Cerebellopontine angle meningioma

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The meningioma of the cerebellopontine angle may arise from any area of the dura on the posterior surface of the petrous bone (see Posterior petrous bone meningioma), or originate at the level of the internal auditory canal (IAC), and may be mistaken for vestibular schwannoma.

Epidemiology

Among intracranial meningiomas, 5%-10% are located in the cerebellopontine angle (CPA) \(^1\) \(^2\), thus comprising the second most common tumors in this anatomic region.

They usually occur in patients in the fifth decade of life, with a female predominance. Pathologically, 80%-90% of these tumors are benign (WHO Grade I) and the majority are the meningothelial subtype.

Consistent with these observations, in 193 patients, the mean age was 50.2 years and most patients were female (n = 154). The majority (94.8%, 183/193) of the tumors were grade I and the most common pathological subtype was meningothelial (31.6%, 61/193) \(^3\).

Categories

According to some authors, CPA meningiomas are mostly posterior petrous bone meningiomas arising from the petroclival region \(^4\).

Posterior petrous bone meningiomas were classified into 3 types following the Desgeorges classification, \(^5\) which is based on the tumor's dural attachment to the posterior petrous bone surface:

Type A (anterior) meningiomas originate from the petrous apex

Type M (middle) meningiomas originate at the level of the internal auditory meatus

Type P (posterior) meningiomas develop from the posterior part of the petrous bone, between the posterior wall of the IAC and the groove of the sigmoid sinus.

CPA meningiomas were classified into three groups, according to the relation of the tumor with respect to the IAC and labyrinth: meningiomas located anterior to the IAC, centered at the IAC, and located posterior to the IAC \(^6\).

Meningiomas are typically classified based on their alteration of the IAC \(^7\) \(^8\).

Their site of dural origin and their relationship to surrounding neurovascular structures of the CPA is variable.

At operation four general categories of tumor are found, depending on where they arise and their relationship to the seventh and eighth nerve complex:

Anterior to the internal auditory meatus, displacing the seventh and eighth nerves posteriorly and inferiorly
Between the internal auditory meatus and the jugular foramen, displacing the seventh and eighth nerves superiorly.

Superior to the internal auditory meatus, displacing the seventh and eighth nerves anteriorly in the large tumors.

Surrounding the internal auditory meatus, with the seventh and eighth nerves engulfed in the tumor.

Although enlargement and involvement of the internal auditory canal (IAC) is a common sign of schwannomas of the CPA these features can also be seen in meningioma.

Meningiomal involvement of the IAC occurs in two forms: meningioma truly originating in the IAC and meningioma extending into the IAC from an adjacent location. While the first form is very rare, the second is relatively common.

Tumors exhibiting partial IAC intrusion are being largely ignored. These latter tumors were included in the study of Gao et al., which is also the first to explore the extent of surgical resection of CPA meningiomas with respect to IAC involvement.

**Clinical Features**

The clinical presentation and outcome after surgical resection are different because of the diversity of this tumor entity.

The most common symptoms at the time of the initial evaluation were from the eighth cranial nerve (unilateral hearing loss–24 patients, vertigo or imbalance–19 patients, tinnitus–11 patients), and the fifth cranial nerve (altered sensation–9 patients, facial pain–5 patients). On examination, the most common findings were absent caloric response (19 patients), nystagmus (16 patients), diminished facial sensation (14 patients), ataxia (13 patients), reduced hearing (9 patients), and facial weakness (9 patients). There was often a long interval from the onset of symptoms to the correct diagnosis of a tumor.

**Diagnosis**

The MRI scan usually defines those tumors that arise posterior to the internal auditory meatus but will not distinguish the first three categories. The diagnosis of meningioma is indicated by the flat surface of the tumor against the petrous bone and the durat “tail” extending from the tumor.
In the past it was often utilized angiography when a cerebellopontine angle meningioma was suspected. However, for most of these meningiomas it is now not necessary, because the MRI usually gives all the information needed and in most patients the blood supply comes primarily through the dural attachment.

**Differential diagnosis**

Findings in 20 patients who underwent removal of a meningioma were compared to those in 131 patients who had an *vestibular schwannoma* removed during the same period. They found that in patients with meningiomas the tumors frequently are large at presentation, the otologic symptoms and audiometric findings are less dramatic.

In meningioma the characteristic finding is a broad-based mass aligned with the *petrous ridge*, not
centered over the internal auditory canal [11].

Dynamic contrast-enhanced perfusion MRI can potentially differentiate VS and CPA meningiomas. Meningiomas are reported to have very high mean regional cerebral blood volume ratios that are statistically different than schwannomas [12].

Perfusion imaging has also been shown to differentiate between other types of enhancing CPA masses, including lymphoma or abscess vs. metastasis and hemangioblastoma vs. lymphoma.

Proton MR spectroscopy is another advanced MRI option. Specific signs of CPA meningiomas include a combination of elevated glutamate/glutamine and a characteristic presence of alanine at 1.5 ppm [13].

A myo-inositol peak in schwannomas at 3.55 ppm has also been demonstrated [14].

**Treatment**

Embolization has not been a consideration.

In patients with mild or minimal symptoms, an initial period of clinical evaluation and repeat scans may be indicated to determine whether there are progressive symptoms and an enlarging tumor. This is especially true in the elderly.

The indications for operation are a worsening neurological deficit due to brainstem compression or cranial nerve compression. In a few patients headache or the continued presence of a stable deficit such as diplopia or hearing loss may be the indication. In the series of the MGH Harvard two patients were asymptomatic. One had documented enlargement on follow-up scans and the other was concerned about the presence of the tumor. Radiation therapy has been used when there is regrowth after subtotal or radical subtotal removal and with small tumors which start to enlarge in older patients who are being observed [15].

see Retrosigmoid approach for cerebellopontine angle meningioma.

see Fully endoscopic resection of cerebellopontine angle meningioma.

**Outcome**

The location of a CPA meningioma affects clinical outcome.

Patients with CPA meningiomas with IAC involvement often present with hearing loss and abnormal facial motor function. Resection of these tumors is compatible with hearing restoration.

Nonetheless, the anatomical location and intimate relationship between meningiomas involving the IAC and the cranial nerves are such that skilled surgical management is needed. Despite advances in neurosurgery, the surgical procedure for CPA meningiomas involving the IAC remains challenging.

**Case series**

2015

The pre- and postoperative MR images of 193 consecutive patients with pathologically diagnosed meningioma centered around the IAC were analyzed, focusing on changes in the IAC, maximal axial tumor volume, peritumoral brain edema, and postoperative residual tumor. Patient age, sex, tumor
volume, postoperative residual tumor, and pathological subtype were compared in patients with and without IAC involvement by the tumor and among the different types of IAC involvement. The results showed that the 71 patients (36.8%) with IAC involvement had a higher ratio of peritumoral edema ($\chi^2=5.922$, $P=0.015$), postoperative residual tumor ($\chi^2=22.183$, $P<0.001$), and a predominance of the meningothelial subtype ($\chi^2=5.89$, $P=0.015$). Peritumoral edema was a risk factor for IAC involvement ($P=0.016$, OR=2.186). Radiologically, IAC involvement could be distinguished as intruding (31%, 22/71), filled (29.6%, 21/71), and dilated (39.4%, 28/71). Patients with intruding IAC were significantly older (54.5±9.54 years, $P=0.021$) and had the lowest postoperative residual tumor values (42%, $\chi^2=7.865$, $P=0.005$), while those with filled IAC were more likely to be female (95%, $\chi^2=9.404$, $P=0.009$). The observations provide the basis for a morphological classification of IAC involvement by CPA meningiomas and further insight into the clinical features of these tumors [16].

2005

Among 421 patients, the charts of 347 patients with complete clinical data, including the history and audiograms, imaging studies, surgical records, discharge letters, histological records, and follow-up records, were reviewed retrospectively. Data about preoperative and postoperative facial nerve function were available in 334 patients, and audiometric analysis was conducted in 333 patients. Patients with neurofibromatosis Type 2 were excluded from the study.

There were 270 women and 77 men, with a mean age of 53.4 years (range, 17.6-84 yr). Among these patients, 32.9% of the tumors originated at the petrous ridge anterior to the inner auditory canal (IAC) (Group 1), 22.2% showed involvement of the IAC (Group 2), 20.2% were located superior to the IAC (Group 3), 11.8% were inferior to the IAC (Group 4), and 12.9% were posterior to the IAC, originating between the IAC and the sigmoid sinus (Group 5). Patients presented with disturbance of Cranial Nerves V-VIII, the lower cranial nerves, and ataxia, depending on the main tumor location. Tumor resection was performed through a suboccipital-retrosigmoidal approach in the semisitting position in 95% of the patients. A combined supratentorial-infratentorial presigmoidal approach was performed in 5%. Total tumor removal (Simpson Grade 1 and 2) was achieved in 85.9% and subtotal removal in 14.1%. The best initial postoperative facial and auditory nerve function was observed in tumors belonging to Groups 3 and 5. Recovery from preoperative deafness was observed in 1.8% of patients. On long-term follow-up, good facial nerve function (House-Brackmann Grade 1 or 2) was observed in 88.9% of patients. Hearing preservation among patients with preoperative functional hearing was documented in 90.8% on long-term follow-up.

Although the outcome of facial and cochlear nerve function is different in CPA meningiomas, depending on the topographic classification of these tumors, preservation of the cochlear nerve is possible in every tumor group and should be attempted in every patient with CPA meningioma. It has to be kept in mind that recovery of hearing was also observed in patients with preoperative profound hearing deficits [17].

There were 57 patients with cerebellopontine angle meningiomas, 37 women and 20 men, ranging in age from 38 to 89 years, with 10 over 70 years of age (Table 17.16). In the 42 patients listed in Table 17.16, Anterior, the tumor arose anterior to the internal auditory meatus in 27 and anterior-inferior in 10 and grew diffusely in five. The extent of the tumor removal is recorded in Table 17.16. In only 14 patients could I be sure of a total removal but in another 10 a radical subtotal removal was done. On follow-up, 34 of the 42 patients had a good result, three had a fair result because of postoperative disability, four had poor results because of severe preoperative neurological disabilities that did not significantly improve, and there was one postoperative death. This occurred in a 89-year-old man who had been well until he started to develop worsening ataxia. CT showed a 4-cm tumor with
hydrocephalus. A shunt was placed, with improvement, but several months later symptoms of brainstem compression worsened. A subtotal removal was done but the patient died of a cardiopulmonary complication.

None of the patients with radical subtotal removal has shown recurrence and all have been followed by scans. Most of the patients with subtotal removal had large tumors (>3 cm). Three were growing anteriorly into the middle fossa. After subtotal removal, 15 of 18 had follow-up scans, which showed no growth in eight, slight growth in five (three of whom have been given radiation therapy and two of whom are being observed after showing no further growth on subsequent scans), and moderate growth in two of the disabled patients, where nothing further has been done.

Postoperative complications included permanent increased ataxia in three, one of whom had to have a cerebellar infarction removed, one patient with wound infection and meningitis, and one with a cerebrospinal fluid leak requiring repair. Several patients had temporary increases in ataxia, incoordination, or swallowing problems which improved. Four patients had a shunt for hydrocephalus at some time in their course.

Of the 15 patients with meningiomas arising posterior to the internal auditory meatus, 13 had a total removal (Table 17.16). One had a radical subtotal and one a subtotal removal because of involvement of the lower cranial nerves. All had a good result. There were no postoperative neurological complications and no recurrences.

Yasargil et al. reported that 27 of 30 patients had a good result and in 27 the tumor was “radically excised.”

Sekhar and Jannetta reported total removal in 14 of 22 patients, with no operative mortality and a good outcome in 16.

Samii and Ammirati reported total removal of all 24 tumors located posterior to the internal auditory meatus, with a good outcome for 22 patients. Of 32 patients with tumors anterior to the internal auditory meatus, 29 had the tumors totally removed and 28 had a good outcome.

2000

Retrospectively review in 40 patients the common clinical presentations were hearing loss, unsteadiness, and dysequilibrium. Findings upon physical examination included hearing loss (73%), cerebellar signs (32%), trigeminal neuropathy (16%), and facial nerve dysfunction (16%). The most common site of dural origin was the petrous ridge (anterior to the IAC [26%], posterior [21%], superior [18%], and inferior [16%]). Less common sites of dural origin included the tentorium (31%), the clivus (15%), the IAC (10%), and the jugular foramen (8%). Site of dural origin determined the direction of displacement of the facial/vestibulocochlear nerve bundle. The most common microsurgical complication was facial nerve dysfunction (30%). Gross total resection was achieved in 82% of cases, whereas 18% underwent subtotal resection. Two patients died. Follow-up ranged from three months to 13 years with three recurrences.

1985

20 patients who underwent removal of a meningioma were compared to those in 131 patients who had an acoustic neurinoma removed during the same period. They found that in patients with meningiomas the tumors frequently are large at presentation, the otologic symptoms and audiometric findings are less dramatic, and roentgenograms of the skull and tomograms of the petrous apex
rarely show erosion of the internal auditory canal. Computerized tomography is the most useful method for differentiating a meningioma from a neurinoma: when a meningioma is present the characteristic finding is a broad-based mass aligned with the petrous ridge, not centered over the internal auditory canal \[^{19}\].

32 patients with surgically confirmed CPA meningiomas are analyzed. The most common symptoms at the time of the initial evaluation were from the eighth cranial nerve (unilateral hearing loss–24 patients, vertigo or imbalance–19 patients, tinnitus–11 patients), and the fifth cranial nerve (altered sensation–9 patients, facial pain–5 patients). On examination, the most common findings were absent caloric response (19 patients), nystagmus (16 patients), diminished facial sensation (14 patients), ataxia (13 patients), reduced hearing (9 patients), and facial weakness (9 patients). There was often a long interval from the onset of symptoms to the correct diagnosis of a tumor. Brain stem auditory evoked potentials, blink reflex testing, posterior fossa myelography, computerized tomographic scanning, and angiography were abnormal in all patients in whom the test was done, but all tests were not performed on all patients. Computerized tomographic scanning and angiography are important for definitive diagnosis and for planning surgical treatment. The histopathology of the temporal bone was studied in three patients with meningiomas in the region of the internal auditory meatus \[^{20}\].

## Case reports

Entezami et al., describe the case of a 49-year-old male with a recurrent right CPA meningioma arising from the petrous bone that was previously treated with a subtotal resection and postsurgical radiosurgery, presenting with acute left-sided hemiparesis secondary to intratumoral hemorrhage. Although surgical evacuation and decompression was recommended, the patient declined operative intervention and was managed medically.

Meningiomas can cause subarachnoid, intraparenchymal, and rarely intratumoral hemorrhage. Symptomatic hemorrhage can worsen the prognosis, with increased morbidity and mortality. Several etiologies have been proposed for this phenomenon, including rupture of aberrant vasculature, intratumoral necrosis, and tearing of stretched bringing veins.

Only two prior cases of CPA meningioma have been reported in the literature. Recognition of CPA meningioma hemorrhage as a clinical entity can help in future diagnoses and management \[^{21}\].

## References


