

**TITLE: Cerebellopontine Angle Masses**

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## **Introduction**

A complete discussion of masses involving the cerebellopontine angle is beyond the scope of this paper. Therefore, this summary will focus on masses which arise in or are centered within the anatomic confines of the cerebellopontine angle (CPA), as well as address their radiologic diagnosis and options for treatment. Masses arising outside but extending into the CPA will be listed for completeness; but an appropriate discussion of these is not possible in this short presentation.

The cerebellopontine angle is a rather small area located in the posterior fossa near the origins of several vital cranial nerves. The medial and lateral borders of this space are the brainstem and the petrous portion of the temporal bone, respectively. Superiorly, it is bounded by the middle cerebellar peduncle, and inferiorly by the arachnoid tissue of the lower cranial nerves. Posteriorly, the cerebellar tonsil limits this space, as does the clivus anteriorly. Within this space lie several cranial nerves, including portions of VII – XII, as well as the CSF of the quadrimenial cistern, the arachnoid tissue of the above cranial nerves, and several blood vessels, most notably the anterior inferior cerebellar artery (AICA).

## **Symptoms**

The symptoms of CPA masses arise from compression of local structures within or at the boundaries of the region. A patient may present with symptoms from ataxia and visual loss due to brainstem compression and hydrocephalus, to a mild decrease in hearing noted on a routine audiogram. An easy way to remember the symptoms is to proceed logically through the structures of the CPA. Many of the details below are given for acoustic neuroma, which is by far the most common CPA mass.

### **Cochlear**

Compression of the fibers of the cochlear nerve may result in sensory hearing loss. This may be the most common presenting symptom, present in up to 85% (of acoustic neuromas).

Usually this loss is gradual and noted as a progressive unilateral hearing loss. However, in up to 26% of cases (of AN), this hearing loss may be sudden. This loss is believed to be a result of compression of the labyrinthine artery. One must remember though, that although 26% of tumors may present with sudden loss, only 1-2.5% of those who present with a sudden loss will be found to have a CPA mass causing their symptoms. It may also be noted that steroid therapy, which is indicated for sudden SNHL may result in resolution of hearing loss even if a CPA mass is present. When the hearing loss is gradual, the rate of progression has not been found to correlate with size or specific location of the tumor. Tinnitus may be the second most common symptom (56%). This may be secondary to the sensory hearing loss. Other audiologic disorders which may be found include a reduced understanding of audible sounds (discrimination), and increasing difficulty in understanding sounds when presented at higher intensities (rollover).

### **Vestibular**

Compression of the vestibular nerve may result in symptoms of disequilibrium or vertigo. If the tumor is slowly enlarging, these symptoms may be mild due to compensation. It should be noted that at the time of diagnosis, the most common vestibular symptom from a CPA mass is disequilibrium, not vertigo.

### **Facial**

Compression of the facial nerve may result in hemifacial weakness or unilateral facial paralysis. Spasm of the facial nerve with a new onset of facial twitching has also been noted. Hitselberger's sign, a decrease in sensation of the external auditory canal, is due to compression of the sensory component of the facial nerve. Although acoustic neuroma is the most common CPA mass, it is uncommon for AN to present with facial weakness or paralysis, and this symptom should alert to the possibility of another diagnosis.

### **Other cranial nerves**

Involvement of the trigeminal nerve may result in either unilateral hypo- or hyperesthesia, as well as trigeminal neuralgia. Involvement of the abducens nerve, while rare, may cause diplopia on lateral gaze away from the side of the lesion. Compression of the glossopharyngeal nerve can result in dysphagia, while vagal compression can result in hoarseness and aspiration, and spinal accessory compression can result in shoulder weakness.

### **Axial structures**

When tumors grow large enough to compress the cerebellum, a wide based gait may result. The patient also tends to fall towards the side of the lesion. Brainstem impingement may progress to symptoms of hydrocephalus including visual loss and headaches.

### **Differential**

Masses involving the CPA may be thought of as primary or secondary. Primary masses arise within the CPA and have a somewhat limited differential. Secondary masses are those that may arise from outside the anatomic confines of the CPA but extend into the area as they grow or metastasize. The differential for these is rather extensive, and is listed later.

For lesions arising within the CPA, the differential is comprised of three main tumors: acoustic neuroma, meningioma, and epidermoids. These make up approximately 95% of primary CPA lesions. Acoustic neuroma alone has been shown to account for 60-92% of CPA lesions. Many studies which show a relatively low percentage of AN (60%) also include petrous bone masses and paragangliomas in their cohort. Therefore, it is likely that when evaluating primary CPA masses, AN is by far the most common. Other masses which account for a small minority of the remaining lesions include arachnoid cysts, non-vestibular schwannomas, vascular anomalies, lipomas, melanoma, and dermoids.

## **Acoustic Neuroma**

Acoustic neuroma or vestibular schwannoma is the most common mass of the cerebellopontine angle. It typically presents in the middle to late decades of life, and is usually unilateral. Bilateral disease or disease early in life should lead to suspicion of neurofibromatosis, a diagnosis which has implications on the treatment of choice for these lesions. Acoustic neuromas are benign masses. These masses tend to grow slowly over several years, with growth rates commonly from 0.2 – 2 mm per year. However, some tumors have been found to progress at rates up to 10 mm per year. These lesions may also remain stable for multiple years with no signs of growth on long-term follow-up. The masses themselves occur with equal frequency on the superior and inferior vestibular nerves. Schwann cells are more numerous in the region of Scarpa's ganglion, and as a result, more tumors are found near this location. This is also why AN, as opposed to several other CPA masses, almost always has an intracanalicular component. While the vestibular portion of the vestibulocochlear nerve commonly is involved, the cochlear portion is rarely involved with tumor. However, when the schwannoma arises from this nerve, the lesion may extend into the cochlea, due to the glial-schwann junction occurring distally at the modiolus. Pathologically, acoustic neuromas are composed of two types of tissue, called Antoni A and Antoni B. The A type is comprised of compact tissue with spindle cells arranged in palisades. The B type is loose tissue with cystic components. Type A tissue is much more common, and likely results in the uniform, smooth enhancement of the lesion on MRI.

The radiologic features of acoustic neuroma have been well documented. The anatomic location of the tumor is usually centered about the porus acousticus. The tumors usually have an intracanalicular component, and may extend into the CPA. Other lesions may arise completely within the CPA, and have no extension into the IAC. These lesions may grow to a larger size before diagnosis due to their lack of compression of the nerves within the IAC. Smaller lesions are becoming more common as new techniques for imaging are being found. The range of size at diagnosis therefore varies from mere millimeters up to 6-8 centimeters. As the tumors grow they generally enlarge the porus of the IAC. Although the diameter of the IAC varies greatly from individual to individual, the diameter of the contralateral IAC has been found to be nearly equal in normal patients. Therefore, an enlargement of the porus greater than 2 mm in comparison to the contralateral side is a relatively good predictor of AN. Since the lesion commonly arises near the area of the porus and enlarges in a spherical fashion, it is considered a concentric mass. Therefore, as the tumor enlarges, it will have acute angles to the remainder of the petrous portion of the temporal bone. Masses such as meningiomas, which are eccentric and tend to spread along the petrous area will have obtuse angles. Other features that may favor AN in the differential are: lesions rarely extend anteriorly and superiorly, they almost never penetrate into the middle cranial fossa, and they lack prominent vasculature, as well as dural tails.

CT scans of acoustic schwannomas tend to show the above features: a porous-centered mass, acute angles, IAC involvement. They also demonstrate the homogeneous nature of the mass. The majority of lesions, excluding previously treated tumors and very large tumors, will show uniform density on CT. Calcifications and central necrosis are rare, however, central clearing has been noted in some larger lesions. The density of AN on CT is similar to that of nearby brainstem, and more dense than surrounding CSF. If given IV contrast, the tumor will most likely show homogeneous uptake and turn very bright. Again, non-homogeneous uptake may be seen with previously treated lesions and large tumors.

MRI is the study of choice if the diagnosis of AN is in question. The T1 weighted exam with Gadolinium contrast has been shown not only to be 100% sensitive for the diagnosis of AN, but also to have the highest negative predictive value for the lesion as well. On standard T1 images, the tumor should be relatively isointense to pons but more intense than CSF. On T2 images, the lesion should be mildly brighter than pons, but darker than CSF. After Gadolinium, the T1 sequence should show a very intense lesion, brighter than all other surrounding structures.

## **Meningioma**

Meningioma is the second most common diagnosis of a primary CPA lesion. These lesions arise from the cap cells of arachnoid villi which are in high concentration near the foramen of cranial nerves and dural venous sinuses. The CPA contains a large number of cranial nerves (V-XII), and is also bonded near its anterior superior aspect by the superior petrosal sinus. Any of these may give rise to a CPA meningioma. Whereas AN is strictly a primary CPA mass, meningioma can arise within the CPA, extend from the middle fossa to the CPA, or extend from the CPA into the middle fossa. Most tumors of this area though, arise from the posterior portion of the petrous bone, and extend along its medial boundary. These lesions uncommonly enter the IAC, and thus, may be large at the time of diagnosis. As opposed to AN, the meningioma is a vascular tumor, and may show flow voids of the pial blood vessels. Meningiomas are also not homogeneous masses, and may show central clearing. Calcifications can be present in up to 25%, and when present, greatly lean towards the diagnosis of meningioma over AN. Another significant finding is that of a “dural tail”. This tapering of the mass along the dura is present 50-72% of the time, and is considered a major diagnostic feature. Also, as a result of this tapering effect, the angles formed between the tumor and the surface of the petrous bone are generally obtuse.

On CT scan, meningiomas may appear isodense to surrounding structures. The widening of the porus noted in acoustic schwannomas should be absent. CT scan may also show calcifications within the tumor, which are highly suggestive of the diagnosis. MRI is the study of choice. T1 images will show a lesion near the intensity of pons, however, it is may not be homogeneous, and may have a central hypointensity in larger lesions. On T2 images the lesion is between pons and CSF in intensity. After administration of gadolinium, the T1 image should show an intense lesion, although not as intense as AN. This sequence is also of value in evaluation for the dural tails. Flow voids caused by the tumor’s vasculature may be noted on any sequence, but is easier to see on T1 post-gad due to the brightness of the tumor after contrast.

## **Epidermoid**

Epidermoids are the third most common masses of the CPA, comprising 2-6% of the total lesions. These lesions are identical to the cholesteatomas of the middle ear, and may arise within the CPA or within the petrous portion of the temporal bone (congenital cholesteatomas). They are composed of a stratified squamous epithelium lining which surrounds a mass of keratin debris. They are generally benign and slow growing, but transformation of the capsule into a squamous carcinoma has been reported. Unlike most other CPA tumors, epidermoids are associated with facial weakness, paralysis, and spasm. Radiologically they are found to expand into nearby structures. They can cause erosion of the petrous bone, they may extend into the middle fossa with a “dumbbell” appearance, or they may cross midline and expand in the contralateral cistern. Surgically, these tumors are noted to be adherent to adjacent structures, and are may be difficult to remove.

On CT, epidermoids are close to CSF in density and generally do not enhance. MRI is the study of choice for the diagnosis of epidermoid, because the lesions often are difficult to differentiate from arachnoid cysts. T1 images show a very low intensity mass which approximates CSF. T2 images show a very bright mass which again approximates CSF in intensity. These findings are identical to arachnoid cysts. Special MRI protocols such as constructive interference steady state (CISS) or diffusion weighted imaging are required for positive identification. On CISS the epidermoid appears moderately intense, while the arachnoid cyst is hypointense. The same is true for diffusion weighting. This differentiation is crucial, since arachnoid cysts can often be medically managed with diuretics.

## **Other Primary CPA masses**

In addition to acoustic neuroma, meningioma, and epidermoids, there are a number of other primary lesions of the CPA. These include arachnoid cysts, non-acoustic cranial nerve schwannomas, vascular malformations, dermoids, teratomas, and lipomas. Together, these make up less than 5% of all CPA lesions.

The arachnoid cyst is important to remember for two reasons. First, it is difficult to distinguish radiologically from an epidermoid. Both epidermoids and arachnoid cysts appear dark on T1 weighted MRI and are very bright on T2. The best method of differentiation may be through diffusion weighted MRI or CISS sequence MRI, which would show the epidermoid as a solid tissue mass, and the arachnoid cyst as a fluid filled structure. Second, the treatment for epidermoids and cysts are very different. Surgery is the mainstay for epidermoid lesions of the CPA, where symptoms of arachnoid cysts may be controlled without surgery through the use of diuretics.

Although acoustic schwannomas may constitute up to 90% of CPA lesions, schwannomas of other cranial nerves probably only account for 2%. Trigeminal schwannomas more commonly arise in the middle fossa and later extend into the CPA, where they may cause typical vestibular or acoustic symptoms. These lesions may present with facial numbness or trigeminal neuralgia. The lesions are radiographically similar to acoustic neuromas on CT and MRI, except for their location. When presenting with vestibulocochlear symptoms they are commonly found to arise within the region of Meckel’s cave and the dumbbell into the CPA.

Any neuroma lesion found within the CPA and middle fossa is most likely to originate from the middle fossa and extend to CPA (CN V lesions) rather than progressing the other direction. These lesions often require a combined surgical approach due to their involvement of more than one cranial fossa. Facial schwannomas are important to remember because of their similarity in appearance to acoustic schwannomas. Radiographically, they can be indistinguishable from acoustic neuromas when they arise between the CPA and geniculate ganglion. The only time the diagnosis can be made with confidence is when extension beyond the geniculate ganglion into the tympanic or mastoid segments of the facial nerve occurs. When resecting these lesions, resection of the facial nerve with an interposition graft (usually the greater auricular) is required. Neuromas of cranial nerves IX, X, and XI may present with isolated cranial nerve symptoms, or when involving the foramen, present with a constellation of symptoms known as the “jugular foramen syndrome.” This includes dysphagia (CN IX), hoarseness and possible aspiration (CN X), and shoulder weakness (CN XI). Radiographic densities and intensities of neuromas are found on CT and MRI, respectively, as well as widening of the jugular foramen. Hypoglossal neuroma may present with hemiatrophy of the tongue and enlargement of the hypoglossal canal on CT. Surgical resection is the mainstay of treatment for these lesions.

There are several types of vascular malformations of the CPA. The most common is vertebrobasilar dolichoectasia which is an abnormal dilation and elongation of a segment of the vertebrobasilar artery. Symptoms may include facial twitching or trigeminal neuralgia. The abnormality can be diagnosed on CT, MRI, or conventional vs. MR angiography. Another common vascular abnormality is an aberrant loop of the anterior inferior cerebellar artery (AICA). The position of this artery is highly variable, and may pass over, under, or between the facial and vestibulocochlear nerves. It also common for the artery to loop into the IAC for a short distance. This finding is usually incidental and rarely causes symptoms, but may be confused as an intracanalicular lesion, and has been implicated by some as a cause for vertigo. Giant aneurysms can occur in the CPA region, but are rather rare. These, as the VBD above can be diagnosed by their flow voids on MRI, their enhancement with IV contrast, or by conventional or MR angiography.

### **Secondary CPA masses**

There are a large number of lesions which may extend into the CPA as they grow, or compress structures of the CPA, giving a false clinical diagnosis. These may include lesions of the petrous apex, intra-axial posterior fossa tumors, or paragangliomas.

Petrous apex lesions may include cholesterol granulomas, epidermoid cysts (congenital cholesteatomas), carotid artery aneurysms, or chondrosarcomas, with cholesterol granulomas being the most common. Intra-axial lesions may include astrocytoma, ependymoma, medulloblastoma, hemangioblastoma, choroids plexus papilloma, or metastasis.

### **Treatment**

Treatment for the majority of CPA masses is surgical. This is true for the common lesions including AN, meningioma, and epidermoids. Exceptions to this rule may include recurrent tumors after prior resection, patients medically unsuitable for surgery, or small non-symptomatic or non-progressing lesions. For acoustic neuromas, there is often much debate on

exact indications for surgery and when observation may be appropriate. In part, this is due to the slowly progressive nature of the tumor. Growth rates for AN range from 0.2mm to 2mm per year, although rates of up to 1 cm per year have been reported. There are also a large number of lesions that are found to remain stable over time with long follow-up. Obviously, for a young, healthy patient with progressive symptoms and a small localized tumor, surgical therapy would be warranted. However, several indications for observation have been proposed. The first is advanced age. Some may state that patients over 65 (or 75 depending on author preferences) do not tolerate the surgical treatments well, and observation may be indicated. Others argue this may delay a large group of patients who need surgery until they are older and less fit to undergo the procedure. Poor overall health, lack of symptoms, or a stable clinical or radiographic tumor are also reasons cited to observation before surgery. For patients with AN in an only hearing ear, some also favor postponing surgery until hearing is non-functional. It must also be noted that non-surgical treatment for AN is now possible, and several authors recommend stereotactic radiosurgery on patients who are not fit for standard surgical procedures.

For patients who are candidates for surgical therapy, several factors should be evaluated. These include cochlear, vestibular, and facial nerve function, size of the lesion, extension of the lesion (middle fossa, medial, inferior, etc.), compression of brainstem, extent of IAC involvement, and function of the contralateral ear. Each of these considerations will be discussed in their appropriate sections below.

### **Translabrynthine Approach**

The translabyrinthine approach is the approach of choice for otologic surgeons. It provides wide exposure to the CPA, as well as excellent exposure of the lateral portions of the IAC. It is useful when the tumor involves the IAC and has significant involvement of the CPA, since the CPA portion is less amenable to a middle fossa approach, and the IAC component is not well suited for a retrosigmoid approach. This approach also provides good exposure of the facial nerve along the course of the tumor and has a good rate of facial nerve preservation. A consideration for the translabyrinthine approach is the hearing status in both ears. Most authors would advise against a translabyrinthine approach if the patient has functional hearing. This is generally defined with the “50/50” rule, which is a speech reception threshold less than 50 db with a discrimination score greater than 50 percent. Another consideration is involvement in an only hearing ear. Some advise against a translabyrinthine approach in these patients until all hearing is lost. Complications of the translabyrinthine approach may include dizziness, vertigo, facial paralysis, CSF leak, and meningitis. The rates of these complications are generally low. The CSF leak rate may be higher due to communication with the middle ear. Facial nerve paralysis may be less common with this method. The main drawback of this approach, however, is permanent loss of hearing and vestibular function on the operated side. Most patients compensate their vestibular function adequately for normal daily activity.

### **Middle Fossa Approach**

The middle cranial fossa approach involves drilling out the IAC from its superior aspect through a wide exposure in the floor of the middle fossa. This obviously would provide excellent exposure to the lateral aspect of the IAC, but would also limit visualization and dissection within the medial portions of the CPA. Therefore, the indications for a middle fossa

approach include obvious extension into the middle fossa, small intracanalicular tumors when hearing is serviceable, IAC based tumors with minimal CPA extension, and very large tumors which require wide exposure for adequate resection. Due to the nature of the dissection, several unique complications are possible, these include temporal lobe stroke and injury to the middle meningeal artery or petrosal sinus. Some authors recommend against using this procedure in the elderly, since they often do not tolerate retraction of the temporal lobe well. Many also consider extension into the CPA beyond 0.5 – 1 cm as a contraindication due to lack of visualization of this area provided by this exposure.

### **Suboccipital Approach**

The suboccipital or retrosigmoid approach is a common neurosurgical approach to CPA lesions. This allows exposure of the CPA from the posterior aspect. It allows for visualization of the tumor's inferior and medial extent. This approach is preferred when lesions are large due to the wide exposure provided. It has a good rate of hearing preservation, and is indicated in patients with serviceable hearing. It is also preferred when compression of brainstem structures is noted. Contraindications to this approach would be intracanalicular tumors, especially if the extension is far lateral. Some drilling of the IAC from the posterior approach may be performed, however, as the drill-out and dissection of tumor is carried more lateral, identification and preservation of non-involved nerves becomes more difficult. This procedure is also not indicated as the sole method of treatment for tumors with middle fossa extension. The usual complications of CSF leak and meningitis may occur. Facial nerve paralysis may be slightly higher in this group. Cerebellar injury may occur with traction on the cerebellar peduncle, or if a portion of the peduncle is resected to provide exposure of a medial tumor.

### **Stereotactic Radiosurgery**

In contrast to conventional radiotherapy which delivers wide-field radiation to tumors, the stereotactic approach uses multiple small beams of radiation and targets them on a precise point for more targeted therapy. Several studies have been conducted on this new method in the past few years, and several indications have been formulated. These include patients medically unsuited for surgery, patients with prior surgical resections, small lesions, and functional hearing status. Noted contraindications are large tumors (greater than 3 cm), a prior dose of radiation to the CPA or surrounding structures, or compression of the brainstem. Using these guidelines, local control of acoustic neuromas has been reported up to 94% in large studies. This number includes tumors which are "cured" as well as those which are rendered non-progressive on follow-up. Hearing preservation has been adequate, between 47-77% for all lesions. This new method of treatment has shown promising results, and may be especially beneficial for patients that were previously considered inoperable or those who do not wish to have the morbidity of surgery. Noted complications of this technique include facial paralysis of 5-17%, injury to the trigeminal nerve in 2-11%, and post-radiation hydrocephalus requiring ventriculo-peritoneal shunt up to 3%.

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