The treatment of giant acoustic neuromas

António Cerejo¹,²

Abstract

It is widely accepted that giant acoustic tumors (4 cm or more in diameter) require surgical treatment. However, this is a demanding technical procedure, with possible postoperative neurological dysfunction and complications. Giant tumors are frequent in specialized centers, representing about 15% of the total number of acoustic neuromas. The size of the tumor is an important factor concerning the difficulties of surgery. Learning is time-consuming, and the results are highly dependent on surgeon's skill and on a well trained team. The details of the procedure are briefly explained, given attention to the more important steps and possible pitfalls.

In my personal view, total removal of the tumor in a single operation should be the goal of the surgery. Since experience is crucial, I defend the referral between Neurosurgery departments. The importance of anatomical training and the role of excellent intraoperative monitoring is emphasized. Excellent intraoperative monitoring and postoperative management in a Neurocritical Care Unit are important factors for a good outcome.

For conclusion, with adequate training and experience, total removal of the tumor in a single operation is possible, with acceptable rates of morbidity and no mortality.

Keywords: Giant acoustic neuroma, Surgery.

¹Department of Neurosurgery, São João Hospital Centre, Porto, Portugal
²Faculty of Medicine, University of Porto, Porto, Portugal

Correspondence: António Cerejo
Faculty of Medicine, University of Porto
Alameda Prof. Hernâni Monteiro, 4200-319, Porto, Portugal
Email address: a.cerejo@sapo.pt

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**Introduction**

From the beginning of Neurosurgery, the treatment of acoustic neuromas has been a controversial issue for neurosurgeons. If small and medium-sized tumors may be observed, or treated either by surgery or radiosurgery, giant acoustic neuromas (4 cm or more in diameter) require surgical treatment [1]. The problem is that the surgical removal of giant acoustic neuromas is a challenging task, since it remains a demanding technical procedure, with possible postoperative neurological dysfunction and a significant rate of complications [1-5].

**Giant acoustic neuromas**

**Clinical presentation**

In our experience, as in other authors series, patients with giant acoustic neuromas have neurologic symptoms due to the involvement of the cochlear nerve (hypoacusia), but also due to dysfunction of other cranial nerves (like the trigeminal and lower cranial nerves) and compression of the cerebellum and brainstem. They may also present intracranial hypertension. So, besides hearing loss, patients may complain of ataxia, trigeminal neuralgia or facial dysesthesia, and swallowing difficulties.

The incidence of giant tumors represents approximately 2% of all acoustic neuromas diagnosed at Western countries [4], but admissions due to this pathology in specialized centers may be much more frequent, due to patient referral.

At our hospital, giant tumors represent about 15% of the total number of acoustic neuromas. According to our experience, it is also possible to speculate that tumors may enlarge in a particularly fast manner, due to intratumoral hemorrhage (sometimes present in pathologic examination of large lesions) or cyst enlargement, since there are cases of overwhelming signs of intracranial hypertension and cerebellar dysfunction in patients that noticed the hypoacusia only a few weeks before admission.

**Surgical treatment**

In the majority of cases, microsurgical removal of the tumor is the only treatment option in giant acoustic neuromas. Surgeons must be aware that this is a dangerous procedure, with possible mortality and considerable morbidity, with a significant rate of complications. Learning is difficult and time-consuming, and the results are highly dependent on surgeon’s skill and on a well trained team.

The size of the tumor is an important factor concerning the difficulties of surgery, in terms of the possibilities of total removal, the rate of complications and neurological dysfunction. In giant tumors, the brainstem is compressed and displaced. In some cases, the “protective” arachnoid membrane may be disrupted, which may make dissection of the tumor from the brain stem very difficult. Other major problems of the surgical procedure are related to the distortion and stretching of the cranial nerves and vascular structures [5].

In my opinion, some details of the surgical procedure are very important if the surgeon has the objective of a safe, complete removal of the tumor. First, the use of semi-sitting position, that brings considerable advantage when dealing with these lesions. There is a real risk of air embolism, but, with appropriate positioning, and frequent check of venous rupture by applying jugular compression, this risk may be minimized, and the surgeon may take advantage of the absence of blood in the dissection of the tumor from the nearby structures. Besides, the surgeon may use both hands for tumor dissection, since irrigation is enough to keep the field clear.

Very important is the use of retrosigmoid approach. A craniotomy 3-4 cm in diameter is enough to give access to all the structures from the tentorium to foramen magnum. Special care must be taken in the careful closure of mastoid air cells, avoiding post operative cerebrospinal fluid fistula.

Also crucial is neuromonitoring. In giant acoustic neuromas the facial nerve is severely stretched, especially as it leaves the cerebellopontine angle cistern at the entrance of the internal auditory canal. The nerve may be so thin that “spreads” around the tumor. Any manipulation of the facial nerve may cause loss of function, and the possibilities of recovering are variable [1, 2]. The preservation of the cochlear nerve in patients with preoperative hearing is very difficult in giant tumors, due to the extreme susceptibility of this nerve to injury during manipulation [1]. Having this facts in mind, intraoperative monitoring of the facial nerve (with electromyography and, more recently, facial nerve motor evoked potentials) and cochlear nerve (with auditory evoked potentials) is nowadays an imperative tool in the preservation of the function of these nerves. The caudal cranial nerves have to be dissected from the tumor in most cases, and swallowing disturbances may arise after surgery [2, 4]. Monitoring of these nerves may also be used whenever possible.

The details of microsurgical removal are another important issue. There are some aspects that may anticipate an “easier” surgery, or a more time consuming procedure. Using the semi-sitting position, tumors that extend superiorly may be less difficult to dissect, since the lesion tends to fall due to the effect of gravity. The same may happen when there is a visible arachnoid cistern anterior to the tumor, since it may provide a much better idea, intraoperatively, of the dimensions of the lesion.

In my experience, some steps must be considered: always open the cisterna magna before placing the cerebellar retractor, allowing the exit of a large amount of cerebrospinal fluid and cerebellar relaxation. After that, a retractor is placed on the cerebellum, just close to the tumor surface. This retractor is 1cm large, and I try to keep it as fixed as possible, avoiding cerebellar contusion. Then, an extensive “debulking” of the tumor is started (in general, no coagulation is necessary inside the tumor, since the intra
tumoral bleeding will spontaneously stop in most cases). After debulking, we start the dissection in the superior portion of the lesion, even if this maneuver may carry a risk of damaging the facial nerve. Taking advantage of gravity, with limited use of coagulation, continuous irrigation of the surgical field and the help of neuromonitoring, it is possible to identify and dissect the facial nerve from the tumor. Keeping the arachnoid membrane intact is very important, since this membrane will provide a safe dissection from brain stem. I always try to find the facial nerve at the brain stem, to have control over the position of the nerve. The internal auditory canal is then opened with diamond drill under continuous irrigation and the nerves VII and VIII are identified in the internal acoustic canal. The dissection of the tumor from the nerves proceeds in four directions (superior, inferior, lateral and, medial) until the removal is completed.

The risk of complications, such as brainstem infarction, posterior fossa or brainstem hemorrhage, facial nerve paralysis, hearing loss, dysfunction of the caudal cranial nerves, cerebrospinal fluid leakage, and infection are considerable in the surgical treatment of these tumors [1, 2]. These complications may be life threatening without appropriate treatment, and the management of these patients in a Neurocritical Care Unit in the postoperative period is mandatory.

The ultimate objectives in the treatment of these tumors are the complete resection, with preservation of facial and hearing function, no neurological deficits, and return of the patient to normal life. In the present times, the management of giant acoustic neuromas is either complete microsurgical tumor removal in one or more stages, or subtotal removal followed by radiosurgery of the residual part. The need for staged removal may be considered when the procedure is too time-consuming, which may be a problem, since the procedure may take several hours, although, with increased experience, the surgery tends to be much faster. In my experience, all the problems of a single procedure remain the same in staged surgery, and dissection may be more complicated due to scar tissue after the first surgery.

Subtotal removal followed by radiosurgery is possible, but the problem is the tendency to consider the extent of the tumor removal greater than it really is, from the point of view of the surgeon, peroperatively. Besides, the overall results of this strategy, comparing with total removal, remain to be established [2, 4], due to considerable rates of tumor regrowth and new postoperative neurological dysfunction. In my opinion, this strategy may be considered if the surgeon is not comfortable enough to proceed to total removal, or if the risk of lesion of neurovascular structures seems to be unacceptable in particularly difficult tumors.

In my personal view, total removal of the tumor is possible in a single operation, without mortality and with an acceptable rate of serious complications. The experience of the surgeon is crucial, and therefore, the team must be familiar with the procedure. Of course, it is not easy to gain the necessary experience in a department that deals with an average number of cases. So it is advisable, in my opinion, that the surgery of giant acoustic tumors is “restricted” to a few people in each department. More than that, the referral between Neurosurgery departments should be considered, at least in relatively small countries or neighbor regions. The major problem is how to improve the surgical results in a single operation with radical tumor removal, rather than follow alternative strategies (even if valuable in some circumstances) that do not provide the cure of the patient.

The results, in terms of facial function, hearing preservation and post operative complications, still have to improve, and several aspects are crucial for this goal: first, the importance of anatomical training and improvements in personal surgical technic must always be emphasized when dealing with this pathology. More than recognize the different vascular and neuronal structures, the surgeon must be able to predict the location of these structures and take extreme care until he is able to get an accurate perspective of the surgical field. This skill comes with training and experience. Second, major advances must be pursued in intraoperative monitoring. Electromyography of the facial nerve, currently used in acoustic neuroma surgery, is very important for intraoperative localization of facial nerve, but its value for determination of postoperative facial function is not well established [6, 7]. Computerized analysis of the intraoperative facial nerve electromyogram, with real-time monitoring of pathologic patterns during surgery, and facial motor evoked potentials may have a predictive value concerning postoperative facial function, and its application during surgery may have an important contribution for facial nerve outcome improvement. The same may happen if improvements in the evaluation of the peroperative data of brain stem auditory potentials can be achieved.

Of course, excellent postoperative management in a Neurocritical Care Unit is an important factor in the outcome of these patients, since a significant complication rate is expected, but a very good evolution is possible with appropriate care.

**Conclusion**

With appropriate tools and adequate training and experience, I believe that there is an open field for improvement in the surgical treatment of giant acoustic neuromas. As in other fields of human knowledge, we must be able to keep our mind open to better solutions, but we must also believe that the way of getting better results is by training to improve our skills.

**Competing interests**

The author declares no conflict of interest.
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