

Frontal subcutaneous lipoma associated with interhemispheric lipoma, lipomeningocele, and corpus callosal dysgenesis in a young adult: CT and MRI findings

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Abstract

Intracranial lipomas are rare incidental lesions. Rokitsky first described a corpus callosal lipoma on the posterior part of the corpus callosum in 1856. Since then many cases have been reported. We report the imaging findings of an interhemispheric lipoma extending into subcutaneous plane associated with spinal lipomeningocele and corpus callosal dysgenesis. Computed tomography and magnetic resonance imaging findings are characteristic. All cases have been reported in pediatric age group till now. Such a case in this age group has not been reported earlier.

Key words: Callosal dysgenesis; interhemispheric; lipomeningocele; meninx; subcutaneous lipoma

Introduction

Intracranial lipoma is a rare disease accounting for 0.06–0.46% of all intracranial tumors. It frequently develops in the supratentorial midline region, mostly in the pericallosal region (30–50%). Intracranial lipoma with a large subcutaneous component is a rare association. Intra-extracranial lipoma associated with spinal dysraphism and lipomeningocele is even more rare, not been reported earlier. We report here the multimodality approach of such a case in a 22-year-old female.

Case History

A 22-year-old female patient was referred to our department with a scalp swelling since birth. Patient also had complains

of urinary incontinence since birth. On physical examination, the patient had a huge soft scalp swelling. There were no signs of facial anomalies. Higher mental functions were within normal limits. Developmental milestones were attained at appropriate age. Urine routine examinations revealed pus cells, suggestive of infection. Other laboratory investigations were within normal limits.

Patient was referred for computed tomography (CT) scan of head. Noncontrast head CT showed a fat attenuating mass lesion (–80 to –120 HU) in the interhemispheric fissure, extending to the subcutaneous region through cranium bifidum [Figures 1 and 2]. The lesion was also seen extending into bilateral lateral ventricles and through the tentorial

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Cite this article as: 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.

Access this article online

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DOI:
10.4103/ijri.IJRI_280_17

incisura infratentorially. The lesion was causing mass effect in the form of compression of adjacent brain parenchyma laterally and cerebellum inferiorly. The intracranial part of the mass had a central linear calcification extending to the bone defect and peripheral curvilinear calcification. Corpus callosum could not be assessed. On contrast-enhanced CT, the lesion did not enhance, but vessels were seen traversing through the mass [Figure 3]. Three-dimensional volume rendered and multiplanar reformatted images showed the subcutaneous scalp mass and the frontal bone defect clearly [Figures 4 and 5].

CT abdomen revealed thickened trabeculated urinary bladder wall with diverticulae formation, suggestive of chronic urinary incontinence [Figure 6]. A fat-attenuating mass with internal soft tissue membranes was seen extending from spinal canal to subcutaneous region through the defect in posterior element of sacrum [Figure 7]. These findings were suggestive of spinal dysraphism with lipomeningocele.

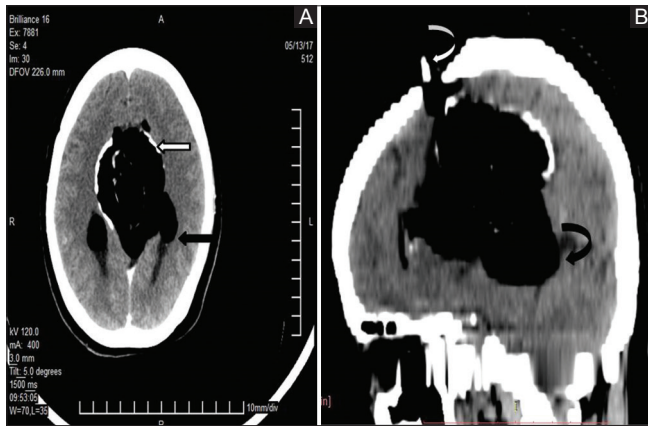


Figure 1 (A and B): Axial (A) and sagittal (B) NCCT images showing huge fat density lesion with peripheral curvilinear calcification (arrow) in the interhemispheric fissure. The lesion is seen extending extracranially through cranium bifidum (curved white arrow). It is also extending into atria (black straight arrow) and frontal horns of bilateral lateral ventricles, causing minimal compression of cerebellum infratentorially (curved arrow)

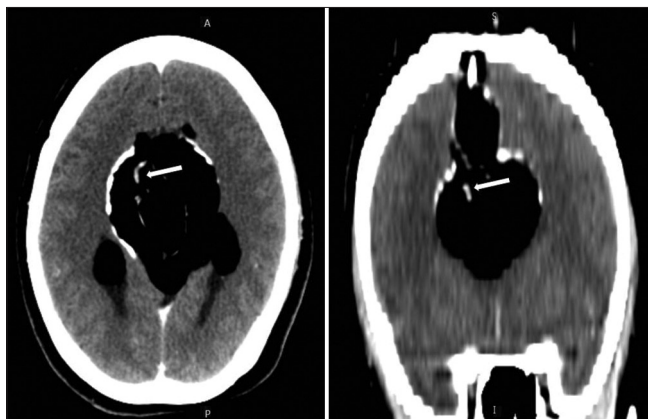


Figure 3: Axial and Coronal CECT BRAIN showing branches of Anterior cerebral artery encased by the interhemispheric mass (arrows)

Magnetic resonance (MR) images revealed hyperintense signals in the interhemispheric lesion on T1-, T2-, and FLAIR-weighted images [Figure 8]. The lesion showed suppression of signals on fat-suppressed T2-weighted image [Figure 9B], consistent with lipoma. There were peripheral hypointense signals on T1, T2-weighted image, and T2 GRE sequence [Figure 9C], suggestive of calcification. Posterior part of body and splenium of corpus callosum were absent, suggestive of partial agenesis of corpus callosum [Figure 9A].

Considering the risks related to surgery, patient was advised follow-up for the interhemispheric lipoma. However, patient was advised surgery for the subcutaneous component and spinal lipomeningocele which she refused.

Discussion

Intracranial lipomas are a rare type of congenital malformation, forming 0.06–0.46% of intracranial lesions^[1] with 30–50% occurring in pericallosal region. They are assumed to result from the persistence or mal-differentiation

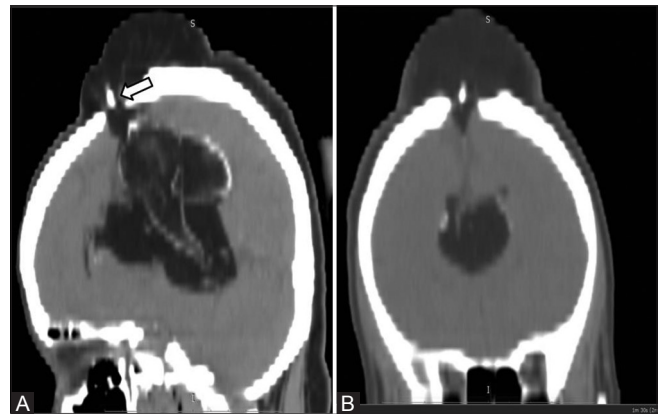


Figure 2 (A and B): Sagittal (A) and coronal (B) NCCT image (window width of 580) showing extension of the interhemispheric fat attenuating lesion through the cranium bifidum into the subcutaneous region. There is central (arrow) as well as peripheral calcification

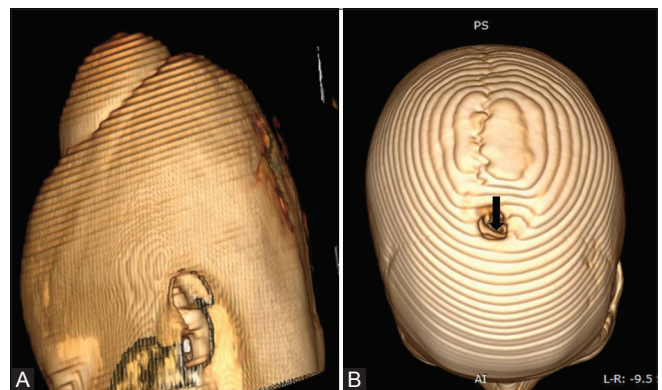


Figure 4 (A and B): 3D VRT reformatted CT images (A) sagittal image showing scalp swelling. (B) axial image showing focal defect in the region of anterior fontanelle (black arrow)

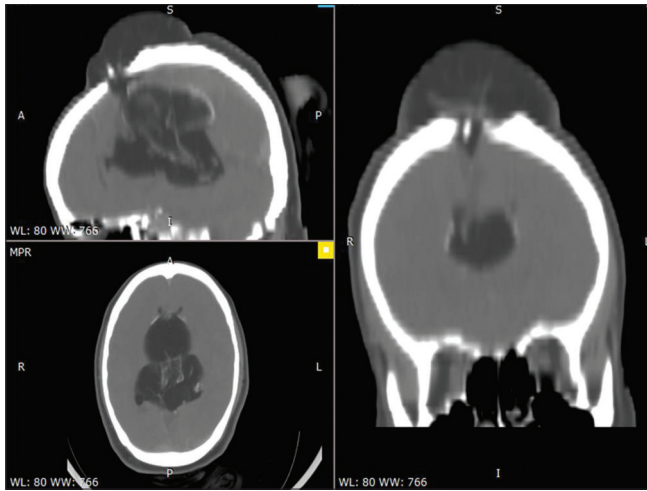


Figure 5: 3D MPR reconstruction showing intra and extracranial extension of the lipoma

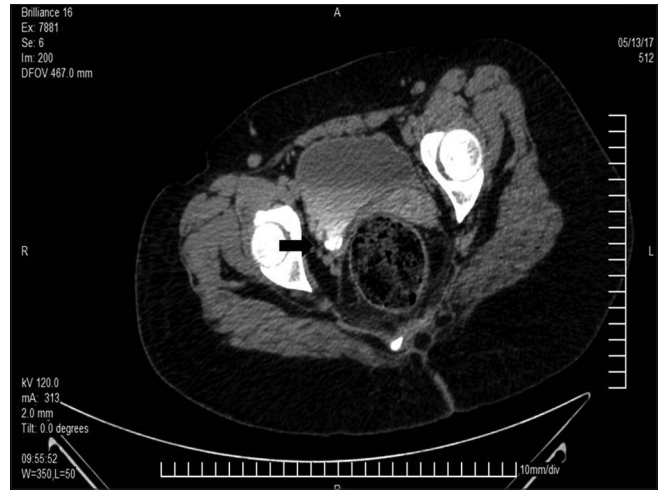


Figure 6: Delayed phase CT pelvis showing trabeculated urinary bladder wall (arrow)

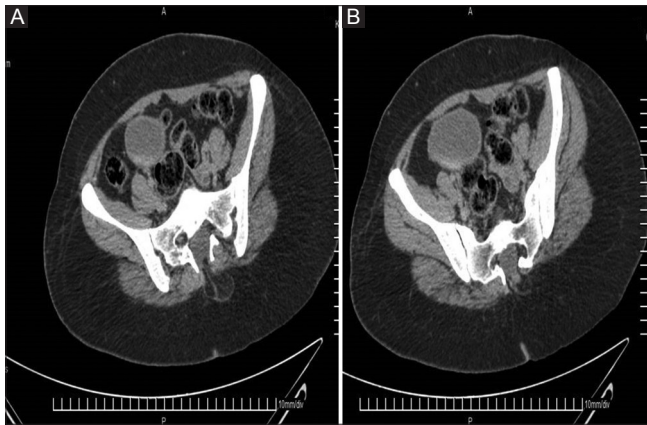


Figure 7 (A and B): Sequential axial non contrast CT images (A) and (B) showing fat attenuating lesion with internal membranes extending from spinal canal through the spina defect into the subcutaneous fat

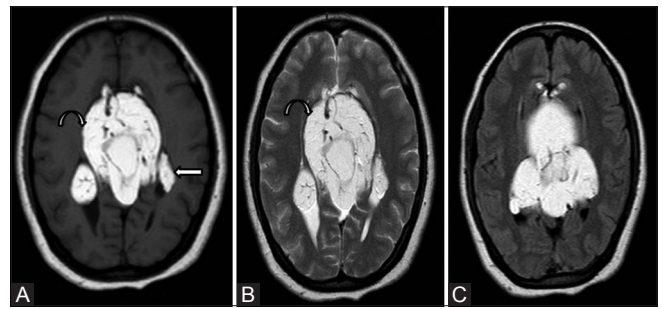


Figure 8 (A-C): (A) Axial T1WI and (B) axial T2WI, (C) axial FLAIR image showing hyperintense mass lesion in interhemispheric fissure, extending into bilateral lateral ventricles (arrow). T1 and T2WI both show hypointense signals at the periphery (curved arrow)

of the primitive meninx into lipomatous tissue during the development of the subarachnoid cisterns.^[2]

Most are asymptomatic and found incidentally. If symptomatic, the most frequent clinical presentation is seizure.^[3] Our case presented with swelling in scalp and lumbosacral region since birth. She also had complaints of urinary incontinence since birth, now presenting with altered behavior since 1month.

Interhemispheric lipoma associated with subcutaneous component is extremely rare. There have been only three case reports of isolated intracranial interhemispheric lipoma with subcutaneous component as reported by Delphine Mitilian *et al.*^[4] and Chen *et al.*,^[1] associated with sagittal sinus fenestration and falcine sinus by Ahmetoglu *et al.*,^[5] however, association with spina bifida and lipomeningocele has not been reported in any case report.

Different communication patterns are observed between intracranial and extracranial components of the lipoma

ranging from no connection,^[6] fibrous-lipomatous stalk^[7,8] to direct continuity through cranium bifidum.^[9] The explanation for direct extension through cranium bifidum is explained by Truwit and Barkovich^[2] as secondary dehiscence of the cranium with potential evagination of a small tuft of meninx primitiva. The scalp defect if persists leads to direct continuation of intracranial lipoma with the subcutaneous component as was found in our case.

According to Tart and Quisling,^[10] intracranial lipomas are divided into curvilinear and tubulonodular types. Curvilinear lipomas tend to be posterior, small, and linear, while tubulonodular lipomas to be anterior, round, and cylindrical. The tubulonodular type, i.e. anterior pericallosal lipomas more commonly form a frontal subcutaneous mass via the interhemispheric tract and frontal bone defect. Our patient had tubulonodular type of interhemispheric lipoma with extension into the subcutaneous tissue through the frontal bone defect.

A variety of vascular abnormalities have been described in association with intracranial lipoma, including distension, kinking, or narrowing of arteries and veins; engulfment

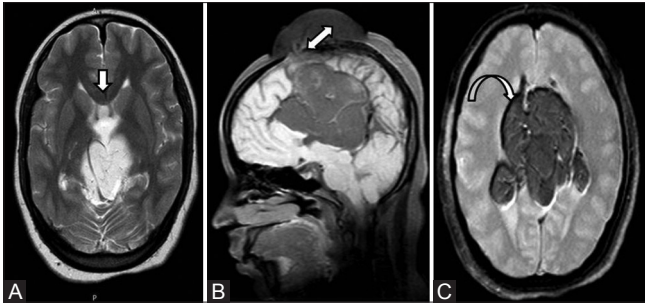


Figure 9 (A-C): (A) Axial T2WI showing hyperintense mass in interhemispheric fissure. Anterior part of corpus callosum is seen (arrow). (B) Sagittal Fat Sat T1WI showing extracranial subcutaneous extent of the lesion through the frontal bone defect (double headed arrow). (C) Axial T2WI GRE showing intermediate intensity mass with peripheral hypointense signals (curved arrow) suggestive of calcification

of the cerebral arteries; arteriovenous malformation; and aneurysm.^[11] Our patient had encasement of pericallosal branches of anterior cerebral artery by the lipoma.

Zettner and Netsky^[12] describe the tumor as replacing the corpus callosum or lying on its dorsal surface actually “outside” the brain. About half of the pericallosal interhemispheric lipomas are associated with dysgenesis of corpus callosum.^[13,14] A normal corpus callosum implies insult after its formation at a later embryonic stage,^[15] while severe degree of hypoplasia implies insult at an early stage. Other associated anomalies include lipomas at other sites, hypoplastic fornix, absent septum pellucidum, spina bifida, myelomeningocele, frontal bone defects, encephaloceles, heterotopic gray matter, agenesis of the vermis, and cleft lip.^[16] In our case, there was partial agenesis of corpus callosum with nonvisualization of posterior part of body and splenium. There was associated spina bifida with lipomeningocele in the sacral vertebrae.

Calcification and bone formation are also common. As stated by Zettner and Netsky^[11] that “the bony elements are foci of osseous metaplasia following calcification, a process frequently encountered elsewhere in the body.” In our case, the lesion showed central as well as peripheral rim calcification.

CT is diagnostic, demonstrating fat density attenuation (–80 to –110HU), calcification, its location, extent, and associated anomalies. MRI is used to assess the anatomy of the corpus callosum, monitor growth of the mass, and to determine if any invasive characteristics indicate more aggressive treatment. MRI shows a homogeneous well-circumscribed lesion displaying the characteristic short-T1 and T2 signal of fat.

Both CT and MRI findings are characteristic of the lesion, so biopsy confirmation is not required for the diagnosis. These are “leave me alone” lesions. According to Gerber *et al.*,^[17] the surgical removal of these lesions has not been

gratifying. The vascularity of the tumor, the encasement within the lesion of the anterior cerebral artery and its branches, as well as the adhesions of the collagenous capsule to the brain tissue, all add to the surgical risk. In fact, attempts at resection have had relatively high morbidity with little benefit. Surgical shunting may be beneficial if pressure symptoms or progressive dementia are secondary to hydrocephalus. The extracranial mass can be partially removed if cosmesis is an issue. Epilepsy, the commonest symptom produced by the tumor, is unlikely to be relieved by surgery and should be treated by anticonvulsant therapy. Our patient was also planned for conservative management and was put on regular follow-up.

However, recently some surgeons plan for extirpation of the tumor by microsurgical technique sparing the diencephalic, cingulate vessels, and dissecting the tough tumor by means of the CO₂-Laser.

Conclusion

To conclude, a patient presenting with scalp lesion should be evaluated thoroughly as it may be a pointer to an intracranial brain mass, irrespective of the age group. This was a rare presentation of a benign lesion presenting in such an age group probably because of ignorance.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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