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Case Report

Primary congenital intracranial lipoma with extracranial extension in a pediatric patient: A case report and literature review

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ABSTRACT

Background: Congenital intracranial lipomas are rare, representing only 0.1–0.5% of brain tumors, and are typically asymptomatic. An extracranial component is exceedingly rare, usually reported in slow-growing adult cases. This report presents a unique case of a rapidly growing congenital intracranial lipoma with extracranial extension in a pediatric patient.

Case Description: A 3-year-old girl with a progressively enlarging forehead mass, present since birth, was found to have a large interhemispheric lipoma (12 × 10 × 9 cm) extending into the left lateral ventricle and connected to an extracranial mass (17 × 10 × 10 cm) through a frontal bone defect. Additional findings included corpus callosal agenesis and venous drainage from the scalp lesion into the superior sagittal sinus. The microsurgical intervention involved the excision of the extracranial lipoma and subtotal resection of the intracranial component, preserving critical neurovascular structures.

Conclusion: This case of rapid lipoma growth in a pediatric patient is atypical, differing from slow-growing, asymptomatic presentations in adults. High-magnification microsurgery and Doppler guidance facilitated safe resection with a positive outcome. This case highlights the need for further research into developmental factors and rapid growth mechanisms in pediatric intracranial lipomas with extracranial components, potentially representing a distinct clinical entity.

Keywords: Extracranial extension, Microsurgery, Outcome, Pediatric intracranial lipoma

INTRODUCTION

Intracranial lipomas are rare, constituting only 0.1–0.5% of all primary brain tumors.[19] Even more uncommon are those presenting with an extracranial component, a phenomenon sparsely documented in the literature.[11,17,19] These lesions are congenital and attributed to the atypical persistence of primitive meningeal tissue within the reticuloendothelial elements of the meninges. Their development is often associated with the midline's embryological derangements, specifically

where meningeal anlage remnants persist abnormally. This genesis is especially intriguing as it represents an overlap with brain dysraphism, with both intracranial and extracranial elements involved.

Intracranial lipomas tend to be positioned near midline structures, frequently appearing in cerebral cisterns such as the quadrigeminal, superior cerebellar peduncle, suprasellar, cerebellopontine angle, and Sylvian cisterns. Their morphology can vary; posterior lesions, typically smaller, are often curvilinear and situated near the corpus callosum, whereas larger, tubulonodular forms are more likely found anteriorly.[11,17] In most cases (>50%), these lipomas are asymptomatic, often detected incidentally on imaging for unrelated issues.

Several hypotheses have been proposed to elucidate the genesis of intracranial lipomas, though much remains uncertain, especially regarding cases with both intracranial and extracranial components.[6,19] Management strategies for these lipomas, mainly when they present in complex forms with intraventricular extension or extensive subcutaneous components, are challenging and necessitate careful consideration. Surgical intervention, especially for more giant lipomas with intracranial extension, has yielded suboptimal results with a high risk of complications. This highlights the critical need for advancing understanding and management approaches, particularly in cases warranting long-term follow-up for effective monitoring and evaluation.

CASE DETAILS

A 3-year-old girl presented with a progressively enlarging forehead mass that had been present since birth. The lesion was initially the size of a lemon; however, it increased to the current size of presentation within the previous 1 year. Clinical examination showed a firm, subcutaneous lesion over the frontal region, accompanied by distinct craniofacial features, including hypertelorism (increased distance between the eyes), a flattened nasal bridge, and low-set ears [Figures 1a-c]. A thorough evaluation for additional congenital anomalies revealed no other abnormalities.

Initial imaging with a non-contrast computed tomography scan of the head identified a well-defined, fat-density mass in the subcutaneous plane of the forehead [Figures 2a and b]. This lesion extended through a small (1.5 cm) defect in the frontal bone and was connected through a thin lipomatous stalk to an intracranial lesion in the interhemispheric region. The scan further revealed agenesis of the corpus callosum, while the paranasal sinuses appeared normal [Figures 2c-f].

A subsequent brain magnetic resonance imaging (MRI) provided greater detail, showing a non-enhancing, lobulated mass measuring 12 cm × 10 cm × 9 cm within the anterior interhemispheric fissure, extending into the left lateral ventricle. The lesion displayed a characteristic fat signal across all sequences, with peripheral blooming seen on susceptibility-weighted imaging. A large fat-density subcutaneous lesion (15 cm × 10 cm × 10 cm) was also observed in the frontal region, connected to the interhemispheric lesion by a thin lipomatous stalk. The MRI also confirmed corpus callosal agenesis and venous drainage from the scalp lesion into the superior sagittal sinus. The distal A2 and A3 segments of both anterior cerebral arteries were observed traversing the interhemispheric lipoma [Figure 2f]. A separate fat-density lesion with vascular voids was also detected in the right temporal sulcal space. Based on these findings, a diagnosis of corpus callosum lipoma with an associated extracranial component was made, and further screening showed no major congenital anomalies.

Figure 1: (a) Preoperative clinical photograph of a 5-year-old male presenting with a frontal subcutaneous lipoma. The lesion, initially noticed after the 1st year of life, has rapidly grown in the past year, impairing vision and causing progressive headaches. The right lateral view shows the lesion's extracranial portion, measuring approximately 15 cm × 10 cm × 10 cm, with a similar dimension intracranially, connected by a lipofibromatous band, as observed intraoperatively in subsequent images. (b) Preoperative frontal photograph illustrating the patient's hypertelorism and forehead swelling, with no focal neurological deficit. (c) Preoperative image from the cranial end, showing external measurement of the lipoma dimensions (15 cm × 10 cm) with a scale.

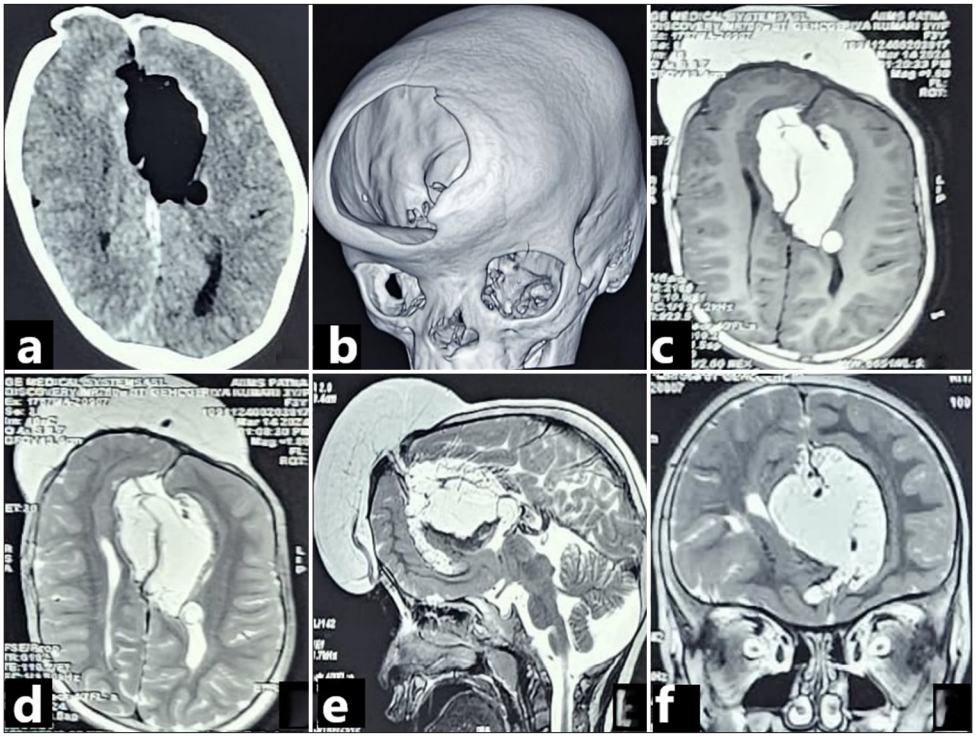

Figure 2: (a) Preoperative noncontrast computed tomography (NCCT) axial head scan, showing a hypodense mass in the anterior interhemispheric fissure extending anteriorly, with associated bony defect. (b) Preoperative 3D reconstruction of NCCT showing a midline calvarial defect with bony deformation. (c) Magnetic resonance imaging (MRI) T1-weighted axial view showing a hyperintense mass in the anterior interhemispheric fissure, compressing the frontal horns of the bilateral lateral ventricles and extending into the body of the left lateral ventricle. (d) MRI T2-weighted (T2W) axial view revealing a hyperintense mass in the anterior interhemispheric fissure with flow voids, including an intralesional vessel traversing the lesion anteriorly and another flow void in the extracranial portion. (e) MRI T2W sagittal section demonstrating the intracranial interhemispheric lipoma, with a connecting stalk to the extracranial scalp lesion in the subcutaneous plane in the frontal region. Agenesis of the corpus callosum is noted posteroinferiorly. (f) MRI T2W coronal section showing the anterior cerebral arteries passing through the lesion.

The surgical approach involved excising the frontal subcutaneous lipoma and the affected bone, resecting the connecting lipomatous stalk, and performing a subtotal excision of the interhemispheric lipoma [Figures 3a-d]. Microsurgical excision of the interhemispheric component was carefully performed, leaving a cuff of lipomatous tissue around the vessel to avoid injury to pericallosal vessels. At places, we had difficulty excising the lipoma as it adhered to the nearby subarachnoid membranes and sellar-suprasellar spaces. A lipomatous mass extending into the frontal horn of the lateral ventricle was also excised, leaving a small cuff adherent to the choroid plexus, achieving near-total excision. Careful microneurosurgical techniques, aided by Doppler guidance, were employed to preserve critical neurovascular structures.

Reconstruction was completed with titanium mesh cranioplasty. The procedure and postoperative course were smooth, and the patient experienced no neurological deficits [Figures 4a-d]. At her 18-month follow-up, she showed excellent recovery and stable progress, with no further residual intracranial or extracranial component growth.

DISCUSSION

Primary congenital multiple intracranial lipomas with an extracranial component are rare, and very few case reports are available[5-7,9-15] [Table 1]. Most lipomas are asymptomatic and incidentally detected when patients are investigated for other illnesses.[11,16,17,19] The growth of lipomas into large intracranial lesions with extracranial growth defines rapid growth. The giant lipoma with a subcutaneous component (12 cm × 10 cm) was reported with a similar interhemispheric fissure component and agenesis of the corpus callosum in 26-year-old females. Still, she refused surgery as she was only

having minor cosmetic problems. This lesion grows very slowly, and fast growth, as present in our patient, had not been reported earlier. All the previously reported cases with giant subcutaneous lesions were adults and predominantly female, but the patient in our study was in the pediatric age group.[9,14] The pediatric patients presented with smaller extracranial components of the reported size of <6 cm. Presentation at such an early stage with large lesions indicates the impending progression of neurological illness, as in our case study. It has been postulated earlier that it arises from the reticuloendothelial component of meninx primitivae. In the present case, the inclusion of meninx primitive at the different cisternal locations and secondary dehiscence of the anterior cranial vault at the time of development of anterior neuropore can be suggested, which have been earlier reported in a few cases. Such developmental anomaly and its genesis had been reported in earlier studies, but its fast growth in the present cases compared to other reported cases is challenging to explain. It needs to be established as there was no evidence of malignant transformation caused by fast growth compared to other reported studies. There are a few case reports where the patient has intracranial lipoma with extracranial component connected with lipomatous stalk and calvarial defect and features of spinal dysraphism.[1,5] Such findings were not present in our patient. Still, they suggested developmental anomalies similar to spinal dysraphism, where nondisjunction between cutaneous and neuroectodermal tissues had been reported when such a lesion was found, as in the present case.[5]

Most patients in the presenting age group are asymptomatic, and such rapid growth has not been reported. The condition causes visual difficulties and difficulty breathing due to the projecting frontal mass compressing the root of the nose, causing hypertelorism and obstruction in vision. This patient had no history of seizure, headache, papilledema, or any neurological deficit reported in earlier reported symptomatic cases. In this patient, the involvement of interhemispheric fissure and extension along fissures to lateral ventricle was probably going to cause a mass effect and features of obstructive hydrocephalus, so we planned for microsurgical intervention and correction of cosmetic issues

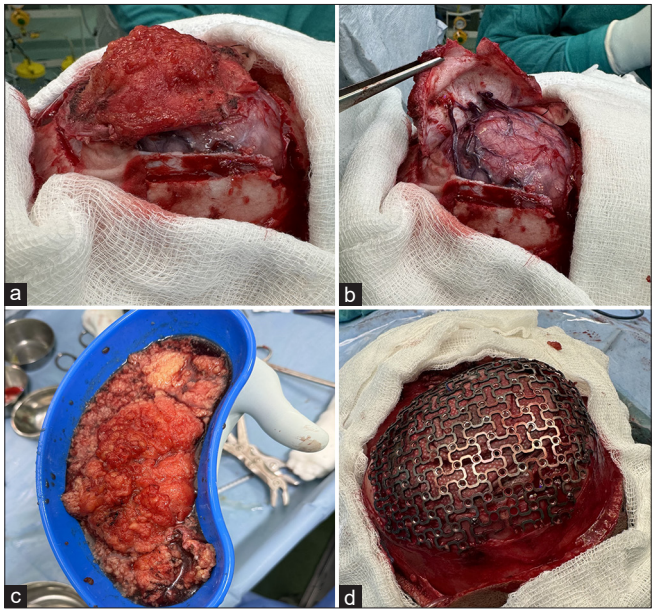

Figure 3: (a) Intraoperative photograph post-scalp incision, revealing a subcutaneous lipomatous mass overlying the dura in the midline. (b) Intraoperative image after dural elevation, showing dilated, tortuous cortical veins in the anterior frontal cortex on the right, mirrored on the contralateral side. (c) Intraoperative photograph displaying the lipomatous mass following excision. (d) Post-excision intraoperative image showing the dura repair with a synthetic graft and titanium mesh cranioplasty following maximal safe resection of the intracranial lipoma and extracranial mass excision.

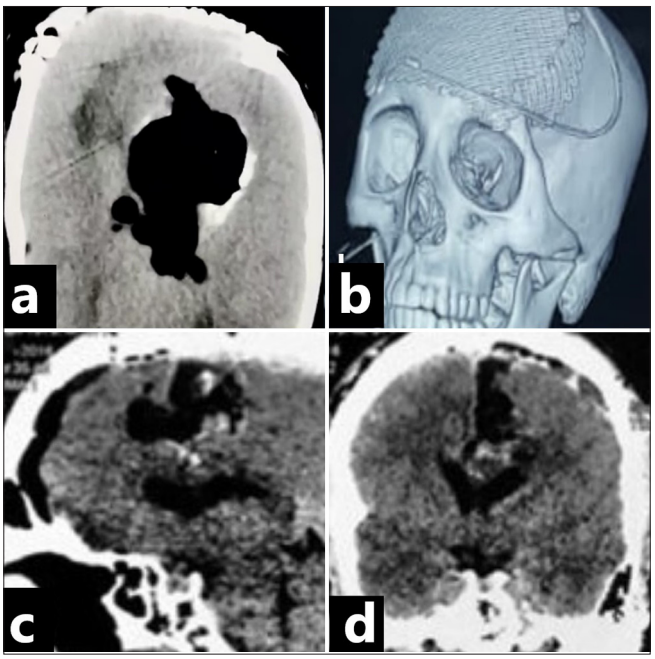

Figure 4: (a) Postoperative axial magnetic resonance imaging brain with contrast, taken at 18 months follow-up, showing successful excision of the lipoma's extracranial and intracranial portions, with no ventriculomegaly or recurrence. (b) Follow-up 3D coronal front view of the head at 18 months, showing no residual bony defect and tissue growth over the titanium mesh, restoring the contour and covering the bony defect. (c) Postoperative sagittal non-contrast computed tomography (NCCT) brain at 18 months follow-up shows excision with postoperative changes, absence of lesion regrowth, and no ventriculomegaly. (d) Postoperative coronal NCCT brain with contrast at 18 months follow-up, revealing no ventriculomegaly or recurrence of the lipoma, with postoperative changes.

Table 1: Previous case reported for corpus callosum lipoma and their comparison with present case

Authors	Age/sex	Clinical features	CT findings	MRI findings	Lipoma size (cm)
El Marrakchi, *et al.*, 2024[5]	3 months/m	Bluish round lumbosacral mass Incidental diagnosis on workup	Large CCL associated hypogenesis of the corpus callosum with bilateral choroid plexus lipoma	Polymicrogyria with hypertense T1 and fluid-attenuated inversion recovery (FLAIR)	NA
Kazutoshi Konomatsu *et al.* 2024[11]	30 y/f	Seizures	Low-density lesion without calcification at the pericallosal space	Linear hyperintense lesion over the corpus callosum on T1-weighted and T2-weighted linear high signal intensity lesion over the corpus callosum on MP RAGE and fluid-attenuated inversion recovery (FLAIR) hypoplasia of the corpus callosum	NA
Dhara Rana *et al.* 2021[14]	30y/f	Headache Seizures Mass over anterior aspect of scalp	Large interhemispheric partially calcified lipoma communicating with a large scalp lipoma 3 small defects in the left frontal lobe lipoma extension into the body of the lateral ventricles bilaterally agenesis of the corpus callosum	interhemispheric lipoma extending anterosuperiorly from the midline through the frontal bone and into the left and right frontal scalp soft tissues.	6 x4.7 (Intracranial)
Thapa Ashish Jung *et al.*, 2021[18]	42y/f	Incidental diagnosis on imaging for head injury Occasional brief episodes of seizures	Hypodense lesion around the corpus callosum and was confused with pneumocephalus	Curvilinear shaped area of altered signal intensity surrounding the splenium of the corpus callosum which appeared hyperintense on both T1 and T2 weighted images. thin rim of signal loss around this lesion on all sequences, consistent with calcification	NA

(Contd...)

Table 1: *(Continued)*

Authors	Age/sex	Clinical features	CT findings	MRI findings	Lipoma size (cm)
				on fat-suppressed T1 weighted images [Figure 2] in which the lesion showed signal loss confirming	
José D. Charry *et al*. 2021[4]	27y/f	Headache seizures	Rounded hypodense lesion with irregular edges, located in the frontal midline supratentorial level and slight compression effect of adjacent parenchyma	High intensified lesion in a sequence of T1 and T2 in supra-callose midline surrounded by bilateral circumflex arteries Spectroscopy NMR which showed a maximum peak of lipid-lactate, with decreased peak of NAA, Cho, and Cre, compatible with lesion of fat content dysgenesis of the callosum corpus	NA
Hamza Karabağ *et al*; 2014[9]	26y/f	Mass over anterior aspect of scalp	NA	Lipoma on corpus callosum, filling interhemispheric fissure subcutaneous lipoma, closure defect of the anterior fontanelle irregular azygos anterior cerebral artery	12 x 15 (extrcranial)
Omer Karakas *et al*. 2013[10]	3 months/m	Mass on anterior aspect of head Seizures Wide open anterior fontanelle Low-set ears, ocular hypertelorism, broad nasal root, depressed nasal bridge	Colpocephaly and elevation in the third ventricle reflecting corpus callosum agenesis anterior interhemispheric calcified lipoma and subcutaneous lipoma anterior cranium bifidum occultum	Corpus callosum agenesis, midline interhemispheric cyst interhemispheric tubulonodular calcified lipoma, midline subcutaneous lipoma	NA

(Contd...)

Table 1: *(Continued)*

Authors	Age/sex	Clinical features	CT findings	MRI findings	Lipoma size (cm)
				no evident connection between the two formations	
Jiménez Caballero *et al*; 2012[8]	23y/f	Headache seizures	Hypodense image measuring 5 cm×3 cm in the area of the corpus callosum surrounded by mural calcifications on both sides. Its density is indicative of fat content. Agenesis of the corpus callosum.	Entire corpus callosum from the genu to the splenium was occupied by tissue that was hyperintense on all sequences, with the exception of the fat suppression sequence.	5 x 3 cm
Delphine Mitilian *et al*; 2009[13]	1 months/m	Progressively increasing Mass on anterior aspect of head	NA	Interhemispheric lipoma extending to the choroid plexus of the lateral ventricles, agenesis of the corpus callosum bilateral polymicrogyria of frontal cortex. Frontal subcutaneous lipoma	3 x 2 (extracranial)
Aftab Alam *et al*; 2006[2]	22y/m	Recurrent seizures	NA	Calcified curviliner corpus callosum lipoma over posterior aspect	NA
Given CA. *et al*; 2005[7]	2 months/m	Mass on anterior aspect of head	NA	Interhemispheric (pericallosal) lipoma with associated agenesis of the corpus callosum intracranial component connected to subcutaneous portion by lipomatous stalk pericallosal branches of ACA seen within the interhemispheric component	NA

(Contd...)

Table 1: *(Continued)*

Authors	Age/sex	Clinical features	CT findings	MRI findings	Lipoma size (cm)
Hiroshi Kudoh, *et.al*; 1984[12]	3 months/f	Progressively increasing Mass on anterior aspect of head	Lipoma with calcifications in genu and body of corpus callosum in both lateral ventricles and in interhemispheric fissure Corpus callosum dysgenesis A defect of the frontal bone was also noticed in the midline. pericallosal arteries dilated and penetrated the mass	NA	5 x 3.5 (extracranial)
Present case	3y/f	Progressively increasing Swelling over forehead since birth	Fat density is seen in the anterior interhemispheric fissure with extension into body of left lateral ventricles Calcific foci in the periphery of the lesion fat density scalp lesion in frontal region	Lobulated nodular non-enhancing mass of size 7.6×4.1×4.7 cm (APxTRxCC) is seen in the anterior interhemispheric fissure with extension into body of left lateral ventricles blooming is seen at the periphery of the mass on SWI follows fat signal on all sequences with fat saturation. another scalp lipoma 3.3x 9.2×7.5 cm (APxTRxCC) which is communicating with the interhemispheric lesion via a thin lipomatous stalk. corpus callosal agenesis	Intracranial :7.6×4.1×4.7 cm Extracranial: 3.3x 9.2×7.5 cm
Authors	**IC-EC connection**	**Type**	**Management**	**Follow up**	
El Marrakchi, *et al*., 2024	absent	TN	conservative	1y	

(Contd...)

Table 1: *(Continued)*

Authors	IC-EC connection	Type	Management	Follow up
Kazutoshi Konomatsu *et al*. 2024	absent	curvilinear	AED	
Dhara Rana *et.al*., 2021	present	TN	AED Planned for surgery later	
Thapa Ashish Jung *et al*., 2021	Absent	curvilinear	conservative	
José D. Charry *et al*. 2021	absent	TN	AED	
Hamza Karabağ *et al*; 2014	NA	TN	--	
Omer Karakas *et al*. 2013	absent	Tubulonodular calcified	Medical management with AED	
Jiménez Caballero *et al*; 2012	absent	TN	AED	
Delphine Mitilian *et al*; 2009	present	TN	Extracranial lipoma excision	After 3 months asymptomatic
Aftab Alam *et al*; 2006	absent	curvilinear	NA	
Given CA. *et al*; 2005	present	TN	subcutaneous component of the lipoma removed	NA
Hiroshi Kudoh, *et.al*; 1984	absent	TN	Surgical excision	NA
Present case	present	TN	Complete resection of scalp lipoma+maximal safe resection of interhemispheric lipoma+mesh cranioplasty	6 months

m: male, f: female, Y: years, IC-EC: Intracranial- ExtracranialCCL: corpus callosum lipoma, NA: Not available, TN: Tubulonodular, AED: Anti epileptic drug, MP RAGE: Magnetization prepared rapid gradient echo, NMR: Nuclear magnetic resonance, NAA: N acetyl aspartate, Cho: Choline, Cre: creatine, ACA: Anterior cerebral artery

caused by the projection of subcutaneous mass and the defect in the frontal bone. Different studies have reported poor results for surgical excision.[3,20,21] In our study, we used the help of high magnification while using a microscope to identify vessels and confirm the localization of vessels by intraoperative Doppler, which helped minimize vascular injury. As the reported studies suggested slow growth of such lesion, we left a cuff of lipomatous lesion adherent to it and the surrounding critical structure, which helped in near total excision of the lesion without causing the neurological deficit.

We want to convey that even though we operated on our patient as she showed rapid growth of lipoma as per description by the patient's parents, who gave a history of extracranial component size increase as the reason they decided to seek medical opinion, based on previous studies that we opted for maximal safe resection of lipoma to avoid potential long-term deficits arising out of vascular injury.

CONCLUSION

This case highlights the rare presentation of primary congenital multiple intracranial lipomas with an extracranial component in a pediatric patient, contrasting with the typical slow-growing, asymptomatic nature of such lesions predominantly observed in adults. The rapid growth and large size of the lipoma underscore the possibility of atypical developmental pathways, suggesting a need for further research into the embryological origins and molecular mechanisms driving accelerated growth in pediatric lipomas. Microsurgical excision was essential not only to alleviate potential mass effects and

prevent complications like obstructive hydrocephalus but also to address cosmetic concerns associated with extracranial extension. High-magnification microscopy and intraoperative Doppler guidance proved instrumental in achieving safe, near-total resection with excellent outcomes and no neurological deficits. This case underscores the importance of precise surgical techniques and may inform future management strategies, especially for pediatric patients with large congenital lipomas with extracranial extensions.

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