Case report

Medical Ultrasonography
2010, Vol. 12, no. 3, 249-252

Interhemispheric lipoma associated with agenesis of corpus callosum in an infant: case report

Roxana Tania Popa[1], Diana Feier[1], Otilia Fufezan[2], Ligia Blaga[3]

[1] Emergency County Hospital Cluj,
[2] 3rd Pediatric Clinic, Cluj-Napoca,
[3] 2nd Gynecology Clinic Cluj-Napoca

Abstract

Agenesis of the corpus callosum is an anomaly that may occur as isolated or in association with other central nervous system or systemic malformations. We report the case of an infant antenatal diagnosed with ventriculomegaly referred in the postnatal period to our department for imaging evaluation. Ultrasonography showed the absence of the corpus callosum and an interhemispheric lesion highly suggestive for a cerebral lipoma. The diagnosis was confirmed through MRI.

Keywords: cerebral lipoma, agenesis of corpus callosum, ultrasonograpyh, infant

Rezumat

Agenezia de corp calos este o malformație care poate aparea izolată sau in asociație cu alte anomalii ale sistemului nervos central. Prezentăm cazul unui sugar diagnosticat antenatal cu ventriculomegalie. Evaluarea postnatală efectuată ecografic in serviciul nostru a evidențiat lipsa corpului calos si o leziune interemisferică sugestivă pentru un lipom cerebral. Diagnosticul a fost confirmat de investigația prin rezonanță magnetică.

Cuvinte cheie: lipom cerebral, agenezie de corp calos, ultrasonografie, sugar

Introduction

Intracranial lipomas (ICL) are rare intracranial lesions. They represent a group of congenital malformations of the brain parenchyma, mainly occurring in the region of the corpus callosum. ICL are associated with other parenchymal or brain vascular malformations in up to half of cases. Surgical removal is not recommended because of high complications rate and benign course of these lesions.

The most frequent intracranial abnormality associated with ICL is the agenesis of corpus callosum (ACC). Normally, the corpus callosum begins to develop at about 12 weeks of gestation and can be sonographically appreciated by 18 to 20 weeks of gestation. Since ACC usually occurs in combination with other malformations, children with ACC mostly present with intellectual retardation, seizures, hydrocephalus and cerebral palsy. Isolated ACC mostly results in pleiotropic, rather mild clinical manifestations that are mostly recognized at school age but is rarely entirely asymptomatic.

Received 05.06.2010 Accepted 25.06.2010
Med Ultrason
2010, Vol. 12, No 3, 249-252
Address for correspondence: Roxana Tania Popa
Emergency County Hospital,
1-3 Clinicilor str
400006, Cluj-Napoca, Romania
e-mail: roxanatania@yahoo.com

Case report

A 12 day old male infant was referred to our department for sonographic evaluation of antenatal diagnosed ventriculomegaly.

The patient was born on term, by C-section, with a body weight of 4200 g and an Apgar score of 9. The neurological exam showed no abnormalities. No seizures were reported and no facial dysmorphism.

Cerebral ultrasound examination with a SA 8000 EX machine, using a microconvex probe (4-9 MHz) revealed colpocephaly: the asymmetric dilatation of the occipital and temporal horns of the lateral ventricles, more expressed on the right in this case, and frontal horns of

normal caliber, but more separated from the midline than normal. The corpus callosum was completely absent.

A slightly inhomogeneous, echogenic mass of 4.5 x 3 x 2,5 cm was depicted on the midline, centered on the interhemispheric fissure, situated anterior in the callosal region. The mass showed posterior acoustic attenuation (fig 1, fig 2, fig 3). At color Doppler examination blood vessels were depicted mainly in the posterior region of the mass (fig 4).

The ultrasound exam conclusion drawn was of cerebral lipoma, tubulonodular form, associated with agenesis of corpus callosum.

These findings persisted unchanged at follow-up over the next couple of months.

MRI confirmed the diagnosis. The interhemispheric lesion showed signal intensity of fat on all sequences: markedly hyperintense signal on T1 weighted images, with signal attenuation on fat suppression sequences and high signal, but marginally lower than CSF on T2 weighted images with signal attenuation on fat suppression sequences but not on FLAIR (fig 5).

Angio-MRI showed no cerebral vascular malformations.

The patient was referred to the pediatric neurology department for observation and conservative treatment.

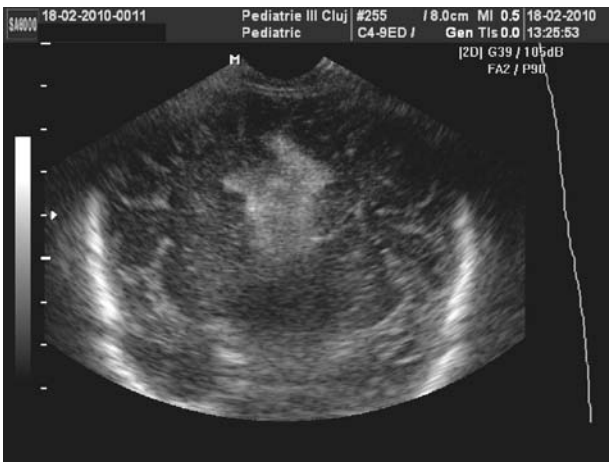


Fig 1. Ultrasound cerebral scan – coronal anterior view shows a hyperechogenic mass in the interhemispheric area with posterior acoustic enhancement.

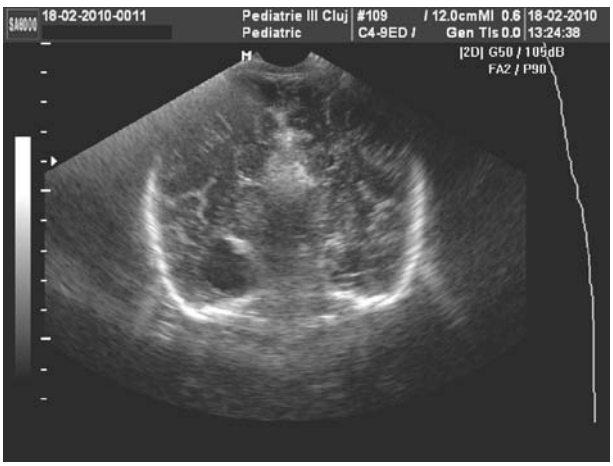


Fig 2. Ultrasound cerebral scan – coronal view slightly posterior than the previous image reveals that the distance between the anterior part of the lateral ventricles is larger than normal and shows the dilatation of the temporal horns, more expressed on the right side.

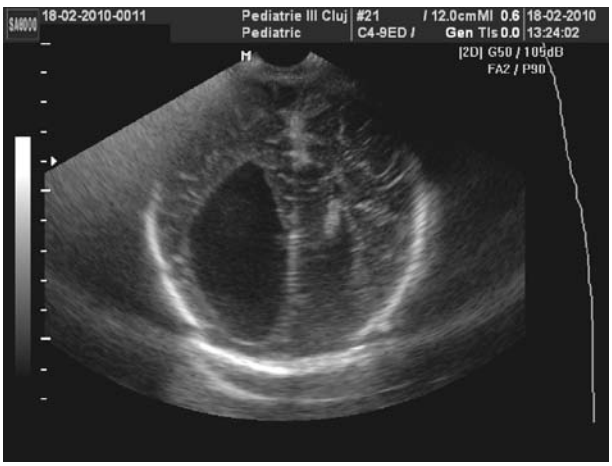


Fig 3. Ultrasound cerebral scan – coronal posterior view depicts asymmetric enlargement of the occipital horns, especially the left one (colpocephaly).

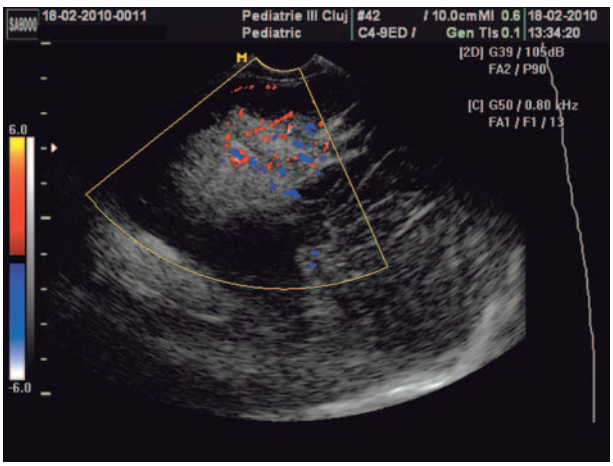


Fig 4. Ultrasound color Doppler cerebral scan – median sagital view shows the absence of the corpus callosum, the interhemisferic echogenic lesion (lipoma) with vessels inside, especially in the posterior area.

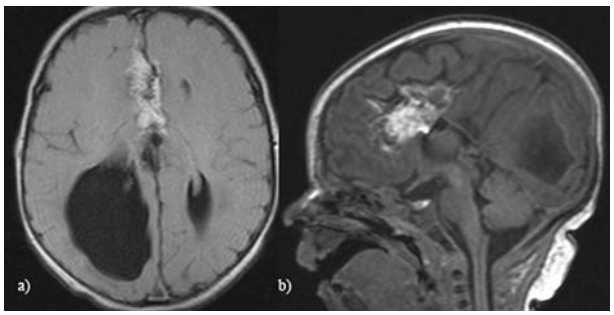


Fig 4. MRI: a) T1 Sagital image at the midline level demonstrates the absence of the corpus callosum and the high signal intensity lesion; b) Axial FLAIR reveals colpocephaly (mainly on the right) and intehemispheric high signal intensity mass (lipoma).

The outcome was favorable, without neurological symptoms in the first year of life.

Discussion

ICL are infrequent Central Nervous System (CNS) lesions with an incidence that ranges between 0.08 to 0.2% of autopsied cases and between 0.06 to 0.3% among those found during imaging, but according to others this frequency can rise up to 0.4% [1].

According to the World Health Organization these lesions are considered grade I mesenchymal, non-meningothelial tumors. Their origin is not clear and the idea of a primary malformation is still accepted. In fact, their origin may arise in the abnormal persistence and maldiffirentiation of the primitive meninx during the development of the subarachnoid cisterns. This theory is concurrent with the fact that both vessels and nerves course through the lipoma, instead of being displaced by it [2].

So far ICL have been divided into two groups, each with different morphologies and associated brain anomalies. The tubulonodular ones consist of anteriorly situated round or cylinder-shaped lipomas, generally greater than 2 cm in diameter. They have high association of corpus callosum dysgenesis, frontal lobe anomalies, and frontal encephaloceles. The curvilinear lipomas are thin, posteriorly situated, curving around the splenium and are less than one centimeter in size. The curvilinear lipomas are generally associated with a normal corpus callosum and otherwise have a low incidence of associated anomalies [3].

Diagnosis accidentally occurs during diagnostic procedures in case of an encephalic disorder. Ecographic prenatal diagnosis is possible at 26 weeks of gestation. Ultrasound demonstrates the characteristic appearance of fat: a hyperechoic midline mass in the region of the corpus callosum, with posterior acoustic attenuation.

CT demonstrates fat density mass (-80 to -110HU). Additionally the tubulonodular variety may demonstrate peripheral curvilinear calcification sometimes referred to as the bracket sign on coronal reformatted images.

The anterior cerebral vessels can be seen coursing through or above the mass, and may have associated vascular malformations or aneurysm formation. CTA may thus be indicated.

MRI is the modality of choice to fully characterize the extent of the lipoma, but also the frequently associated agenesis / dysgenesis of the corpus callosum. ICL follow the signal intensity of fat on all sequences. Again, as vascular abnormalities are associated with these lesions, careful examination of the vessels is essential (best seen on T2 FSE sequences).

ICL are usually asymptomatic. Although several reports mention seizures as an associated or presenting feature, this may only represent a higher incidence of intracranial abnormalities compared with the general population. Surgical removal is not recommended because of high complications rate and benign course of these lesions, except shunt operation for hydrocephaly if needed [4].

The corpus callosum is a white matter structure located in the midline and composed of fibers that connect both cerebral hemispheres. The estimated prevalence of ACC is 0.3–0.5% in the general population and 2.3% in developmentally disabled individuals [5].

The development of the corpus callosum begins during the fifth week of fetal life with the formation of the primitive lamina terminalis, which thickens to form the commissural plate. The development takes place from anterior to posterior with the successive formation of the genu, rostrum, body and splenium. The cavum septum pellucidum forms at the same time [6].

Depending on the time of injury during pregnancy, the anomalies vary from complete to partial agenesis of the corpus callosum.

ACC may be an isolated finding; however, it is frequently associated with other malformations and genetic syndromes including chromosomal aberrations and inborn errors of metabolism. Associated central nervous system (CNS) abnormalities include interhemispheric cyst, Chiari malformations, anomalies of neuronal migration including lissencephaly, schizencephaly, pachygyria and polymicrogyria, encephaloceles, Dandy-Walker malformations, holoprosencephaly, and olivopontocerebellar degeneration. Extracranial malformations include abnormalities of the face and of the cardiovascular, genitourinary, gastrointestinal, respiratory, and musculoskeletal systems.

The diagnosis can be suspected with intrauterine fetal sonography, when colpocephaly and the absence of the septum pellucidum between the 17th and 20th week of gestation is seen [6].

The third ventricle is usually dilated and dislocated towards the interhemispheric fissure. In coronal sections, it is not possible to visualize the corpus callosum. ACC is better documented in coronal and sagittal sections of the fetal brain; therefore, it may be necessary to perform a transvaginal examination.

MRI can be performed both antenatal and postnatal and clearly demonstrates the exact extent of callosal dysgenesis.

There are currently no specific medical treatments for callosal disorders, but individuals with ACC and other callosal disorders may benefit from a range of developmental therapies, educational support, and services [7].

A possible pathophysiological explanation of the association of cerebral tubulonodular lipoma with ACC is the persistence of meninx at the site of lamina reuniens, interfering with the formation of the massa comissuralis. Depending on the timing when this process occurs, the corpus callosum may not develop at all. Furthermore the maldifferentiation of this persistent meninx evolves into either a lipoma of the cistern of the lamina terminalis or a bulky anterior interhemispheric lipoma [2].

In conclusion we presented the case of two rare, but frequently associated cerebral malformations, ICL and ACC, in an infant without clinical abnormalities or neurological disorders.

References

1. Fandino J, Bermudes J. Lipoma de la cisterna cuadrigemina y de la cisura calcarina: caso clinico y revision de la literatura. Neurocirugia 2005; 16:173–176.
2. Truwit CL, Barkovich AJ. Pathogenesis of intracranial lipoma: an MR study in 42 patients. AJR Am J Roentgenol 1990; 155:855–864.
3. Tart RP, Quisling RG. Curvilinear and tubulonodular varieties of lipoma of the corpus callosum: an MR and CT study. J Comput Assist Tomogr 1991; 15:805–810.
4. Maiuri F, Cirillo S, Simonetti L, De Simone MR, Gangemi M. Intracranial lipomas: diagnostic and therapeutic considerations. J Neurosurg Sci 1988; 32:161–167.
5. Jeret JS, Serur D, Wisniewski K, Fisch C. Frequency of agenesis of the corpus callosum in the developmentally disabled population as determined by computerized tomography. Pediatr Neurosci 1985–1986; 12:101–103.
6. Anca IA. Ecografia transfontanelara la nou-născuți și sugari. Ed Medicală, București 2007.
7. Ramelli G, Zanda N, Wyttenbach M, Bronz L, Schnider A. The prognosis of agenesis of the corpus callosum might mostly be favourable. Swiss Med Wkly 2006; 136:404–405.