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# LIPOMA OF THE CEREBELLOPONTINE ANGLE

## CASE REPORTS AND LITERATURE REVIEW

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SUMMARY - 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 vality of attempting complete excision in all cases.

KEY-WORDS: cerebellopontine angle, lipoma, surgery.

**Lipoma do ângulo pontocerebelar: relato de casos e revisão da literatura**

RESUMO - São estudados dois pacientes com lipoma no ângulo pontocerebelar (APC), submetidos a tratamento cirúrgico. A literatura disponível foi revisada, tendo sido identificados 29 casos submetidos a tratamento cirúrgico, com a mesma lesão. As manifestações clínicas, possibilidades dos métodos diagnósticos, indicações cirúrgicas e estratégicas operatórias são discutidas. Chama-se a atenção para as peculiaridades dos lipomas do APC e a discutível validade da tentativa de remoção completa em todos os casos.

PALAVRAS-CHAVE: ângulo pontocerebelar, lipoma, cirurgia.

Intracranial lipomas do not occur frequently. Vonderahe and Niemer[32] observed 4 cases (0.08%) in 5000 routine autopsies. Budka[2] identified a higher incidence (0.3%) in 4290 specimens, only ones of which was symptomatic and located in the cerebellopontine angle (CPA). Zimmerman et al.[35] diagnosed 12 cases (0.08%) in 14000 tomographic exams, 700 of which were intracranial tumors. Until a short while ago, lipomas were classified as neoplasms, congenital in origin[27]. In the International Classification of Tumours[36] they were included among other malformative tumors and tumor-like lesions. Lipomas are usually asymptomatic and generally found at autopsy, CT or MRI scan. The most frequently symptomatic ones are located in the CPA, and, therefore, most often encountered in surgical cases. Despite this, not more than 39 cases are reported in literature, 29 of them found at surgery[1-4,7,8,10-12,15,18,19,21-26,28,29,31,33,34]. In this communication we describe two cases with clinical manifestations of a process located in the CPA, submitted to surgical treatment.

## CASE REPORTS

Patient 1. LN, 28-year-old woman with a history of left-sided facial pain since the age of 5. During this period she was submitted to three tooth extractions, in an attempt to solve the pain. In 1973, she underwent an angiographic examination with normal results. The pain persisted with characteristics of neuralgia of the trigeminal nerve, in territories of the first and second rami. One month before hospitalization the pain intensified, and marked fluctuations occurred in the hearing on the left side, to the point of deafness, associated with tinnitus and spontaneous vertigo. At examination, severe involvement of hearing was found, persistent tinnitus and vertigo, and trigeminal pain affecting the territories of the first and second rami, on the left. X-ray showed a widening of the internal acoustic

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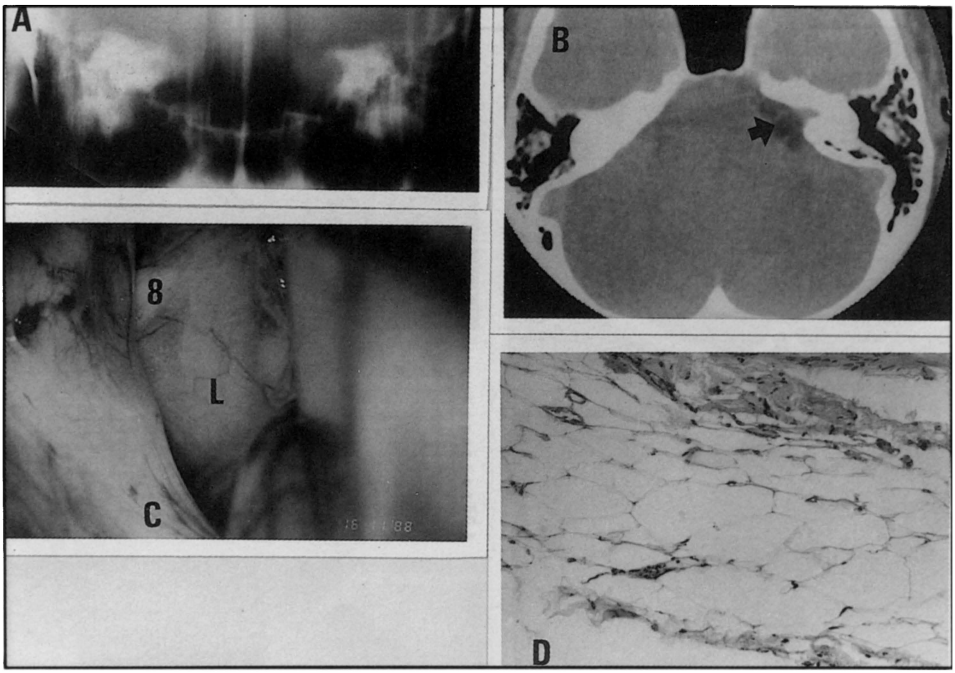


*Figure 1. Patient 1. A, politomography shows enlargement of the internal acoustic meatus and erosion of the petrous bone. B, CT scan shows hipodense lesion in CPA transversed by 7th and 8th cranial nerves (arrow). C, operative field: L, lipoma; 8, 8th cranial nerve. D, photomicrograph of the surgical specimen shows mature fat tissue with thin fibrous septa and eventual nervous filaments (hematoxylin and eosin, x40).*

meatus (IAM) and the amputation of the apex of the petrous bone (Figura 1A). Computed tomography (CT) showed (Figura 1B): in the left CPA, a hypodense lesion with density equal to that of fatty tissue, and approximate dimensions of 2.15/1.3/1.9 cm, causing deformity by extrinsic compression of the apex of the petrous bone, as well as impression on the brain stem, without any displacement of the fourth ventricle. The lesion did not change when intravenous iodine contrast was used. On November 16, 1989, the patient was submitted to left retrosigmoid craniectomy, under general anesthesia, in a seated position. In the cistern of the CPA, a yellowish mass was identified, with a thin translucid capsule which contained nerves eight, seven, six and five and the regional blood vessels (Figura 1C). The capsule was incised, and fragmentary, careful removal of the fatty-looking tissue ensued. The fatty tissue was between trabeculae of fibrous tissue. The nerves and vessels in the CPA were included, but not displaced, by a bleeding fibroadipose mass, which adhered strongly to these structures, thus preventing its complete removal. The fifth, sixth, seventh and eighth nerves were decompressed and the arteries were preserved, despite manipulation. There was strong adhesion of the mass to the brain stem. No abnormalities occured during the postoperative period. At the time the patient was discharged from hospital, she presented deafness, paresis of the sixth and seventh nerves, and intense hypoesthesia and hypoalgesia in the left trigeminal territory. The facial pain and tinnitus had disappeared. Anatomopathological exam: mature fat tissue (Figura 1D). Exams performed 1-3 years after surgery indicated absence of facial pain, disappearance of the paresis of the sixth nerve, slight facial asymmetry and sligth hipoesthesia in the territory of the trigeminal nerve. CT showed a mass with a smaller volume and the same tomographic features.

Patient 2. AA, a 57-year-old man with a complaint of persistent frontal headache, occasional dizziness and diminished hearing on the right side during the last ten years. Examination indicated a very slight facial paresis and hypoacusia on the right and dizziness (Figura 2D). The CT (Figura 2A) showed a mass with density appropriate to fatty tissue located in the CPA, measuring 1.8/1.5/1.7 cm, without enhancement with contrast and or changes in the bone. On October 7, 1992, the patient was submitted to retrosigmoid craniectomy on the right side, under general anesthesia in the seated position. After displacement of the right cerebellar hemisphere, an yellowish mass was identified, with a translucid capsule adhering to the lateral aspect of the inferior portion of the pons and to the medulla including nerves seventh and eighth in the entry zone to the neuroaxis (Figura 2B). The vessels and nerves

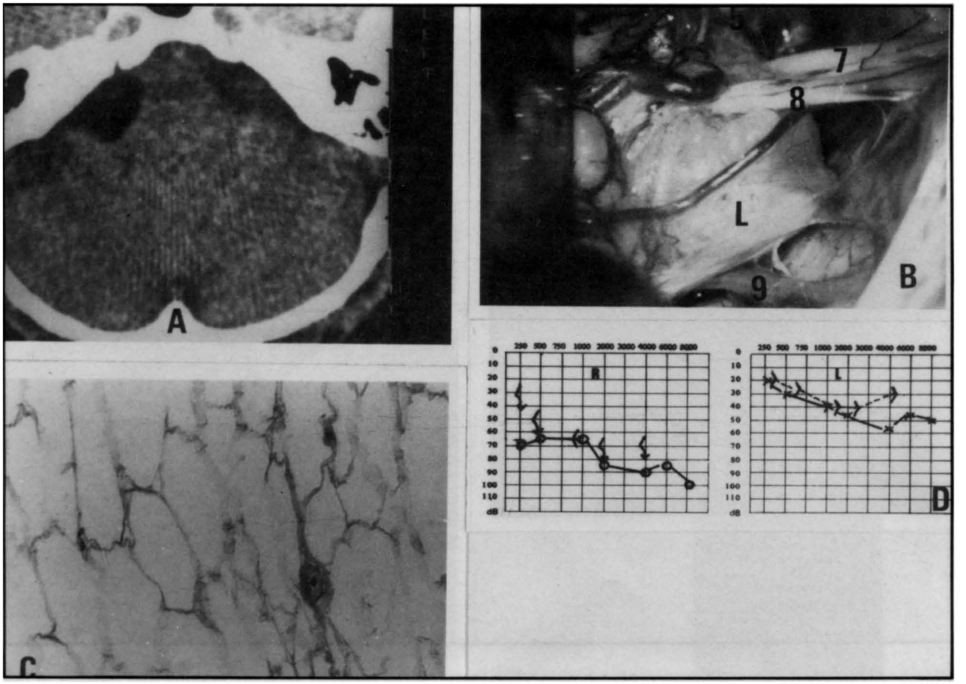


*Figure 2. Patient2. A. CT scan shows hipodense lesion in CPA. B, operative field: 5th (5), 7th(7), 8th(8), 9th(9) cranial nerves and the lipoma (L). C, photomicrograph of the surgical specimen shows mature fat tissue (hematoxylin and eosin, x100). D, right moderate to severe sensorineural hearing loss.*

in the region had not been displaced by the mass. The capsule was incised with fragmentary, partial removal of the tumor. The fatty-looking tissue was trabeculated, and was strongly attached to the neighboring anatomical structures, and this prevented ample removal without lesion. Anatomopathological exam: mature fat tissue (Figura 2C). No abnormalities occured during the postoperative period. Control of auditory function showed the same deficits of the preoperative period, but no more dizziness (Figura 2D).

## COMMENTS

Intracranial lipomas are rare in surgical series. Thomsen[30] did not find any lipoma in 157 cases of surgery of masses in the CPA. The first reports refer to autopsy material, because they rarely cause symptoms. Vonderahe and Niemer[32] identified 4 cases in 5000 autopsies (0.08%). Budka[2], studying 1956 neuropathological autopsies, found 9 cases (0.46%), and in 4290 neurosurgical biopsies found a single case (0.02%) located in the CPA. Matson[17] observed only one case of lipoma in 750 brain tumor surgeries in children. In neuroradiological material, Zimmerman et al.[35] diagnosed 12 lipomas in 14000 tomographic exams (0.08%), and the same number in 700 cases of intracranial tumors (1.7%). Kazner et al.[11] found 11 cases (0.06%) in 17500 patients studied with CT scans, corresponding to 0.34% (11/3200) of the intracranial masses. More recently, Eghwrudjakpor et al.[5], using CT and MRI scans, diagnosed 3 cases of lipoma (0.27%) among 1100 intracranial tumors.

Truwit and Barkovich[31] extensively reviewed the theories described to explain the pathology of the intracranial lipomas. They concluded that intracranial lipomas are not hamartomas nor true neoplasms, but malformations. The findings support the hypothesis that the formation of an intracranial lipoma is the result of the abnormal persistence and maldifferentiation of the meninx, during the development of the subarachnoid cisterns. The authors found an association of congenital anomalies detectable at radiological exams in 24 (60%) of their cases, which reinforces the hypothesis

of a developmental defect. However, no reports of intracranial malformation associated with CPA lipomas are identified in the literature[14].

The most frequent topography of intracranial lipomas is interhemispheric. Truwit and Barkovich[31], studying 42 cases with MRI scans, observed the followings distribution: interhemispheric 45%, quadrigeminal-supracerebellar 25%, suprasellar-interpeduncular 14%, cerebellopontine angle 9% (3 cases) and Sylvian cistern 5%. Maiuri et al.[16], reviewing 200 cases reported in literature up to that time, found the following topographic distribution: pericallosal cistern and corpus callosum 65%, chiasmatic and interpeduncular cistern 13.5%, ambient cistern 13%, cistern of the cerebellopontine angle 6.5% and Sylvian cistern 3.5%. Using the above mentioned data, the possibility of finding a lipoma of the CPA in skull studies using CT scan is 0.004%, and, among the cases of intracranial expansive process diagnosed, 0.02%, agree with Budka's autopsy findings.

Maiuri et al.[16] review literature and find that intracranial lipomas produce symptoms in approximately 42% of the cases; those of the corpus callosum in 50%, those of the ambient cistern in 20%, of the CPA in 80% and the Sylvian cistern in 50%. Lipomas located in the chiasmatic and interpenduncular cisterns have proved asymptomatic. Signs and symptoms caused by CPA lipomas are related to the nerve structures of this region: dizziness, loss of hearing, trigeminal neuralgia and sensory impairment in the distribution of the fifth cranial nerve[1-4,7,8,10,18,19,21-26,28,29,33,34]. In patient 1 there was trigeminal neuralgia during 23 years, fluctuations of hearing and severe hypoacusia, persistent tinnitus and vertigo. In patient 2 there was hypoacusia, vertigo and slight facial dysfunction. Kitamura et al.[12] described 2 cases of fluctuating hearing loss, and 1 case exhibited a Menière-like syndrome. The heat tests or electronystagmography and audiometry may present abnormal results. Mattern et al.[18] in one case observe a delay of Waves I and V, with increased I to V interwave latency in a study of evoked potentials in the brain stem. One case is found in literature of CPA lipoma causing facial hemispasms with a surgical finding[14]. The caudal nerves may be involved[8,26,33], but there are no reports symptoms related to these nerves.

X-ray examination of the skull may demonstrate widening or erosion of the internal auditory canal or defect in the petrous bone[2,7,23,33,34]. In patient 1 there was a widening of the internal acoustic meatus (IAM) and erosion of the apex of the petrous bone. In cases inside the CPA without affecting the IAM, no bone changes are observed (case 2). Pneumoencefalography shows a mass which occupies the cerebellopontine cistern, or a filling defect in the internal acoustic meatus[7]. Cisternography may demonstrate a mass in the CPA[23,33]. Angiography is often normal in CPA lipomas,due to the lack of displacement of the contiguous vascular structures. In case 1, the angiographic study was normal. With routine studies using CT and MRI scans, the number of diagnoses in living patients has increased during the last few years. The tomographic expression of lipomas at CT is a lesion with regular margins, homogenous, with low density and attenuation values between -60 and -200 Hounsfield Units (HU) and no contrast enhancement[4,5,16]. In both cases studied, the tomographic image presented these characteristics. Sometimes it is possible to identify anatomical structures passing through them (Figura 1B). MRI scan shows a high signal on T1 weighted images and low signal on T2 images. The administration of gadolinium does not produce any mass enhancement[4,24], however, Saunders et al.[28] observed enhancement, using gadolinium, in three of his cases. Lesions with a fatty component meriting differential radiological diagnosis in relation to lipomas are epidermoid and dermoid cysts and teratomas. The epidermoid cysts have mean attenuation values of 0 to -20HU. The dermoid cysts usually are less homogeneous masses due to the presence of hairs or calcification. Their mean attenuation values at CT scan (-20 to -80UH) are higher than those of lipomas and their signal on T1 is longer on magnetic resonance[11,13,20,31,35]. Teratomas contain many different tissues, including fat, cartilage, muscle and bone, and very inhomogeneous lesions, which makes it easier to establish differential diagnosis using CT and MRI scans[13,34].

When CPA lipoma is diagnosed, surgery is indicated for cases in which its excision will benefit the patient, or when there is doubt as to diagnosis. This management is related to the great difficulty and occasional impossibility of complete removal. There is a greater tendency to operate on lipomas located in the CPA, considering the symptoms related to the fifth and eighth nerves[3,4,7,23,26,29]. In case 1 the surgical indication was due to persistent, uncontrollable symptons of

trigeminal neuralgia, and, in the second, to progressive loss of hearing. The possible benefícts for the auditory function have not been stressed in the patients who underwent surgery and some authors even suggest biopsy with decompression and partial excision, in an attempt to preserve hearing[2,3,8]. This was the management in case 2. Saunders et al.[28] favor the translabyrinthine approach, since it is impossible to preserve hearing even in small intrameatal masses when the intention is to perform complete removal.

The approachs used, have been the middle fossa approach[23] in intrameatal processes, translabyrinthine[23,24,28] in intrameatal masses and in cases of deafness, in patients treated by an otologist, and the classic retrosigmoid approach by neurosurgeons[1,3,4,10,15,19,22,24-26,28,29]. The surgical aspect of CPA lipomas is typical and can not be confused with any other type of expansive lesion in the area: a yellowish mass with a tenuous translucid capsule containing mature adipose tissue. Special attention should be given to the extent of removal. The involvement and strong adhesion of the lipoma to the anatomical structures of the CPA favor a biopsy or partial removal with decompression of the symptom-related structures. Attempts at complete excision led to the excision of the lipoma with nerve structures inside, or heavy trauma, causing inconvenient postoperative dysfunctions[3,21,24,29,33]. Christensen et al.[3], studying surgical specimens, identified mature adipose tissue traversed by peripheral nerves ranging from individual nerve fibres to medium-sized fascicles surrounded by perineurium. In one case of autopsy, reactive fibrosis and gliosis were identified in the adherent adjacent brain. In the CPA, the lipomatous tissue entraps nerve fascicles and nerves fibres. Olson et al.[23], in intrameatal lesions, achieved complete excision without functional impairment in one case. In patient 1 total removal and broad decompression of the nerves involved was attempted. This caused complete and persistent dysfunction of the eighth and partial dysfunction of the fifth and seventh nerves, besides transient dysfunction of the sixth. In patient 2 a sparing excision and partial decompression of the seventh and eighth nerves was performed without vascular manipulation. The symptoms did not become worse during the postoperative period (Figura 2D). Of 29 patients treated surgically and reported in literature, in 19 craniectomy of the posterior fossa was perfomed, 9 were approached translabyrinthically and one was approached through the middle fossa. In those approached from the posterior fossa, only two[3,25] underwent complete removal. In patients in whom the translabyrinthine and middle fossa approach were used, all had complete excision except one[23,24,28]. The possibility of complete removal without great sequelae depends on the extent of the lesion. Complete excision was most often achieved in the intrameatal ones, which were the smallest.

It is believed that the growth of lipomas is insidious or extremely slow, thus justifying waiting in asymptomatic patients with lesions suggesting CPA lipomas at CT and/or MRI studies. The long duration of symptoms in the cases described, and tomographic review 3 years after surgery in case 1 support this statement.

CPA lipomas are masses which may or not cause symptoms. The review of information found in literature allows the management of asymptomatic patients whose diagnosis was perfomed by CT and/or MRI by waiting. In symptomatic patients, the surgical approach is indicated, taking into account the clinical manifestations and close relationship between the amount of excision and resulting sequelae. We do not agree that systematic indication of the translabyrinthine approach is a good strategy, considering the resulting auditory sequelae.

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