PERICALLOSAL-INTERHEMISPHERIC LIPOMA ASSOCIATED WITH BILATERAL VENTRICULAR CHOROID PLEXUS LIPOMAS WITH CORPUS CALLOSAL DYSGENESIS: CASE REPORT

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Abstract: BACKGROUND: Pericallosal lipomas (PCL) are rare non-neoplastic lesions, accounting for approximately 0.1% to 0.5% of all intracranial tumors and constitute 50% of all intracranial lipomas. PCL may have calcification and may continue into lateral ventricles. Neurological manifestations, including epilepsy, hemiparesis/hemiplegia, headache, and behavioral disorders, can also occur. Computed tomography (CT) and magnetic resonance image allow the diagnosis of intracranial lipomas with a high degree of reliability.

CASE DESCRIPTION: A five-year-old boy was admitted to the hospital with neurological complaints. Further radiological investigation showed a pericallosal-interhemispheric lipoma associated with choroid plexus lipoma and corpus callosum dysgenesis.

CONCLUSION: Pericallosal lipomas are rare mesenchymal lesions, usually asymptomatic and frequently incidental findings. However, they can progress with varying intensity headaches, signs of focal deficits, and changes in the level of consciousness. Its diagnosis can be established with CT and MRI, and usually does not require interventional methods. Surgery is reserved for symptomatic cases.

Keywords: Choroid plexus lipoma; Intracranial lipoma; Pericallosal lipoma.

INTRODUCTION

Pericallosal lipoma (PCL) was first described by Rokitansky in 1856 [1]. These lesions are rare, benign and consist in a slow growing congenital anomalous maldevelopment that accounts of 0.1 – 0.5% of all primary brain tumor. PCL account for 50% of all intracranial lipomas [2,3,4,5]. Hassan describes that the pathogenesis of a pericallosal lipoma is considered to be the result of an abnormal resorption of the primitive meninges [6].

About half of PCL cases are asymptomatic and they are conservatively managed [10]. An
increase in the recent diagnosis of intracranial lipomas is observed thanks to advances in neuroradiological examination modalities with high-resolution imaging [4,14]. CT and MR findings of intracranial lipomas are very characteristic [3,15,16,17].

CASE REPORT

A five-year-old boy was admitted to the hospital with complaints of aphasia and gait disturbance. Clinical and neurological examination were unremarkable. We performed a radiological evaluation including computed tomography (CT) and magnetic resonance imaging (MRI) scanning.

CT image revealed a hypodense image in the localization of corpus callosum within calcifications, without enhancement after intravenous contrast and associated with dysgenesis of corpus callosum. The lesion was compatible with a lipoma and measured approximately 5.7 x 4.6 x 2.4 cm. Left ventricle was collapsed and the right ventricle was dilated.

Cerebral MRI showed a marked dysmorphism of corpus callosum, highlighting in its topography an oval lipomatous formation with lobulated contours, measuring approximately 5.5 x 3.6 x 2.5 cm, with peripheral lobulated components that project into the lateral ventricles (tubulonodular lipoma); Fig 1. the lesion was characterized by being hyperintense in T1W, T2W and FLAIR, and hypointense to gray matter in T1 Fat-Sat image. As well as CT image, MRI showed the collapsed aspect of left ventricle and the right ventricle dilated and the dysgenesis of corpus callosum. Fig 2. In addition, A3 segments of the anterior cerebral arteries were closely related to the upper border of the interhemispheric lipoma described above.

Follow up MRI, performed 13 months later, did not demonstrate lipoma growth, but the emergence of a pseudoaneurysm, measuring 1.4 cm, in the middle third of the right pericallosal artery, in contact with the upper part of the lipoma. Fig 3.

DISCUSSION

Pericallosal lipoma (PCL) was first described by Rokitansky in 1856. Since that time, numerous reports detailing the clinical, radiographic, and pathologic features have been published [1]. These lesions are rare, benign and consist in a slow growing congenital anomalous maldevelopment that accounts of 0.1 – 0.5% of all primary brain tumor. Usually, they are either midline or adjacent to midline structures. PCL account for 50% of all intracranial lipomas [2,3,4,5].

Hassan describes that the pathogenesis of a pericallosal lipoma is considered to be the result of an abnormal resorption of the primitive meninges. Commonly, this resorption occurs between the eighth and the 10th week of development to create sub-arachnoid spaces. When the primitive meninx persists longer, instead of being resorbed, it differentiates into mature lipomatous tissue. Because of their malformative origin, pericallosal lipoma usually do not become hyperplasic [6].

In 20 to 50% cases, PCL may occur with choroid plexus lipoma (CPL). Generally during development of choroid plexuses, a portion of interhemispheric cistern and tela choroidea will invaginate through choroidal fissure and infolding primitive meninx will attach to developing choroid plexuses [3]. PCL may occur with hypogenesis/agenesis of the corpus callosum in most of the cases (90%) of anterior PCL and some of the cases (30%) of posterior PCL. The association of corpus callosal agenesis with PCL is common, because, maldevelopment of primitive meninx occurs before the interhemispheric commissural fissures develop [7]. They may be also associated with others severe congenital anomalies such
as midline anomalies, dysmorphic changes and some vascular variants [8].

In about 50% of cases the detection of lipoma is incidental [9]. About half of PCL cases are asymptomatic and they are conservatively managed. Surgery is ordinarily not indicated due to high vascularity and close attachment of lipoma to adjacent structures, however surgical intervention may be considered due to associated central nervous system abnormalities or due to pressure on adjacent structure causing symptoms or disability [10]. The most frequently reported symptoms are seizures, headache, mental retardation, emotional lability, and hemiparesis [3,10]. But, mental/behavioral disturbances have also been described [8,11]. Signs of raised intracranial tensions are usually absent due to slow-growing nature of the lipoma [9]. Unlike true neoplasms, they do not multiply; however, they do hypertrophy like other fat cells when patients gain weight but almost never exert mass effect [11].

Truwit and Barkovich described two subgroups of interhemispheric lipomas: curvilinear and tubulonodular. They proposed that the tubulonodular is a result of a more severe insult that occurs at an early embryonic stage and, therefore, interferes with the normal development of the corpus callosum. Commonly, the tubulonodular type is located anteriorly, with the epicenter at the genu in 83% of cases [12,13], and it is associated with high incidence of facial defects, frontal masses, and/or encephaloceles. Curvilinear lipomas are thin and located posteriorly around the splenium. This type is generally associated with normal corpus callosum and has a low incidence or associated anomalies [12].

An increase in the recent diagnosis of intracranial lipomas is observed, with lesions being identified in various locations. Its diagnoses have become possible thanks to advances in neuroradiological examination modalities with high-resolution imaging. Computed tomography (CT) and magnetic resonance imaging (MRI) findings are capable of accurately characterizing the fatty nature of the lesion, allowing for the diagnosis of intracranial lipomas with a high degree of reliability without biopsy confirmation [4,14].

CT and MR findings of intracranial lipomas are very characteristic. On CT, lipomas are sharply demarcated areas of marked hypodensity that do not demonstrate enhancement after intravenous contrast. They usually have CT density between −40 HU and −100 HU. Calcification is often present in interhemispheric lipomas, most commonly within the fibrous capsule surrounding the lipoma. The calcification may be curvilinear, extending around the periphery of the lipoma, or may be nodular within the center of the lesion. The MR appearance of a lipoma is that of a hyperintense mass on T1-weighted sequences, becoming hypointense on long-TR images as the TE increases. On fast saturation pulse sequences lipomas are isointense to gray matter [15]. They show a moderately high signal rate in a T2-image [16,17] as well as FLAIR image [3].

**CONCLUSION**

Pericallosal lipomas are rare mesenchymal lesions, usually asymptomatic and frequently incidental findings. However, they can progress with varying intensity headaches, signs of focal deficits, and changes in the level of consciousness. Its diagnosis can be established with CT and MRI, and usually does not require interventional methods. Surgery is reserved for symptomatic cases.
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