SKULL BASE SURGERY OF NON VESTIBULAR SCHWANNOMAS OF THE POSTERIOR CRANIAL FOSSA

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ABSTRACT

Objective: *To study the surgical approaches and results of surgery in twenty one patients with non vestibular schwannomas operated upon at the neurosurgery department*

Methods: *We retrospectively analyzed a series of 21 patients with histologically verified non-vestibular schwannomas, treated at the Neurosurgery Department, Alexandria University in the period between 2003 and 2008.*

The age in this group of patients ranged from 20 to 65 years (Mean 38 years). All patients underwent detailed general and neurological examination and preoperative gadolinium-enhanced magnetic resonance imaging (MRI). Computerized tomography (CT) scan with thin slices was used in 16 cases to study the bony anatomy. The mean follow-up period was 29 months (range 10 to 61 months).

Results: *Twenty one patients with intracranial schwannomas arising from cranial nerves other than the vestibulocochlear were surgically treated in the Neurosurgery department, Alexandria University, in the period between 2003 and 2008. There were 14 males and 7 females and the mean age was 38 years. Seven of our patients underwent surgery elsewhere for partial resection before being referred to our facility; one of these cases underwent fractionated stereotactic radiotherapy for residual tumor after the first surgical intervention.*

Conclusion: *Non-vestibular schwannomas are rare tumors that are best treated by total surgical resection. The location and size of the tumor dictates the surgical approach, however skull base approaches offer better tumor exposure and therefore result in better total tumor resection rate.*

Key words: *Skull base, schwannomas, surgical approaches.*

INTRODUCTION

Schwannomas are usually slowly growing benign tumors that arise from schwann cells.[5,24] Although they can be located along any cranial, spinal, or peripheral nerves, 25% of schwannomas are found in the head and neck. Intracranial schwannomas arising from sites other than the vestibular nerve are rare, constituting 2.9 to 4% of all schwannomas.[8, 11] In this article we reviewed the surgical outcome of 21 consecutive cases of non-vestibular schwannoma that had been surgically treated in the Neurosurgery Department, Alexandria University.

METHODS

We retrospectively analyzed a series of 21 patients with histologically verified non-vestibular schwannomas, treated at the Neurosurgery Department, Alexandria University in the period between 2003 and 2008.

The age in this group of patients ranged from 20 to 65 years (Mean 38 years). All patients underwent detailed general and neurological examination and preoperative gadolinium-enhanced magnetic resonance imaging (MRI). Computerized tomography (CT) scan with thin slices was used in 16 cases to study the bony anatomy

The mean follow-up period was 29 monthes (range 10 to 61 months).

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RESULTS

Twenty one patients with intracranial schwannomas arising from cranial nerves other than the vestibulocochlear were surgically treated in the Neurosurgery department, Alexandria University, in the period between 2003 and 2008. There were 14 males and 7 females and the mean age was 38 years. Seven of our patients underwent surgery elsewhere for partial resection before being referred to our facility; one of these cases underwent fractionated stereotactic radiotherapy for residual tumor after the first surgical intervention.

Imaging characteristics

MR images were available for all patients. The tumor had a low to isointense signal on T_1-weighted images, and a high signal on T_2-weighted images. CT scans with detailed bone algorithms were available for 16 patients Pre-operative embolization was not attempted in any of our patients.

Trigeminal nerve schwannoma

There were 14 cases of trigeminal nerve schwannoma in this series. There was a slight male predominance (8 males and 6 females). All these patients presented with varying degrees of trigeminal nerve dysfunction (mostly facial hyposthesia, but few cases presented with trigeminal neuralgia and weakness in the muscles of mastication). Other clinical findings included other cranial nerve dysfunction (III, IV, VI, VII and VIII), cerebellar manifestations and hemiparesis.

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Six patients had prior surgical intervention at other institutions and presented with re-growth of their post-operative residual tumors. One of these patients underwent fractionated stereotactic radiotherapy for tumor residual after the first surgery, but presented to our facility with tumor progress one year after radiotherapy.

A variety of surgical approaches was used to access these tumors. In the first 5 cases of this series only conventional approaches were used, namely frontotemporal or retrosigmoid approach, depending on the main bulk of the tumor being lying in the middle fossa or in the posterior fossa; respectively. In these 5 cases total resection was possible in one case, subtotal resection in another case, and in the remaining 3 cases only partial resection was carried out. In the last 9 procedures, cranio-orbitozygomatic (COZ) was used in one case, subtemporal approach with zygomatic osteotomy in one case, Kawase approach in 3 cases, translabyrinthine approach in one case and retrosigmoid approach with suprameatal extension in 3 cases. In these 9 cases, total resection was achieved in 6 cases Fig 1, and only subtotal resection was possible in the remaining 3 cases.

In this group of 14 patients with trigeminal schwannoma, total resection was achieved in 7 cases (50%).

Facial nerve schwannoma

There were 2 cases of facial nerve schwannoma. Both patients presented with facial nerve palsy (grade 3 and 5 on House Brackmann grading) and diminished hearing on the tumor side. One patient also exhibited lower cranial nerves dysfunction and ataxia. This patient underwent subtotal resection elsewhere and came to our facility with a large recurrence.

In both cases the tumor was located in the cerebellopontine angle and a retrosigmoid approach was used. Total resection was possible in one case, and only subtotal resection was possible in the other because the tumor was adherent to the anterior inferior cerebellar artery (AICA) and to the brain stem.

Jugular foramen schwannoma

Four patients with lower cranial nerves schwannoma were included in this series. Three cases presented with lower cranial nerves palsy in association with other pressure symptoms as diminished hearing and cerebellar manifestations. The 4th case presented only with diminished hearing and ataxia. All except one patient underwent complete tumor excision. In 2 cases the mass was purely cisternal and we used the retrosigmoid approach to access the tumor. In the remaining 2 cases, lesions extended into the area of the foramen magnum, and therefore we used the far-lateral approach Fig 2.

Hypoglossal schwannoma

A single case was surgically treated for hypoglossal schwannoma. The tumor was discovered during MRI examination for cervical myelopathy. Neurological examination revealed tongue atrophy. The tumor was mainly cisternal in location and was totally excised through a transcondylar approach Fig 3.

Overall results and complications

There was no post-operative mortality in this series. Total resection was achieved in 12 cases (57%). There was no tumor recurrence or re-growth during the follow-up period. Post operative complications included pneumonia in 3 cases, cerebrospinal fluid (CSF) leak in 2 cases, meningitis in 2 cases, hemiparesis in one case and hydrocephalus in one case, which was treated by a ventriculoperitoneal shunt. Post operative CSF leak was treated by lumbar drainage in one case, while the other case required surgical repair after he developed post operative pseudomeningocele.

Additional cranial nerves deficits were found in 8 cases, but they recovered in 6 cases. Only 2 patients had permanent post-operative new cranial nerve dysfunction (one case with trochlear nerve palsy and the other with abduscens nerve palsy).

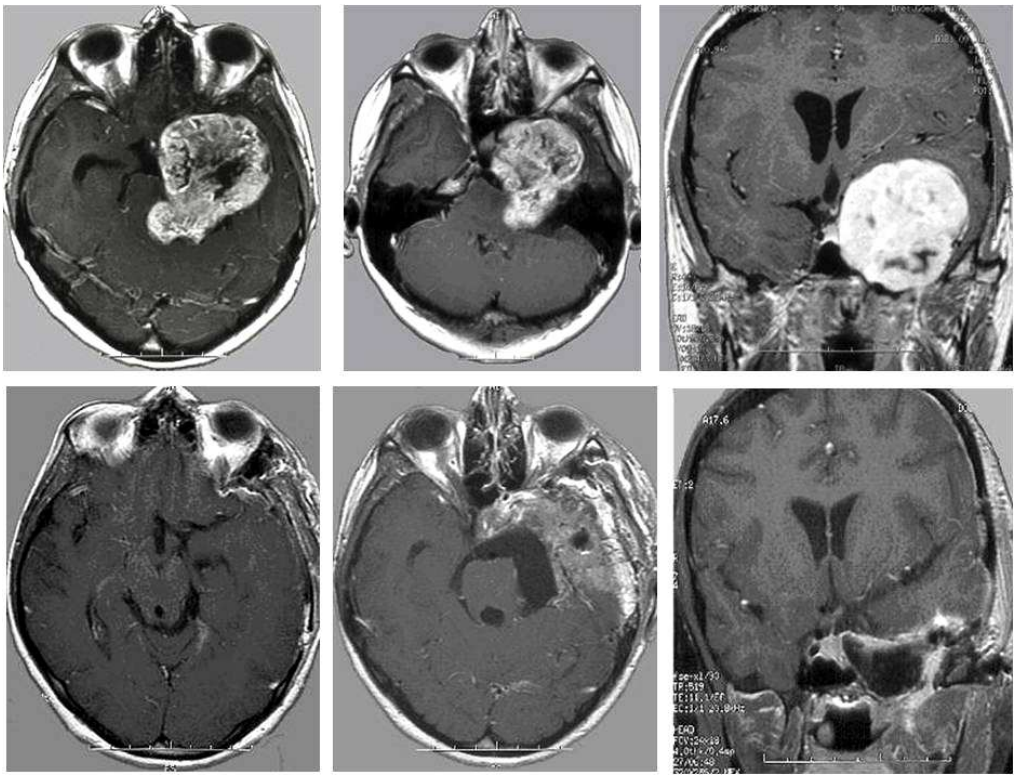

Fig 1: Pre and post operative MRI of a case of trigeminal schwannoma. Notice the total removal

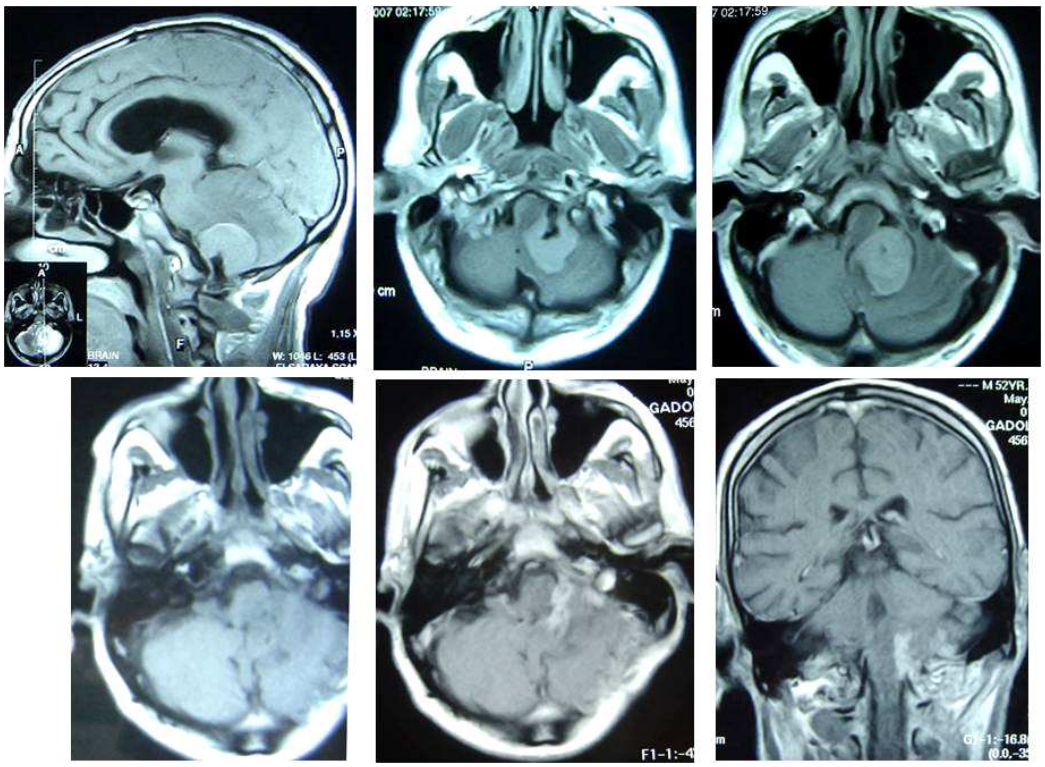

Fig 2: pre and postoperative MRI of a case of jugular foramen schwannoma. Notice the total renoval.

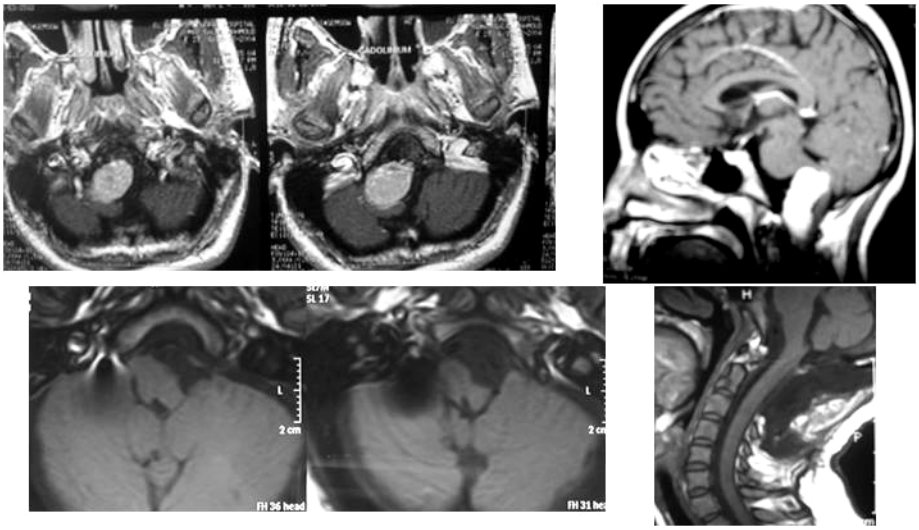

Fig 3: Pre and postoperative MRI of a case of hypoglossal schwannoma. Notice the complete tumor excision.

DISCUSSION

Non-vestibular schwannomas are uncommon brain tumors. They have characteristic features on MRI, which make them easily recognized pre-operatively; schwannomas are iso- or slightly hypointense on T_1-weighted images with significant enhancement after gadolinium injection. MRI offers additional advantages including the ability to obtain sagittal views, demonstration of extracranial extension, and visualization of different neural and vascular structures in relation to the tumor.[9,15,24,26] High resolution CT clearly demonstrates the location and extent of tumor growth as well as bony changes.[17, 26]

Trigeminal nerve schwannoma

Trigeminal nerve schwannomas are the most common type of non-vestibular schwannomas of the brain, representing 0.8 to 8% of all intracranial schwannomas. They tend to occur in the middle age without significant sex variation.[7,12,21,24] Because the early symptoms of trigeminal schwannoma can be mild, these tumors frequently reach large sizes before surgical intervention.[7, 12, 18]

For middle fossa tumors, a variety of approaches were used in this series: frontotemporal transsylvian, COZ and subtemporal with zygomatic osteotomy. Retrospectively, we think that the last approach is ideal for these tumors. It has the advantages of simplicity, minimal temporal lobe retraction and adequate extradural exposure of the tumor. This is similar to the recommendation of Goel and Muzumdar, Al-Mefty et al and Zhang et al.[1,6,30] We believe that Kawase approach is best suited for accessing middle fossa tumors with smaller posterior fossa extensions located above the level of the internal auditory canal.[29] It is usually easy to perform anterior petrosectomy in these cases, as the tumor usually erodes a considerable part of the petrous apex. Chang et al.[3] studied the extent of exposure of Kawase approach in an anatomical study, and concluded that it is not suitable for tumors extending below the level of the internal auditory canal.

The retrosigmoid approach affords excellent exposure of the tumor in the cerebellopontine angle.[6,21] However, a suprameatal extension to this approach increases the accessibility to the trigeminal porus and allows mobilizing the trigeminal nerve to improve the access toward the middle fossa.[3] A single case with complete preoperative hearing loss was operated through a translabyrinthine approach. In this case the tumor was large with extensions into both posterior and middle fossae.

The rate of total resection in this series was 50%. The major impediments to complete resection were inadequate exposure due to the use of conventional approaches and tumor adhesion to vital structures especially in recurrent cases. Guthikonda and colleagues [7] reported total resection in 9 out of 15 patients (60%) surgically treated for trigeminal schwannoma. Goel and Muzumdar[6] reported an impressive total resection rate of 70% in their large series of trigeminal schwannoma. Zhang and Colleagues[30] reported total resection in 16 out of 22 patients (72.7%) operated through skull base approaches, while in 12 cases operated using conventional approaches, total resection was achieved in only 4 cases (33.3%).

Facial nerve schwannoma

Facial nerve schwannoma is a rare tumor with less than 500 cases reported in the literature. The tumor may occur along any portion of the nerve and may involve several nerve segments. It may even arise from one of facial nerve branches as chorda tympani or nervus intermedius.[5,16,27] Sekhar.[24] classified these tumors according to their anatomic features into 3 types: cerebellopontine angle, geniculate and tympanomastoid tumors. In this series both cases

with facial nerve schwannoma had cerebellopontine angle tumors.

The clinical presentation is variable and depends on the tumor location.[5,16,29] Our cases presented with facial nerve palsy, diminished hearing, lower cranial nerves dysfunction and ataxia. It was not possible to distinguish the tumor from acoustic neuromas preoperatively.

The ideal treatment of facial nerve schwannoma is complete microsurgical resection with preservation of nerve continuity; however this is not possible in every case.[16,25] In this study, we were able to completely excise the tumor with preservation of nerve continuity in one case. In the second case, which had a previous surgical intervention, only subtotal resection was possible because the tumor was adherent to vital structures (Brain stem and AICA). On long term follow-up, both patients maintained their pre-operative facial nerve function.

Jugular foramen schwannoma

Four cases with schwannomas arising from cranial nerves IX, X and XI were included in this series. Jugular foramen schwannomas present with jugular foramen syndrome, other cranial nerves palsies and cerebellar dysfunction.[2,4,11,19,20,24] Schwannomas differ from meningiomas located within the jugular foramen because they compress rather than invade the neurovascular structures. Therefore they could be excised through simple approaches. In this series all 4 tumors had no extracranial extension. In 2 cases the tumor was mainly cisternal and was totally removed in both cases through a retrosigmoid approach. In the other 2 cases, the tumor extended to the foramen magnum region and we used the far lateral approach. This is in accordance with the recommendations of several authors.[4,11,14,20,22] Excellent total resection rates were reported in the literature ranging from 81 -100%.[2,4,11,14,22,23,24] In this series, total resection was achieved in 3 out of 4 cases. The small residual in this case did not progress during a follow up period of 30 months.

Hypoglossal nerve schwannoma

Hypoglossal schwannomas can be completely intracranial, intracranial/extracranial, or completely extracranial (28). The most common clinical presentation reported in the literature is hypoglossal nerve palsy and unilateral tongue atrophy followed by manifestation of cerebellar or brain stem compression. Involvement of other cranial nerves has been reported.[10,13,24] Intracranial tumors are best approached through a far lateral or a partial transcondylar approach which provide early identification of the vertebral artery and adequate exposure of the hypoglossal canal.[10,13,20,24]

Conclusion

Non-vestibular schwannomas are rare tumors that are best treated by total surgical resection. The location and size of the tumor dictates the surgical approach, however skull base approaches offer better tumor exposure and therefore result in better total tumor resection rate.

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