Cerebellopontine angle lipoma

Cerebellopontine angle (CPA) lipomas are rare, benign, slow-growing intracranial lipomas.

Characteristically lipomas of the CPA have the facial nerve and vestibulocochlear nerve coursing through it on their way to the IAM. They are associated with intravestibular lipomas and sensorineural hearing loss.

**Epidemiology**

They account for ~10% of all intracranial lipomas.

**Diagnosis**

Radiographic features

MRI brain Signal characteristics are those of a lipoma

T1: high signal

https://images.radiopaedia.org/images/2107232/e3e8537790364cc8a2404efc00cdd4_gallery.jpg

T2: high signal

ture FISP/FIESTA: low signal margin due to chemical shift artefact fat saturated sequences: shows signal dropout.

http://www.otosurgery.org/DJL_NEWSITE%20images/MRI_cpa-lipoma.gif

**Differential diagnosis**

Cerebellopontine angle tumors (CPA) are frequent; vestibular schwannomas and cerebellopontine angle meningiomas represent the great majority of such tumors. However, a large variety of unusual lesions can also be encountered in the CPA. The site of origin is the main factor in making a preoperative diagnosis for an unusual lesion of the CPA. In addition, it is essential to analyze attenuation at computed tomography (CT), signal intensity at magnetic resonance (MR) imaging, enhancement, shape and margins, extent, mass effect, and adjacent bone reaction. CPA masses can primarily arise from the cerebellopontine cistern and other CPA structures (arachnoid cyst, nonacoustic schwannoma, aneurysm, melanoma, miscellaneous meningeal lesions) or from embryologic remnants (epidermoid cyst, dermoid cyst, lipoma). Tumors can also invade the CPA by extension from the petrous bone or skull base (cholesterol granuloma, paraganglioma, chondromatous tumors, chordoma, endolymphatic sac tumor, pituitary adenoma, apex petrositis). Finally, CPA lesions can be secondary to an exophytic brainstem or ventricular tumor (glioma, choroid plexus papilloma, lymphoma, hemangioblastoma, ependymoma, medulloblastoma, dysembryoplastic neuroepithelial tumor). A close association between CT and MR imaging findings is very helpful in establishing the preoperative diagnosis for unusual lesions of the CPA.
The differential for lesions with high T1 signal includes:

- haemorrhagic vestibular schwannoma
- neurenteric cyst
- thrombosed berry aneurysm
- white epidermoid
- ruptured intracranial dermoid

**Treatment**

They are not associated with malformations and can become symptomatic in a slowly progressive way by affecting the more susceptible of the cranial nerves in this region. As a result of the rare occurrence, the experience in treating and managing these tumors is limited. Consequently, the recommendations for treatment and for the role of surgery are very variable.

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Resections are considered in symptomatic patients who are refractory to targeted medical therapies, but at those stages the lipomas have often reached considerable sizes and encompass critical neurovascular structures.

**Scoring system**

The objective of a study is to develop and to evaluate the utility of a scoring system for CPA lipomas. The hypothesis is that CPA lipomas with lower scores are probably best managed with early surgery.

The PubMed database was searched using relevant terms. Data on patient and lipoma characteristics were extracted and used to design a scoring system. CPA lipomas were stratified by scores with corresponding managements and outcomes analyzed.

One hundred and seventeen patients with CPA lipomas were identified and 40 CPA lipomas were scored. The remaining CPA lipomas were deficient in data and not scored. No lipomas were scored as 1. Score 2 lipomas (n = 12; 30%) most often underwent serial surveillances (n = 5; 41.6%), with the majority of symptoms remaining unimproved (n = 2; 40%). Patients with score 2 CPA lipomas treated with medical therapies (n = 3; 25%) often experienced symptom resolution (n = 2; 66.6%) (p = 0.0499). Patients with score 2 CPA lipomas undergoing surgical resections (n = 3; 25%) all experienced symptom resolution (n = 3; 100%) (p = 0.0499). Score 3 was most common (n = 16; 40%) and these lipomas were often surgically resected (n = 10; 62.5%). The majority of patients with score 3 CPA lipomas having undergone surgical resections (n = 10; 62.5%) experienced symptom improvement (n = 1; 10%) or resolution (n = 4; 40%).

Score 2 CPA lipomas are smaller and would be deemed non-surgical in general practice. However, the data of Lagman et al., suggest that these lipomas may benefit from either medical therapies or early surgical resections. The advantages of early surgery are maximal resection, decreased surgical morbidity, and improved symptom relief.
Case series

2014

Of 15 patients with CPA lipomas, six were female and nine were male, with an average age at presentation of 50.2 years (range, 31.7-76.4 yr) and an average follow-up time of 51.7 months (range, 6-216 mo). The lipomas were unilateral in all cases, nine on the right (60%) and six on the left (40%) side. None of the lipomas increased in size. All patients were treated conservatively. Sensorineural hearing loss was the main presenting symptom (80%) followed by tinnitus (46.7%) and vertigo (20%). None of the patients suffered from facial nerve dysfunction. There was no correlation between weight gain and tumor growth.

CPA lipomas can be diagnosed accurately with appropriate magnetic resonance imaging techniques and be managed conservatively with safety. Cochleovestibular are the most common presenting symptoms, whereas facial nerve involvement is rare. CPA lipomas do not tend to grow and can be monitored on a less regular basis.

2013

Between 1996 and 2012, 15 patients were diagnosed with a CPA or IAC lipoma at the authors' institution and were included in the analysis. The mean duration of radiological and clinical follow-up was 3.4 years and 5.1 years, respectively. Eight lesions were confined to the IAC, while seven involved the CPA. The median tumor size at diagnosis was 7.2 mm; one patient demonstrated tumor growth on serial MRI while the remaining subjects did not have radiological progression. The most common presenting symptoms were sensorineural hearing loss (40%) and tinnitus (33%); five patients were diagnosed after incidental discovery on MRI. Fourteen patients were managed with observation, while one subject underwent subtotal resection. None of the observed patients reported worsening symptoms at last follow-up.

While rare, lipomas should be included in the differential diagnosis of CPA and IAC lesions. Owing to a generally benign clinical course and high morbidity associated with resection, microsurgery should only be considered in cases of definite tumor enlargement with intractable symptoms from mass effect. Careful radiological evaluation is critical for establishing an accurate diagnosis in order to prevent unnecessary morbidity associated with resection.

2006

A healthy 42-year-old woman who presented with left-sided hearing loss and facial synkinesis. T1-weighted magnetic resonance imaging revealed an enhancing lesion of the left CPA with no signal on fat suppression sequences. Despite conservative therapy, the patient developed progressive hemifacial spasm, and a suboccipital craniotomy approach was used to debulk the tumor, which encased cranial nerves V, VII, VIII, IX, X, and XI. Surgical histopathology demonstrated mature adipocytes, consistent with lipoma. Two years after surgery, the patient remains free of facial nerve symptoms. Cerebellopontine angle lipomas are rare lesions of the skull base and are reliably diagnosed with T1-weighted and fat suppression magnetic resonance sequences, which we recommend in the routine radiologic workup of CPA tumors. Accurate preoperative diagnosis is crucial because most CPA lipomas should be managed conservatively. Partial surgical resection is indicated only to alleviate intractable cranial neuropathies or relieve brainstem compression.

2002

Tankéré et al., report four new cases of CPA lipomas diagnosed in the Department of...
Otorhinolaryngology-Head and Neck Surgery of Hôpital Pitié-Salpêtrière and review 94 cases reported previously in the literature.

Lipomas represented 0.14% of CPA and internal acoustic meatus tumors. Localization was on the left side in 59.9%, on the right side in 37%, and bilateral in 3.1% of the patients. The diagnosis was confirmed radiologically in 33 of 98 patients, surgically in 60 patients, and by autopsy in 5 patients. The most frequent associated symptoms were of cochleovestibular origin, such as hearing loss (62.2%), dizziness (43.3%), and unilateral tinnitus (42.2%). Other associated symptoms involved the facial nerve (9%) or the trigeminal nerve (14.4%). Complete resection was performed in only 32.8% of the patients with frequent cranial nerve involvement. Frequent cranial nerve involvement was seen in 95.4% of all patients. After surgery, patient symptomatology was unchanged in 9.2% of the patients, and 50% were improved; however, new postoperative deficits occurred in two-thirds of the patients. Overall, 72.2% of the patients experienced new postoperative deficits such as hearing loss (64.8%). Preservation of hearing was possible in only 26% of the patients. Only 18% of patients were improved after surgery without any new postoperative deficits.

Preoperative diagnosis of internal acoustic meatus/CPA lipomas is based on magnetic resonance imaging. The aim of surgery in these cases is not tumor removal but cranial nerve decompression or vestibular transection, and surgery is performed only in patients with disabling and uncontrolled symptoms.

1998

17 IAC/CPA lipomas, bringing the total number of documented cases to 84 in 1998. There appears to be a nearly 2:1 male to female predominance. Sixty percent were left-sided lesions, and three were bilateral. Hearing loss, dizziness, and tinnitus were the most common presenting symptoms. Surgical resection was performed in 52 (62%) of these lesions; however, total tumor removal was accomplished in only 17 (33%), which is most likely because of the fact that these tumors tend to have a poorly defined matrix and a dense adherence to neurovascular structures. Sixty-eight percent of patients experienced a new deficit postoperatively, 11% were unchanged, and only 19% improved with no new deficit. Only one documented case of tumor growth was identified; however, the reported follow-up was short (average, less than 3 years).

With the magnetic resonance imaging techniques now available, lipomas can be reliably differentiated from other masses within the CPA and IAC, so histopathologic diagnosis is rarely necessary. Because of the potential for significant morbidity with resection of these lesions, we believe that conservative follow-up is the best treatment option for patients with these rare lesions. Surgery is indicated only when significant progressive or disabling symptoms are present.

Case reports

2012

A 5-year-old boy was evaluated for recurrent primary generalized seizures of 20 days duration. He had preceding headache, vomiting for which he was subjected to a Magnetic resonance imaging (MRI) scan of the brain. Imaging studies revealed a hyperintense mass in the right CPA suggestive of lipoma/epidermoid. He was referred to us for further management.

Basic routine blood investigations were essentially normal. His vital parameters were stable. He was conscious, oriented, and obeying commands. Child was active and had no motor/sensory/cranial nerve deficits. Plantars were bilaterally flexor with normal deep tendon reflexes. MR imaging has now achieved a very high sensitivity and specificity for detecting lipomas. On T1-weighted MR images,
Lipomas typically appear hyperintense compared with brain tissue and hyperintense on T2-weighted MR images. MRI scan of the brain was done and the findings is given below.

Multiplanar, multisequences, MR imaging, including SE T1 axial, flair axial, FSE T2 axial were done. Postcontrast T1-weighted multiplanar sequences were also performed.

A 17mm (trans) ×15mm (AP) × 15mm (CC) well-encapsulated mass lesion noted in the right CP angle which was hyperintense on T1, T2, and FLAIR with inversion on fat suppression sequence. The lesion was situated inferior to the right Vth nerve and indenting upon right lower pons and medulla. The right VIIth and VIIIth nerves and superior cerebellar artery were encased by the lesion. No evidence of tumor extension into IAC was noted. There was neither significant shift nor hydrocephalus. Right cerebello pontine angle tumor–lipoma/epidermoid.

A right retro sigmoid suboccipital approach was chosen to expose the tumor. The lesion was extra-axial, yellowish and surrounding the seventh and eighth nerve complex. Branches of the AICA were embedded in the tumor. The lesion was partially decompressed. Neurovascular structures were preserved. There were no postoperative deficits.

Histopathological evaluation revealed it to be lipoma-right CP angle

These tumors can cause symptoms related to the VIII nerve involvement, such as hearing loss, tinnitus, and vertigo. However, trigeminal symptoms such as neuralgia, paresthesia or headache, can also occur with CPA lipomas extending to the trigeminal cisterns.

This patient presented with headache, and seizures. Neuroimaging revealed a hypodense mass in the right CPA suggestive of lipoma/epidermoid. He underwent surgical exploration and decompression of the tumor. Histopathology confirmed it as lipoma.

2009

A 13-year-old female patient was evaluated due to a 1-year history of headache and hearing loss. The physical examination was unremarkable. The audiometric evaluation demonstrated a discrete sensorineural hearing loss on the right side. The CT scan revealed a markedly hypodense non-enhancing mass in the right CPA. The MR imaging showed a lesion measuring 2.1 × 2.0 × 1.7 cm in the right CPA cistern. The mass was hyperintense on T1-weighted images and isointense with hypointense halo (chemical-shift) on T2-weighted images, with very low signal on T1-weighted images with fat suppression (Figs 1 and 2). The VII and VIII cranial nerves were seen as linear images with low signal inside the CPA mass. The diagnosis of CPA lipoma was suggested and the surgical treatment was chosen once the patient was young and the chance of lesion growing and future complications was considerable. A craniotomy with posterior fossa approach was performed, the lesion was partially removed, and the histological examination confirmed the diagnosis of lipoma. Six months after the surgery the patient remains asymptomatic. The parent signed the informed consent agreeing with the study.

Case 2

A 35-year-old woman presented with a six-month history of vertigo, without significant abnormalities on physical examination. A CT scan revealed a left-sided hypodense non-enhancing CPA mass. The MR imaging showed a left CPA cistern hyperintense lesion on T1-weighted images and isointense with hypointense halo (chemical-shift) on T2-weighted images, measuring 1.4 × 1.3 cm and showing no enhancement after contrast administration (Fig 3). The diagnosis of CPA lipoma was suggested and the patient was managed conservatively. The symptoms were controlled with medical therapy.
follow-up MR imaging performed one year later showed no significant modifications\textsuperscript{10}.

### 1997

The case of an extensive lipoma of the cerebellopontine angle (CPA) represents 0.05\% of all CPA tumors operated on in a department from 1978 to 1996. The lipoma constitutes an important differential diagnosis because the clinical management differs significantly from other CPA lesions. The clinical presentation and management of the presented case are analyzed in comparison to all previously described cases of CPA lipomas. The etiology and the radiological features of CPA lipomas are reviewed and discussed. CPA lipomas are maldevelopmental lesions that may cause slowly progressive symptoms. Neuroradiology enables a reliable preoperative diagnosis. Attempts of complete lipoma resection usually result in severe neurological deficits. Therefore, we recommend a conservative approach in managing these patients. Limited surgery is indicated if the patient has an associated vascular compression syndrome or suffers from disabling vertigo\textsuperscript{11}.

### 1994

Two patients with cerebellopontine angle (CPA) lipoma were studied. They were submitted to surgical treatment. Available literature was reviewed and 29 cases with same lesion were identified which had been treated by surgery. Clinical manifestations, possibility of diagnostic methods, surgical indications and treatment strategies are discussed. Attention is called to the peculiarities of CPA lipomas and the doubtful validity of attempting complete excision in all cases\textsuperscript{12}.


8\textsuperscript{) Bigelow DC, Eisen MD, Smith PG, Yousem DM, Levine RS, Jackler RK, Kennedy DW, Kotapka MJ. Lipomas of the internal auditory canal and cerebellopontine angle. Laryngoscope. 1998
