Case Report

A giant interhemispheric lipoma and corpus callosum agenesis

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ABSTRACT

The lipomas of the central nervous system are rare lesions and are often located in the midline and in the corpus callosum. They are rarely symptomatic and account for 0.06-0.46 % of intracranial lesions. We report a 5 year old girl who 'was admitted to the pediatrics clinic due to an intractable complex partial seizure. Biochemical tests for seizure etiology did not reveal any specific cause. Electroencephalography (EEG) showed minimal epileptic activity. Magnetic resonance imaging (MRI) showed that there was a 50x36 mm in diameter mass, and corpus callosum agenesis. Her seizures were controlled with carbamazepine (10mg/kg/day). Although intrahemispheric lipomas are rare, they should be considered in the differential diagnosis of non-febrile seizures in children.

KEY WORDS: Intrahemispheric lipoma, Corpus callosum agenesis, Children.

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INTRODUCTION

Lipomas are the most common benign tumors of the adipose tissue. Although these tumors are frequently observed elsewhere on the body, lipomas are rare benign tumors of the central nervous system that accounts for 0.06-0.46% of intracranial tumors.[1] The lesions are often asymptomatic; they are frequently diagnosed incidentally during radiological investigations or at autopsy.[2] In this report management of a child presenting with the diagnosis of a giant interhemispheric lipoma and corpus callosum agenesis is discussed with the relevant literature.

CASE REPORT

A five years old female child was admitted to pediatric emergency clinic with the complaint of intractable partial seizures. On physical examination there was no other motor and sensory deficit, except anisocoria (i.e. left pupil minimally midratic). Systemic examination was unremarkable. Routine hematologic and biochemical analysis were within normal limits. The sleep electroencephalogram (EEG) demonstrated that there were isolated sharp spiked slow waves and minimal abnormal epileptic activity. On cranial MRI, posterior and splenium of corpus callosum were not present; in addition there was 50x36 mm hyperdense lipomatous mass at the anterior of corpus callosum on T1-weighted images. Bilateral anterior cerebral arteries were coursing through the mass and were giving their branches within the lipoma (Figure-A, B). The patient was examined in the neurosurgery and ophthalmology department. On ophthalmic examination there were neither papilledema nor visual field defects but left pupil was anisocoric. The patient was treated with carbamezepime (10mg/kg/day), her seizures were controlled.

DISCUSSION

Intracranial lipoma was first described in 1856 by Rokitansy after a necropsy but diagnosis of the lesion in living patient was not made until 1939.[3] There is no sex predilection but cerebellopontine angle lipomas appear twice as common in males.[4]

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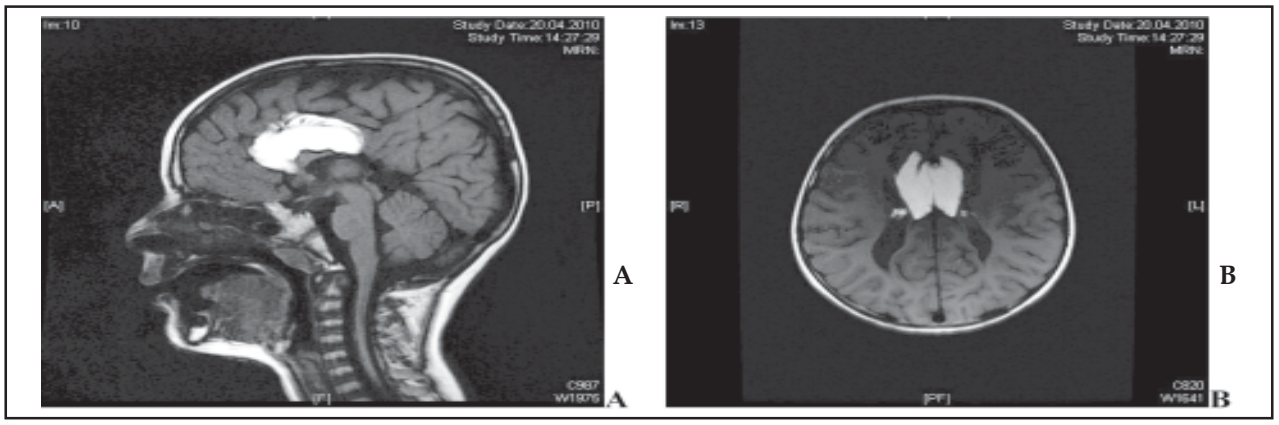

Figure-(A): Precontrast Sagittal and axial (B) T1-weighted cerebral magnetic resonance imaging scan shows a pericallosal lipoma. The corpus and splenium of corpus callosum is invisible with partial agenesis.

Although the etiology and pathogenesis of intracranial lipomas are nor clearly understood, derivation from the embryological "meninx primitiva" theory is the most plausible theory that it suggests an abnormal, persistent focus of meninx primitiva differention into adipose tissue and maturation into a lipoma.[1] Histopathological examination of lipomas reveals that they are composed of mature adipocytes, fibromuscular tissue, bone and calcifications, vessels and nerves passing through the lipoma.[1,5,6]

About 50% of the intracranial lipomas are asymptomatic.[4,7] The most common presenting symptom is epilepsy.[3,8] Epileptic seizures may be either grand mal or petit mall, but the seizures are usually severe partial type and they often appear before 15 years age.[8] Headache, vomiting, hemiplegia vertigo, mental retardation or emotional lability are the other findings.[7,9] Due to their localizations intracranial lipomas may lead to unilateral sensorioneural hearing loss[10], precocious puberty[11] or hydrocephaly.[12,13]

The rare occurrence of intracranial lipomas makes the diagnosis difficult. Since lipomas are usually asymptomatic they are diagnosed incidentally on CT or MRI evaluations or at autopsy as in our case. On CT, lipomas are sharply demarcated areas of marked hypodensity that do not demonstrate enhancement after contrast injection. Calcification is often present within the fibrous capsule of the lipoma. On MRI there is a hypointense mass on T1 and T2 -weighted sequences and it appears hypointense on FS (fat saturated) sequences.[14] Most intracranial lipomas occur in the midline in the region of the corpus callosum (45%), the ambient cistern and suprasellar region (23%).[1,6] These lesions are frequently associated with hypogenesis or agenesis of corpus callosum.[4] Intracerebral vessels and cranial nerves may cross through the mass.[1] Similarly our case showed that anterior cerebral arteries were crossing through the mass and giving its branches within the lipoma, visualized by contrast enhanced MRI images.

Because of the rarity of these neoplasms therapeutic experience with primary intracranial lipomas is also extremely limited. Surgical resection should only be considered in patient with intractable seizures, or hydrocephaly or intractable headache.[2,9,10] As intracranial lipomas cannot be treated successfully by surgical resection the ideal treatment consists of conservative approaches.[8] For this reason anticonvulsants can be used in patients with seizures as in our case.

In conclusion, although intracranial lipomas are very rarely seen, these tumors should be considered in the differential diagnosis of the intracranial tumors in children with partial seizures and non-febrile seizure.

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