Epileptic seizures associated with intracranial lipoma in a 13 month old girl

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Abstract

Intracranial lipomas are congenital malformations that are usually incidental but can rarely be symptomatic manifesting as headaches or epilepsy.

We are presenting a case of a previously healthy 13 month-old girl who presented with unprovoked generalized tonic-clonic seizures. She was found to have intracranial pericallosal lipoma with some areas of calcification on brain imaging studies that included an MRI and CT scan. EEG was normal on two occasions. She was started on Keppra. Trileptal then was added due to the continuation of her seizures. However, that was discontinued due to hyponatremia. Onfi was added after which she continues to be mostly seizure free.

The association between intracranial lipomas and seizures has been debated, but has been reported mostly with sylvian more than pericallosal location, as in our patient. Seizures can become refractory to anti-seizure medications. Surgery is associated with morbidity and mortality due to the high vascular nature of lipoma.

Introduction

Intracranial lipomas are congenital malformations of mesenchymal origin. They are frequently located around the midline, mostly in the pericallosal cistern. Other locations include the superior cerebellar peduncle, quadrigeminal plate, sylvian cistern, suprasellar cistern and cerebellopontine angle cistern[1].

More commonly, intracranial lipomas are incidental findings but they can rarely be symptomatic manifesting as headaches or epilepsy[2].

Intracranial lipomas can be radiographically similar to other lesions such as dermoid cysts, hamartomas, and gliomas, and differentiation from these condition is important for appropriate therapeutic planning [3-4].

The Case: History & Physical Exam

This is a 13 month-old female infant with no known significant past medical history who presented with a history of 2 episodes concerning for seizures. The first event happened during sleep and involved stiffening of upper and lower extremities followed by shaking and up-rolling of the eyes that lasted for 1.5 minutes where she was unresponsive.

The second event happened the next morning with a similar description and while the patient was asleep. She was hospitalized at that . There was no history of head trauma, meningitis or encephalitis. There was no concern about her development. Past medical history: sleep terrors since the age of 9 months. Birth history: She was born at tem via normal vaginal delivery without perinatal complications. Birth weight was 6 lb and 8 oz. Family history: was positive for febrile seizures in her mother and history of seizures in the maternal uncle and maternal great grandfather. Her growth parameters and vital signs were normal. Her physical exam was notable for a flat nasal bridge, epicanthal folds, telecanthus, one small cafe au lait spot over the left chest area, and widely spaced nipples.

Investigations & follow up

During her hospital admission, labs including her CBC, electrolytes, liver enzymes, respiratory viral panel, urinalysis, urine toxicology screen were all unremarkable. An EEG done during wakefulness was normal with no focal slowing or epileptiform discharges. Her MRI showed linear, hyperintense T1 (Figure 1), hypointense FLAIR lesion at the superior aspect of corpus callosum and located on both sides in the parasagittal location representing tubulo-nodular pericallosal lipoma. No acute intracranial hemorrhage, mass effect or midline shift. No abnormal contrast enhancement. Due to bright signal intensity on T1 weighted images,CT scan head without contrast (Figure 2) was obtained and showed a linear fat containing lesion in the pericallosal region representing lipoma. Additionally, dense calcification was seen involving falx anteriorly. No definite evidence of acute intraparenchymal hemorrhage, mass effect or midline shift. Patient was discharged home on Keppra. Further workup including Chromosomal Microarray was normal. Epilepsy Gene Panel showed 1 pathogenic variant identified in the CLN3. There was also a variant of unknown significance identified in ALG13. None of these genetic abnormalities was thought to be causing her seizures. Recommendations were parental testing for the ALG13 variant and CDT and N-glycans to assess for evidence of congenital disorder of glycosylation given the ALG13 variant; that was normal. Upon follow up at 17 month she was noted to have subtle motor delays and she was enrolled in the Early On program. Until the age of 21 months, she continued to have some intermittent breakthrough seizures. Keppra dose was subsequently increased. EEG was repeated and was normal. After Keppra, Trileptal was added. However, that was discontinued after she developed hyponatremia; then Onfi was added.

Her seizures continued to be under fairly good control on the combination of keppra and Onfi with only occasional breakthrough seizures. There was no concern about her development during the follow up visit at the age of 2 year.

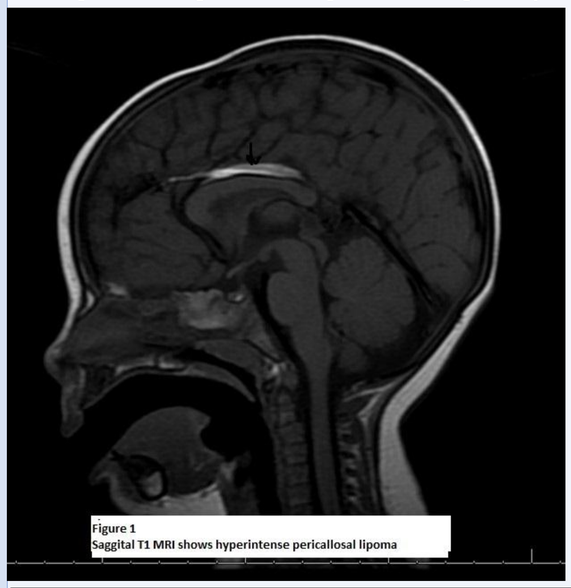

Figure 1
Saggital T1 MRI shows hyperintense pericallosal lipoma

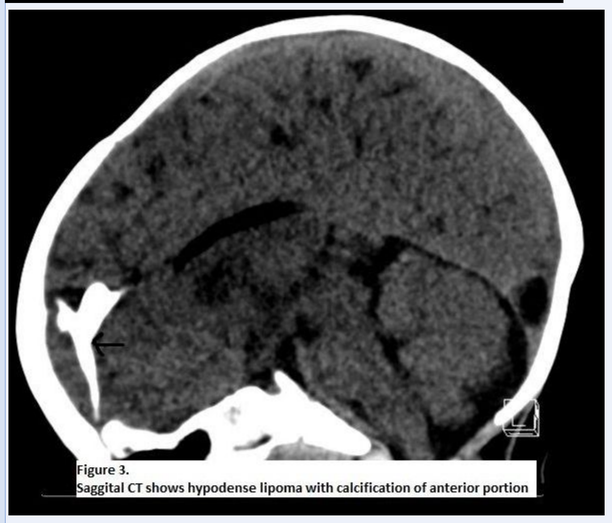

Figure 3.
Saggital CT shows hypodense lipoma with calcification of anterior portion

Discussion

Intracranial lipomas account for only 0.1–0.5% of all primary brain tumors[4]. They were first described in 1856 by von Rokitansky who considered them as benign, slow-growing congenital hamartomatous conditions[5].

Intracranial lipomas are thought to be a result of persistence of the primitive meninx followed by transformation into mature adipose cells during the initial weeks of intrauterine life. lipomas may have secondary or concomitant effects on the development of adjacent cortical structures, which happen between the sixth and twentieth weeks [6]. It may interrupt the migration of gray matter[7]. MR signals for lipomas are usually typical for adipose-rich tissues[1]. Pericallosal lipomas can be associated with corpus callosum agenesis[8]. However, our patient had intact corpus callosum.

The vast majority of intracranial lipomas are incidental findings on brain imaging done for other causes[9]. The presenting symptoms include epilepsy, headache, developmental delay and cranial nerve palsies[10]. Seizures related to lipomas can be generalized or partial seizures with impaired consciousness[11]. Although our patient had subtle dysmorphic features and abnormal epilepsy gene panel, her generalized seizures could not be attributed to her genetic abnormalities as both the identified genes were not known to cause epilepsy. Seizures may become intractable and surgery as a treatment option may be considered. Subtotal resection resulted in complete reversal of the preoperative symptoms as per Shinozaki et al [12].

In conclusion, although rare and commonly asymptomatic, pericallosal lipomas should be considered in the differential diagnosis in children with seizures.

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