Surgery of vestibular schwannomas

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ABSTRACT

Vestibular schwannomas (VS) are the most common tumors of the cerebellopontine angle (CPA). Their surgical management has reached high standards in the last decade. Treatment options for VS are microsurgical removal or Radiosurgery. The following three basic operative approaches are currently utilized: Retrosigmoid approach (RSA), translabyrinthine and middle fossa approach. The following article elaborates the operative technique by the senior author based on his vast experience of VS surgery formed over the last three decades; during which period he has operated more than 3500 of such patients

Key words: Vestibular schwannoma, surgery, operative technique

INTRODUCTION

Vestibular schwannomas (VS) are the most common tumors of the cerebellopontine angle (CPA). Their surgical management has reached very high standards in the last decades due to the evolution of microsurgery, neuroanesthesia, neurological intensive care, neuromonitoring and neuroradiology. The operative mortality has become exceptional and the major morbidity rate is very low. In experienced hands, complete tumor removal is achieved in 80 to 99% of the patients.[1-5] The major goal has become preservation of neural functions, in particular of facial nerve function and hearing, which are the most important predictors of the quality of life.

The wide distribution of MRI facilities allows for earlier or incidental VS detection, when they are still asymptomatic and small in size. The conservative or “wait and scan” strategy is a reasonable initial management option in such VS.[6] The rationale of this strategy is the lack of progress of some tumors even decades after their radiological diagnosis. Moreover, MRI is a reliable means for monitoring of their growth tendency. The decision on the individual management plan should always be taken considering not only the tumor features and the patient’s clinical condition but also his psychological comfort, expectations and occupational activities. Despite our better understanding of VS biology, their natural evolution still cannot be predicted.[6,7] We recommend initial follow-up in the following cases: 1) elderly or somatic unstable patients with small VS and mild stable symptoms; 2) in patients with small tumors and complete hearing loss; 3) in patients with VS on the only hearing side; or 4) those refusing surgery.

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One important consideration, however, is that hearing preservation may be possible only if surgery is performed at an earlier stage - in patients with smaller tumors who still have functional hearing and good brainstem auditory evoked potential (BAEP) response. On the other hand, significant risk of loss of useful hearing during observation exists even in stable non-growing VS.[8] Moreover, hearing deterioration may be of sudden onset. Thus, during the period of follow up, hearing-preserving surgery might become impossible.

Treatment options for VS are microsurgical removal or stereotactic radiosurgery/radiotherapy.[9,10] The following three basic operative approaches are currently utilized: Retrosigmoid approach (RSA), translabyrinthine and middle fossa ones.[5,11-13] Their advantages and limitations have been discussed extensively in the literature. A general conclusion that could be drawn is that the results depend more on the individual surgeon’s experience than on the selected approach. A learning curve has been shown to exist in attaining good standards of nerve preservation and low morbidity.[5]

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OPERATIVE TECHNIQUE

The senior author's (M.S.) concept of VS surgery was formed over the last three decades; during the period he has operated more than 3500 patients with VS.[5,13-16] We favor the RSA as a simple and flexible approach, providing excellent access to the whole CPA. It is related to a very low procedure-related morbidity rate, allows for hearing preservation and provides wide exposure of the tumor, regardless of its size, extension and pathological type [Figure 1].[17-19]

The semi-sitting positioning of the patient has several major advantages: CSF, blood and fluids used for irrigation drain out spontaneously due to gravity. Thus, there is no need of constant suction to keep the operative field clear, the need for frequent coagulation during tumor removal is obviated and the surgeon can work bimanually. Moreover, the venous outflow is enhanced and the venous bleeding is less. The head is extended slightly, flexed and rotated to the involved side under the feedback control from the somatosensory evoked potentials monitoring (SSEP). If the anesthetic and operative principles are followed strictly, the risks related to the semi-sitting position, such as venous air embolism, may be avoided. In experienced hands, the semi-sitting position does not cause any lasting morbidity.[20,21]

Preservation of facial nerve function and hearing requires their monitoring throughout the surgery.[5] BAEP monitoring is utilized to monitor cochlear nerve function. Electromyography recordings of the orbicularis oris and oculi muscles are used to monitor facial nerve function via reusable 12-mm twisted-pair subdermal needle electrodes placed at the tumor site. Anatomical preservation of the facial nerve is assessed at surgery using both visualization and electrical stimulation of the facial nerve at its brainstem origin and in the IAC.

The skin is incised 2.5 to 3.5 cm behind the mastoid;[14] the inferior part of the incision should be slightly curved laterally to allow for an unrestricted by the soft tissues access to the cerebellomedullary cistern and lower part of the CP angle (the lateral-to-medial perspective). The initial burr hole is placed 2-2.5 cm below the superior nuchal line, two-thirds behind and one-third in front of the occipitomastoid suture. In case an emissary vein is seen in the area, the placement of the burr hole should be modified accordingly. Importantly, the asterion is not an absolutely reliable anatomic landmark and is quite variable. The course of the sigmoid sinus, in contrast to that of the transverse sinus, is less variable. It projects along the axis defined by the mastoid tip and the squamosal-parietomastoid suture junction or over the mastoid groove.

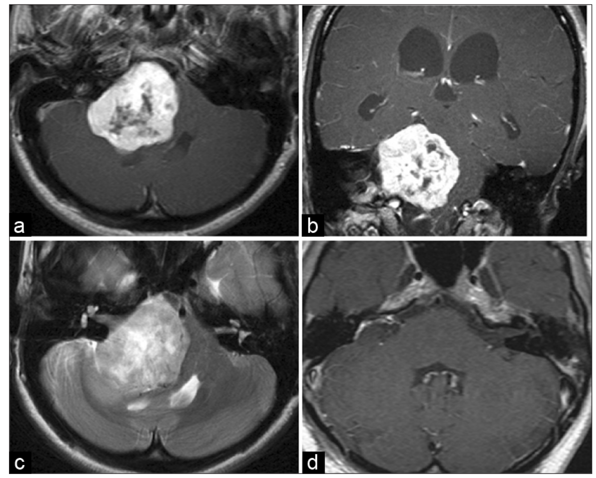

Figure 1: (a-c) Preoperative and (d) postoperative MR images of a patient with very large VS on the right side. The tumor has been removed completely via the retrosigmoid approach. In the postoperative image, the fat tissue in the IAC area is visible as a hyperintense signal

A craniectomy exposing the borders of the transverse and sigmoid sinuses is then made. More extensive exposure of the sinus is not necessary and is dangerous. It may lead to sinus laceration or desiccation, with the risk of later thrombosis. The bone opening has to extend inferior to the floor –the horizontal plane– of the posterior fossa. We avoid making one-piece craniotomy in the RSA because it is related to higher risk of injury to underlying sinuses and tearing of the emissary vein or the dura by the craniotome. The dura is incised curvilineary approximately 1.5-2 mm medial to the sigmoid and inferior to the transverse sinus. Auxiliary dural incisions are avoided. Such opening allows for unrestricted access to the CP angle and IAC and for a primarily watertight closure in most cases.

Major concern is the prevention of retraction-induced injury of the cerebellum. The initial step should be always wide opening of the cerebellomedullary cistern to allow sufficient egress of CSF. The retractor is placed only when the cerebellum relaxes away from the petrous bone. In most VS, the intrameatal tumor part is approached initially. Once the main anatomical landmarks of the CPA are identified, the dura behind the internal auditory canal (IAC) is excised circumferentially and its posterior wall is widely opened. If hearing is to be preserved the inner ear structures – the common crus, vestibular aqueduct and posterior semicircular canal – should be preserved.[22] The nerves in their intracanalicular segment have almost always preserved anatomical shape and less variable location. The facial nerve is displaced almost always anterior to the tumor, superior to the cochlear nerve. These nerves have to be identified and the intrameatal tumor portion should be removed or -in case of more severe adhesions to the nerves - at least debulked. In VS with significant posterior extension along the pyramid, primary IAC opening is difficult. In such rare cases, the IAC is opened after removal of the overlying tumor portion.

The extrameatal tumor removal starts with tumor debulking or internal decompression, regardless of its size. Debulking is best performed with an ultrasonic aspirator, which allows for gentle and controlled removal. Alternatively, a platelet-shaped knife can be used. Tumor debulking should proceed systematically in all directions. Then, the remaining tumor part is pulled slightly into the operative field and its capsule is dissected in the arachnoid plane follows. In larger VS, these steps are repeated alternatively in all directions.[25]

The arachnoid plane should be clearly identified and the dissection should always follow it. The so-called three-hand technique is extremely useful: The surgeon uses both his hands for tumor preparation and removal, while the assistant irrigates and cleans the operative field. In case the dissection is not in the correct plane, the fragile nerves can be destroyed or their vascular blood supply can be interrupted. In the area near the porus of the IAC, the nerves are very adherent and vulnerable. This portion is dissected only when both the proximal and the distal portions of the facial nerve have been freed from the tumor. Final hemostasis is done after tumor removal and identification of all neural structures. The posterior skull base is reconstructed with methyl methacrylate.

RESULTS

In a recently evaluated series of 200 patients, complete tumor removal was achieved in 196(98%).[5] In four patients (2%), the tumor capsule was so adherent to the facial nerve that its removal would inevitably damage the nerve. Small piece of the capsule was therefore left unremoved. Three of these four schwannomas were cystic.

Tumor recurrence was observed in one patient (0.5%). Two years after surgery, this patient presented with an asymptomatic 8-mm intracanalicular recurrent tumor.

Facial Nerve Outcome

The facial nerve anatomical preservation rate was 98.5% (197/200 patients).[5] In VS with extension grades T1, T2 and T3, the rate of facial nerve preservation was 100%. Two weeks after surgery, excellent facial nerve function (House–Brackmann Grades I and II) was present in 59% of patients, good function (House–Brackmann Grade III) in 16%, fair (House–Brackmann Grade IV) in 17%, and poor (House–Brackmann Grade V) or no function (House–Brackmann Grade VI) in 8%. The best functional results were achieved in VS patients with grade T1 or T3: Excellent facial function was achieved in 90 and 96%, respectively. At the last follow-up examination, 81% of patients had excellent or good facial nerve function. Moreover, there were no patients with total facial palsy.

Analysis of the literature shows that the rates of facial nerve anatomical and functional preservation are similar among the middle fossa, translabyrinthine and retrosigmoid approaches. Preoperative tumor size and the experience of the surgeon are the main predictors of facial nerve preservation.[4,11,23,24] In VS larger than 4 cm, good function is achieved in 38 to 75%.[1,3,26]

Hearing Preservation

We attempt cochlear nerve and hearing preservation in all patients with preoperatively available hearing. The rate of cochlear nerve anatomical preservation increased from 79% in the first 1000 cases, operated by the senior author, to 84% in the recent series.[5,15] Functional preservation of the cochlear nerve was achieved in 46% of patients in our previous study and improved to 51% in the current. In cases of smaller VS (extension grades T1 to T3a), the rates of functional hearing preservation were 60, 72, and 56%, respectively.

The most significant parameters correlating to hearing preservation are tumor size and extension, preoperative hearing level, BAEP wave preservation and surgeon's experience.

Operative Morbidity and Complications

In four patients (2%), we observed transient neurological complications, which resolved completely during follow up. Computed tomography scanning after surgery revealed small insignificant intracranial hemorrhages in the CPA in two cases (1%) – they were managed conservatively.

CSF leak following VS removal via the RSA originates most frequently from the air cells located in the petrous bone around the IAC and mastoid air cells, which are opened during the surgical procedure. The prevention of this complication relies on the good sealing of the potential sources of leakage. Initially we used muscle pieces; but in the last few years, we prefer to apply multiple fat pieces, harvested from the incision area. They are placed in the area of bone drilling and fixed with fibrin glue. Due to this modification, we could reduce the CSF leak rate from 5.7% to 2.2%.[27] In case CSF leak appears, the management involves the immediate placement of a lumbar drain for seven days. After removal of the drain, provocation test is made; if it turns out to be positive, the patient should undergo surgical revision.

Three patients had postoperative hydrocephalus and were treated with temporary external ventricular drainage. The postoperative headache rate following VS removal via the RSA is reported to be as high as 54% and has been reported earlier.[28] The methyl methacrylate cranioplasty offers the possibility to achieve better cosmetic result and prevents scar tissue formation between the dura and neck muscles. In the present series, we did not observe any patient with lasting headache after surgery.

Surgery after Failed Radiosurgery

Radiosurgery is an alternative management option of VS, which is becoming increasingly popular. Treatment failure, however, occurs in 2.5% to 9% of the tumors after radiosurgery/radiotherapy.[9,10] Patients with failed radiosurgery are treated either with repeated radiosurgery or surgically.[29-31] Surgery is indicated in patients with large tumors compressing the brainstem, in case of sustained tumor growth, which is documented on serial MR examinations or in case of progression of symptoms.[32]

Surgery after failed radiosurgery is more complex and challenging due to the development of extensive post-radiational changes.[29,31] Still, its goal should be compete tumor removal.[32] It is difficult to assume that a remnant of VS that expanded despite the previous therapy will change its biological behavior and stop growing. As recommended by some authors, partial removal[31,33] may indeed lead to better functional outcome in some cases; but it is a short-term solution.

The surgical concept and the operative technique applied in such cases do not differ from that in non-treated VS of the same size.[32] The major challenges are the dissection of tumor capsule from cranial nerves or brainstem due to the tight adherence or lack of clear arachnoid plane; the increased vulnerability of the facial nerve; and the radiation-induced vasculopathy with the related tendency for delayed rupture and bleeding.

In a recently published analysis of our experience, we found that complete removal of VS after failed previous radiosurgery/radiotherapy can be achieved without mortality and major morbidity.[32] The postoperative facial nerve function is indeed worse than in untreated VS of similar size and the risk of new neurological deficits or CSF leak tended to be higher. However, the differences were not significant.

CONCLUSION

The goal in treating VS should be complete removal because it is the only option that potentially cures the patient. Preservation of neurological functions is essential for the maintenance of the quality of life after surgery. In our experience, these goals can be achieved safely and successfully by using the retrosigmoid approach.

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