REVIEW ARTICLE

Nonvestibular Schwannoma Tumors in the Cerebellopontine Angle: A Structured Approach and Management Guidelines

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ABSTRACT

The most common cerebellopontine angle (CPA) tumor is a vestibular schwannoma, but one in five CPA tumors are not vestibular schwannomas. These tumors may require different management strategies. Compared with vestibular schwannomas, symptoms and signs from cranial nerve VIII are less frequent: other cranial nerve and cerebellar symptoms and signs predominate in patients with these less common CPA tumors. Computed tomography and magnetic resonance imaging often show features leading to the correct diagnosis. Treatment most often includes surgery, but a policy of observation or subtotal resection is often wiser. This review provides a structured approach to the diagnosis of nonvestibular schwannoma CPA lesions and also management guidelines.

KEYWORDS: Cerebellopontine angle, epidermoids, meningiomas, tumors, management guidelines

About 6 to 10% of all intracranial tumors arise in or involve the cerebellopontine angle (CPA) and the vast majority of these (~80%) are vestibular schwannomas.[1–5]

Meningiomas and epidermoids account for ~10% and 6%, respectively, and the remainder consist of an extremely heterogeneous group of tumors that affect the region.[1–5] These tumors often resemble vestibular schwannomas in their clinical presentation and otologists must be aware that approximately one in five CPA tumors is not a vestibular schwannoma. A thorough history and examination will often provide clues leading to a different diagnosis and management strategy. In recent years, advances in neuroradiology have facilitated the preoperative differentiation of CPA lesions and have aided the surgical planning process. Magnetic resonance imaging (MRI) is the investigation of choice (Table 1). This review provides a structured approach to diagnosis of "nonvestibular schwannoma" CPA tumors and management guidelines.

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Table 1 Radiological Characteristics of CPA Tumors

Lesion	CT	CT + Contrast	MRI T1-weighted	MRI T2-weighted	MRI + Gadolinium	MRI Special Features
Vestibular schwannoma	→	+	↓	↓ / ↑	+	
Meningioma	↑	+	→ / (↓)	→ / ↓ / (↑)	+	''Dural tail''
Epidermoid	↓	−	↓	↑	−	
Lipoma	↓	−	↑	→ / ↓	+	Isointense to fatty tissues
Arachnoid cyst	↓	−	↓	↑	−	Isointense to CSF
Glomus tumor	→ / ↑	+	↓	↑	+	''Salt and pepper''
Choroid plexus tumor	→	+	→ / ↓	→ / ↑	+	

CPA, cerebellopontine angle; CT, computed tomography; MRI, magnetic resonance imaging; CSF, cerebrospinal fluid; ↓, hypointense; →, isointense; ↑, hyperintense; +, enhancement; −, no enhancement.

Vestibular schwannomas are usually the cause for the typical symptoms of a CPA syndrome. Audiovestibular disturbances are by far the most common symptoms. The typical natural history of vestibular schwannomas is an insidious hearing loss that develops over a period of several years, a pattern quite different from most of the less common CPA tumors.[6] Larger CPA tumors can inflict functional deficits on any of the cranial nerves that traverse the angle or neural structures that form part of its boundaries, the pons and cerebellum. In this way, these tumors cause altered facial and corneal sensation, nystagmus, ataxia, facial palsy, and so on. The progression and sequence of symptoms depend on individual regional anatomy and compliance, growth rate, and invasive nature of the tumor. The sequence of these symptoms may suggest a nonvestibular schwannoma lesion.[1,3,7] It also must be remembered that at least 30% of patients who present with a CPA syndrome have no diagnosable tumor at all but suffer from cerebrovascular disease, migraine, or other neurological disorder.[8] However, when a tumor is found some aspects of the CPA syndrome are almost always present.[8]

TUMORS DEVELOPING IN THE CPA

The CPA is covered or lined by the meninges and, in addition to cerebrospinal fluid, contains nerves, vessels, and possibly embryologic remnants. Each of these structures can be the tissue of origin of an unusual nonvestibular schwannoma CPA lesion.

Meningiomas

Meningiomas arise from epithelial cells that aggregate in clusters around the tip of the arachnoid villi. They can develop anywhere these cells are located. CPA meningiomas develop from cells within the internal auditory canal (IAC) or meatus, exit foramina of the trigeminal nerve (Meckel's cave), jugular foramen, sigmoid sinus, torcula, the superior or inferior petrosal sinuses, and from the region of the porus acusticus and clivus.[1,3,9] They are generally benign tumors but are locally aggressive invading bone along the haversian canals. This feature may produce radiologically demonstrable hyperostosis. Tumors displace or surround the cranial nerves and vessels rather than invade them and can become strongly adherent to these structures.[1,9] Several classification schemes have been proposed, an identifiable site of dural attachment being the main criterion used, but most were devised prior to modern imaging techniques. Today, each tumor is best classified according to where the bulk of its volume is located and by its relationship to major neurovascular structures—information generally more useful to the surgeon.[5,9,10]

Meningiomas account for ~10 to 20% of all intracranial neoplasms. The incidence increases with age and the average age at the time of diagnosis of posterior fossa meningiomas is 43.5 years.[11] Five

to 10% of all meningiomas are found in the CPA, predominately in middle-aged women. Second to vestibular schwannomas, meningiomas are the most common tumor found in the region and constitute ~10% of these tumors.[1,3,9] Hormonal aspects of the incidence and growth of these tumors has been investigated. Meningioma tumor cells contain a high concentration of progesterone receptors, moderate numbers of androgen receptors, and a low level of estrogen receptors. The potential of hormone therapy is being investigated.[11]

Neurofibromatosis type 2 (NF2) is associated with meningiomas and ~20% of adolescents with meningiomas have some form of neurofibromatosis.[11] There is little evidence of a hereditary predisposition to meningiomas per se in the absence of a neurocutaneous disorder, but chromosomal abnormalities are often present in the tumor cells.[11] Meningioma is thought to result from the loss or inactivation of a putative tumor suppressor gene located on chromosome 22. Meningiomas were among the first solid tumors recognized to have cytogenetic alterations. The most consistent change reported in benign meningiomas is a partial deletion on chromosome 22q12 or total deletion of chromosome 22. Previous irradiation of the head is associated with an increased risk of meningioma.[11]

Meningiomas differ from vestibular schwannomas in both their clinical presentation and treatment strategies. The size of the tumor is probably the most important factor to consider. Compared with vestibular schwannomas, audiovestibular symptoms (hearing loss, dysequilibrium, and tinnitus) are often less marked and of shorter duration in patients with meningiomas. Other cranial nerve symptoms tend to bear the brunt of these tumors. Hearing loss is found at presentation in ~50 to 80% of patients with meningiomas compared with almost all patients with vestibular schwannomas.[1,4,5,7,9,10,12–14] Tinnitus is experienced by anywhere from 15 to 60% of patients with meningiomas compared with 80% with vestibular schwannoma.[1,4,9,12–14] Dysequilibrium troubles ~30 to 60% of patients with meningiomas and is a presenting symptom in ~80% of patients with vestibular schwannoma.[1,4,9,10,12–14] Facial pain is rarely encountered in patients with vestibular schwannomas but is a presenting symptom in ~5 to 30% of patients with meningiomas.[10,12–14]

On examination, involvement of cranial nerve VIII can be demonstrated in ~50 to 70% of patients with meningiomas but is almost always present in the case of a vestibular schwannoma.[1,7,9,12–14] Fifth cranial nerve impairment is found in 20 to 60% of patients with meningiomas and abnormal cranial nerve VII function is observed in 10 to 50%, compared with ~40% and $< 5\%$, respectively, in patients with vestibular schwannomas.[1,4,7,9,12–14] Deficits of one or more of cranial nerves V to XII are found in up to 30% of patients with meningiomas but are relatively rare in vestibular schwannoma patients.[1,4,9,12–14] Cerebellar signs (ataxia, dysdiadochokinesis, etc.), infrequent in vestibular schwannoma patients, are reported in ~30 to 90% of patients with meningiomas.[7,9,10,12–14] Pure-tone thresholds tend to be better at the time of diagnosis than those of patients with vestibular schwannoma, with ipsilateral deterioration in up to 75% compared with almost all in patients with vestibular schwannomas.[1,13,14]

With contrast-enhanced computed tomography (CT), meningiomas are markedly hyperintense, homogeneous masses. CT is superior to MRI for evaluation of bone erosion, hyperostosis, calcification within the tumor, and extension into the middle ear. Hyperostosis and calcifications are more frequently associated with meningiomas than with other CPA tumors and involvement of the IAC is less frequent than in vestibular schwannomas.[3,11] There is usually more uniform contrast enhancement compared with vestibular schwannomas.[1,3] Meningiomas are better seen on MR scans with and without gadolinium administration. The T1-weighted image is heterogeneous and isointense or hypointense relative to brain parenchyma, and a low-signal intensity margin around the tumor is more commonly present than with vestibular schwannomas. Most meningiomas are large, sessile, and oval in shape with a broad dural attachment to the petrous bone as compared with vestibular schwannomas (Figs. 1 and 2).[1,3,11,12] Meningiomas are extremely variable in intensity on

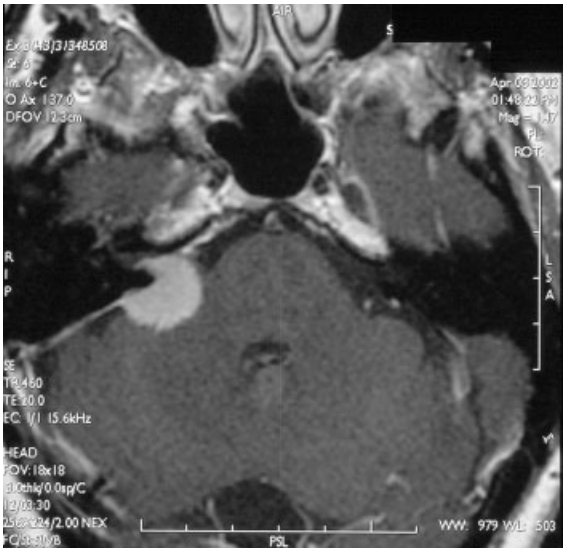

Figure 1 Meningioma. A 52-year-old woman complained of tinnitus and right-sided hearing loss for 6 months. Enhanced axial T1-weighted MRI of the posterior fossa shows a 16- × 11- × 18-mm large, heterogeneous, sessile lesion. The lesion extended into the internal auditory canal, but did not widen it.

T2-weighted images. Most often, they are iso- or hypointense compared with brain parenchyma and vestibular schwannomas and appear more homogeneous and less intensely enhancing after gadolinium administration. A different relaxation increment with gadolinium as compared with vestibular schwannomas may have some utility in differentiating these tumors.[1,3,11,12] A true "dural tail sign" when present is characteristic of meningiomas and seen extending away from the tumor mass along the dural surface on T1-weighted enhanced images (Fig. 2). It is reported to be present in ~50 to 70% of pathologically confirmed meningiomas. It has not been established if this feature represents neoplastic or reactive changes in the meninges adjacent to the tumor. Nevertheless, it is wise to bear this in mind at surgery and to achieve as wide a resection of the dura as possible.[3,11,12] Carotid angiography should be considered before resection of large CPA meningiomas, as they can be extremely vascular and preoperative embolization may reduce morbidity.

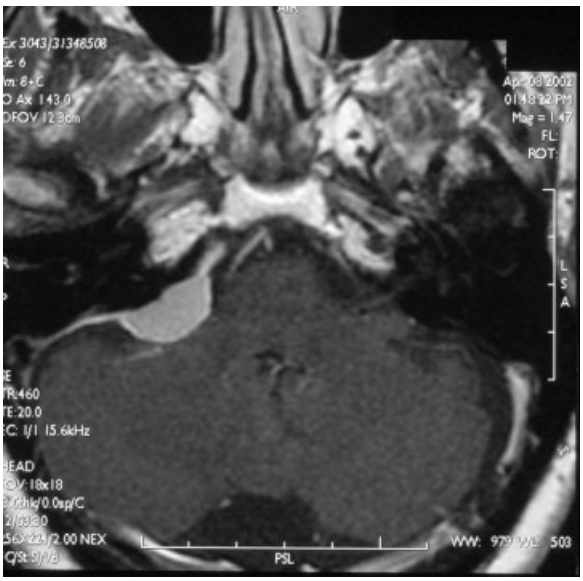

Figure 2 Meningioma. Same patient as in Fig. 1. Enhanced T1-weighted magnetic resonance image shows ''dural tail sign.''

The treatment of choice for meningiomas is surgical excision, and the goal of surgery is complete resection of tumor, involved dura, and hyperostotic bone. Exceptions to this strategy include patients with severe comorbidities and elderly patients with small tumors, where "watchful waiting" or alternative treatment may prevent surgery. As with vestibular schwannomas, a change in philosophy from radical excision to aggressive subtotal excision with preservation of neurological function is now usually advocated.

Excision may be accomplished though several standard approaches that are also employed for vestibular schwannomas. The choice depends on the patient's hearing acuity, the size and location of the tumor, and its involvement with neurovascular structures. The retrosigmoid (suboccipital) approach is the standard neurosurgical approach to the posterior fossa and has the advantage of offering the possibility of hearing preservation. Access to the lateral portion of the IAC may be limited but is usually not needed in the removal of tumors with little IAC involvement. The lateral portion of the IAC can be reached through the middle fossa approach, but this technique is not suitable for tumors that extend more than 1 cm into the CPA as medial exposure is limited and, of course, most

meningiomas are much larger than that. Postoperative facial nerve weakness is not uncommon with the middle fossa approach and is another consideration. The translabyrinthine approach offers direct access to the CPA; a lower mortality and improved facial nerve outcome have been reported compared with the retrosigmoid approach. It also has the advantage of minimal cerebellar retraction. The obvious disadvantage is sacrifice of residual hearing and diminished access to the dural origin on the petrous bone where troublesome bleeding often happens. This approach should be reserved for patients with poor hearing or tumors that fill the entire IAC.[3] A combined translabyrinthine-retrosigmoid approach can be used for removal of tumors greater than 3 to 4 cm. The transcochlear approach offers anterior exposure to facilitate resection of tumors extending into the region of the clivus, Meckel's cave, and the petrous portion of the internal carotid artery. Tumors in these areas may involve the tentorium, and a combination of the transcochlear and transtentorial approach may be necessary for these patients.

With refinements in microsurgical techniques, operative morbidity and mortality have been substantially reduced.[4,5,7,10,12] Mortality and complication rates increase with the age of the patient and size of the tumor. The most common surgical complications are cranial nerve dysfunctions, mainly involving cranial nerves VII, V, and VIII.[4,12] Cranial nerve damage is more common in tumors that arise anterior and inferior to the IAC.[12] Cerebrospinal fluid leak may require treatment with a lumbar drain or wound revision.[7,10,12] Reported mortality and recurrence rates are less than 10% in most series and are probably influenced by the surgeon's view on the appropriateness of radical surgery.[4,7,10,12]

CPA meningiomas involve the IAC to a lesser extent than vestibular schwannomas but may cause diminished hearing preoperatively, presumably by pressure on the cochlear nerve. Therefore, the criteria for attempting hearing preservation should be less stringent than for vestibular schwannoma.[11] Hearing preservation is more successful, when attempted, than in vestibular schwannoma surgery and success rates vary between 33% and 100%, compared with ~35% in vestibular schwannomas. Most studies have failed to document their auditory results adequately.[11]

Complete excision of a CPA meningioma is often difficult because of frequent involvement of essential structures; treatment strategies for residual and recurrent tumors are necessary. The role of radiotherapy has been investigated for both residual and recurrent tumors, and also as primary treatment in high-risk patients. Conflicting outcomes and results have been reported but most would suggest that this can be an effective and relatively safe form of treatment. Further follow-up studies are needed to assess long-term effects.[11,15] Similarly, brachytherapy with iodine^{125} seems to be of value for both recurrent and primary skull base meningiomas.[16]

Epidermoids

Epidermoids are distinguished from dermoids by the absence of skin adnexal components. In the CPA they are thought to develop from sequestrated epithelial cell rests from the laterally migrating secondary optic and otic capsules or from developing neurovasculature at a later state of embryogenesis.[3,5,17] They increase in size slowly by accumulation of keratin and cholesterol produced by desquamation of the squamous epithelium lining the mass. Their rate of growth is similar to that of normal skin, not exponential as in most neoplasms.[3,5,17] They encase and surround nerves and vessels, and irritate rather than displace them.[1,2]

Epidermoids account for ~0.2 to 1.8% of all intracranial neoplasms and an equal or slightly male-dominated incidence has been reported.[3,5,17,18] Thirty to 40% of epidermoids are found in the CPA, where they account for ~5 to 9% of all tumors.[5,17] Epidermoids are not associated with other congenital abnormalities.[17]

Because of their slow growth, CPA epidermoids may be asymptomatic for many years. The average duration of symptoms ranges from about 1 to 8 years.[5,7,17] As with meningiomas, hearing loss

is less frequent (50 to 80%) compared with vestibular schwannomas and early progressive facial nerve symptoms predominate.[1,4,7,17,19–21] Other less common symptoms are trigeminal neuralgia, facial numbness or spasm, cerebellar signs, or signs of elevated intracranial pressure. There is great variability in the reported rates of these symptoms and signs.[4,7,17,19–21] Signs involving cranial nerve VIII are found in 50 to 80% of patients. Signs relating to cranial nerves V and VII are less frequent, reported in ~30 to 50% of patients. Lower cranial nerve signs are even more rare.[4,7,17,19–21]

Plain radiographs are of little use, but may show lysis in the region of the petrous apex.[1,17]

On CT scans, CPA epidermoids appear homogeneous and hypodense with characteristically irregular margins. In general no enhancement is seen after contrast injection due to the avascularity of the tumor.[1,3,17] On CT scans, epidermoids may be difficult to distinguish from a subarachnoid cyst, which is also homogeneous and hypodense, but which usually has a more sharply defined margin.[3,17] Rare hyperdense epidermoids have been reported.[3,17] MRI has greatly facilitated the diagnosis of epidermoids and is superior in defining the extent and relationship to the surroundings.[3] The lesions have a characteristic heterogeneous, low-signal intensity appearance with no gadolinium enhancement on T1 images (Fig. 3). T2 scans typically demonstrate an inhomogeneous, high-signal intensity lesion, and are best to define the full extent of the tumor (Fig. 4).[3,5,17] Clinically it is important to distinguish the lesion from a cholesterol cyst, which appears hyperintense on both T1- and T2-weighted images.[1] Distinguishing an epidermoid from an arachnoid cyst on MRI can be difficult because the signal intensity of the epidermoid is similar to cerebrospinal fluid but heterogeneity within the tumor is more frequent with epidermoids.[3] Special fluid-attenuated inversion recovery and diffusion sequence MRI scans have proven useful in differentiating between arachnoid cysts and epidermoids.[22]

Microsurgical removal of epidermoids is the treatment of choice and the retrosigmoid approach

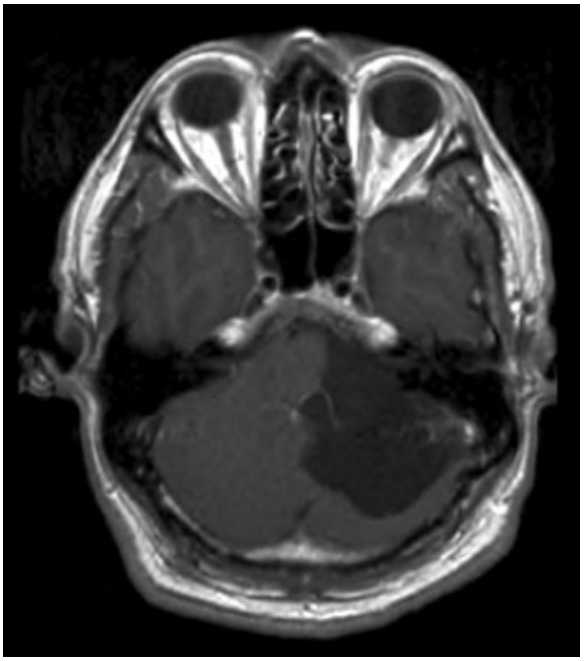

Figure 3 Epidermoid. A 55-year-old man complained of left-sided tinnitus and hearing loss for 2 years. He also experienced severe recurrent dizziness and chronic headache. Enhanced axial T1-weighted magnetic resonance imaging showed a large heterogeneous low-signal intensity tumor with no enhancement. The tumor was removed by the retrosigmoid approach.

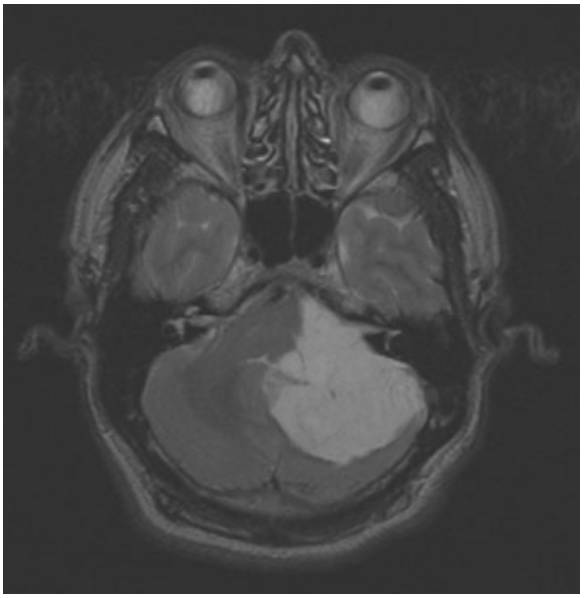

Figure 4 Epidermoid. Same patient as in Fig. 3. T2-weighted magnetic resonance image shows an inhomogeneous high-intensity lesion.

is the standard technique. As with meningiomas, alternative approaches such as the translabyrinthine or middle fossa can be employed if required.[1,3–5,7,19,20,23] The goal of surgery is decompression of the cyst and removal of the capsule. The interior of the tumor is soft and caseous and can be easily removed with suction or curettage. The capsule, however, is more difficult to remove. Because of the pattern of growth, neurovascular structures are often engulfed in the tumor and total excision can be difficult without increased mortality and morbidity.[1,3–5,7,19,20,23] Recent series show that the rates of radical resection range from 0 to 97%. Most authors advocate subtotal removal of the tumors instead of sacrificing functional nerves and, because of the slow growth rate, reoperation may be required only every decade or so.[1,3–5,7,17,19,20,23] Higher mortality and increased morbidity can be expected than from vestibular schwannoma surgery. Typical complications include deficits of cranial nerves VIII and VII. Aseptic meningitis may develop in up to ~40% of patients and is almost unique to this condition. Treatment includes steroids and antibiotics. Complications may be avoided by thorough removal of the lesion, minimal intraoperative spillage of debris, and the prophylactic use of steroids.[3,19,23] Spontaneous meningitis without surgery has been reported and an intracranial epidermoid should be suspected in a patient with repeated episodes of aseptic meningitis.[17] Malignant transformation of an epidermoid into a squamous cell carcinoma has been reported with more severe and rapidly progressive symptoms.[17] Even in modern series recurrence rates up to 35% are reported.[1,3–5,7,19,20,23] In such cases, CT and MR scans should be used to evaluate the evolution of the lesions, keeping in mind that the tumors may not become symptomatic during the patient's normal lifespan.[19,20,23]

Lipomas

The incidence of intracranial lipomas based on autopsies and CT screening is ~0.08%, but these are rare in the CPA where they represent only 0.05% of tumors. Only ~50 cases have been reported in the literature.[24] They are not considered true neoplasms but rather congenital malformations—an abnormal persistence and maldifferentiation of the meninx primitiva.[24,25] Blood vessels and nerves pass through the tumor and they are not displaced. These tumors grow slowly and the symptoms they cause are produced by mass effect.[24,25]

There is a 2:1 male predominance.[25] The mean age at diagnosis is ~50 years.[3] Lipomas of the posterior fossa are not associated with brain malformations as are other intracranial lipomas.[3,24]

The natural history of these tumors is often insidious and much longer compared with vestibular schwannomas. They cause cranial nerve VIII symptoms and in some cases symptoms in cranial nerve VII or V. Other cranial nerve and cerebellar symptoms have also been reported.[24,25]

Lipomas rarely cause bone erosion.[18] On CT scans lipomas are homogeneous, hypodense, and fail to enhance with contrast, similar to epidermoids and arachnoid cysts.[3,24,25] On T1-weighed MR scans lipomas appear hyperintense, similar to subcutaneous fat, and therefore enhancement is difficult to distinguish (Fig. 5). As with other fatty tissues, lipomas appear iso- or hypointense on T2-weighted images. The MRI appearance of lipomas is diagnostic, since no other lesion is hyperintense before enhancement on T1-weighted images and hypointense on T2-weighted images, with the extremely rare exception of atypical epidermoids, cholesterol cysts, or mucoceles. A definitive diagnosis can be made with fat suppression on an unenhanced T1-weighted scan.[3,25]

Because of the tight adhesion to the brainstem and cranial nerves and the slow progression of these tumors, a conservative approach should be adopted. Surgery may be indicated for patients with intractable trigeminal neuralgia, facial spasm, or dysequilibrium. Malignant degeneration has not been reported. Both the retrosigmoid and translabyrinthine approach have been employed when resection has been indicated. Significant morbidity has been reported and most authorities have

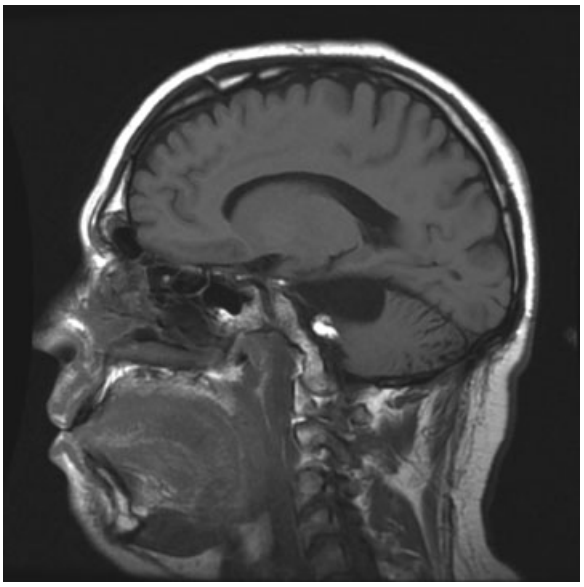

Figure 5 Lipoma. A 28-year-old man was referred with a recurrent cyst in the left cerebellopontine angle. He complained of headache, left-sided hearing loss, dizziness, and vision and balance problems. He had a left-sided abducens palsy. Cyst removal had been performed three times previously, but after all operations a small tumor in relation to the cyst was visible on control magnetic resonance imaging. Unenhanced T1-weighted magnetic resonance imaging showed a large cyst in the left CPA and a hyperintense tumor in relation to the cyst and pons. The cyst and tumor were removed by the retrosigmoid approach.

advocated partial resection with preservation of intact nerves.[3,24,25]

Arachnoid Cysts

Arachnoid cysts are presumably congenital malformations of the arachnoid and are histologically characterized by a cyst wall made of arachnoid or ependyma and a cystic space filled with cerebrospinal fluid or xanthochromic fluid. They are quite heterogeneous in their appearance and it is difficult to regard them as a homogenous group with just one pathology.[1,26,27]

They compose ~1% of all intracranial lesions and the CPA is the second most common location, second to the sylvian fissure. Strangely, only ~50 cases of CPA arachnoid cysts have been reported in the literature and the average age at diagnosis is ~30 to 40 years.[3,26,27] The etiology remains unknown but abnormal cerebrospinal fluid flow, trauma, or inflammation have been proposed.[3,26,27]

The cysts cause symptoms similar to other CPA lesions. They are often difficult to diagnose as symptoms are nonspecific, intermittent, and vague. The time interval between the onset of symptoms and diagnosis is often several years.[3,26,27]

On CT scans the lesions appear cystic with the same low density as cerebrospinal fluid. They fail to enhance after contrast administration and can be confused with epidermoids. On MRI they also appear similar to cerebrospinal fluid, that is, hypointense on T1-weighted images and hyperintense on T2-weighted images. No enhancement is seen.[1,3,26,27]

Asymptomatic cysts require no treatment but should be followed by serial MR scans. Microsurgical decompression and fenestration via the retrosigmoid approach is the most commonly recommended procedure.[1,3] There are sporadic reports of cyst recurrence after inadequate fenestration and some authors advocate complete cyst resection.[27] Others suggest cystoperitoneal shunts.[1,3,26,27]

Other Cranial Nerve Lesions

Schwannomas appear to be the primary lesion of cranial nerves V, VII, IX, X, and XI. Their presenting symptoms reflect the nerve involved. Overall they constitute 2 to 3% of all CPA tumors.

Facial schwannomas are slow-growing tumors and may involve any part of the nerve. The posterior fossa and/or CPA is involved in ~50%. Symptoms most often include dysfunction of cranial nerves VII or VIII.[1–3,18] On CT and MRI the mass appears identical to a vestibular schwannoma and the key to diagnosis is the neuroanatomic location of the tumor.[1–3,18] Features include widening of the fallopian canal in the temporal bone.[3,18] In patients with mild or no facial dysfunction, a conservative attitude with observation and, if necessary, only debulking surgery are appropriate, in the attempt to spare facial nerve function. The surgical approach depends upon tumor extension and location, and

the surgeon should be prepared to operate on the entire length of the nerve. Rerouting of the nerve is preferred when possible but cable grafting is often necessary.[1,3,18] Malignant facial schwannomas have been reported and may develop as part of NF2.[18]

Trigeminal nerve schwannomas tend to involve the ganglion, the nerve root, or both. Symptoms of nerve V tend to dominate over dysfunction of nerve VIII and the patients present with facial pain or numbness. Other symptoms of an expanding CPA tumor may also be present.[1,18] CT scans demonstrate enlargement of Meckel's cave or foramen lacerum and the tumors tend to be hypo- or isodense and show contrast enhancement. On MRI, the tumors appear iso- or hypointense on T1-weighted images and isointense on T2-weighted images. They enhance as brightly as other schwannomas. Tumor removal is often possible through combined posterior and middle fossa approaches with lateral opening of Meckel's cave.[1,18]

Lower cranial nerve schwannomas account for less than 1% of all CPA lesions. Symptoms reflect the nerve involved. Treatment, when necessary, is usually surgical.[18] The future role of stereotactic radiotherapy or gamma knife for these schwannomas remains to be seen. No doubt with time more and more cases will be treated this way and will probably have much the same outcome as with vestibular schwannomas.

Miscellaneous

Dermoids are midline cysts and rarely invade the CPA. They contain skin adnexal components, unlike epidermoids.[18] Other extremely rare CPA congenital rest lesions include respiratory epithelial cysts and enterogenous cysts. They may mimic arachnoid cysts, lipomas, epidermoids, or cystic vestibular schwannomas on MRI.[18] Hamartomas are most uncommon and rarely found in the CPA as are salivary gland heterotopias and teratomas.[18] The vertebral and basilar arteries and some of their branches pass through the CPA and although not true neoplasms may cause mass effect on neural structures.[1,2,18] Melanomas may develop in the CPA as either primary or metastatic tumors, the latter being more common. The prognosis is poor.[2]

TUMORS ORIGINATING FROM THE SKULL BASE

Lesions originating from the structures of the skull base, especially the temporal bone, may reach the CPA by direct extension.

Paragangliomas (Glomus Tumors)

The largest of these tumors (Fisch type D) may present in the CPA by extension from minute bodies present in the jugular foramen, along the vagus or Jacobson's nerve. They are usually benign but locally aggressive and destroy the petrous bone to enlarge to the CPA. Symptoms include pulsatile tinnitus, headache, or hearing loss. On otoscopy a red pulsatile mass can be seen behind the tympanic membrane with increased vascularization of the floor of the external auditory canal. Most patients have conductive hearing loss but with tumors that occupy the CPA either a mixed loss or profound sensorineural loss is not uncommon. Arteriography should be performed to demonstrate the extent of the tumor and source of its blood supply.[1,2] On CT, tumors appear well defined with adjacent bone erosion and marked enhancement after contrast injection. Paragangliomas have a characteristic "salt and pepper" appearance on both T1- and T2-weighted MR images because of intratumoral blood vessels and hemorrhages. They enhance intensely after gadolinium administration.[1,2] Treatment options include radiotherapy but only surgery is curative. Subsequent cranial nerve deficits are common.[1]

Miscellaneous

Cholesterol granulomas arise from the apex of the petrous bone and may enlarge to reach the posterior

fossa to produce cranial nerve dysfunction.[2] Chondroma and chondrosarcoma are rare and develop from embryonic cartilaginous remnants enclosed in the bones of the skull base.[2,18] Chordomas, remnants from the notochord, endolymphatic sac tumors, pituitary adenomas, and craniopharyngiomas may also involve the CPA.[2,18]

INTRA-AXIAL TUMORS WITH CPA IMPINGEMENT

Rarely, intra-axial or intraventricular tumors invade the CPA. Their diagnosis is difficult, but subtle image signs such as narrowing of the cisterns, edema at the site of tumor origin, and also the clinical history of the patient are sometimes helpful.

Choroid Plexus Papillomas

Choroid plexus papillomas are rare tumors. They represent less than 1% of intracranial neoplasms. Derived from the epithelial cells of the choroid plexus, they have the same microstructure of normal choroid plexus when benign. Most often affecting children, they arise typically in the lateral ventricles. In adults the most common site is the fourth ventricle. Malignant forms are very rare. Patients with choroid plexus papillomas may present with signs of a CPA tumor and, if of significant size, raised intracranial pressure is almost always present. CPA choroid plexus papillomas should be evaluated using MRI and angiography. Their prognosis is excellent when total surgical excision is possible. Radiotherapy has been employed when surgery is not possible but the results are variable.[2,28,29]

Primary Central Nervous System Neoplasms

As few as 0.3 to 2% of lesions in the CPA originate from primary brain tissue. Presenting symptoms may be typical of a CPA lesion but an intra-axial lesion may be suspected because of the rate of progression of symptoms and the degree of non-acoustic cranial nerve symptoms.[2,30] Primary central nervous system tumors of any type can present as a CPA lesion but the most common types are medulloblastomas, astrocytomas, ependymomas, and other brainstem gliomas. It may not be possible to distinguish these lesions from vestibular schwannomas by CT. MRI should demonstrate their intra-axial status more reliably. Stereotactic biopsy has an accuracy rate of ~75% and treatment may include surgery, radiotherapy, and in some cases chemotherapy.[2,18,30]

Miscellaneous

Only 13 cases of CPA lymphomas have been reported in the literature. They develop in the CPA as either an extra-axial lymphoma, an extension from an intra-axial lymphoma, or as a leptomeningeal lymphoma. Aggressive removal is not necessary, but additional radiotherapy should always be considered.[31] Reports of metastatic lesions from either intracranial or extracranial sources are occasionally published. The most common sources of extracranial origin are breast, prostate, lung, and gynecological tumors.[18]

CONCLUSIONS

Consider diagnoses other than vestibular schwannoma when a CPA lesion is suspected and hearing loss is not the predominant symptom or when scans are not typical. All these patients should have a complete MR study with contrast and if necessary fat suppression sequences. The complete removal of extensive meningiomas and epidermoids is often neither necessary nor wise. A policy of subtotal resection minimizes morbidity. Stereotactic radiotherapy or gamma knife radiosurgery is often an effective alternative therapy for residual

meningiomas that continue to grow. Similarly, staged surgical interventions for residual, symptomatic epidermoids can delay the onset or acquisition of serious neurological deficits.

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