
A Clinical Case Report About an Intracranial Lipoma

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Abstract: Intracranial lipoma is a uncommon congenital malformation, represent less than 0.1% of all intracranial tumors. Result from abnormal persistence and mal-differentiation of meninx primitiva, during embryogenesis. The cases in which the tumor presents an extracranial component are caused by a secondary dehiscence of the skull with evagination of a small tuft of primitive meninges. These lesions are frequently located in the inter-hemispheric fissure in the pericallosal region in 50% of cases, other locations as ambient or quadrigeminal cisterns present an incidence about 20-25%. In half of cases the patients present associate midline brain malformations of varying severity, the most frequent is the agenesis of the corpus callosum. The most accepted theory states that in embryonic phases, mesenchymal alterations at the level of the cranial sutures in the sagittal plane can cause anomalies of the SSS, sinus rectum, tentorium, and parietal bones that would explain the associated anomalies. Majority of intracranial lipomas are detected incidentally, being a third of the asymptomatic cases. As in all other pathologies certain localizations cause specific symptomatology of the involved area. The most common clinical manifestations are headache, epilepsy, mental dysfunctions and cranial nerve deficits. The clinical diagnosis is unclear and it is necessary release complementary exams in every suspected cases. Neuroimaging studies such as computed tomography (CT) or magnetic resonance imaging (MRI) make possible identify these lesions. We describe a clinical case about a 79-year-old woman with headache without any other symptoms. Was done CT and MRI that showed a inter-hemispheric lesion located in the pericallosal region, without any other abnormality associated. The development, during the last decades, of neuroimaging and histogenesis techniques increased the knowledge about this pathology and modified the management in recent times. The surgery is contraindicated in the majority of the cases, however still play a role in some cases especially in presence of hydrocephalus, uncontrollable seizures, and bony affectation with cosmetic influence. The medical control of the epilepsy is mandatory. We present a clinical case about pericallosal intracranial lipoma and review the current literature. Taking in account the clinical situation and the imaging findings, we decide for a conservative approach with clinical/neurological and radiological follow-up.

Keywords: Intracranial Tumor, Lipoma, Congenital Malformation, Computed Tomography, Magnetic Resonance

1. Introduction

Intracranial lipomas (IL) are rare lesions, representing 0.1%-0.5% of all intracranial tumors.

They are congenital lesions composed of adipose tissue resulting from abnormal persistence as well as altered differentiation of the primitive embryonic meninges during the development of the subarachnoid cisterns. The altered differentiation of the meningeal tissue during embryogenesis is its most likely origin.

IL are formed by normal adipose tissue, thus being considered a congenital anomaly rather than a true neoplasm.

The most frequent location is interhemispheric fissure, usually located on the corpus callosum or pericallosal area (50%). These lesions are commonly associated with callosal hypogenesis or agenesis. Lipomas that develop in the interhemispheric fissure prior to fusion of the marginal sinuses to form the Superior Sagittal Sinus (SSS) result in a fenestrated superior SSS. The falcine sinus is a normal accessory sinus that is usually obliterated at birth. It provides alternative venous drainage, important in cases where there is no development of a straight sinus, as occurs in the presence of IL in the posterior pericallosal region. [1]

These lesions can arise with an incidence of 20-25% in

ambients or quadrigeminal cisterns, cerebellopontine cisterns, suprasellar, Sylvian fissure, prepontine cisterns and rarely on the cerebral hemispheres. [2]

Half of IL are related to midline brain malformations. They have variable severity, including hypoplasia/aplasia of the corpus callosum and vascular abnormalities. These alterations include arterial and venous distention, twisting or narrowing, arteriovenous malformations, and aneurysms. [3]

Most IL are detected incidentally, as in our clinical case. Symptoms, if present, are associated with increased intracranial pressure, obstructive hydrocephalus, or seizures. They can manifest with headache, mental dysfunction, and cranial nerve deficits. [3, 4, 5]

Clinical manifestations depends on the location of the lesion and mainly are asymptomatic, represent incidental lesions, discovered on Computed Tomography (CT) or Magnetic Resonance Imaging (MRI), performed for other reason such as head trauma or headaches. The typical findings in both imaging techniques are the presence of a lesion with appearance and characteristics compatible with fat. On CT scan, IL appear as a non-enhancing lesion with uniform fat density, in some cases may exist peripheral calcification. The CT and MRI features are often pathognomonic for intracranial lipomas. [5]

The IL on the CT imaging present as homogeneous and hypoattenuating lesions with fat-density (-70 Hounsfield Unit (HU)), sharply circumscribed.

On MRI these lesions frequently present homogeneous T1 and T2 hyperintensity, as observed in our clinical case.

In the fat saturation images diminish their signal. Other important characteristic in IL is that typically not enhance contrast and the edema is not present around the lesions. The presence of calcification is uncommon, but are described in some clinical cases. [5, 6]

Currently MRI with and without fat saturation, is the gold standard technique for the the diagnosis.

The typical characteristics of the lesions signal in T1 images are strength high, in T2 the images present also signal strength high. In the images T1 with gadolinium the lesions don't present enhancement. The SWI can produce blooming due to susceptibility artifact. High-resolution sequences are the most favorable since lipomas are often crossed by cranial nerves and vessels. The differential diagnosis be do it with intracranial dermoid, in this case if occur a rupture will often have multiple droplets scattered through the subarachnoid space, frequently in the midline.

Should be also include in the differential diagnosis list the intracranial teratoma, lipomatous transformation of neoplasm: PNET, ependymoma and glioma. Other option are rare but should also be include in the differential diagnosis list is epidermoid, that present restriction on DWI. [5, 6, 7]

The treatment are manly conservative, and surgical procedure is indicated in situations with cosmetic defects that require resection of the extracranial portion of the lipoma, obstructive hydrocephalus with indication for a cerebrospinal fluid (CSF) diversion procedure or ventriculostomy, uncontrollable seizures. [7]

When indicated the resection could be extremely difficult and potentially risky and dangerous. Is especially related with the location of the mass and because the huge adhesion with surrounding brain parenchyma, neurological and vascular structures. [8]

2. Case Report

A 79-years-old female who consulted with headaches of 6 months of duration, no other associated symptoms. No seizures and no visual alterations. Clinical and neurological evaluations were unremarkable. Laboratory analysis and electroencephalography yielded normal results. Was released a radiological evaluation with CT and MRI.

A non-contrast CT showed a slightly rounded hypodense lesion with regular edges, supratentorial midline location with slight compression effect of adjacent parenchyma, no other malformations. The lesion was superior to the corpus callosum occupying the entire anteroposterior extension. Peripheral calcifications were not noted, the corpus callosum did not present changes. The lesion was more hypodense than CSF. (Figures 1 and 2).

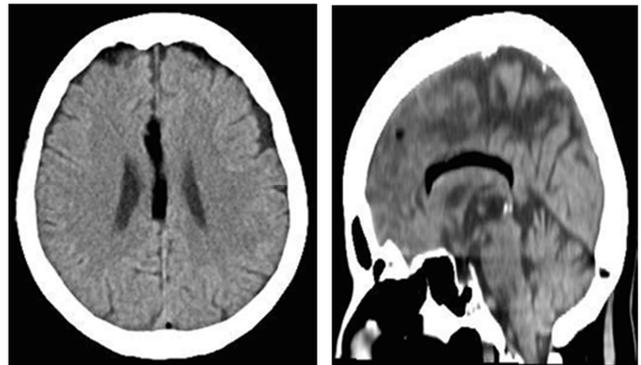


Figure 1. Computed tomography (CT) brain, axial an sagittal slices, showing supratentorial slightly rounded hypodense lesion, located in interhemispheric fissure in pericallous area, without compression effect in adjacent parenchyma.

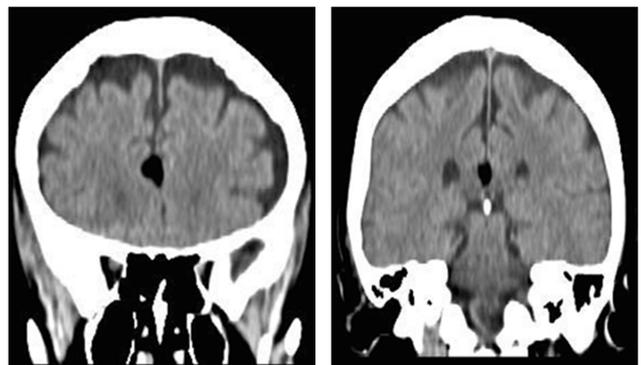


Figure 2. Computed tomography (CT) brain, coronal slices respectively anterior and posterior part of the lesion. showing supratentorial slightly rounded hypodense lesion, midline. Pineal gland with calcifications, frequently in older patients.

The study was complemented with simple and contrasted MRI. Was identified a linear inter-hemispheric lipoma above

the corpus callosum measuring approximately 6 cm longitudinal, diameter 1,2 cm in anterior part and 0,8 cm in posterior part. The lesion located in the supracallosal midline, predominantly on the right, presented hyperintensity on T1 and T2, without surrounding edema. The cerebral parenchyma, cerebellum and brainstem without evidence of significant

lesions. Ventricular system of normal size and morphology, centered in the midline. There are no signs of bleeding and no vascular malformation or other space-occupying lesions. Basal cisterns was free. There is no skull base malformation. Flow in venous sinuses, basilar trunk, and internal carotid arteries (Figures 3, 4).

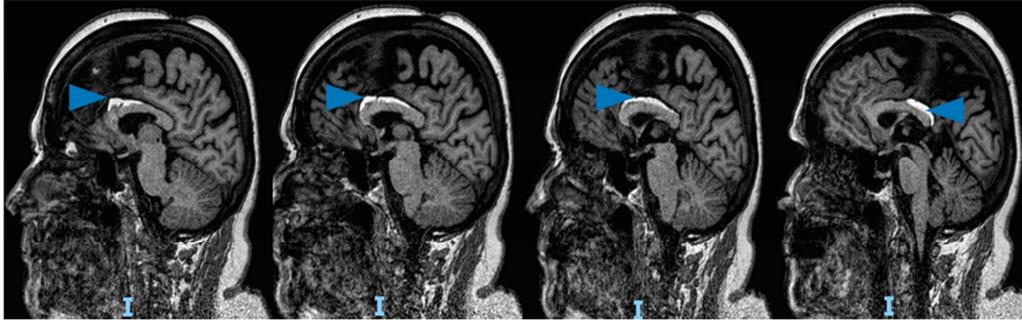


Figure 3. Sagittal T1-weighted MRI, hiperintense lesion in supra-callosal midline (arrowhead). No edema and no mass effect in surrounding parenchyma.

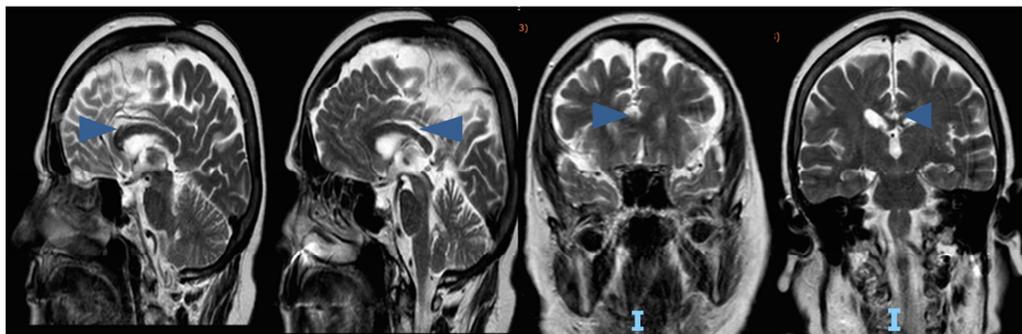


Figure 4. Sagittal and coronal T2-weighted MR image reveals hyperintense pericallosal lipoma (arrowheads).

The Diffusion weighted imaging (DWI) is actually a standard sequence. This sequence permit the analyses of the movement in the extracellular water molecules within tissue. This Brownian motion at a cellular level is evaluated using phased defocusing and refocusing gradients. Apparent diffusion coefficient (ADC) value, obtained from DWI, allows an apparent measure of tissue water diffusion in units of mm²/s per voxel to be achieved.

The relationship of values obtained is inversely proportional, a lower ADC implies a greater restriction. This can be a measure of cellular density, anisotropy and

potentially molecular function.

The advanced MR imaging techniques such as DWI can be potentially useful guiding clinical decisions. This technique can provide improvement in the qualitative and quantitative information on changes at the cellular level, providing data on tumor cellularity and cell membrane integrity.

In gradient echo images (T2wFFE) identified a supratentorial slightly rounded hypointense lesion in interhemispheric fissure laterally in right side. In DWI and ADC images the lesion don't present restriction of the normal diffusion of the water molecules (Figure 5).

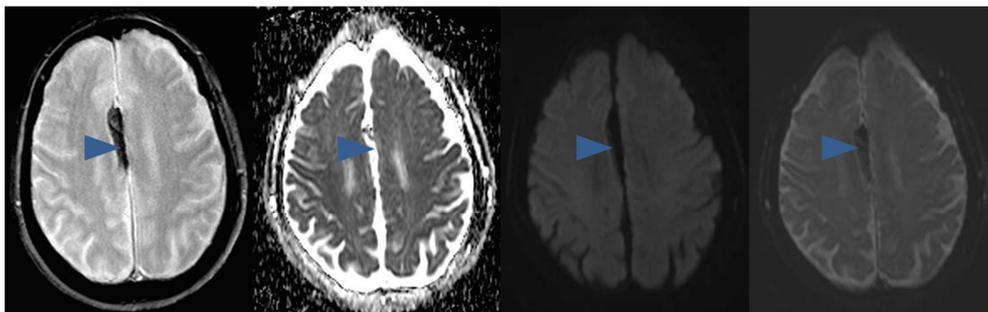


Figure 5. Axial MRI weighted; T2wFFE; DWI ADC MAP; SDWI HR FACTOR1000; SDWIHR FACTOR0 respectively. All images show the midline lesion compatible with Lipoma (arrowheads).

The CT and MRI findings were consistent with a incidentally intracranial lipoma. Was decided maintain

clinical and imagiological follow up. During this period the patients was asymptomatic and free of symptoms particularly seizures. We present a case of intracranial lipoma, a rare tumor, which is the greatest value of our work.

3. Discussion

Intracranial Lipoma (IL) is a rare intracranial tumor. Rokitsansky was the first author to called attention to this lesion in 1856. [9]

Some authors described that IL represent less than 0.1% of all intracranial tumors. [10]

These very uncommon lesions are defined as congenital malformations result from the abnormal persistence and maldifferentiation of the meninx primitiva. The majority of IL occur near the midline, generally are pericallosal, around 55% are accompanied by additional intracranial congenital malformations with varying severity. Most of which are agenesis or dysgenesis of the adjacent structures. The vast majority encountered as incidental findings on imaging studies, symptomatic intracranial lipomas are sometimes observed. [11, 12, 13]

The most frequent clinical manifestations are headache, epilepsy and mental dysfunctions, one third of the cases being asymptomatic. The different locations can cause specific symptomatology of the involved area.

Diagnosis based on symptoms or clinical data needs to be complemented with neuroimaging studies such as computed tomography or magnetic resonance imaging. Nonetheless, the definitive diagnosis as in all pathologies is achieved exclusively with the histological study. [14-16]

Frequently can be identified structural differences at the level of the cell density between benign and malignant lesions.

This study propose is determine the ADC role by comprehensively evaluating DWI and histologic features of a wide variety of pathology proven lesions.

The final goal of the study is identify specifically the added value of ADC mapping to conventional 1.5 T MRI to distinguishing the lesions into benign and malignant. In some cases this two techniques can avoid unnecessary surgical resection and help in management guide. The different conventional sequences are useful for lesion detection. However, the DWI helps to achieve the diagnosis, providing information on the grade and type of the tumor, it also allows monitoring the response to treatment. [17]

Currently DWI should be included in MRI protocols for the diagnostic study of neoplasms. The role of DWI weightings lies in the "functional" data obtained by evaluating the free diffusivity of water molecules in intracellular and intercellular spaces, which in tumors depend mainly on its cellularity. The DWI is a non-invasive tool that should be used routinely in clinical practice and is still a subject of research: it is still used in almost all types of cancer to differentiate malignant from benign lesions, as well as to distinguish different types of cancer, histotypes or tumor grades. It is equally useful for predicting or evaluating responses to treatment. It can also be used to determine the presence of residual or recurrent tumors

during the follow-up period. [17]

Other studies complement the information and evaluate the usefulness of adding DWI with ADC mapping to conventional 3.0-T MRI to differentiate between benign and malignant superficial soft-tissue masses. In all cases was determined the usefulness of this techniques. [18]

Pericallosal lipomas (PCLs) are classified and divided into two groups, anterior and posterior, indicating their location in relation to the corpus callosum. Those that are related to the Genu of the corpus callosum, or anterior, PCLs are tubulonodular in shape, are usually larger than 10 mm, and are commonly associated with various intracranial malformations, including those that affect the corpus callosum itself, such as agenesis. However, those posterior related with the splenium of the corpus callosum are usually elongated, slender, and less than 10 mm. The incidence of malformations of the corpus callosum itself or other intracranial malformations is significantly lower in this group. [19]

Generally, IL are benign masses that do not require surgery, except in cases that present the previously mentioned alterations. Hydrocephalus is a uncommon consequence of these tumors and is usually due to obstruction of the cerebral aqueduct. In our clinical case, even though it was a large lesion, due to its location and the lack of compression on the Sylvius aqueduct or Monro's foramina, no hydrocephalus was observed during the follow-up period. [19]

In case of seizures the electroencephalogram can reveal an epileptic focus associated with IL, anticonvulsant agents proved effective for controlling epileptic seizures.

IL surgical resection is extremely difficult, hazardous and potentially dangerous, however permit decompression and obtain samples for histological diagnosis. [20]

The surgical approach is very rarely indicated because may result in high morbidity and mortality. These tumors frequently present highly vascular nature, with adhesions to vessels and surrounding neural tissue in general and the cranial nerves in particular. Some authors described that in 36% of cases the vessels and nerves coursed through the lesion, the approach and resection of the tumor is more complicated. Other authors reported, small series of cases, with subtotal resecting resulted in complete reversal of the symptoms. Especially in this potential hazardous cases we don't never forget that IL are a benign lesions. [21]

When the IL affect the cranial bone and the surgery solved cosmetic reasons in this cases is recommended obliterate the bony defect by opposing the edges of the pericranium surrounding the defect. Are more common in infancy or childhood. [22, 23]

Usually lipomas do not cause mass effect on surrounding brain tissue, specially in this cases the surgery is unnecessary as also in asymptomatic patients. [24]

As a summary, surgery in the IL is generally contraindicated. We must not forget that ILs are a type of benign lesion. However, the surgical management has an important role in certain cases, especially in cases with hydrocephalus, uncontrollable epileptic seizures, and bone involvement with cosmetic complications. If was detected

seizures, anti-seizure medications are the first line of treatment.

4. Conclusion

Intracranial lipomas rare benign disease and frequently present a slow growing. The symptoms depend of the location of the lesion. The CT and particularly the MRI are the gold standard for the image test diagnosis. The use of complementary techniques such as DWI and ADC mapping complement the traditional techniques. The differential diagnosis must be done with epidermoid cyst, teratoma, craniopharyngioma and epidermoid tumors. The behavior of these different entities is not similar, the prognosis of IL is good even without treatment.

In many cases the the diagnosis is incidental and the pharmacological control is favorable, as described in our work.

The surgical management is infrequent and could result in high morbidity/mortality due to the highly vascularity of these lesion and the high adhesion to the surrounding tissue. In some cases the vascular and nervous structures going thought the lesion.

The surgery should be considered when seizures are related with the tumor and not respond to medical treatment, hydrocephalus caused by the tumor and in those cases with aesthetic changes related to tumor growth.

The development of neuroimaging techniques and the greater understanding of histogenesis have increased knowledge about intracranial lipomas. In this way, its prognosis has been modified and, above all, the surgical indications have been limited to the previously reported cases.

Is recommended maintain the follow up with MRI annually at least in the 5 years following the diagnosis and whenever any symptom related to the presence of the tumor occurs.

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