EDITORIAL

Defining the Optimal Management for Patients with Large Vestibular Schwannomas

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Vestibular schwannomas (VS), also known as acoustic neuromas, are benign, slow growing tumors that arise from the vestibular-cochlear nerve. Patients diagnosed with vestibular schwannomas typically present with hearing loss, tinnitus, or dizziness. The best management of patients with small- to moderate sized vestibular schwanommas is controversial.[1-3] Although it is estimated that vestibular schwanomas account for 6-10% of primary brain tumors and affect 17.4 per one million people annually, the size of newly diagnosed vestibular schwannomas has decreased secondary to the widespread application of magnetic resonance imaging.[4] As such, in most higher income countries, large or giant (>4 cm) vestibular schwanomas are rare and this hinders most attempts at good research in this area.

These giant tumors cause considerable brainstem compression, may involve or compress the 5th, through 10th cranial nerves and may involve associated vessels. Gait ataxia, long tract signs, signs of intracranial hypertension such as papilledema are frequent; hydrocephalus can be seen in more than half of these patients prior to treatment.[5] These pathoanatomic facts may complicate any attempt at treatment but also alter the balancing of risks and benefits of treatment compared to smaller tumors. Although patients with small and medium sized tumors can often be observed for years before progression leads patients to seek treatment, those with larger tumours have a much smaller margin of reserve before developing serious complications caused by the associated mass effects.

Surgery for the mass effect seen with giant tumors can be life saving and is rarely debated. The best results from surgery for giant tumours include resection rates of better than 90%, preserved facial nerve function in 75%; lower cranial nerve palsies of 2-19%, CSF leak in 2%, meningitis in 5% and rarely death due to postoperative hemorrhage or ischemic stroke.[5,6] In an attempt to improve postoperative results, strategies such as intentional subtotal resection, staged resection, and, intentional partial removal followed by radiosurgery have been advocated with reasonable results reported thus far.[7-9]

Like surgery, most of the literature on radiation techniques such as radiosurgery (RS) and fractionated stereotactic radiation therapy (FSRT) for the treatment of VS focus on outcomes for small and medium sized tumors and have significant methodologic shortcomings. Despite the proposed radiobiologic benefits and technical differences of each approach, little evidence has supported that one technique is superior.[10] The small number of series reporting results of radiosurgery or stereotactic radiotherapy for VS over 3cm, show that outcomes are not as good as they are for smaller tumors. For example, Mendl et al reported actuarial five-year rates of: maintenance of pre-treatment hearing level probability of 30%, facial nerve preservation probability of 80%, trigeminal nerve preservation probability of 85%.[11] They showed that tumor progression occurred in 4 of 25 (16%) patients regardless of SRS or FSRT. The overall five-year tumor control probability was 82%.They concluded that in patients with tumor volume above 18.6 cm^3 or tumor diameter above 3.3 cm other treatment strategies should be considered in the management of large VS because of an increased complication rate. So size matters for considering any sort of treatment for VS.

In this issue of the Journal, Zeiler et al have made an important addition to this literature.[12] In 25 patients with VS between 3 and 4 cm (average 3.28 cm) treated at the Winnipeg Gamma Knife (GK) Unit, they showed that gamma knife treatment at a marginal dose of 13Gy or lower, could achieve reasonable short-term outcomes. Although transient complications occurred in 80% of patients, these were manageable and all cleared by the time of follow-up (average 34.5 months). Sixteen percent of the patients developed hydrocephalus that required shunts, and, one patient had permanent worsening of hemifacial spasm. There were no reported cases of permanent worsening of ataxia, long tract signs, trigeminal, facial or lower cranial nerve function. Ninety-two percent had tumour control with 56% showing a decrease in tumour size and two patients' tumours grew. The authors acknowledge the significant limitations of such a small retrospective series with short follow-up.

I believe that the report by Zeiler et al is particularly important because it raises so many new questions that require further study.[12] Will these preliminary results persist in the long-term? Should we treat hydrocephalus before embarking on GK? Will surgery for the patients who eventually fail give worse results than if they had undergone surgery in the first place and reserved GK as a secondary treatment? Who are the best patients with large tumours to offer GK as a primary treatment? Our approach in Toronto has been to offer surgery or fractionated stereotactic radiotherapy to these patients and not GK. This state of affairs offers clinicians in Canada an opportunity. By developing collaborative efforts between centres in Canada and beyond, we can begin to answer some of these important questions through properly designed studies and prospective collaborative registries.

So where do we stand when it comes to managing patients with VS larger than 3 cm? Clearly, size matters. Patients have a narrow margin of safety with observation and regardless of which treatment they choose. All treatment modalities have higher complications and failure rates as the tumour enlarges.

Clinicians must individualize treatment to the needs of the patient through a careful review of their history, physical, radiological and audiological data. Clearly those suffering significant mass effect and raised intracranial pressure will need to strongly consider the benefits of surgery. For those with no or minimal signs of mass effect, clinicians will have to carefully work with patients to come to a rational management plan. Ultimately, we will need to perform well controlled collaborative studies to resolve these issues.

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