

# Interhemispheric Lipoma, Callosal Anomaly, and Malformations of Cortical Development: A Case Series

Tetsu Niwa<sup>1,2</sup> Linda S. de Vries<sup>3</sup> Gwendolyn T. R. Manten<sup>3</sup> Maarten Lequin<sup>4</sup> Inge Cuppen<sup>5</sup>  
Jun Shibasaki<sup>6</sup> Noriko Aida<sup>2</sup>

<sup>1</sup> Department of Radiology, Tokai University School of Medicine, Shimokasuya, Isehara, Japan

<sup>2</sup> Department of Radiology, Kanagawa Children's Medical Center, Mutsukawa, Minami-ku, Yokohama, Japan

<sup>3</sup> Department of Perinatology, Wilhelmina Children's Hospital, University Medical Center Utrecht, Utrecht, The Netherlands

<sup>4</sup> Department of Paediatric Radiology, Wilhelmina Children's Hospital, University Medical Center Utrecht, Utrecht, The Netherlands

<sup>5</sup> Department of Paediatric Neurology, Wilhelmina Children's Hospital, University Medical Center Utrecht, Utrecht, The Netherlands

<sup>6</sup> Department of Neonatology, Kanagawa Children's Medical Center, Mutsukawa, Minami-ku, Yokohama, Japan

Address for correspondence Tetsu Niwa, MD, PhD, Department of Radiology, Tokai University School of Medicine, 143 Shimokasuya, Isehara 259-1193, Japan (e-mail: niwat@tokai-u.jp).

Neuropediatrics 2016;47:115–118.

## Abstract

### Keywords

- ▶ interhemispheric lipoma
- ▶ callosal anomaly
- ▶ malformations of cortical development
- ▶ magnetic resonance imaging
- ▶ infant

Intracranial lipomas are rare congenital malformations. The most common type of intracranial lipoma is the interhemispheric lipoma, which is frequently associated with callosal anomalies such as hypogenesis or agenesis of the corpus callosum. In contrast, interhemispheric lipomas are less often accompanied with malformations of cortical development (MCD). We report magnetic resonance imaging findings of three infants with an interhemispheric lipoma, associated with a callosal anomaly, and MCD: two infants with nodular interhemispheric lipoma, agenesis of the corpus callosum, and polymicrogyria, and one infant with interhemispheric curvilinear lipoma, hypoplasia of the corpus callosum, and heterotopias. An association was suggested regarding the occurrence of these malformations.

## Introduction

Intracranial lipomas are rare congenital malformations, which are composed of adipose tissue and are considered to be a result from the abnormal persistence and maldifferentiation of the primitive embryonic meninx, the mesenchymal precursor of the leptomeninges, during development of the subarachnoid cisterns.<sup>1–4</sup> Common locations of intracranial lipomas include the deep interhemispheric fissure, quadrigeminal cistern, prepontine cistern, and sylvian cistern.<sup>3,5</sup> They are rarely found on the surface of the cerebral cortex.<sup>1,2,5–7</sup>

The most common type of intracranial lipoma is the interhemispheric lipoma. Interhemispheric lipomas tend to be located above the corpus callosum, typically seen as either large and lobulated lipomas in the anterior callosal region or thin and curvilinear in the posterior callosal region.<sup>8</sup> Interhemispheric lipomas are frequently accompanied with hypogenesis or agenesis of the corpus callosum.<sup>1,2,8</sup> They, however, may less frequently be associated with malformations of cortical development (MCD). We present magnetic resonance imaging (MRI) findings of three infants with an interhemispheric lipoma, callosal anomalies, and cortical developmental abnormalities.

received  
August 5, 2015  
accepted after revision  
November 9, 2015  
published online  
January 25, 2016

© 2016 Georg Thieme Verlag KG  
Stuttgart · New York

DOI <http://dx.doi.org/10.1055/s-0035-1570752>.  
ISSN 0174-304X.

## Case Report

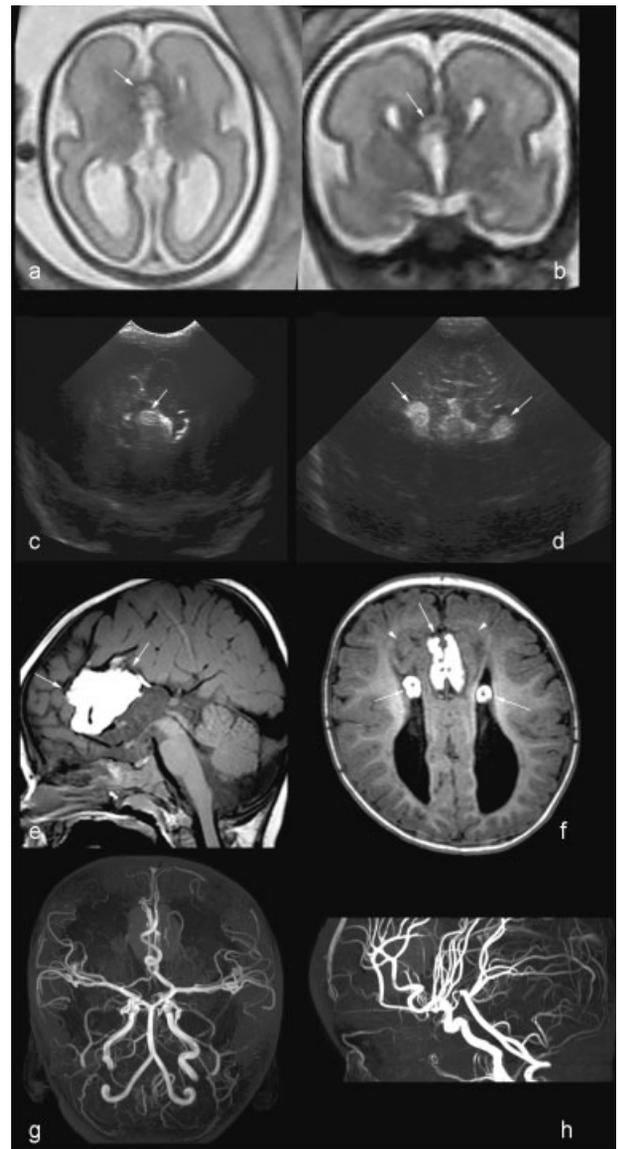
### Case 1

This male infant was born to a 36-year-old woman, who had one previous pregnancy, resulting in a healthy male infant. The mother was referred at a gestational age of 25 weeks, due to suspicion of a fetal brain malformation seen on a routine fetal biometry scan. Fetal neurosonography showed an interhemispheric lipoma associated with agenesis of the corpus callosum with dilatation of the posterior horn (colpocephaly). These findings were confirmed by a fetal MRI performed at a gestational age of 26 weeks (►Fig. 1a, b). During pregnancy, the size of the lipoma remained the same; however, there was an increase in ventricular size with advanced gestational age. The infant was delivered by elective cesarean section at a gestational age of 39 weeks and 3 days with a birth weight of 3,755 g (p 50–80), a head circumference of 36.5 cm (p 50–98), and length 49 cm (p 3–50). Apgar scores were 9, 10, and 10 at 1, 5, and 10 minutes, respectively. The general physical findings of this newborn were unremarkable. The infant fed well and showed no circulatory or respiratory problems. An electroencephalogram was performed that did not show any ictal discharges and normal background activity. Cranial ultrasonography showed an irregular echogenic midline mass as well as round echogenic lesions bilaterally in the ventricles suggestive of lipomas (►Fig. 1c, d). MRI performed on day 2 showed an interhemispheric lipoma extending into the lateral ventricles, agenesis of the corpus callosum, as well as thick, irregular cortex in the midline of the frontal lobes consistent with polymicrogyria (►Fig. 1e, f). The MRI was repeated at 9 months, as there was frontal bossing and an increase in size of the lipoma, but no clinical signs or raised intracranial pressure. MR angiography showed dilatation and tortuosity of the anterior cerebral artery (►Fig. 1g, h).

At 9 months, he had a pincer grasp and was able to roll over and sit without support. The child was last seen at 18 months, showing good progress in his development. He had started to walk without support at 17 months and he did not develop epilepsy. His head circumference remained on the 50th percentile.

### Case 2

This male infant was born to a 26-year-old woman, who had three previous pregnancies, all electively terminated. The pregnancy was followed up by the midwife at a maternity home. Fetal ultrasonography showed ventricular dilatation and a hyperechoic lesion at the interhemispheric fissure at a gestational age of 35 weeks. The infant was born by vaginal delivery at a gestational age of 38 weeks and 2 days with a birth weight of 2,702 g ( $p = 10$ –25), a length of 44.8 cm ( $< p 3$ ), and a head circumference of 35.5 cm ( $p = 3$ –10). Apgar scores were 8 and 9 at 1 and 5 minutes, respectively. An MRI of the brain was performed on day 1, showing an interhemispheric lipoma extending into the lateral ventricles as well as agenesis of the corpus callosum. T2-weighted image showed a thick cortex around the interhemispheric lipoma (►Supplementary Fig. S1,



**Fig. 1** Images of an infant with interhemispheric lipoma, agenesis of the corpus callosum, and polymicrogyria (case 1). (a and b) Single-shot T2-weighted images of the fetal brain show an interhemispheric mass (arrows) and agenesis of the corpus callosum. (c and d) Cranial ultrasonography at birth showed an irregular echogenic midline mass as well as round echogenic lesions bilaterally in the ventricles (arrows), suggestive of lipomas. Follow-up MRI was performed at the age of 9 months. (e and f) T1-weighted images show an interhemispheric lipoma extending into both lateral ventricles (arrows) as well as agenesis of the corpus callosum. (f) T1-weighted images also show a thick, irregular bumpy cortex (arrowheads) adjacent to the interhemispheric lipoma, consistent with polymicrogyria. (g and h) MR angiography shows dilatation and tortuosity of the anterior cerebral artery adjacent to the interhemispheric lipoma. MRI, magnetic resonance imaging.

online-only). The general physical findings were unremarkable and the neonatal course was uncomplicated. A repeat MRI was performed at the age of 3 years and 8 months. The MRI showed a slight increase in size of the interhemispheric lipoma. The MRI also showed thick, irregular, and bumpy cerebral cortex adjacent to the callosal lipoma consistent with polymicrogyria. The T2-weighted image showed dilatation of the anterior cerebral

artery around the interhemispheric lipoma (► **Supplementary Fig. S1**, online-only). His assessment at 13 months suggested a development of mild bilateral spastic diplegia. At 18 months of age, his height and body weight were both on the  $-2$  standard deviation. When he was last seen at the age of 3 years and 8 months, there were no longer signs of cerebral palsy and he had motor milestones within the normal range. His developmental score was just 83, just below the normal range (Tanaka-Binet intelligence scale, normal  $\geq 85$ ).

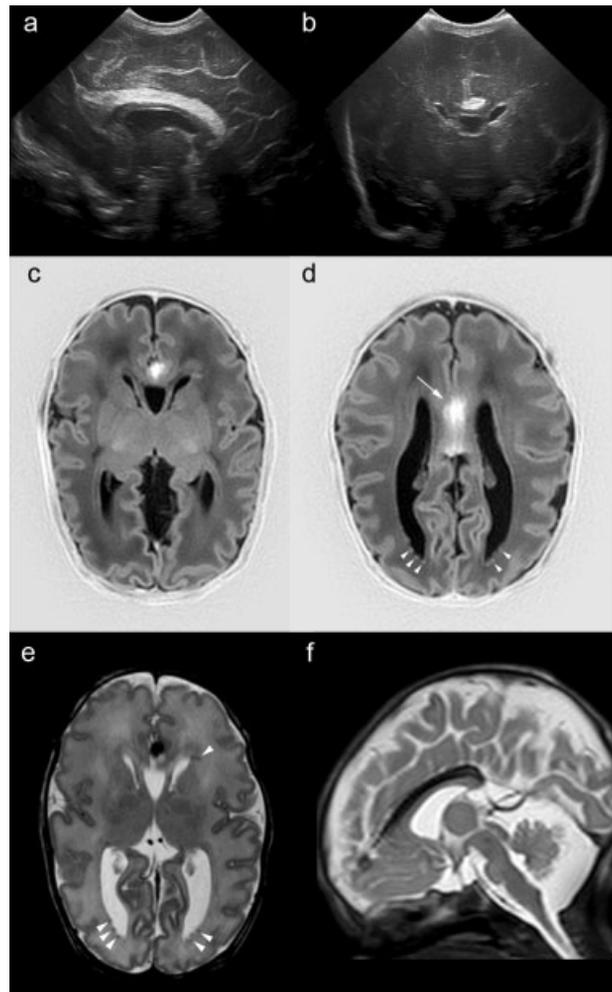
### Case 3

This female infant was born by cesarean section for intra-uterine growth retardation at a gestational age of 34 weeks and 6 days. Her birth weight was 1,590 g ( $< p 3$ ), length 43 cm ( $< p 3$ ), and head circumference 31 cm ( $p = 3-50$ ). The mother's only previous pregnancy produced a developmentally normal child.

Fetal ultrasonography at a gestational age of 20 weeks did not detect any abnormalities. Apgar scores were 8, 8, and 8 at 1, 5, and 10 minutes, respectively. There were no respiratory problems. She was noted to be pale at delivery and her hemoglobin concentration was 4.3 mmol/L. Because of the severe anemia, cranial ultrasonography was performed, which showed a hyper-echogenic lesion overlaying the corpus callosum (► **Fig. 2a, b**). No cause was identified for the anemia. She had a skin tag on the nose and a large umbilical hernia. Subsequently, an MRI was performed on day 10, showing a curvilinear interhemispheric lipoma, overlaying a hypoplastic corpus callosum (► **Fig. 2c-f**). Nodular lesions isointense to the cerebral cortex were identified at the peripheral portion of the lateral ventricles, consistent with subependymal heterotopias. The MRI also showed a small pons and cerebellum, suggesting hypoplasia. The single-nucleotide polymorphism (SNP) array showed a deletion in 5p15.33p15.31, which has previously been reported.<sup>9</sup> She was last seen at 6 months of age. Eye contact was short but present. She was not able to roll over at the time of this study. Her tone was normal and symmetrical.

### Discussion

We report three infants with an interhemispheric lipoma, callosal anomalies, and MCD. Interhemispheric lipomas tend to be associated with other brain malformations of varying severity. Among these, agenesis or dysgenesis of the corpus callosum is the most frequently associated brain anomaly. An occurrence of a lipoma is suggested to mechanically inhibit the development of the corpus callosum.<sup>10</sup> Curvilinear lipomas are known to be associated with adjacent hypoplastic corpus callosum.<sup>8</sup> These observations are in agreement with our MRI findings. Other malformations associated with intracranial lipomas include absence of the septum pellucidum, hypoplasia of the vermis, cranium bifidum, encephalocele, mid face defect, spina bifida, and myelomeningocele.<sup>1,11</sup> On the contrary, intracranial lipomas are less frequently accompanied with MCD. Several cases were reported regarding the association between sylvian or temporal gyral lipoma and MCD.<sup>1,2,6,7</sup> The MCD in these cases include cortical dysplasia, polymicrogyria, subcortical nodular heterotopia, and pachygyria-like



**Fig. 2** Images of a preterm infant with an interhemispheric lipoma, hypoplasia of the corpus callosum, periventricular heterotopias, and infratentorial abnormality (case 3). (a and b) Cranial ultrasonography at birth showed a curvilinear interhemispheric lipoma (arrows), overlaying a hypoplastic corpus callosum. (c and d) Inversion recovery T1-weighted and (e and f) fat-suppressed T2-weighted images on day 10 show a curvilinear interhemispheric lipoma (arrows) and hypoplasia of the corpus callosum. (c-e) T1- and T2-weighted images also show subependymal heterotopias (arrowheads). (f) Sagittal T2-weighted image shows a small pons and cerebellum.

abnormalities. These MCD are thought to occur as a result of interference with growth of cortical tissue by lipomas.<sup>7</sup> In our case series, polymicrogyria was noted in two cases, and heterotopia was identified in one case. In two of our three infants, polymicrogyria was found in the cortex adjacent to the interhemispheric lipoma. Interhemispheric lipomas are considered to develop between the 8th and 10th weeks of development.<sup>11</sup> Thus, interhemispheric lipomas may develop before neuronal migration and postmigrational cortical development, and affect the occurrence of polymicrogyria in these children. On the contrary, subependymal heterotopias noted in case 3 were remote from the interhemispheric lipoma. Therefore, there does not seem to be a direct effect of the interhemispheric lipoma to the occurrence of these heterotopias. Furthermore, the MRI of this infant also showed infratentorial abnormalities, suggestive of a more complex etiology in this infant, and indeed

a deletion (5p15.33p15.31) was found with the SNP array. To the best of our knowledge, there has only been one case with an interhemispheric lipoma, callosal hypoplasia, and polymicrogyria in familial lipomatosis,<sup>4</sup> and no case with interhemispheric lipoma and heterotopias. Taking into account the high frequency of interhemispheric lipoma among intracranial lipomas, the occurrence of MCD in cases with interhemispheric lipoma may be less common, compared with those in cases with sylvian or temporal gyral lipomas.

Intracranial lipomas are also known to be associated with vascular abnormalities including aneurysm, arteriovenous malformation, abnormal branches, dilatation and tortuosity of the feeding arteries, and veins with abnormal drainage.<sup>1,2,7</sup> As intracranial lipoma and arterial development co-occur during the same gestational weeks, an association between them has been suggested. The possible etiologies of these vascular abnormalities include congenital vessel structural deficiency, eliminating nutrition to the arterial wall from the cerebrospinal fluid, and some secretion of the lipoma to affect arterial smooth muscle.<sup>11</sup> In our series, dilatation and tortuosity of the anterior cerebral arteries was found around the interhemispheric lipoma in case 1 and case 2. This dilatation may be related to the development of interhemispheric lipomas.

Intracranial lipomas are usually asymptomatic and stable lesions, and are rarely accompanied by headaches, convulsions, psychomotor retardation, and cranial nerve defects.<sup>1,3</sup> We, therefore, would not suggest routine repeat imaging for patients with intracranial lipomas once the diagnosis has been made.

## Conclusion

We presented the rare association of an interhemispheric lipoma and callosal anomaly, with associated MCD in three newborn infants.

## References

- 1 Jabot G, Stoquart-Elsankari S, Saliou G, Toussaint P, Deramond H, Lehmann P. Intracranial lipomas: clinical appearances on neuroimaging and clinical significance. *J Neurol* 2009;256(6): 851–855
- 2 Kakita A, Inenaga C, Kameyama S, et al. Cerebral lipoma and the underlying cortex of the temporal lobe: pathological features associated with the malformation. *Acta Neuropathol* 2005; 109(3):339–345
- 3 Yilmaz N, Unal O, Kiyamaz N, Yilmaz C, Etlik O. Intracranial lipomas –a clinical study. *Clin Neurol Neurosurg* 2006;108(4):363–368
- 4 Nunes JC, Martins RF, Bastos A, et al. Brain lipoma, corpus callosum hypoplasia and polymicrogyria in familial multiple lipomatosis. *Clin Neurol Neurosurg* 2013;115(7): 1157–1159
- 5 Yildiz H, Hakyemez B, Koroglu M, Yesildag A, Baykal B. Intracranial lipomas: importance of localization. *Neuroradiology* 2006;48(1): 1–7
- 6 Thakur S, Sood RG, Jhobta A, Makhaik S, Thakur C. Sylvian fissure lipoma with angiomatous component and associated brain malformation: a case report. *Iran J Neurol* 2013;12(4): 169–171
- 7 Saatci I, Aslan C, Renda Y, Besim A. Parietal lipoma associated with cortical dysplasia and abnormal vasculature: case report and review of the literature. *AJNR Am J Neuroradiol* 2000;21(9): 1718–1721
- 8 Barkovich J, Raybaud C. Intracranial lipomas. In: Barkovich AJ, Raybaud C eds. *Pediatric Neuroimaging*. 5th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2011:520–524
- 9 Elmakky A, Carli D, Lugli L, et al. A three-generation family with terminal microdeletion involving 5p15.33-32 due to a whole-arm 5;15 chromosomal translocation with a steady phenotype of atypical cri du chat syndrome. *Eur J Med Genet* 2014;57(4): 145–150
- 10 Utsunomiya H, Ogasawara T, Hayashi T, Hashimoto T, Okazaki M. Dysgenesis of the corpus callosum and associated telencephalic anomalies: MRI. *Neuroradiology* 1997;39(4): 302–310
- 11 Sommet J, Schiff M, Evrard P, Blanc R, Elmaleh-Bergès M. Pericallosal lipoma and middle cerebral artery aneurysm: a coincidence? *Pediatr Radiol* 2010;40(8):1417–1420