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James Ayokunle Balogun

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Intracranial lipoma associated with a subcutaneous lipoma. A rare entity

Toyin Ayofe Oyemolade[1], James Ayokunle Balogun[2]

[1] Division of Neurosurgery, Department of Surgery, Federal Medical Centre, PMB 1053, Owo, NIGERIA
[2] Division of Neurosurgery, Department of Surgery, College of Medicine, University of Ibadan and Department of Neurological Surgery, University College Hospital, Ibadan, NIGERIA

ABSTRACT

Intracranial lipomas are rare, frequently asymptomatic, congenital malformations. They are most commonly located in the pericallosal region and are often detected incidentally during neuroimaging studies or postmortem examinations. While other associated brain malformations, most notably callosal agenesis, are frequently reported, association with a subcutaneous scalp lipoma is extremely rare. We present a case of pericallosal lipoma associated with callosal agenesis and subcutaneous lipoma over the anterior fontanelle in a 6-month-old female infant who had excision of only the extracranial mass and has remained asymptomatic from the intracranial mass for the 3 years of follow up.

Keywords
intracranial,
lipoma,
subcutaneous

Corresponding author:
Toyin Ayofe Oyemolade

Department of Surgery, Federal Medical Centre,
Owo, Nigeria

toyinmolade@yahoo.com

INTRODUCTION

Intracranial lipomas are rare benign congenital lesions[1]. They are most commonly located in the deep interhemispheric fissure especially in the pericallosal region[2]. Intracranial lipomas are frequently asymptomatic and are therefore usually detected incidentally during neuroimaging studies or postmortem examinations[3, 4]. They are often managed conservatively because the surgical risks outweigh the benefits[1, 5, 6, 7]. They are frequently associated with other brain malformations, agenesis or dysgenesis of corpus callosum being the most common[7]. Extremely rare however is the association of intracranial lipoma with a subcutaneous lipoma[8, 9]. We present a case of an asymptomatic giant intracranial lipoma associated with corpus callosal agenesis and a midline frontoparietal subcutaneous lipoma.

CASE PRESENTATION

A 6-month old female infant was referred to us with a midline frontoparietal subcutaneous mass. The mass was noticed at birth and was increasing in size progressively. The child was otherwise well; the developmental milestones were within normal limits. The neurological examination findings were also essentially normal. The mass measured

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8cm by 6cm by 5cm with the anterior half over the anterior fontanelle (Fig. 1). Other systemic examination findings were normal. Brain MRI showed a large inter-hemispheric pericallosal and a fronto-parietal subcutaneous mass, hyper intense on T1 (Fig. 2) and T2 weighted images but hypointense on T2* and fat suppression sequences (Fig 3) consistent with lipoma. There was no connection between the intra and extra-cranial masses. The extracranial mass was excised completely. No cranial defect was found at surgery, confirming no connection between the extra-cranial and intra-cranial masses. The excised subcutaneous mass was confirmed to be a lipoma on histology. She is currently being followed up at the outpatient clinic. She remains asymptomatic 3 years after surgery and her developmental milestones are within normal limits.

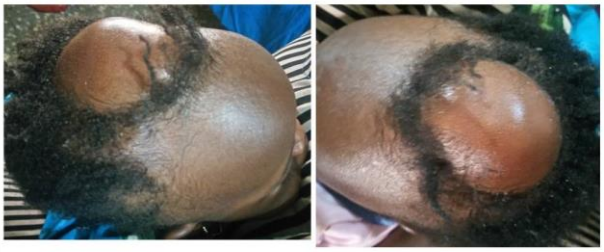

Figure 1. Clinical photograph showing the frontoparietal subcutaneous lipoma.

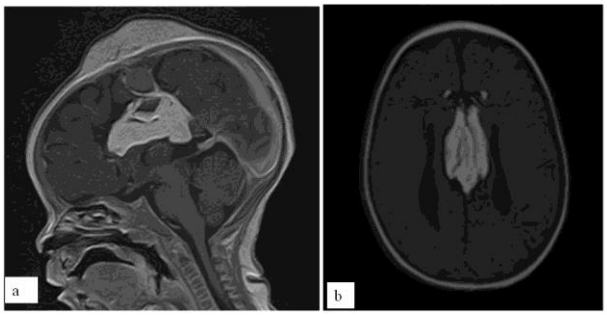

Figure 2. A sagittal (a) and axial (b) T1weighted MRI showing hyperintense frontoparietal subcutaneous and an interhemispheric pericallosal masses.

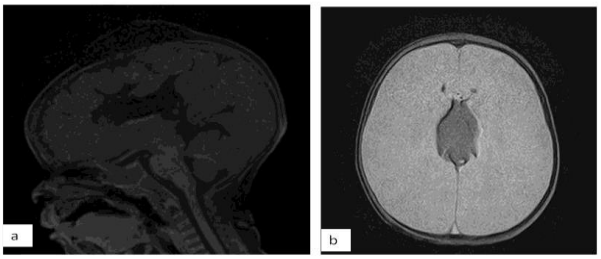

Figure 3. Sagittal T1 fat suppressed (a) and axial T2* weighted MRI images showing hpyointense lesion consistent with lipoma.

Discussion

Intracranial lipomas are rare benign congenital lesions accounting for 0.06% to 0.46% of all intracranial tumours[1]. They result from abnormal persistence and mal-differentiation of the meninx primitive during the development of the subarachnoid cisterns and hence are congenital anomalies rather than true neoplasms[10]. About 45% of cases occur in the pericallosal region, 25% in the quadrigerminal cistern and 15% in the suprasellar cistern. Other locations include cerebellopontine angle cisten (9%), Sylvian cistern (5%) and rarely on the surface of the cerebral hemispheres[8, 11, 12, 13]. The callosal lipomas can be divided into two types: a bulky tubulonodular anterior variety which is associated with forebrain and rostral callosal anomalies and a ribbon-like curvilinear posterior lipoma with a normal or nearly normal corpus callosum[14].

More than half of intracranial lipomas are associated with other malformations of the central nervous system including callosal agenesis or hypogenesis (the most common), encephalocele, spinal bifida, vermian hypoplasia, absent septum pellucidum and cortical malformation[1, 13]. Interhemispheric lipoma associated with a subcutaneous lipoma as in our patient is however extremely rare[8]. In these cases, the intra and extracranial masses may have no connection with each other, may be connected by a fibrous lipomatous stalk or may be continuous with each other through cranium bifidum[6, 15, 16, 17].

Intracranial lipomas are usually asymptomatic, often discovered only incidentally on neuroimaging[3, 4]. When symptomatic, symptoms depend on the location of the lipoma and may include epilepsy, persistent headache, ataxia, psychomotor retardation and cranial nerves deficits[7, 18, 19, 20]. The imaging findings in intracranial lipomas are characteristic. On cranial CT scan, lipomas are markedly hypodense (density of -50 to -100 HU) with frequent areas of calcifications, the latter being more common in the pericallosal lipomas[4, 7]. On MRI, they have high intensity on T1 and T2 weighted images and low intensity on T2* weighted and fat suppression images without contrast enhancement[21, 22].

Radical surgical excision is usually contraindicated because of attendant high morbidity and mortality due to high vascularity and strong adherence to surrounding tissues[6, 7]. Stable or asymptomatic lesions are managed conservatively[1, 5]. In patients with epileptic seizures, anticonvulsant therapy is the treatment of choice[23]. Our patient had excision of only the subcutaneous fat and is being followed up at the outpatient clinic.

Conclusion

Intracranial lipomas are rare congenital malformations and are often asymptomatic. Intracranial lipoma associated with subcutaneous lipoma is extremely rare. Children with subcutaneous scalp lipoma should have brain imaging to look for possible associated intracranial lipoma and other associated anomalies.

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