy and clinical observation with radiological follow-up (magnetic resonance).
Several aspects must be taken into account such as tumor size, clinical conditions of the patient, age, and hearing status. Each therapeutic modality has its indications, limitations and morbidity. Microsurgical resection is indicated in cases of large tumors, usually more than 3 cm, which compress the brainstem. Asymptomatic elderly patients with small tumors can be observed with the periodic application of magnetic resonance.

**Evolutionary and technical aspects of radiosurgery in the treatment of vestibular schwannomas**

The first article on radiosurgical treatment of vestibular schwannomas was published by Leksell, in 1971, 20 years after his pioneering conceptual paper on stereotactic radiosurgery. Since then, radiosurgery has evolved sharply and among the factors that have contributed to this evolution is the advance in medical imaging methods (tomography and magnetic resonance), which has resulted in better definition of anatomical structures and tumors. This has also contributed to more conformational dosimetric planning, and emergence of automated robotic systems that provide more precise dose delivery and optimize the prescription dose, leading to fewer adverse effects without compromising the effectiveness of the radiosurgical method.

Radiosurgery can be performed with the gamma-knife, linear accelerator (Linac), cyberknife or proton beam systems.

In our service, we use the linac (2100-Varian). The stereotactic frame is fixed under local anesthesia to the patient’s head, and after followed by brain tomography. The fusion of tomography and magnetic resonance (previously performed) is achieved through planning software (Brainlab Corp.), where the contour of the tumor and anatomical structures of interest, including the cochlea, are traced in order to maximize dose delivery to the tumor, sparing normal adjacent structures. Treatment lasts approximately 20 minutes per isocenter, and at the end, the frame is removed and the patient sent home.

Current prescription dose for vestibular schwannoma is between 12 and 13 Gy, which is associated with a low incidence of complications without compromising the high rate of tumor control.

The expected result of radiosurgical treatment of vestibular schwannomas is the stabilization of tumor growth, and possible reduction in their volume. These tumors rarely disappear after irradiation.

**RESULTS**

Gabert & Regis reported a case series of more than 1500 patients with vestibular schwannomas treated with gamma knife, using a 12 Gy dose, obtaining tumor control in 97%, without facial paralysis and a hearing preservation rate of 60%.

Kondziolka et al. published a case series of 162 patients with a 5-10-year follow-up, treated with gamma knife, and obtained a tumor control rate of 98%. There was a reduction in tumor volume in 62% of the cases, 33% remained unchanged and 6% exhibited a slight increase. Some tumors grew slightly (1-2 mm) in the first 6-12 months after treatment and then regressed.

Lunsford et al. reported a case series of 829 patients treated with gamma knife, obtaining a tumor control rate of 97%, in a 10-year follow-up. There was no improvement in the control of tumor growth with doses of more than 13 Gy when compared to doses between 12.5-13 Gy.

In a case series of 216 patients treated with gamma knife, using a marginal dose of 12-13 Gy, in a 12-year follow-up, the authors reported a tumor control rate of 98% and hearing preservation of 71%, with no additional facial motor compromise after radiosurgery.

Hasegawa et al. published a case series of 440 patients treated with gamma knife and a follow-up of more than 10 years. They reported 79% of the patients who underwent radiosurgery as the first choice and 21% who were submitted to prior microsurgical resection. Tumor control rate at 5 and 10 years was 93% and 92%, respectively. The rate of facial nerve preservation was 97% for the group of patients that received a marginal dose of more than 13 Gy, and 100% for the group that received a marginal dose of less than 13 Gy. One patient exhibited malignant transformation.

Friedman reported a case series of 390 patients treated with the linac system, 63 of whom were followed-up for more than 5 years. The tumor control rate at 2, 5 and 10 years was 98%, 90% and 90%, respectively. The rate of facial and trigeminal nerve compromise was 4.4% and 3.6%, respectively.
1994, when the dose decreased to 12.5 Gy, the incidence of trigeminal and facial nerve compromise has been 0.7%.

In patients with type 2 neurofibromatosis, radiosurgery has been less effective in controlling tumor growth when compared with sporadic vestibular schwannoma. Mathieu published a case series of 62 patients, with a mean follow-up of 53 months, in which the local control rate at 5, 10 and 15 years was 85%, 81% and 81%, respectively.

**COMPLICATIONS**

Since the early 1990s, when the marginal dose decreased to 12-13 Gy, the estimated risk of facial paralysis has been less than 1%, hearing loss between 21% and 29% (up to 2 years of treatment), while for intracanalicular tumors the risk is lower (around 10%). The risk of trigeminal neuropathy is estimated at between 3%-8%.

The estimated risk of malignant tumor transformation after radiosurgery is 1:20,000.

**CONCLUSION**

Stereotactic radiosurgery is a safe and effective method, widely used throughout the world in the treatment of vestibular schwannomas in the control of tumor growth associated with a high rate of hearing and facial nerve preservation. The results published in the literature show that the method was reproducible at different centers worldwide.

**REFERENCES**


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