

## Original Article

## Diagnostic and surgical challenges in resection of cerebellar angle tumors and acoustic neuromas

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### Abstract

**Background:** Cerebellopontine angle (CPA) lesions can mimic more common tumors through nonspecific symptoms and radiologic findings.**Methods:** To increase the preoperative diagnostic accuracy for CPA pathologies, the authors review the full spectrum of reported CPA lesions.**Results:** A wide spectrum of lesions mimics vestibular schwannoma (VS) in the space of the CPA.**Conclusion:** The presence of any suspicious clinical and radiographic finding uncharacteristic of VS makes it necessary to maintain a broad differential diagnosis list. Differentiation of CPA lesions, although challenging, may be best achieved by incorporating the clinical history, physical exam findings, audiometry results, and multi-modality imaging studies to construct a comprehensive preoperative knowledge of the lesion. This knowledge will allow improved operative execution and outcomes.**Key Words:** Acoustic neuroma, cerebellopontine angle tumor, ependymoma, neurosurgical procedures**Access this article online****Website:**  
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### INTRODUCTION

Housing 6–10% of all intracranial tumors,<sup>[7,8,19,22,36]</sup> the cerebellopontine angle (CPA) represents a space bounded by the pons, anterior cerebellum, and petrous temporal bone.<sup>[4]</sup> CPA lesions most commonly present with symptoms related to the brainstem or cranial nerves compressed by or involved with the lesion and are difficult to differentiate based on clinical presentation alone.<sup>[4]</sup> Vestibular schwannomas (VSs) comprise 70–90% of all mass lesions within the CPA space.<sup>[2,19]</sup> After VSs, the next two most common lesions found within the CPA are meningiomas (5–10%) and epidermoid cysts (3–7%).<sup>[6,15,17,24-26,30]</sup>

The remaining abnormalities found within the CPA account for less than 1% of all lesions in this region, are exceedingly rare, and may be challenging to preoperatively diagnose with accuracy. Although the introduction of various magnetic resonance imaging (MRI) sequences has allowed for an earlier and more accurate diagnosis of intracranial masses, CPA lesions may be misdiagnosed as they can mimic VS and other more common tumors in the area through their nonspecific presenting signs, symptoms, and radiologic findings.

Inaccurate diagnosis may lead to suboptimal treatment strategies. Therefore, to potentially increase the preoperative diagnostic accuracy of clinicians involved

with the care of patients with CPA pathologies, the authors attempt a comprehensive review of the full spectrum of reported CPA lesions.

### SPECIFIC TUMOR TYPES

The broad spectrum of unusual lesions within the CPA includes dermoid cysts, lipomas, neurenteric cysts, neuroepithelial cysts, cholesterol granulomas, paragangliomas, chordomas, chondrosarcomas, pituitary adenomas, endolymphatic sac tumors, arachnoid cysts, metastasis, non-vestibular schwannomas, gliomas, lymphomas, ependymomas, papillomas, hemangioblastomas, and medulloblastomas, among others.<sup>[1,3,4,6,7,30]</sup> The authors provide a brief overview of a few selected lesions. The features of other tumors are summarized in Table 1.

### Vestibular schwannomas

VSs are benign, slow-growing schwann cell tumors that typically arise from the inferior vestibular portion of the VIII cranial nerve.<sup>[4]</sup> VSs do not typically invade surrounding cerebrovascular tissues. Most VSs originate from the VIII cranial nerve in the lateral third of the internal auditory canal, where they grow medially along the line of least resistance, remodeling and expanding the canal and extending into the CPA. In 90% of patients, the initial presenting symptoms entail unilateral sensorineural hearing loss, tinnitus, and imbalance. Atypical symptoms include headache, facial numbness, facial weakness, nausea, vomiting, otalgia, dysarthria, and dysphagia. Facial weakness does not usually appear until the tumor has grown quite large.<sup>[9]</sup>

Resection of large VS may be challenging due to the adherence of the tumor to the surrounding

**Table 1: Partial list of tumors found within the cerebellopontine angle**

Lesion	MRI			Suggestive features
	T1	T2	Enhancement	
Lesions arising from the cerebellopontine cistern				
Meningioma	Isointense to hypointense	Hypointense to variable	Yes	Broad base, hemispheric shape; "dural tail"; may appear as lytic or osteoblastic lesions on plain X-ray; usually forms an obtuse angle with the petrous ridge
Epidermoid cyst	Hypointense	Hyperintense	No	Hyperintense on diffusion-weighted imaging; lobulated margins; produces no reaction of the adjacent bony structures
Dermoid cyst	Hyperintense	Hypointense	No	Midline lesions that rarely invade the CPA laterally; contain elements from all skin layers; T1 images suggestive with fat, calcification, and fat-fluid levels
Arachnoid cyst	Hypointense	Hyperintense	No	Isointense to CSF on all sequences; hypointense on diffusion-weighted imaging; erodes adjacent bone structures
Lipoma	Hyperintense	Isointense to fat	No	Exactly matches signal intensity of fat with all sequences
Facial schwannoma	Hypointense	Hyperintense	Yes	Plain X-rays may show widening of the IAC and fallopian canal expansion; extends much further laterally than acoustic neuromas
Trigeminal schwannoma	Hypointense	Hyperintense	Yes	Follows the anatomical course of the nerve; plain X-ray may demonstrate enlargement of the foramen lacerum/rotundum/ovale or erosion of Meckel's cave
Aneurysm	Hypointense	Hypointense	Possible	High-flow aneurysms appear as oval or round masses that demonstrate flow void; low signal intensity on T2-weighted imaging is suggestive; thrombosed aneurysms demonstrate high signal intensity on T1
Melanoma	Hyperintense	Isointense or hypointense	Yes	Variable signal intensity on T1- and T2-weighted images in proportion to the amount of melanin
Lesions arising from the skull base				
Cholesterol granuloma	Hyperintense	Hyperintense	No	Hypointense rim on T1- and T2-weighted images; arises from the petrous apex and expands into the posterior cranial fossa and produces nerve disturbances; homogenous central high signal intensity on T1-weighted images is very suggestive
Paraganglioma	Hypointense	Hyperintense	Yes	Salt-and-pepper appearance, flow voids

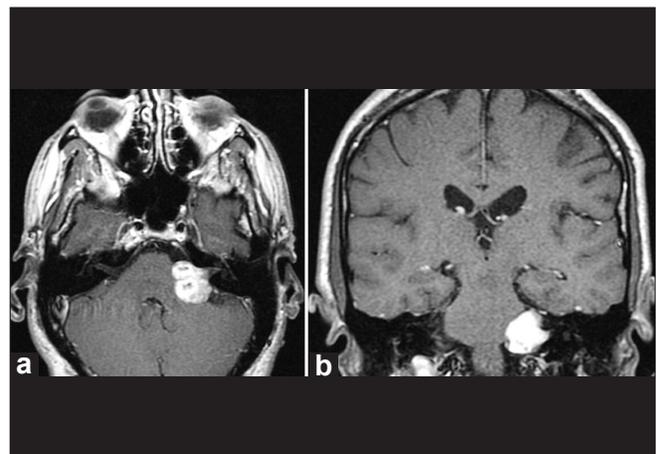
Contd...

**Table 1: Contd...**

Lesion	MRI			Suggestive features
	T1	T2	Enhancement	
Chondroma	Hypointense	Hyperintense	Variable	Lytic bone erosion; calcifications present; enhances poorly due to hypovascularity
Chordoma	Hypointense	Hyperintense	Yes	Intratumoral septa; arises in the midline and destroys the clivus; irregular bony erosion with slight calcification at the periphery of the tumor
Endolymphatic sac tumor	Variable	Hyperintense	Yes	Hyperintense cysts on T1- and T2-weighted images; destroys retrolabyrinthine petrous bone
Pituitary adenoma	Hypointense	Hyperintense	Yes	Hx of pituitary adenoma
Apex petrositis	Hypointense	Hyperintense	Yes	Hx of otitis media
Intra-axial or intraventricular lesions invading the CPA				
Glioma	Hypointense	Hyperintense	Yes	Arises from the brainstem with asymmetric expansion; hyperintense adjacent edema noted on T2-weighted imaging
Choroid plexus papilloma	Hypointense	Hyperintense	Yes	Arises from the fourth ventricle and extends through the foramen of Luschka; usually no edema in the brainstem and no erosion of adjacent bony structures
Lymphoma	Hypointense	Hyperintense	Yes	Nonspecific imaging characteristics though edema and mass effect associated with lesion suggest intra-axial origin; restricts on diffusion-weighted imaging
Hemangioblastoma	Hypointense	Hyperintense	Yes	Solid or cystic with nodule; hypervascular; may be associated with Von Hippel-Lindau disease frequently a stippled pattern of calcification and contrast enhancement noted on CT
Ependymoma	Hypointense	Hyperintense	Yes	Irregular; heterogeneous due to calcification, hemorrhage, necrosis, and cystic components; extends through foramen of Luschka
Medulloblastoma	Hypointense	Hyperintense	Yes	Round or ovoid tumor with smooth margins; exophytic type growth into CPA; tends to arise from cerebellar hemisphere; irregularity of some portions of the tumor-brain interface suggests intra-axial origin of these lesions
Dysembryoplastic neuroepithelial tumor	Hypointense	Hyperintense		Cystic component, bone erosion

cerebrovascular structures. However, the tumor does not typically invade the arachnoid membranes and those considered subarachnoid tumors are characterized by the arachnoid membrane remaining on the tumor surface after moving the arachnoid fold.<sup>[16]</sup> These membranes provide a great opportunity to protect the nerves and vasculature along the brainstem and avoid operative injury. Samii<sup>[28]</sup> has recommended that if the tumor capsule alone is held with a forceps and tumor dissection is performed at the level of the arachnoid plane, such inadvertent injuries might be avoided.

Radiographically, VS are round or oval lesions that typically enhance after contrast administration and are located either in the internal acoustic canal or protrude into it [Figure 1]. Computed tomography (CT) images usually demonstrate widening of the canal and other bony changes caused by the lesion. Such bony changes confirm the slow growth rate and pressure characterized



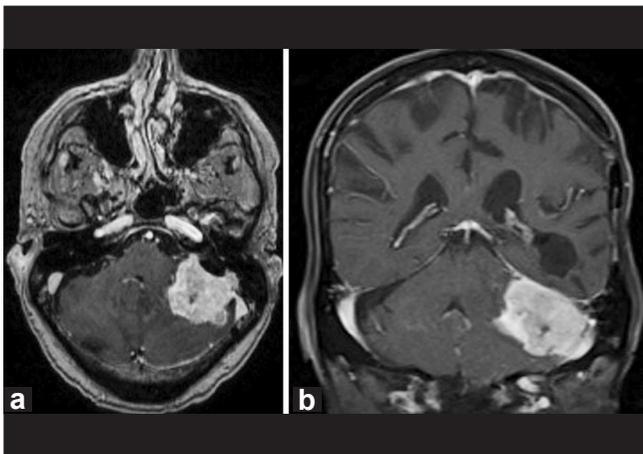
**Figure 1: Vestibular schwannomas are round or oval lesions that typically enhance after contrast administration (a: axial contrast-enhanced magnetic resonance imaging (MRI)) and are located either in the internal acoustic canal or protrude into it (b: coronal contrast-enhanced MRI)**

by this tumor type. Audiometry classically demonstrates sensorineural impairment with high-frequency hearing loss, resultant poor speech discrimination, and abnormal auditory brainstem responses. None of these radiologic or audiometry findings are specific for VS.

### Meningiomas

Meningiomas originate from the cells lining the arachnoid villi that are predominately found along the major venous sinuses of the petrous bone.<sup>[11,22]</sup> Meningiomas that occupy the CPA are usually based at the posterior surface of the petrous bone or the petrotentorial junction.<sup>[11,22,25]</sup> Because the anatomical location of meningiomas differs from that of VS, hearing loss tends to occur later in the disease process and other cranial nerves are more frequently involved. Furthermore, facial nerve symptoms are more likely to occur with meningiomas than with VSs when the lesion size is small<sup>[22]</sup> due to the higher chance that meningiomas can originate from the dura anterior to the porus acousticus and affect the anteriorly located facial nerve in the VII/VIII complex.

Radiographically, CT scan may demonstrate either bony destruction or hyperostosis due to the higher tendency of meningiomas invading the surrounding skull base bone as compared with VS. Meningiomas may present as homogenous masses with a flat base off-center from the internal auditory canal (IAC) although they can invade the IAC and cause its expansion.<sup>[23]</sup> The presence of calcification and dural tails may be helpful in distinguishing meningiomas from VSs. Both VSs and meningiomas are isointense to hypointense on T1-weighted MRI and are of variable signal intensity on T2-weighted sequences [Figure 2]. Furthermore, both lesions often brightly enhance with contrast, making radiographic differentiation of the two lesions difficult at times.<sup>[9,11,22]</sup>



**Figure 2: Magnetic resonance imaging scans (a: axial and b: coronal enhanced images) demonstrate a homogenous mass with a flat base off-center from the the internal auditory canal (IAC). Meningiomas can also invade the IAC and cause its expansion**

### Epidermoid cysts

Epidermoid cysts are the third most frequent tumor of the CPA.<sup>[22]</sup> They encase and surround nerves and arteries rather than displace them.<sup>[12,21]</sup> Facial twitching, pain, spasms, and weakness tend to occur earlier and with a relatively higher incidence in epidermoid cysts than with VSs.<sup>[22]</sup>

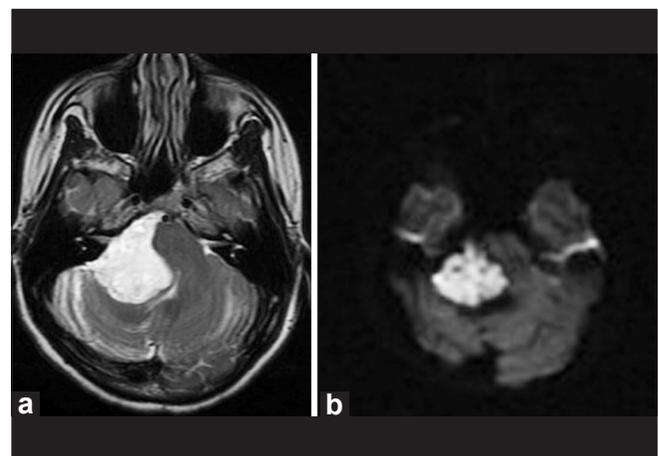
Radiographically, epidermoid cysts present as hypodense lesions with irregular margins eccentric to the IAC, which do not enhance with administration of gadolinium and may cause erosion of the petrous apex. They appear hypointense on T1-weighted and hyperintense on T2-weighted MRI. As opposed to arachnoid cysts, which are the primary differential diagnosis for this lesion, epidermoid cysts demonstrate high signal intensity on diffusion-weighted imaging<sup>[3]</sup> [Figure 3].

### Metastatic malignancies

The most common malignant neoplasms in the CPA include breast, lung, skin (melanoma), gastrointestinal, and genitourinary cancers.<sup>[22,26,27]</sup> The rapid progression of symptoms, including facial nerve involvement and weight loss, are strongly suggestive of malignancy.<sup>[27]</sup> Edema of the adjacent brain parenchyma may be seen in metastatic lesions to the CPA [Figure 4]. Multiplicity of the lesion leads the clinician to consider metastasis as a strong consideration of a mass in the CPA. In addition, leptomeningeal involvement on imaging can be another feature of metastatic tumors. The presenting symptoms vary based on the particular location of the mass in the CPA.

### Non-vestibular schwannomas

Non-vestibular schwannomas rarely present in the CPA.



**Figure 3: Epidermoid cysts present as hypodense lesions with irregular margins eccentric to the the internal auditory canal, which do not enhance with administration of gadolinium and may cause erosion of the petrous apex. They appear hypointense on T1-weighted and hyperintense on T2-weighted magnetic resonance imaging (a). As opposed to arachnoid cysts, which are the primary differential diagnosis for this lesion, epidermoid cysts demonstrate high signal intensity on diffusion-weighted imaging (b)**

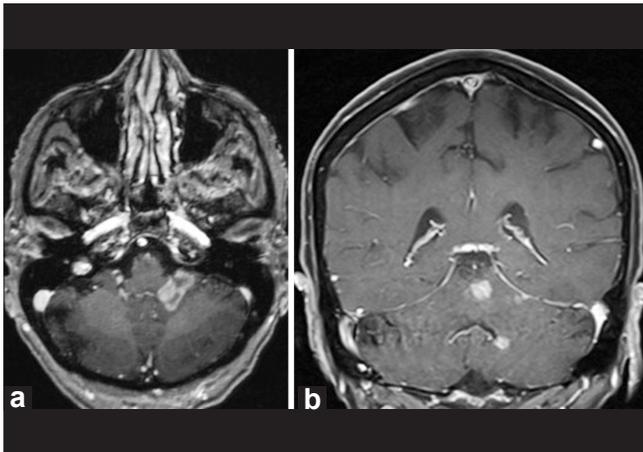
These lesions are usually easily distinguished from VSs based on different symptom patterns, neuroanatomic locations, shapes, and relationships within the skull base foramina and canals.<sup>[4]</sup>

### Trigeminal nerve neuromas

Trigeminal neuromas are exceedingly rare tumors.<sup>[22]</sup> Patients usually present with facial numbness or pain. On imaging, most trigeminal neuromas are located cephalad to VSs, have an anterior–posterior direction in the CPA cistern, and may extend into the Meckel's cave<sup>[4]</sup> [Figure 5]. MRI reveals an isointense or hypointense mass on T1 and isointense image on T2-weighted sequences.<sup>[25]</sup>

### Facial nerve neuromas

Eighty percent of facial neuromas involve two adjacent nerve segments and are often dumbbell shaped.<sup>[4]</sup> Facial nerve schwannomas may be difficult to distinguish from VSs based on their very similar clinical and radiographic



**Figure 4:** Axial magnetic resonance imaging (MRI) with contrast (a) reveals a mass in the fissure of the cerebellopontine angle. Multiple other lesions (b: coronal MRI with contrast) confirm the diagnosis of metastatic disorder (lung cancer)

presentation.<sup>[4,22]</sup> Intratemporal facial neuromas may demonstrate fallopian canal expansion.

### Lipomas

Lipomas rarely occur within the CPA.<sup>[22,29,34]</sup> They tend to surround and encase normal adjacent neurovascular structures with dense adhesions and produce progressive focal symptoms.<sup>[22]</sup> They may present clinically like an acoustic neuroma with asymmetrical hearing loss and tinnitus. Radiographically, they appear as homogeneously hypodense lesions on CT and may cause widening of the IAC. The characteristic feature of a lipoma is its high signal intensity on the unenhanced T1-weighted MRI sequence [Figure 6].<sup>[3,22,29,34]</sup>

### Dermoid cysts

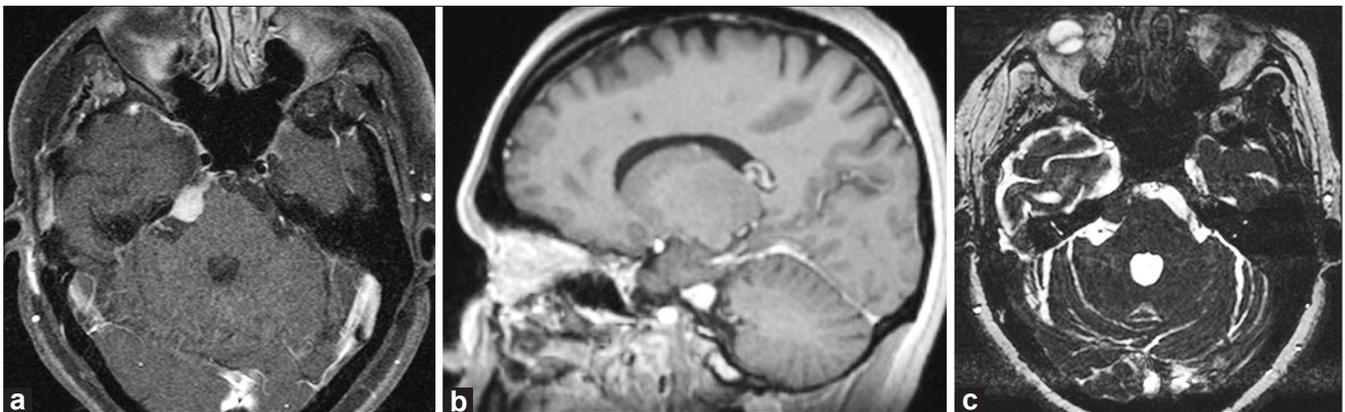
Dermoid cysts may contain fat, hair, or sebaceous glands in addition to squamous epithelium. They are usually midline lesions that rarely invade the CPA laterally. They demonstrate high signal intensity on T1-weighted MRI sequences due to their fatty content and may have a very suggestive fat-fluid level.<sup>[3,12,31,36]</sup>

### Arachnoid cysts

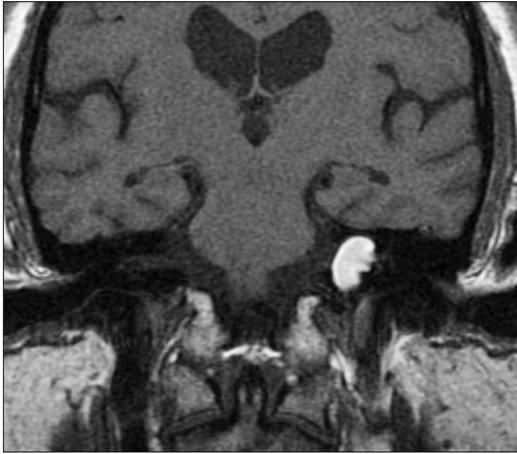
Arachnoid cysts form as a result of splitting and duplication of the arachnoid layers. These lesions can develop secondary to congenital malformation, infection, trauma, increased intraventricular pressure, or embryonic rests.<sup>[18,22]</sup> These lesions usually present with hearing loss, though atypical cases can present with facial weakness. On imaging, the attenuation and signal intensities for an arachnoid cyst match the CSF almost exactly<sup>[3]</sup> [Figure 7]. As opposed to epidermoid cysts, which have a high signal intensity on diffusion-weighted imaging, arachnoid cysts, like CSF, have a low signal intensity on diffusion-weighted imaging.<sup>[22,23]</sup>

### Glomus jugulare tumors

Only the largest group of glomus jugulare tumors is found in the CPA, and when they occur, they present



**Figure 5:** Trigeminal neuromas are located cephalad to vestibular schwannomas (a: axial magnetic resonance imaging (MRI) with contrast). They have an anterior–posterior direction in the cerebellopontine angle cistern and may extend into the Meckel's cave (b: coronal MRI with contrast). Close association with the trigeminal nerve is noted (c: T2-weighted axial MRI)



**Figure 6:** The characteristic feature of a lipoma is its high signal intensity on the unenhanced T1-weighted MRI sequence (coronal)

with pulsatile tinnitus, headaches, hearing loss, and disequilibrium along with dysphonia and dysphagia. Conductive hearing loss and a red pulsatile mass behind the tympanic membrane are characteristic signs for this tumor type. In addition to VII and VIII cranial nerve palsies, IX, X, XI, and XII cranial nerve palsies may occur.<sup>[22]</sup> Arteriography demonstrates the rich, complex blood supply from the external and internal carotid arteries. With selective catheterization of the ascending pharyngeal artery, a dense tumor blush can be seen.<sup>[3,4,22,23]</sup>

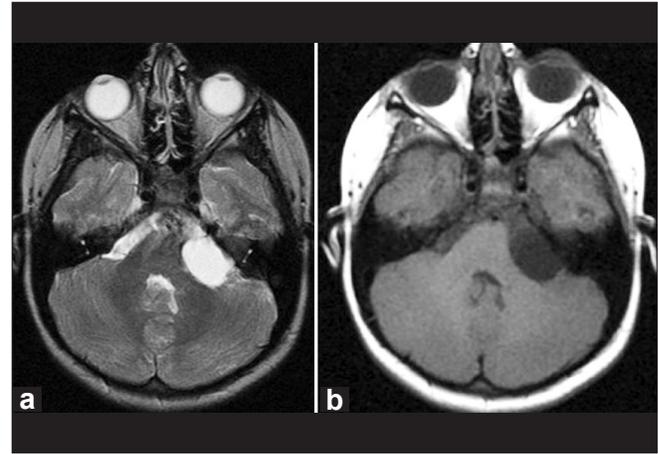
Enhanced MR images reveal an intense enhancement pattern which is most commonly homogenous.

### Vascular lesions

Vascular lesions are rare within the CPA but include distal anterior inferior cerebellar artery (AICA) aneurysms,<sup>[35]</sup> posterior-inferior cerebellar artery (PICA) aneurysms, vascular loops, a distal AICA mycotic fusiform pseudoaneurysm,<sup>[10]</sup> an intracanalicular arteriovenous malformation, and a cavernous hemangioma.<sup>[5,22]</sup> On imaging, vascular lesions are often visualized secondarily due to their flow voids; otherwise, the thrombosed lesion may be overlooked. Contrast administration will enhance these lesions when the blood is fast moving, but the lesion will not enhance if the blood is slow moving.<sup>[10,24,35]</sup>

### Inflammatory lesions

Inflammatory lesions of the cranial nerves have demonstrated both focal and diffuse enhancement patterns on neuroimaging and have mimicked VS. These inflammatory lesions have been associated with Bell's palsy<sup>[33]</sup> and Ramsey-Hunt syndrome.<sup>[13,14,33]</sup> Patients usually present with sudden onset of symptoms that localize to cranial nerves V, VII, and/or VIII. Conservative treatment with anti-inflammatory medications, including steroids, will reduce symptoms even though radiographic



**Figure 7:** On imaging, the attenuation and signal intensities for an arachnoid cyst match CSF almost exactly (a:T2- and b:T1-weighted images) and may cause remodeling of the petrous bone

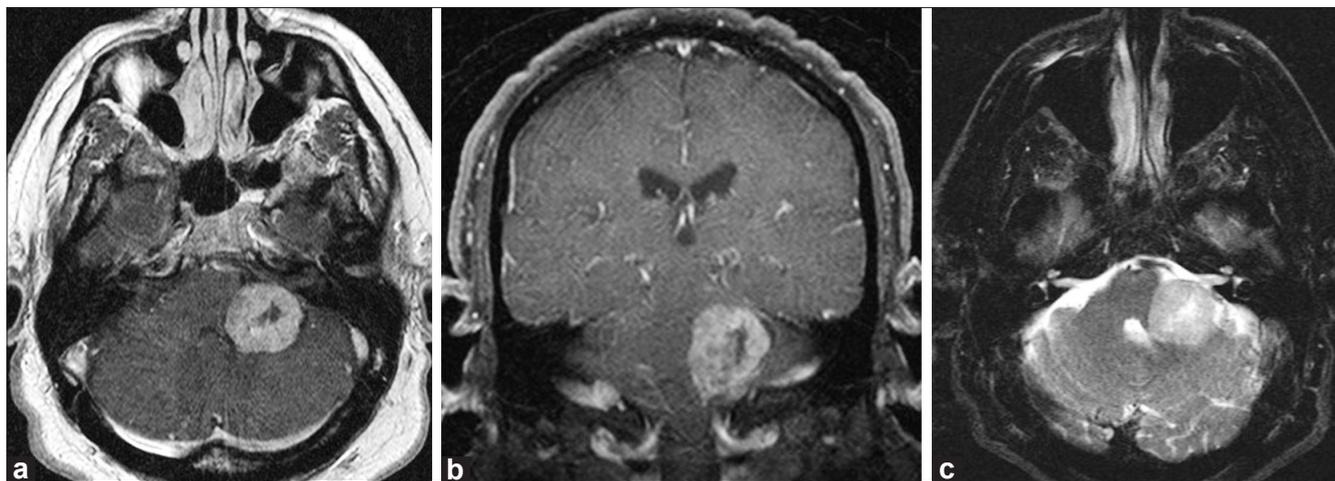
enhancement remains. In most of the reported cases, diffuse enhancement was noted along the course of the nerves. Lesions such as HIV-associated neurosyphilis may mimic an acoustic neuroma.<sup>[5]</sup>

### Infectious lesions

Numerous case reports have documented infectious etiologies for CPA lesions that mimic VS. For instance, a necrotizing granulomatous inflammatory lesion cultured positive for acid-fast bacilli,<sup>[20]</sup> sterile abscesses following resection of a VS 13 years earlier,<sup>[32]</sup> gummas from neurosyphilis, neurocysticercosis in the desert Southwest, and bacterial abscesses secondary to chronic suppurative otitis media have all masqueraded as CPA lesions.<sup>[1,32]</sup> These diverse infection-related lesions display unique imaging characteristics native to their individual etiology. Generally, abscesses demonstrate hypointensity on T1-weighted sequences due to central fluid collection and reveal minimal hyperintensity on T2-weighted imaging with surrounding edema. The striking feature, which is unique to abscesses, is the rim of intense contrast enhancement in the region of the capsule.<sup>[1,32]</sup>

### Ependymomas

Ependymomas usually arise from the fourth ventricle and its lateral recess and extend into the CPA by means of exophytic growth.<sup>[3]</sup> However, ependymomas of an extra-axial origin also exist and may grow directly into the CPA. They appear irregular and may invade the cerebellar parenchyma. Radiographically, ependymomas appear as hypointense lesions on T1-weighted imaging and as hyperintense lesions on T2-weighted imaging [Figure 8]. Ependymomas are markedly heterogeneous due to calcification, hemorrhage, cystic components, or necrosis.<sup>[3]</sup>



**Figure 8:** Ependymomas usually arise from the fourth ventricle and its lateral recess and extend into the cerebellopontine angle (CPA) by means of exophytic growth. However, ependymomas of an extra-axial origin also exist and may grow directly into the CPA. They appear irregular and may invade the cerebellar parenchyma. Ependymomas appear as hypointense lesions on T1-weighted imaging and as hyperintense lesions on T2-weighted imaging (a and b:axial and coronal contrast-enhanced images and c: axial T2-weighted sequence)

## MANAGEMENT RECOMMENDATIONS

Except for the inflammatory and infectious lesions, operative intervention remains a reasonable option for other lesions that are causing mass effect on the brainstem and cause corresponding compressive signs and symptoms. Particularly for VSs and other solid lesions, if the lesion is adherent to the important neighboring cerebrovascular structures, a radical resection is contemplated, with a small piece of the tumor left behind to avoid the associated morbidity. With the availability of radiosurgery for the treatment of residual lesions, postoperative morbidity should be minimized.

## CONCLUSIONS

A wide spectrum of lesions mimics VS in the space of the CPA. The presence of any suspicious clinical and radiographic findings uncharacteristic of VS makes it necessary to maintain a broad differential diagnosis list. Differentiation of CPA lesions, although challenging, may be best achieved by incorporating the clinical history, physical exam findings, audiometry results, and multi-modality imaging studies to construct a comprehensive preoperative knowledge of the lesion. This knowledge will allow improved operative execution and outcomes.

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