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Case Report

Association of limited dorsal myeloschizis and corpus callosum lipoma: A case report and literature review

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ABSTRACT

Background: Intracranial lipomas are a rare clinical entity. These lesions are frequently asymptomatic and originate in the pericallosal area. As they are fat-containing lesions which are intimately attached to the surrounding structures, surgery is not recommended. In some individual reports, subtotal resection is recommended to lessen complications. There have been no previous reports of corpus callosum lipoma (CCL) associated with limited dorsal myeloschizis (LDM).

Case Description: We describe the case of a combination of CCL and bilateral choroid plexus lipoma discovered incidentally during the investigation of LDM in a 3-month-old male child. Given the asymptomatic behavior of the lipoma and the vascular elements of the pericallosal area, it was decided to monitor it regularly. Thus, the patient underwent surgery only for LDM. Histological examination confirmed the diagnosis, and postoperative follow-up 1 year after showed good evolution. To the best of our knowledge, this association has never been described in the literature.

Conclusion: This case suggests a possible developmental relationship between CCL and spinal dysraphism.

Keywords: Corpus callosum, Dysraphism, Intracranial, Limited dorsal myeloschizis, Lipoma

INTRODUCTION

Intracranial lipomas (ICLs) are rare fat containing lesions, generally considered a congenital malformation. The first reported case was documented by Rokitansky in 1858.[4,21,24,28] These lesions are typically diagnosed on fetal sonogram, but many are found incidentally following a traumatic event. They represent <0.5% of cerebral tumors, even though they are considered heterotopic or congenital malformations. The most common location is the pericallosal area as well as the quadrigeminal cistern. In most cases, patients are asymptomatic. In this study, we report the incidental discovery of a corpus callosum lipoma (CCL) during the investigation of limited dorsal myeloschizis (LDM) in a young child. To the best of our knowledge, this association has never been described in the literature.

CASE REPORT

A 3-month male child, born term, and youngest of three siblings, fully vaccinated, and of nonconsanguineous parents presented with a lumbosacral mass. This was a well-monitored single fetal pregnancy carried to term without any incident, admitted for a bluish and rounded lumbosacral mass, painless, and of regular contours, measuring approximately 4 cm [Figure 1]. There was no sign of intracranial hypertension, such as full fontanelle, emesis, lethargy, hypotonia, or breastfeeding refusal. Clinical examination showed a conscious and reactive infant with good psychomotor development. The anterior fontanelle was not tense, and the head circumference was at 38 cm (−2 standard deviation). There were no other apparent cutaneous malformations. An initial cerebral computed tomography (CT) scan was performed, which demonstrated a large CCL associated with bilateral choroid plexus lipoma without evidence of hydrocephalus or Chiari malformation [Figure 2]. Complementary cerebral magnetic resonance imaging (MRI) showed polymicrogyria with hypertense T1 and fluid-attenuated inversion recovery (FLAIR) behavior of the described lesion. Radiological features on spinal MRI were consistent with LDM [Figures 3 and 4]. Given the asymptomatic behavior of the lipoma and the vascular elements of the pericallosal area, it was decided to monitor it regularly. Thus, the patient underwent surgery only for LDM. Histological examination confirmed the diagnosis, and postoperative follow-up 1 year after showed good evolution.

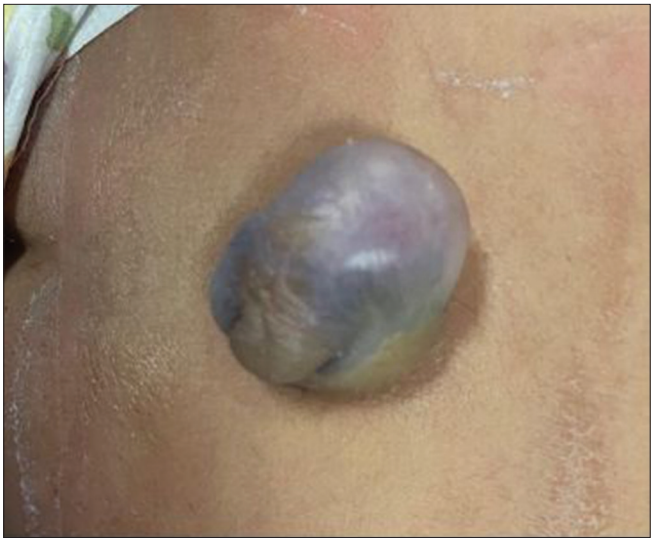

Figure 1: Illustration of the lumbosacral mass.

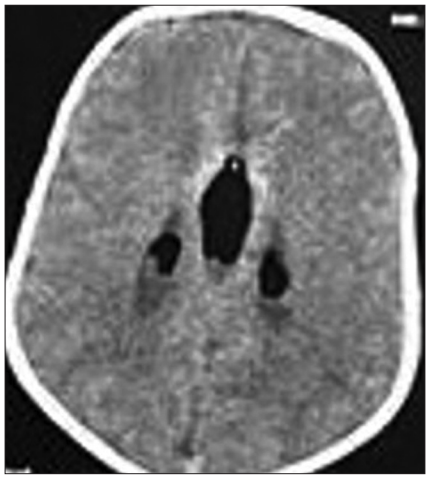

Figure 2: Illustration of the computed tomography scan showing the combination of corpus callosum lipoma and bilateral choroid plexus lipoma associated with hypogenesis of the corpus callosum.

DISCUSSION

ICLs are very rare congenital abnormalities representing <0.5% of cerebral lesions. They are often found in the pericallosal area (40%). However, other locations have been reported in the literature, such as the Sylvian fissure, prepontine cistern, Galen vein system, craniocervical junction, and cerebral convexity.[3,5,6,8,12,14,21,27-29]

As most ICLs are asymptomatic, they are incidentally discovered when assessing for a traumatic brain injury. Otherwise, clinical presentation is mostly related to location. CCL might cause headaches, seizures, psychomotor disorders, or interhemispheric disconnection, resulting in dyspraxia.

Two types of CCL are described in the literature: curvilinear and tubulonodular. While curvilinear lipomas are thin, slender shaped, and seem to be posterior along the splenium, tubulonodular lesions are thick (more than 2 cm), tubular shaped, and localize more often anteriorly. They are also frequently associated with cerebral abnormalities, mainly hypogenesis or agenesis of the corpus callosum (CC). Yilmaz *et al*. noticed four cases of CC hypoplasia within ten patients presenting CCL. Three of them had curvilinear lesions, which challenged this idea.[2,13,20,29]

On CT scans, CCL have a fat density and appears homogeneous with no enhancement when injecting the contrast product. When they are calcified, they can be misdiagnosed and taken for a dermoid cyst or teratoma. On the MRI, these lesions are hyperintense on T1 sequences and FLAIR, hypointense on T2, and fat suppression images.[7,9-11,26,29]

Most CCL is unique asymptomatic slow-growing benign lesions. However, they usually include neurovascular components justifying why surgery is generally avoided except in symptomatic patients. In these cases, subtotal resection is recommended for less morbimortality. On the other hand, the planning of the approach depends essentially on the location and dimensions of the lipoma.[2,27,29]

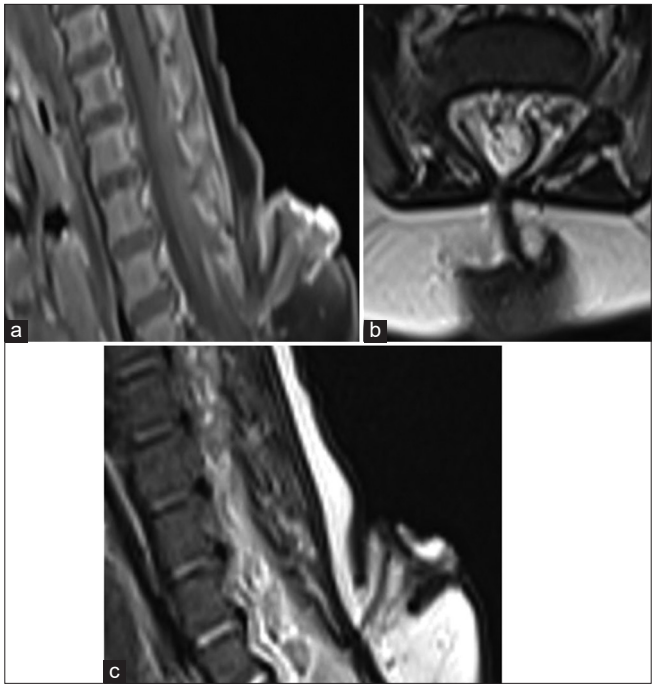

Figure 3: Illustration of (a) sagittal T1, (b) axial, and (c) sagittal T2 spinal MRI spinal magnetic resonance imaging showing limited dorsal myeloschizis.

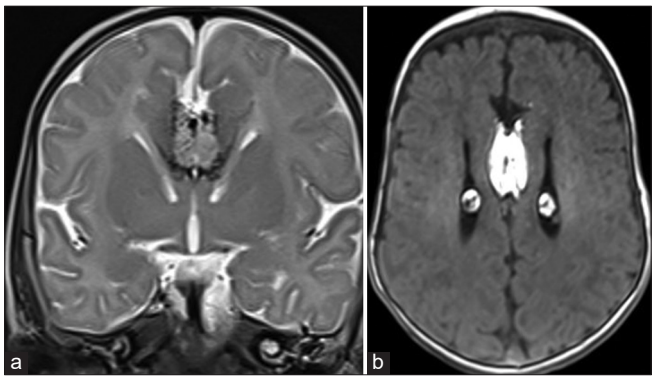

Figure 4: Illustration of (a) coronal T2 and (b) axial FLAIR cerebral MRI showing lipoma located in the pericallosal area and choroid plexus.

CCL can occur as a part of complex malformations such as Pai syndrome or encephalocraniocutaneous lipomatosis. Our patient does not meet major criteria, making these diagnoses less probable. Another pathology that could have been considered is familial multiple lipomatosis, also called symmetrical multiple lipomatosis or Madlung's disease. However, it preferentially occurs in men in the 3rd decade and has never been described in children. In addition to that, it frequently has a metabolic component resulting in systematic signs and other criteria missing in our patient. Considering polymicrogyria associated with hypogenesis of the CC, Aicardi syndrome was also a possible diagnosis. Nevertheless, it primarily affects females. Moreover, the classic triad, as well as the modified criteria necessary to make the diagnosis, are missing [Table 1].[16,17,25]

Table 1: Conditions associated with lipomas.

Syndrome	Associated abnormalities
Pai syndrome	Facial and nasal mucosa polyps, median cleft lip
Encephalocraniocutaneous lipomatosis	Psychomotor disorders, seizures, subcutaneous unilateral tempo-frontal lipomatosis, ipsilateral cerebral and leptomeningeal lipomas, calcifications, and extracranial lipomas
Familial multiple lipomatosis	Multiple subcutaneous lipomas

The association of CCL and spinal dysraphism has been described in the literature. Aggarwal *et al*. described the only case of lipomeningocele associated with interhemispheric lipoma. This suggests a possible continuum in the etiopathogenesis of ICL and spinal dysraphism.[1]

In fact, the pathogenesis of CCL is not fully elucidated. A lot of theories have been advanced. The most accepted one is that they originate from abnormal differentiation of mesenchymal cells of meninx primitive. Other authors evoke a defective fusion of the ectoderm when recovering the mesoderm during neural development. Considering LDM is the result of incomplete disjunction between cutaneous and neural ectoderms, we believe that this case supports this theory and suggests a possible developmental relationship between CCL and LDM.[18]

On the other hand, during the 9th week of gestation, the cells from lamina reunians (LRs) migrate rapidly to the sulcus medianus telencephalic medium (SMTM), allowing the CC to grow anteroposteriorly, with the constitution of the genu first followed by the body and the splenium. CCL interferes with the fusion of LR and SMTM, resulting in hypogenesis or agenesis of the CC.[16]

CONCLUSION

CCL could be part of complex malformations. It is usually discovered incidentally. Association with spinal dysraphism is very rare. A good understanding of the pathophysiology of each entity is the key to comprehend the developmental relationship leading to this association. Subtotal resection of CCL is recommended in symptomatic patients for less morbimortality.[15,19,22,23]

Ethical approval

The Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

REFERENCES

1. Aggarwal N, Gehlot KB, Kumar SD, Khan NK. Frontal subcutaneous lipoma associated with interhemispheric lipoma, lipomeningocele, and corpus callosal dysgenesis in a young adult: CT and MRI findings. Indian J Radiol Imaging 2018;28:22-6.
2. Ben Elhend S, Belfquih H, Hammoune N, Athmane EM, Mouhsine A. Lipoma with agenesis of corpus callosum: 2 case reports and literature review. World Neurosurg 2019;125:123-5.
3. Britt PM, Bindal AK, Balko MG, Yeh HS. Lipoma of the cerebral cortex: Case report. Acta Neurochir (Wien) 1993;121:88-92.
4. Clarici G, Heppner F. The operative approach to lipomas of the corpus callosum. Neurochirurgia (Stuttg) 1979;22:77-81.
5. Douira-Khomsi W, Jhaouet I, Lahmar L, Louati H, Ben Hassine L, Ammar I, *et al*. A rare cause of seizures in infants: sylvian fissure lipoma. Arch Pédiatr 2016;23:867-8.
6. Duman O, Goksu E, Akyuz M, Haspolat S, Senol U. Brain stem lipoma presenting as paroxysmal headache with autonomic features. Headache 2004;44:105.
7. Erbay MF, Tecellioglu M. A pericallosal lipoma case with abnormal vasculature mimicking arteriovenous malformation. Neurol India 2019;67:1331.
8. Fandiño J, Bermúdez J, Arán E. Quadrigeminal cistern and calcarine fissure lipoma: Case report and review of the literature. Neurocir Astur Spain 2005;16:173-6.
9. Fu Y, Fang X, Li Y, Zhao X. Teaching neuroimages: Pericallosal curvilinear lipoma. Neurology 2019;93:e212-3.
10. Georgy BA, Hesselink JR, Jernigan TL. MR imaging of the corpus callosum. AJR Am J Roentgenol 1993;160:949-55.
11. Jerbi Omezzine S, Fkih Hassen S, Ben Rhouma K, Younes S, Hamza HA. Imaging of central nervous system lipoma. J Neuroradiol 2014;41:41.
12. Jerbi S, Chouchane N, Salem AB, Farik S, Zayeni O, Hamza HA. Rare locations of head and neck lipoma. J Neuroradiol 2017;44:114.
13. Jiménez Caballero PE. Interhemispheric lipoma associated with agenesis of the corpus callosum. Neurol Barc Spain 2012;27:515-7.
14. Kannuki S, Okajima K, Sato K, Kusaka K, Matsumoto K. Intracranial lipoma of the temporal lobe--report of a case and review of the literature. No Shinkei Geka 1986;14:379-84.
15. Kim TH, Joh JH, Kim MY, Kim YM, Han KS. Fetal pericallosal lipoma: US and MR findings. Korean J Radiol 2002;3:140-3.
16. Nunes JC, Martins RF, Bastos A, Claudino LS, Guarnieri R, Lima DC, *et al*. Brain lipoma, corpus callosum hypoplasia and polymicrogyria in familial multiple lipomatosis. Clin Neurol Neurosurg 2013;115:1157-9.
17. Olivero F, Foiadelli T, Luzzi S, Marseglia GL, Savasta S. Pai syndrome: A review. Childs Nerv Syst 2020;36:2635-40.
18. Pang D, Zovickian J, Oviedo A, Moes GS. Limited dorsal myeloschisis: A distinctive clinicopathological entity. Neurosurgery 2010;67:1555-79; discussion 1579-80.
19. Pashaj S, Merz E. Detection of fetal corpus callosum abnormalities by means of 3D ultrasound. Ultraschall Med 2016;37:185-94.
20. Popa RT, Feier D, Fufezan O, Blaga L. Interhemispheric lipoma associated with agenesis of corpus callosum in an infant: Case report. Med Ultrason 2010;12:249-52.
21. Seidl Z, Vaneckova M, Vitak T. Intracranial lipomas: A retrospective study. Neuroradiol J 2007;20:30-6.
22. Sherer DM, Onyeije CI. Prenatal ultrasonographic diagnosis of fetal intracranial tumors: A review. Am J Perinatol 1998;15:319-28.
23. Shinar S, Lerman-Sagie T, Telleria ME, Viñals F, García R, Quiroga H, *et al*. Fetal pericallosal lipomas - clues to diagnosis in the second trimester. Eur J Paediatr Neurol 2018;22:929-34.
24. Sms Q, Iqbal Km H. Incidental detection of intra-cranial lipoma in patient with quadriparesis. J Clin Diagn Res 2014;8:MD06-7.
25. Szeto C, Tewfik TL, Jewer D, Rideout A. Pai syndrome (median cleft palate, cutaneous nasal polyp, and midline lipoma of the corpus callosum): A case report and literature review. Int J Pediatr Otorhinolaryngol 2005;69:1247-52.
26. Taydas O, Ogul H, Kantarci M. The clinical and radiological features of cisternal and pericallosal lipomas. Acta Neurol Belg 2020;120:65-70.
27. Ventura E, Ormitti F, Crisi G, Sanna M, Bacciu A. Bilateral cerebellopontine angle lipomas. Auris Nasus Larynx 2012;39:103-6.
28. Yildiz H, Hakyemez B, Koroglu M, Yesildag A, Baykal B. Intracranial lipomas: Importance of localization. Neuroradiology 2006;48:1-7.
29. Yilmaz MB, Genc A, Egemen E, Yilmaz S, Tekiner A. Pericallosal lipomas: A series of 10 cases with clinical and radiological features. Turk Neurosurg 2016;26:364-8.

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