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MENINGIOMA OF THE POSTERIOR SKULL BASE

Abstract—Combined intratemporal and cerebellopontine angle meningiomas are relatively rare. There are unsolved problems with the stability of the skull and spine and the lower cranial nerves and there is a marked tendency for the tumor to involve the spinal cord. This article reports on five cases of combined intratemporal and cerebellopontine angle meningiomas.

Meningioma is a common tumor of the central nervous system, accounting for 13 to 18% of all intracranial tumors.¹⁻³ Meningioma confined to the temporal bone is rare.^{3,4} There is a further rare group of meningiomas that involve the temporal bone and adjacent skull base and extend into both the neck and spinal canal.

Five cases of combined intratemporal and intracranial meningiomas reported here were referred to the private practice of one of the authors (P.A.F.) between 1986 and 1989. The purpose of this article is to review the clinical features, growth patterns, and management of these tumors. Some conclusions regarding pathogenesis, behavior, and treatment are drawn.

CASE REPORTS

Case 1

A 53-year-old woman presented in 1974 with right-sided deafness, tinnitus, disturbance of balance, and an opaque tympanic membrane. Middle ear biopsy revealed meningioma.

In 1975 an extensive radical mastoidectomy was performed. The meningioma was attached to the jugular bulb,

but excision was judged to be complete. Follow-up skull x-ray and angiography were negative.

In September 1986 the patient complained of deterioration in her balance. She had a partial lesion of cranial nerves IX and X on the right, a complete lesion of XII on the right, and a positive Romberg's sign. Computerized axial tomography (CT) scan and magnetic resonance imaging (MRI) revealed a contrast-enhancing mass in the posterior fossa and right temporal bone with extension through the foramen magnum (Fig. 1A, B). Digital subtraction angiography (DSA) showed the right sigmoid sinus and internal jugular vein to be occluded.

A ventriculoperitoneal shunt was inserted in May 1987 and her balance improved. No other treatment was undertaken and there has been no change in tumor size over the ensuing 3 years.

Case 2

A 46-year-old woman presented in 1982 with a mass in the right middle ear. CT scan revealed a tumor involving the posterior and middle cranial fossae. Incomplete excision through a posterior fossa craniectomy was carried out. Histopathologic examination revealed a meningioma.

Six years later she developed pain in the right ear,

A

B

Figure 1. A: Case 1; this axial CT slice taken after intravenous contrast through the inferior portion of the posterior cranial fossa shows a large mass lying on the posterior aspect of the right petrous temporal bone, and extending anteromedially onto the clivus into the sphenoid sinus and the right middle fossa. B: Case 1; MRI scan in the coronal plane (TR = 600, TE = 20) giving T₂ weighting. Tumor mass is seen to the right of the brainstem and extending down through the foramen magnum.

hearing loss, and vertigo. CT scan revealed extensive destruction of the major part of the right petrous temporal bone and part of the occipital bone. A conservative course was adopted, with pain and vertigo being well controlled with medication.

One year later (April 1989) facial weakness and some difficulty in swallowing had developed. On examination, she had a partial right lower motor neuron facial palsy, complete right-sided hearing loss, and partial paralysis of cranial nerves IX to XII on the affected side. A further CT

and an MRI scan (Fig. 2A) revealed that the tumor had advanced considerably and there was extension through the foramen magnum. Such was her pain that an extensive combined skull base and neurosurgical procedure was undertaken. The extension through the foramen magnum into the spinal canal proved to be extradural and was cleared after removal of the articular facet of the occipital bone and a large part of the lateral mass of the atlas. The facial nerve was resected along with the tumor and an end-to-end anastomosis performed. The patient developed a

Figure 2. A: Case 2; CT slice through the foramen magnum after intravenous contrast, taken 1 year after the patient first presented. There is considerable further bone destruction at the petrous apex. The rim of the foramen magnum is eroded and distorted. B: Case 2; postoperative CT at the level of the foramen magnum shows a large operative defect filled with fat. There is no evidence of residual tumor.

cerebrospinal fluid (CSF) leak and meningitis in the post-operative period. There were also severe swallowing problems and prolonged bouts of vomiting with aspiration, exacerbated by the need to maintain traction in a horizontal position. An occipitocervical fusion was carried out some 4 weeks later. Following this, the patient was mobilized and began to cope well with a pureed diet. A postoperative CT scan revealed good tumor clearance (Fig. 2B) and the patient remains tumor free.

Case 3

A 73-year-old woman presented in February 1989 with left-sided pulsatile tinnitus. A mass was noted in the left middle ear. High resolution CT scan (Fig. 3A, B) revealed a strongly enhancing mass based on the posterior surface of the left petrous temporal bone, extending into the upper portion of the left jugular foramen, the margin of which was intact. The adjacent petrous bone showed a change in the trabecular pattern and appearances suggested tumor infiltration.

Because the patient's symptoms were minimal and she was 73 years old, no treatment was undertaken. The mass was not biopsied. In October 1989 repeat CT scan showed no increase in tumor size.

Case 4

A 49-year-old woman presented in 1986 to another institution with a conductive hearing loss. A mass was

noted in the right middle ear and a biopsy revealed meningioma. A CT scan at that time showed an extensive lesion in the lower half of the petrous temporal bone involving the jugular foramen. There was an extension into the posterior fossa with extradural spread through the foramen magnum to the arch of the axis (Fig. 4A, B). She was neurologically normal. Treatment with radiotherapy was undertaken. She presented to one of the author's practice (P.A.F.) in 1989. CT showed that the lesion was unchanged in size. As she remained entirely well, an expectant policy was adopted.

Case 5

A 37-year-old woman presented in 1985 to another institution with hearing loss and unsteadiness of gait. A 5 cm cerebellopontine angle mass with temporal bone invasion and extension through the jugular foramen was detected on CT scan. The posterior fossa portion of the tumor was removed with preservation of facial function. The IX, X, and XIth cranial nerves were intimately involved in tumor and were lost. The intratemporal portion of the tumor was removed during a prolonged second procedure in February 1986, carried out according to the method of Fisch.⁵ Removal was judged to be complete and CT examination some 2 years later showed no evidence of recurrence. However, in July 1989 a recurrence was detected in the posterior fossa with extension through the jugular foramen and into the upper spinal canal (Figs. 5A, B). The patient was entirely well and no further treatment was carried out.

Figure 3. A: Case 3; axial CT slice taken through the posterior fossa after intravenous contrast. There is a sessile tumor lying along the posterior border of the left petrous temporal bone. Flecks of calcium can be seen within it. B: Case 3; axial CT slice through the jugular foramina shows a change in the trabecular pattern within the left petrous apex, indicating tumor infiltration.

CHOICE OF SURGICAL APPROACH

When a posterior skull base meningioma has a significant posterior cranial fossa extension, removal of the latter will generally take precedence, particularly if there

is any brainstem shift or raised intracranial pressure (case 5). This is in contradistinction to the routine generally adopted for large glomus tumors in which removal of the neck and skull base portion results in significant decrease in vascularity in the posterior cranial fossa segment, making its subsequent removal a good deal easier.

Figure 4. A: Case 4; an axial CT slice taken just above the level of the foramen magnum after intravenous contrast, shows extensive tumor in the posterior fossa lying along the medial aspect of the right petrous and mastoid temporal bones, extending forward along the clivus and eroding the posteromedial aspect of the right jugular foramen. B: Case 4; an axial CT slice taken after intravenous contrast through the body of axis, shows tumor in the spinal canal lying anterolaterally on the right. Appearances suggest that the tumor is extradural.

Figure 5. A: Case 5; axial CT slice taken in 1989 through the posterior fossa after intravenous contrast shows a large operative defect in the left occipital plate and petrous temporal bone. There is recurrent tumor along the posterior aspect of the residual petrous temporal bone and in the sphenoid sinus. B: Axial CT slice taken from the same study as Figure 5A shows tumor extending through the foramen magnum on the left.

Tumor in the temporal bone itself, and that extending into the neck through the jugular foramen, is best removed by a standard posterior skull base approach, type A of Fisch.⁵ This approach, carried inferiorly and posteriorly, gives access to the lateral mass of the atlas, which may need to be removed, if there is a significant extension

through the foramen magnum, as there was in case 2. The vertebral artery, lying over the tumor mass, has to be identified as the lateral mass of the atlas is drilled away. This procedure is likely to result in marked instability of the neck, especially if the articular process of the occipital bone has been destroyed by tumor.

Craniocervical fusion was carried out subsequent to tumor removal in case 2 because it was considered that fusion prior to the definitive surgery would have made surgical access impossible.

DISCUSSION

The anatomic classification of Rietz et al.³ is used in this article. Meningiomas of the posterior skull base can be divided into intracranial, combined intracranial and intratemporal, and intratemporal.³ Meningioma confined to the cranial cavity is a well-defined entity,^{1,6-8} which is not discussed further here. Meningiomas confined to the temporal bone are rare. Many case reports were published before high-resolution CT was available, and it may well be that many of these were combined intracranial and intratemporal tumors. When these tumors are shown to be truly intratemporal, they can be treated in most instances by standard otologic techniques.

The first extensive review of meningioma involving the temporal bone and the cerebellopontine angle was published by Nager⁹ in 1964, with some 37 cases. Subsequently, there have been similar reports,^{2-4,10-14} but aggressive tumor spread through the foramen magnum to involve the spinal cord is not stressed. We were only able to find three cases where there was extension through the foramen magnum.^{12,13}

It is now well accepted that meningioma arises from arachnoidal cells. Intratemporal meningioma probably arises from arachnoid cells within the internal auditory canal, at the geniculate ganglion, within the jugular fossa, along the groove for the greater superficial petrosal nerve, and in small brain herniations through the tegmen tympani.^{11,15} These may then spread to the cranial cavity.

On the other hand, posterior fossa tumors may spread to the temporal bone through the posterior petrous face, the internal auditory canal, and the jugular fossa.⁹ The most frequent route of extracranial extension is through the jugular foramen and foramen lacerum into the parapharyngeal space.⁹

It is difficult to determine the site of origin of the five cases discussed here. Each showed extensive disease of the petrous bone and adjacent skull base, with spread into the neck and through the foramen magnum in four.

The development of high-resolution CT scanning has made it possible to assess the extent of temporal bone tumors and to monitor progress with great accuracy. Characteristic changes are seen in Figure 1B, with destruction of the jugular foramen and extension into the neck, and in Figure 2A, with involvement of the intrapetrous carotid and the petrous apex.

Extension through the foramen magnum, which proved to be extradural in the one operated case (case 2), is well shown by both CT (Figs. 4B, 5B) and MRI (Fig. 1B).

The exact extent of spinal canal spread is difficult to determine on CT scanning with intravenous contrast be-

cause of the presence of the vertebral plexus of veins, which enhance strongly. This enhancement may be indistinguishable from tumor enhancement. For this reason, we advise correlation with MRI scanning, which will differentiate tumor from venous plexus.

Therapy in these huge tumors presents a dilemma. Surgery is the accepted form of treatment for meningioma.¹⁶ However, in these tumors there is an acknowledged high recurrence rate.¹⁷ Furthermore, complete surgical resection via a combined skull base and neurosurgical procedure, although feasible, may produce paralysis, either temporary or permanent, of cranial nerves VII to XII on the ipsilateral side. This combination of cranial nerve deficits can produce major problems with aspiration, swallowing, nasal regurgitation, speech production, and shoulder movement. These problems, as well as the potential for CSF leak, are compounded when complete excision involves removal of the occipital condyle, thereby rendering the head and neck unstable (case 2). In this situation, the patient cannot sit up, and cervical traction is required to stabilize the neck before occipito-cervical fusion can be carried out. When neck stabilization is required, we recommend using the Ransford loop in conjunction with bone grafting to fuse the occiput to C1 and C2.¹⁸ It is therefore essential during excision of the intraspinal portion of the tumor to leave the posterior arch of C1 and C2 intact, approaching the tumor via the lateral mass of the atlas and axis.

The role of radiotherapy remains controversial. Meningioma was originally thought to be relatively radioresistant;^{3,9} however, recent studies have suggested a significant role for radiation therapy in unresectable, partially resected, or recurrent meningioma.^{16,19,20} We do not recommend primary or preoperative radiotherapy because subsequent surgery, a major undertaking, is made more difficult.

Finally, some tumors of this type exhibit very aggressive behavior with gross bone destruction and invasion of the spinal canal over a 12-month period (case 2). Others, (cases 1 and 4) have remained dormant for a long period. Predicting tumor behavior is impossible. There is controversy as to whether the histology is of any prognostic significance. Cushing and Eisenhardt¹ expressed the opinion that fibroblastic meningiomas are the most aggressive. Crompton and Gautier-Smith²¹ found the highest recurrence rate in the syncytial variety. Rubenstein²² and others identify the angioblastic lesion as the most aggressive. An increased incidence of mitosis and nuclear pleomorphism should raise a suspicion of the increased risk of recurrence. The four cases in this article in which a histologic diagnosis was made have been carefully analyzed for unusual pleomorphism and increased number of cells showing mitoses. No such evidence of increased aggressiveness or increased risk of recurrence was found. Despite this, two cases (cases 2 and 5) showed clinical and radiologic evidence of aggressive behavior over a relatively short period.

We therefore advocate regular clinical review with high-resolution CT or MRI scanning, or both, in those patients treated conservatively, and long-term follow-up in those patients treated surgically.

CONCLUSIONS

1. Meningioma visible in the ear is generally only the tip of the iceberg. High-resolution CT and MRI will demonstrate the true extent of the tumor.
2. Tumor growth is unpredictable but can in some cases be very rapid. Tumor may spread down the posterior face of the petrous temporal bone through the foramen magnum into the spinal canal.
3. Surgery carries considerable risk of morbidity, particularly from injury to cranial nerves VII to XII, and tumor recurrence rates may be high in the long term. Radiotherapy may be a useful postoperative adjunct.
4. We recommend aggressive surgical resection, by a combined skull base and neurosurgical procedure when significant lower cranial nerve morbidity is already present and the patient is young and fit; the tumor is clearly exhibiting aggressive behavior; and there is a large posterior fossa mass and the patient is sufficiently fit to withstand the surgery.

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REVIEWER'S COMMENTS

This article illustrates the fact that therapy needs to be individualized, based on risk factors related to the age and general condition of the patient, the site of the tumor, and the neural structures involved. Radiation therapy is a reasonable consideration if the tumor continues to progress after total excision has been attempted.

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